Neurosurg Rev (1999) 22:67–93

REVIEW

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The scientific history of hydrocephalus and its treatment

Received: 4 August 1999 / Accepted: 12 August 1999

Summary Hydrocephalus cases were regularly described by Hippocrates, Galen, and early and medieval Arabian physicians, who believed that this disease was caused by an *extracerebral* accumulation of water. Operative procedures used in ancient times are neither proven by skull findings today nor clearly reported in the literature. Evacuation of superficial intracranial fluid in hydrocephalic children was first described in detail in the tenth century by Abulkassim Al Zahrawi. In 1744, LeCat published findings on a ventricular puncture.

Effective therapy required aseptic surgery as well as pathophysiological knowledge – both unavailable before the late nineteenth century. In 1881, a few years after the landmark study of Key and Retzius, Wernicke inaugurated sterile ventricular puncture and external CSF drainage. These were followed in 1891 by serial lumbar punctures (Quincke) and, in 1893, by the first permanent ventriculo-subarachnoid-subgaleal shunt (Mikulicz), which was simultaneously a ventriculostomy and a drainage into an extrathecal low pressure compartment. Between 1898 and 1925, lumboperitoneal, and ventriculoperitoneal, -venous, -pleural, and -ureteral shunts were invented, but these had a high failure rate due to insufficient implant materials in most cases. Ventriculostomy without implants (Anton 1908), with implants, and plexus coagulation initially had a very high operative mortality and were seldom successful in the long term, but gradually improved over the next decades.

In 1949, Nulsen and Spitz implanted a shunt successfully into the caval vein with a ball valve. Between 1955 and 1960, four independent groups invented distal slit, proximal slit, and diaphragm valves almost simultaneously. Around 1960, the combined invention of artificial valves and silicone led to a worldwide therapeutic breakthrough. After the first generation of simple differential pressure valves, which are unable to drain physiologically in all body positions, a second generation of adjustable, autoregulating, antisiphon, and gravitational valves was developed, but their use is limited due to economical restrictions and still unsolved technical problems. At the moment, at least 127 different designs are available, with historical models and prototypes bringing the number to 190 valves, but most of these are only clones.

In the 1990s, there has been a renaissance of endoscopic ventriculostomy, which is widely accepted as the method of first choice in adult patients with aquired or late-onset, occlusive hydrocephalus; in other cases the preference remains controversial. Both new methods, the second generation of valves as well as ventriculostomy, show massive deficits in evaluation. There is only one randomized study and no long-term evaluation.

Key words Hydrocephalus · Medical history · CSF shunts · Hydrocephalus valve · Silicone · External CSF drainage · Shunt failures · Shunt infection

History of the discovery of normal and disturbed CSF circulation

Hydrocephalus is such a common illness and has such obvious manifestations in children that early man can hardly have failed to take note of it or realize that it usually ended in death. Richards's [151] survey of the published literature on pathologic findings in skeletons dating from the period 2500 BC to 500 AD mentions numerous hydrocephalic skulls. The most prominent of these may be that of Pharaoh Ikhnaton [59].

The earliest scientific description of hydrocephalus is ascribed to Hippocrates (466–377 BC), who mentions such symptoms as headache, vomiting, visual disturbance, and diplopia, and explains the illness as a liquefaction of the brain caused by epileptic seizures. The Hippocratic body of works also contains the first use of the term "hydrocephalus"[1], although in ancient times its

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[1] The word is constructed from the Greek words "ὕδωρ" (hydor, water) and "κεφαλή" (kefalé, head).

"وكثيرا ماتعرض الرطوبة للصبيان عند الولادة اذا ضغطت القابلة رأس الصبي بغير رفق
او يكون من علة خفية لاتعرف ...... والرأس يعظم كل يوم حتى لم يطق الصبي أن يقلل
عظم رأسه..... والعمل في ذلك، فينبغي أن يشق في وسط الرأس ثلاث شقوق، وبعد الشق
تخرج الرطوبة كلها، ثم نشد الشق بالخرائق والرفائد....."

Fig. 1 Abulkassim Al Zahrawi (936–1013): facsimile in Arabic with clinical description of hydrocephalus and recommended surgical treatment [44]. Original manuscript is in the library of Patna, India

meaning was considerably different from that of today: it referred originally to fluid collections surrounding the brain and thus, in modern terminology, to subdural hygromata, dilatations of the intracranial subarachnoid space, and arachnoid cysts adjacent to the calvaria. It has often been claimed that Hippocrates, who was quite familiar with trephinations of other kinds[2], might well have punctured the subdural space or even the ventricles by way of the fontanel. This remains no more than speculation [6] and seems improbable, in view of the deficient anatomical knowledge of the time and the general preference of a conservative therapy.

Claudius Galen of Pergamon (130–200 AD) acquired relatively good knowledge of ventricular anatomy through animal dissection and vivisection. He gave the first accurate description of CSF as a clear, watery liquid, as well as of the foramen of Magendie [174]. At this early date, he even hypothesized that CSF was produced in the choroid plexus. However, he also assumed further contribution by the "rete mirabile", by which he probably meant the plexus cavernosus, a large structure in animals that reaches down to the clivus. Galen assumed that the CSF flowed either by way of the infundibulum and pituitary gland into the throat or through the cribriform plate into the nasal cavity. He attributed to it the transport of "pneuma," a hypothetical vital force.

Galen was the first to formulate an anatomical–pathological classification of fluid collections in the head: "There are four kinds of hydrocephalus: between the brain and the meninges, between the meninges and the bone, between the bone and pericranium, and between the bone and the skin. We treat hydrocephalus between the skin and the pericranium with two or three free incisions; that between the meninges and the brain is incurable" (translated by Quin 1814) [174].

Galen was allowed to dissect only animals, not human cadavers, and thus he apparently knew only of fluid collections *outside* the brain, which he, like Hippocrates, designated as hydrocephalus. The Byzantine physicians, too, including Paul of Aegina and Aetius, described infantile hydrocephalus in similar terms and did not go beyond this interpretation.

In late antiquity, as medical knowledge deteriorated in the West, Arab and Persian physicians continued to hand down and build upon the discoveries and writings of their ancient authors with great care for more than half a millennium. Abulkassim Al Zahrawi (936–1013), known as Abulcasis and living in Cordoba, devoted one of his 30 treatises to neurosurgical disease, describing in it not only traumatic head injuries, fractures of the spine, and skull tumors, but also infantile hydrocephalus, which he supposed was caused by mechanical compression: "The skull of a newborn baby is often full of liquid, either because the matron has compressed it excessively or for other, unknown reasons. The volume of the skull then increases daily, so that the bones of the skull fail to close. In this case, we must open the middle of the skull in three places, make the liquid flow out, then close the wound and tighten the skull with a bandage" (text modified) [44]. The degree of detail in these instructions suggests that this mode of therapy was actually practiced (Fig. 1).

Beginning in the twelfth century, classical medicine diffused from Bagdad, Isfahan, and Cairo back into Europe. The medical school in Salerno and the universities of Padua and Montpellier played the most important roles in this process. Unfortunately, scholastic canonization of the reappropriated ancient writers, especially Galen, continued to hinder scientific innovation based on anatomic studies and led to no improvement in medical practice.

New discoveries became possible only in the Renaissance, when observation gained the upper hand against speculation and the dissection of human cadavers was first tolerated, then ultimately legalized. The first illustration of the ventricular system drawn from a dissected human brain appeared in 1510 from the hand of none less than Leonardo da Vinci (Fig. 2), who had already made a wax model of the ventricular system of an ox in 1505 [59].

This illustration manifested a new anatomical realism, which, however, was not consistently maintained. Remarkably, the artist's depiction of the cerebral aqueduct is quite accurate, considering that this structure came to be described in medical literature one decade later, by Jacobus Sylvius in 1515 and Berengarius in 1521 [34]; but the infundibulum, actually very small, is depicted as quite large and appears to extend down to the cribriform plate. Apparently, Leonardo – perhaps against his own convictions – had not yet dared to break with the teachings of Galen, who enjoyed official Church sanction, and the fourth ventricle is depicted as ending blindly. Galen's

[2] In one of his writings, Hippocrates notes with regret that he had performed trephination "to no avail" in a wrongly diagnosed case and that the patient died as a result [12].

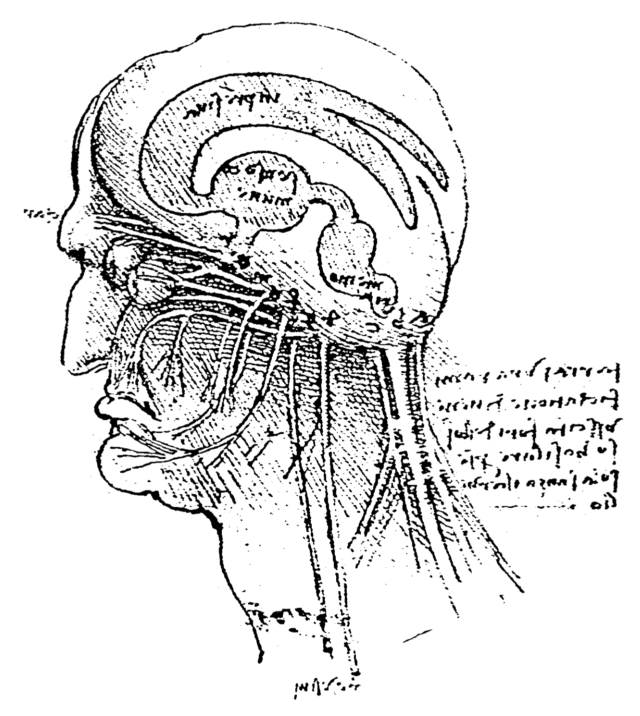


Fig. 2 Leonardo da Vinci (1452–1519): anatomical study of the brain, its nerves, and the male genitalia, 1510 (detail). From the collection of the Schlossmuseum, Weimar, Germany. Note the realistic depiction of the ventricular system, particularly of the aqueduct, which was first described 5 years later in 1515. In Leonardo's drawing, the infundibulum of the third ventricle is depicted in exaggerated proportion and reaches down to the skull base in accordance with the officially sanctioned teaching of Galen

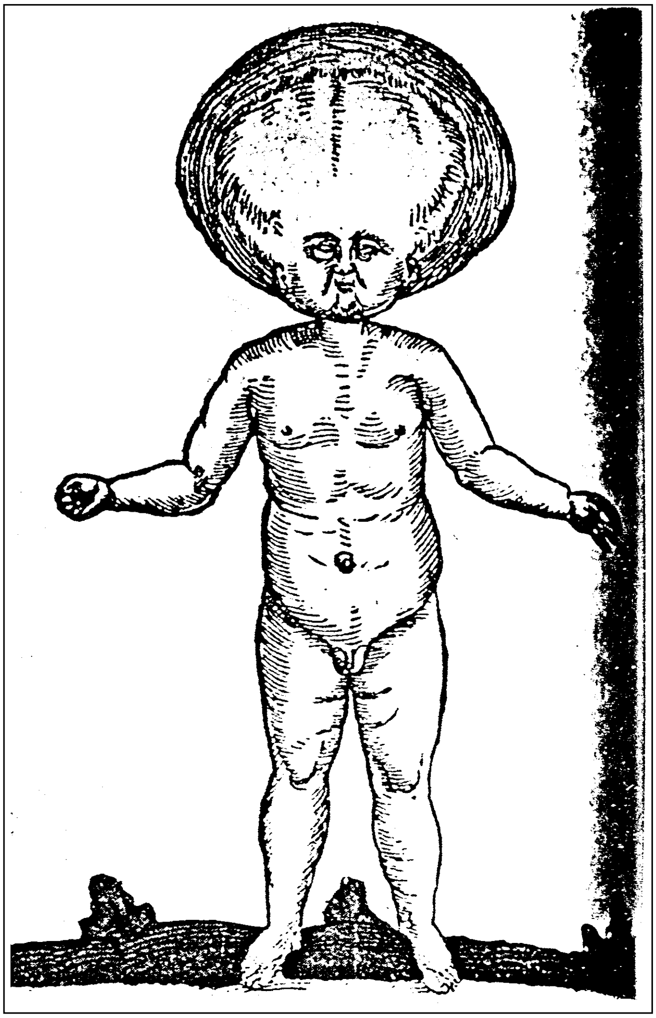


Fig. 3 Hydrocephalus, popular pamphlet, Nürnberg 1556 [120]

description of the foramen of Magendie had been overlooked. Leonardo, like all physicians until the nineteenth century, had no knowledge of the outlets of the fourth ventricle.

The next step was taken a generation later by Andreas Vesalius (1514–1564). He openly questioned Galen's view of the outflow of CSF through the base of the skull, although he himself had not yet discovered its actual outlets [6].

Vesalius's epochal achievement came in 1551 with the first scientific description of hydrocephalus based on a human necropsy (second edition of *De Humani Corporis Fabrica Libri Septem*, 1555):

"I observed [a disease] in Augsburg in a 2-year-old girl whose head had grown in 7 months more or less to a size that was not surpassed in bulk by any man's head I ever saw. This disease was what ancients called *hydrocephalus*, from water which is stored in the head and gradually collects. In this girl's case, however, the water had not collected between the skull and its outer, surrounding membrane or the skin, where doctors' books teach that water is deposited in other cases, but in the right and left ventricles of the cavity of the brain itself. The breadth of these cavities had so increased and the brain itself was so distended that they contained about 9 pounds of water, or 3 Augsburg wine measures, so help me God [174]... Just as the brain itself at the vertex was membrane-like in thinness, indistinguishable from its own membranous covering, so was the skull membranous" [144].

In contrast to the greatly altered cerebral cortex, Vesalius found that "... the base of the skull was in correct proportion to that of the young child before her head took on abnormal proportions. Nevertheless, the cerebellum and entire base of the skull were in their natural state; and so also were the extensions of the nerves". He stated definitively that he "found no water in any other places but the ventricles of the brain, which were enlarged to the extent that I have stated" [174]. These observations laid to rest the 2000-year-old misinterpretation of hydrocephalus as a collection of fluid outside the brain and cleared the way for further study of the circulation of the CSF and its pathophysiology.

Vesalius examined the patient while she was still alive and, like many after him, was fascinated to observe "that the girl had full use of all her senses," and that "such a great force of water had been accumulated for so long in the ventricles of the brain without more extensive symptoms" [175].

Thomas Willis (1621–1675) of Oxford viewed the ventricles as spaces that received the excretions of the brain tissue. His collaborator Richard Lower (1631–1691) had shown in injection studies that the cribriform plate is watertight and thus cannot be the site of exit for CSF from the brain. Knowing this, Willis was the first to postulate that, similarly to blood circulation, which had been discovered by his teacher, William Harvey, CSF must flow into the venous system.

Pacchioni (1665–1726) first described in 1701 the arachnoid granulations named after him, but still assumed they were a site of CSF secretion. As for the resorption of CSF, he supposed that the brain was surrounded by a rhythmically contracting muscle propelling the "lymph" of the brain into the venous sinuses by way of "lymph nodes" [34]. The resorptive function of Pacchionian granulations and the flow of CSF through them into the venous sinuses were discovered by Fantoni in 1738.

In 1769, 120 years before Chiari, Giovanni Battista Morgagni (1682–1771) performed autopsies and published descriptions of several cases of hydrocephalus, including one combining hydrocephalus, low lying cerebellar tonsils, and hydromyelia. Albrecht von Haller (1708–1777) discovered the foramina of Luschka and, in a treatise published in 1747, was the first to present the modern theory of CSF circulation [174, 175], although he could not prove it.

Appearing in 1842, Francois Magendie's (1783–1855) anatomical studies of the pachymeninges in the posterior fossa and spinal canal contained a new description of the caudal opening of the fourth ventricle, which had been discovered by Galen but later went unrecognized by Vesalius, Willis, and others. He proposed a "reverse theory" of CSF circulation in which fluid was produced at the brain surface, flowed into the ventricles through the foramen of Magendie, and was resorbed by the choroid plexus. Modern medical-historical research thus tends to regard him as an obstacle to progress rather than a contributor [175]. Nonetheless, it is to him that we owe the hypothesis that occluded CSF pathways can cause hydrocephalus [182], which was definitively proven by Hilten in 1879. He was also, in 1841, the first to measure CSF pressure, employing suboccipital puncture in a dog.

Although many anatomical and physiological discoveries had been made earlier and many important details added by Magendie, Hubert von Luschka (1820–1875), and Vinzenz Alexander Bochdalek (1801–1883), the "paradigm shift," representing a definitive breakthrough in modern physiological theory of CSF circulation, was first performed in 1875 by Ernst Axel Hendrik Key (1832–1901) and Magnus Gustav Retzius (1842–1919) of Sweden [96]. Their work irrefutably proved that the CSF is secreted by the choroid plexus, flows out of the ventricular system, and is resorbed through the subarachnoid villi and Pacchionian granulations (Fig. 4). Their classical study of CSF circulation remains largely valid today.

The earliest systematic, clinical neurological studies of hydrocephalic patients were performed by Robert Whytt of Edinburgh (1714–1766). Among other things, in 1768 he described differences in the clinical course of the disease that depend on whether the infant patient had open or closed sutures. Diaphany, the transillumination of a fluid-filled skull when the cortical mantle is extremely attenuated, was discovered by Bright in 1831 [34], and the "cracked pot sound" by Macewen in 1893 [1].

Many early reports of ventricular puncture contained rough estimates of CSF pressure, but Quincke was the first to measure it precisely in 1891 with a water column manometer in both the ventricle and the spinal sac (Fig. 5) [134]. Walter Dandy and Kenneth Blackfan of the Johns Hopkins Hospital (Baltimore) created the first animal model of hydrocephalus in 1913 [29] by blocking the aqueducts of dogs with small pieces of cotton (Fig. 6).

They went on to occlude selectively the right and left foramina of Monro. Dandy also demonstrated that animals subjected to such occlusions would not develop hydrocephalus if the choroid plexus had been excised [26]. Further experimental landmark studies were made by Weed in the 1920s and by Bering, Sato, Davson, Pappenheimer, Rubin, Welch, Milhorat, and Raimondi in the 1960s and 1970s.

History of the treatment of hydrocephalus

Once hydrocephalus had been identified as a mechanical, hydraulic disorder, it became clear that its treatment could theoretically be effected by any of three means:

1. Reducing CSF production by deactivating the choroid plexus by surgical removal, pharmacotherapy, or radiation
2. Reopening intracerebral blocked fluid pathways with a bypass or surgical removal of the causative lesion
3. Increasing the resorptive capacity, for example, by shunting CSF into body cavities of normally low pressure

The necessary prerequisites for neurosurgical intervention were provided by the development of general anesthesia (Morton 1846), aseptic technique (Semmelweis 1847, Lister 1867), and neurological localization of brain functions (Jackson, Broca, Wernicke).

Conservative treatment: bandaging, diet, drugs, and radiation

Probably the oldest treatment of hydrocephalus consisted of tightly bandaging the highly deformable infant skull to reduce its size. Such attempts were abandoned at the turn of the century because of their inefficacy and the accompanying danger of increasing intracranial pressure.

Most publications in the eighteenth and nineteenth centuries recommended diets and dehydration cures us-

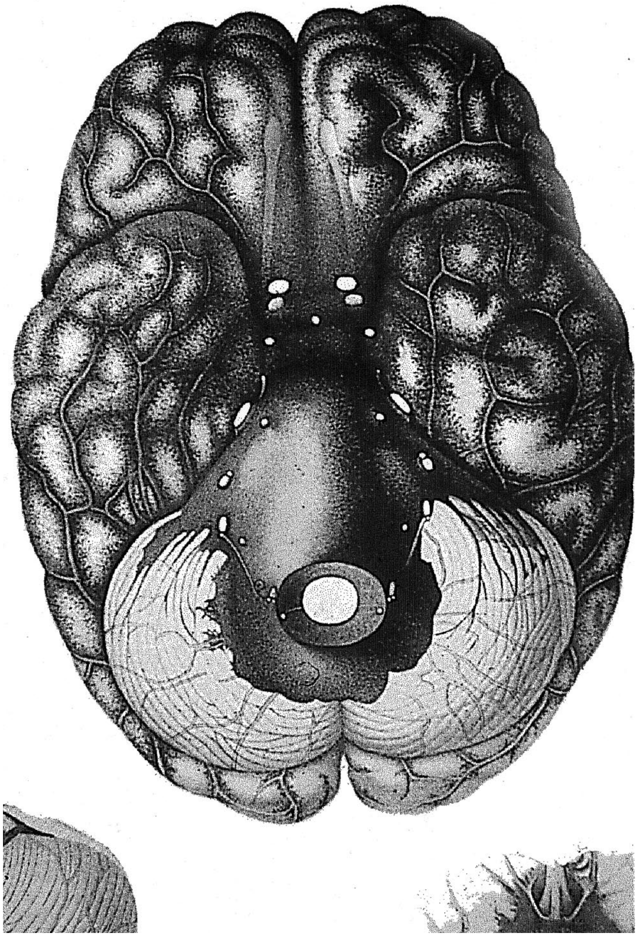


Fig. 4 CSF cisterns, india ink injection. Key and Retzius 1875 [96]

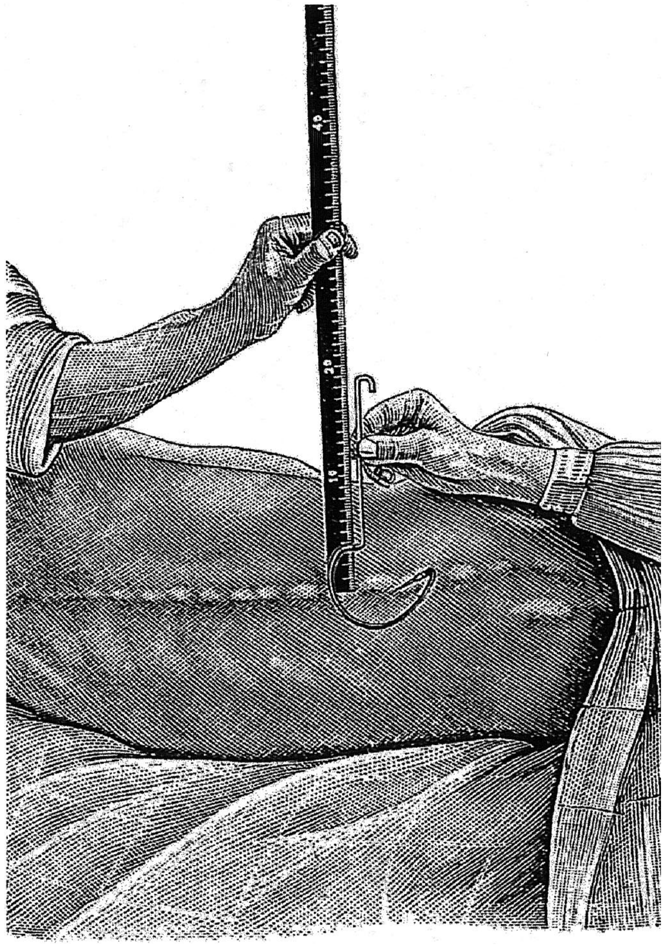


Fig. 5 Quincke, original drawing of a lumbar manometry, 1891 [143]

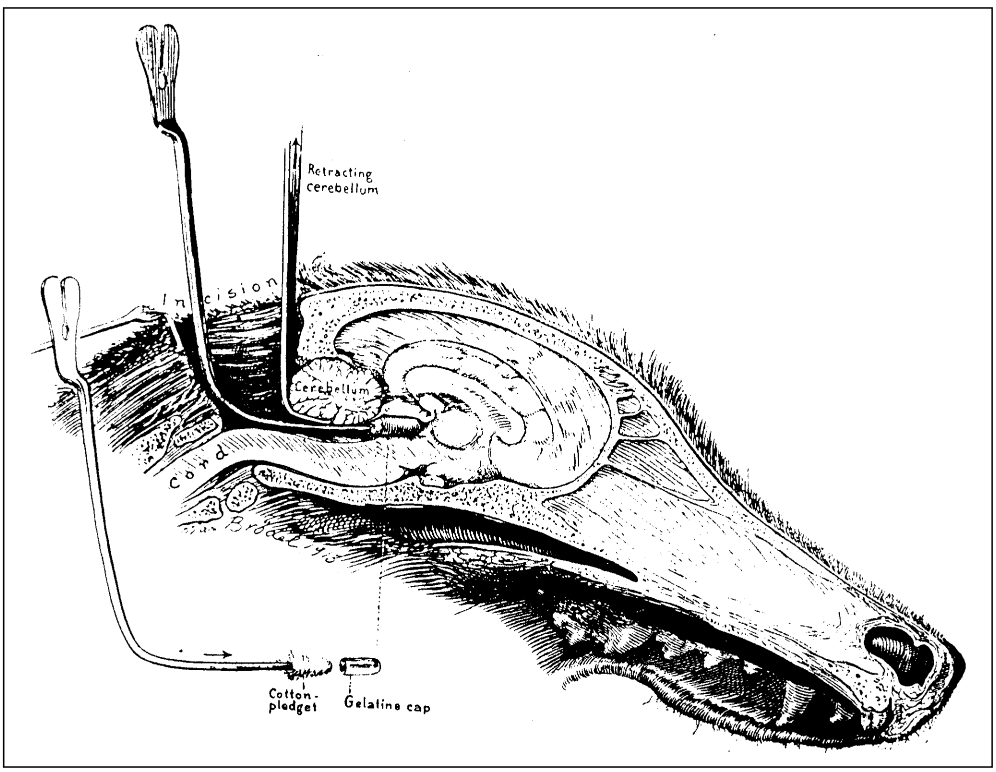


Fig. 6 Occlusion of the aqueduct of a dog with cotton pieces to produce occlusive hydrocephalus. Illustration by Dandy and Blackfan, 1913 [29]

ing laxatives, diuretics, potassium iodide, mercury preparations (calomel), local "drawing ointments," etc. [79, 175]. On rare occasions, patients with hydrocephalus even received (and survived) intraventricular iodine injections. In Henle's [79] summation in 1896: "Regarding the medical treatment of hydrocephalus, one can only say that its benefit is frequently doubtful and, unfortunately, in most cases nonexistent."

Nonetheless, one small kernel of sense remains from all these attempts. In 1954, Tschirgi showed that acetazolamide reduces CSF production in experimental animals. Elvidge began using this substance to treat hydrocephalus in 1957 and it has since remained in use as an adjuvant mode of treatment [35]. Isosorbide was put forward in 1966 by Wise as another means of achieving the same effect, but it never came into general clinical use [35]. CSF production can also be reduced by irradiating the choroid plexus, but this treatment was abandoned because of its severely adverse effects on the central nervous system [179].

In summary, except for the use of acetazolamide as a temporary measure in mild cases, conservative treatment plays no role in hydrocephalus treatment today.

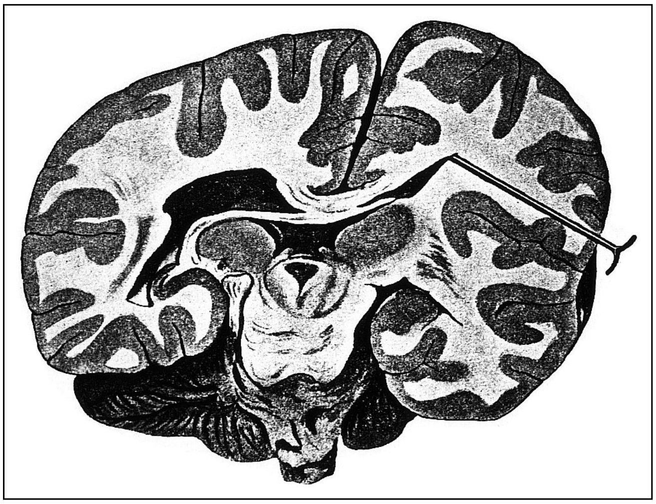


Fig. 7 Split trocar cannula used for VP, EVD, and ventriculosubgaleal drainage [100]

Extracorporeal CSF drainage: punctures of ventricle, lumbar sac, and CSF reservoirs, and external drainage

Ventricular puncture. Trephination of the skull was performed all over the ancient world beginning in neolithic times. The earliest unequivocal evidence of it is reported from the necropolis of Taforalt (Morocco) and has been dated to approximately 10,000 BC [30]. These specimens manifest completed callus formation around the edges of the trephination, which implies that the patients must have survived for a considerable time after the procedure. Similar findings, with survival rates of 80–90%, have also been reached from observing numerous ancient Eurasian and pre-Columbian American skulls [44, 146]. In Anatolia, even osteoplastic trephinations were found [47]. Although the "indications" were probably magical in most cases, such as for releasing evil spirits from the head, there have always been rational, medical grounds for such procedures, such as elevating depressed skull fractures and evacuating hematomata or epidural empyemas.

The worldwide use of trephination makes it seem likely that there were early attempts to treat hydrocephalus by punctures or open surgery. This has, however, not definitively been proven. The same qualification applies with regard to the abovementioned treatise of Abulkassim Al Zahrawi.

The first historically documented ventricular puncture (VP) was performed on 23 October 1744 by Le Cat. Furthermore, he left a wick in place for some time after the procedure and can be regarded as the inventor of external ventricular drainage [74].

Nearly all the VPs mentioned in publications from the eighteenth and nineteenth centuries ended fatally [74, 79, 128], except for one published by Fantoni in 1769, which is stated to have led to a permanent cure [175].

The history of VP under modern, aseptic conditions begins with Wernicke, who punctured the trigone with a trocar using a lateral approach in 1881 [190]. With a pioneer's enthusiasm, he wrote in his textbook: "When performed with aseptic precautions, this operation is intrinsically perfectly safe". This trigonal approach was simultaneously taken up by Broca, Keen, and Quincke in 1891 and still has certain applications today, whereas the transorbital VP performed by Langenbeck in 1850 was not pursued further. The frontopolar approach described by von Bergmann is still used occasionally. The coronal puncture originated by Kocher and the occipital puncture described by Krause and Dandy [25, 100] are the most commonly used today.

By the turn of the century, numerous studies of VP in cases of hydrocephalus had been published. Permanent therapeutic benefit was sometimes reported, but there were often severe complications, mainly infections. In 1902, Oppenheim responded with unconcealed skepticism to Henschen's review of 63 cases of ventricular puncture: 24 of the patients died in the aftermath of the operation, 12 derived no benefit from the treatment, and only a few showed improvement [128].

After technical improvements and the introduction of silver, atraumatic, split stylet cannulae by Krause in 1911 (Fig. 7) [100] and of puncture cannulae with rounded tips by Dandy and Cushing, VP became a relatively safe procedure and began to be practiced around the world.

With the invention of air ventriculography by Dandy in 1918 [25], diagnostic VP became the gold standard for diagnosis of ventricular dilatation and occlusive lesions and remained so until the era of computed tomography (CT). Pneumoencephalography has now been fully replaced by CT and magnetic resonance imaging. Contrast ventriculography is reserved for special cases [103]. In its therapeutic application as well, VP has largely been replaced by continuous drainage, although it retains an important role in emergency treatment.

External ventricular drainage (EVD). Wernicke sometimes left ventricular cannulae in place for a considerable length of time to allow external drainage [184]. His method was taken up by Pollock in 1884, Zrenner in 1886, von Bergmann in 1888, and Broca in 1891 [144]. The drainage devices used include hollow, metal needles (Krause 1911), rubber tubes (Senn 1903), guttapercha, and wicks made of catgut (Cheyne 1899), silk, or horsehair (Keen 1891).

In 1902, Oppenheim expressly warned: "VP and subsequent drainage of the ventricles as first recommended by Wernicke and applied by von Bergmann, Keen, Kocher, Broca, Robsen, and Watson-Cheyne, has proven to be particularly dangerous, as 20 of the 23 individuals so treated died." [128] "... External drainage from the skull, occasionally taken to such an extreme that the scalp, drenched in spirit of turpentine, is actually set on fire, has found advocates again lately but can hardly be considered seriously anymore" [128].

Fedor Krause, in 1911, nevertheless succeeded in draining hydrocephalic ventricles to the outside continually for a period of 8 weeks without causing CSF infection [100]. Apparently, he used his improved EVD technique only perioperatively in a small number of procedures in the posterior fossa, a practice later adopted and systematically applied by Sjöquist in 1937 [168], Poppen, and others. We owe the development away from simple open tubes and toward closed systems with pressure regulation to Ingraham in 1941 [85] and Pampus [130].

In the 1970s, EVD supplanted VP to become one of the most effective and most frequently used therapeutic measures in neurosurgical intensive medicine. Prefabricated systems became increasingly available from the medical products industry [163]. EVD has found wide application because it allows not only continuous drainage of CSF, but also the measurement of intracranial pressure, acquisition of ventricular fluid, and intrathecal administration of antibiotics, chemotherapeutic agents [123], clot-dissolving substances, and contrast agents for positive contrast ventriculography with either conventional x-ray or CT technique. The externalized distal catheterization of infected shunts is a variant of EVD [114].

Spinal external drainage. According to Haynes [74], Paget had removed the laminae of the third and fourth cervical vertebrae and created a CSF fistula to the outside as early as 1873, but this was ineffective. Lumbar CSF drainage, introduced by Vour'ch [183] in 1963, is an important type of external drainage.

Lumbar puncture (LP). The first puncture of the lumbar spinal canal was performed by Corning in 1885 for the purpose of instilling medication [74]. Wynter removed CSF via lumbar puncture in 1889 in a case of acute meningitis [74].

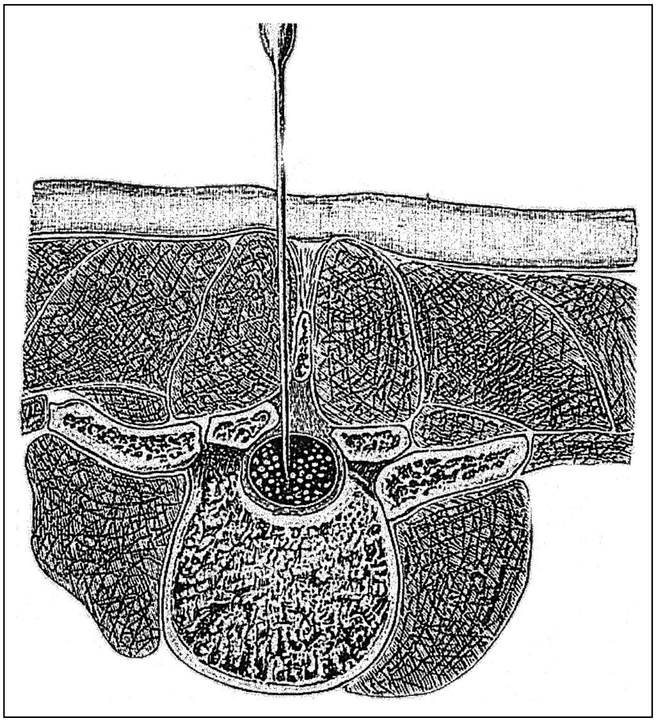


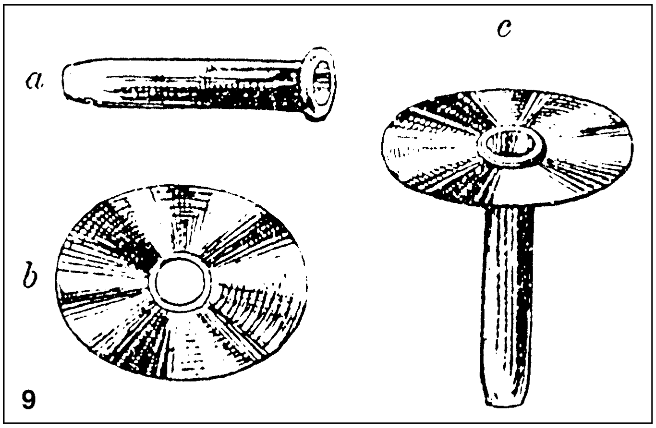
Fig. 8 Quincke, original drawing of a lumbar puncture [143]

The breakthrough, however, was provided in 1891 by Quincke, who substantially improved the technique of lumbar puncture using special, styletted cannulae and was the first to perform it systematically. For Quincke, lumbar puncture was primarily a therapeutic measure and only secondarily a diagnostic procedure (Fig. 8) [143].

One of Quincke's earliest applications of lumbar puncture (often serial punctures) was in the treatment of hydrocephalus. Temporary improvement was observed frequently and lasting benefit only occasionally. This method spread quickly after publication of Quincke's sensational reports. In 1902, Oppenheim referred to 11 publications treating hydrocephalus by lumbar punctures [128].

There were also problematic applications, such as in cases of occlusive hydrocephalus and brain tumor. In 1896, Fleischmann [51] published a rather large series of systematically performed treatments by LP in which he reported numerous deaths during or shortly after the puncture.[3] He referred to three reports entitled "Sudden Death after Lumbar Punctures" or similar. At the turn of the century, these observations gradually led to understanding of the mechanism of iatrogenic impaction of the brain in the foramen magnum, which indeed may also occur in occlusive hydrocephalus.

[3] Fleischmann performed lumbar punctures on a large number of patients with brain tumors and other diseases. One third of these patients died within 24 h, some of them during the LP. In retrospect, this very well documented and thoroughly autopsy-controlled series constitutes possibly the largest ever clinical experiment on quantification of the risk of herniation when LP is performed on patients with brain tumors.



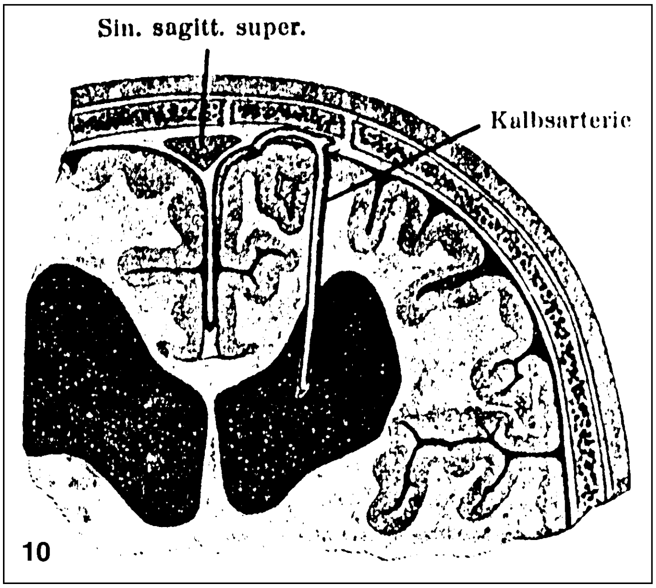


Fig. 9 Mikulicz's second ventriculo-subarachnoid-subgaleal shunt, 1895. Gold plated metal catheter with fixation attachment [79]

Fig. 10 Ventriculosubarachnoid interhemispheric fissure shunt using a formalin-fixed, paraffin-impregnated calf artery, Payr 1908 [132]

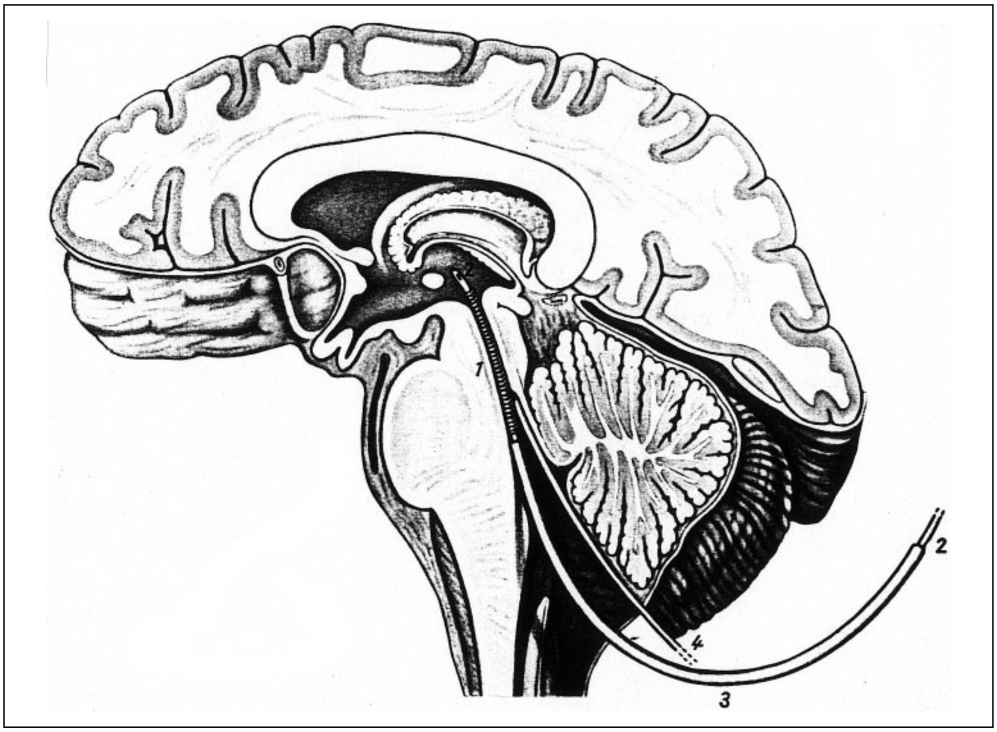


Fig. 11 Interventriculostomy III-IV. A 3 mm tantal helix is introduced over a smaller rubber catheter in Seldinger technique. Drawing by Leksell, 1949 [107]

As differentiation between communicating and noncommunicating hydrocephalus could not be made before the introduction of pneumoencephalography (and noninvasively only with the advent of CT), therapeutic lumbar puncture always presented with the danger of herniation and was often unsuccessful.

Subcutaneous reservoirs. Ommaya introduced the subcutaneous reservoir attached to a ventricular catheter in 1963 [127] originally for applying intraventricular chemotherapy. Such reservoirs enable intermittent, "on demand" puncture of ventricles or cystic lesions. In 1965, Rickham [165] adapted this "antechamber" for use in the treatment of hydrocephalus, making it possible for the reservoir to be integrated into a shunt or simply attached to a ventricular catheter, depending on the surgeon's preference. First descibed by Marlin in 1980, serial puncture of subcutaneous reservoirs became widely used in preterm babies with posthemorrhagic hydrocephalus [109].

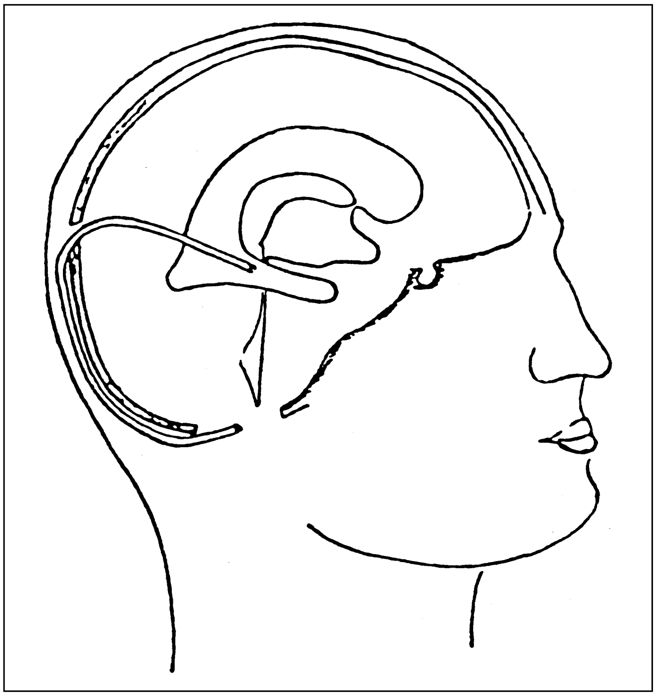


Fig. 12 Ventriculocisternostomy. Original drawing by Torkildsen, 1939 [177]

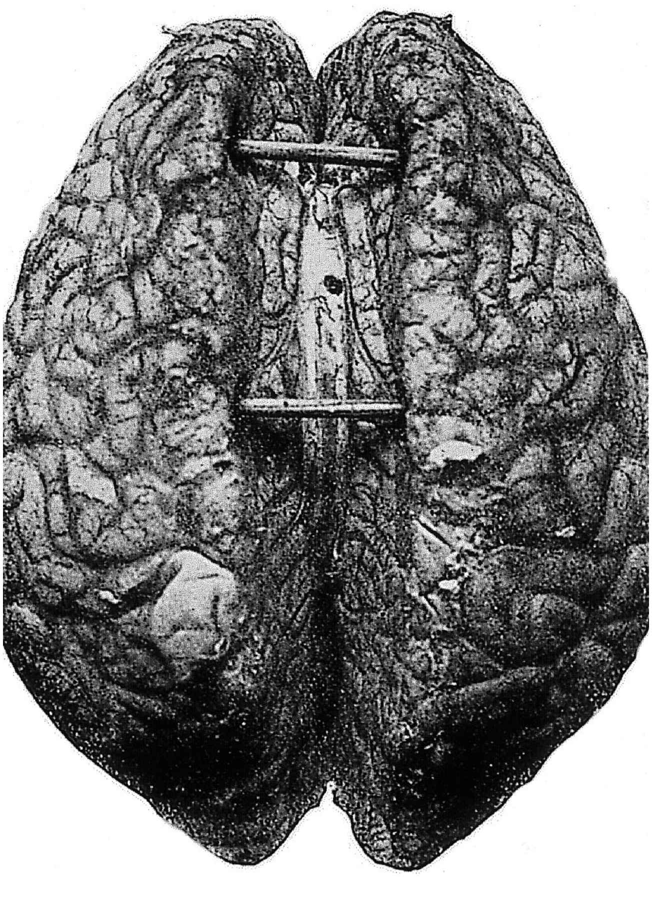


Fig. 13 Anton and Bramann 1908 [5]: autopsy photograph of a patient who had undergone callosal puncture ("Balkenstich"). A perforation measuring ca 4 mm is visible in the midportion of the corpus callosum

Intrathecal CSF bypass methods

Most hydrocephalus patients required and still require permanent, ongoing treatment. In the case of occlusive hydrocephalus, this can be provided by artificial drainage of CSF, either through an intrathecal bypass/recanalization, with or without the use of implants, or through shunts into extrathecal, low pressure compartments such as the venous system and the abdominal or pleural cavities (this applies to all kinds of hydrocephalus).

Drainage with implants into the external subarachnoid space. On 7 February 1893 in Breslau, Mikulicz implanted a wick made of glass wool into the lateral ventricle of a 6-month-old infant, extending outward through the external subarachnoid space and into the subgaleal compartment [79]. This, the first permanent CSF shunt in medical history, was thus simultaneously an intra- and an extrathecal drainage device. Mikulicz's collaborator Henle carefully documented the clinical course over the next 2 years. The head circumference became markedly smaller postoperatively and remained well under control for a year. In 1895, Mikulicz implanted a gold-plated metal tube 3 mm in diameter into a second patient, with an attachment for fixation to the skull (Fig. 9). The procedure was technically successful, but the patient, unfortunately, had an unrecognized brain abscess and died 2 months later [79].

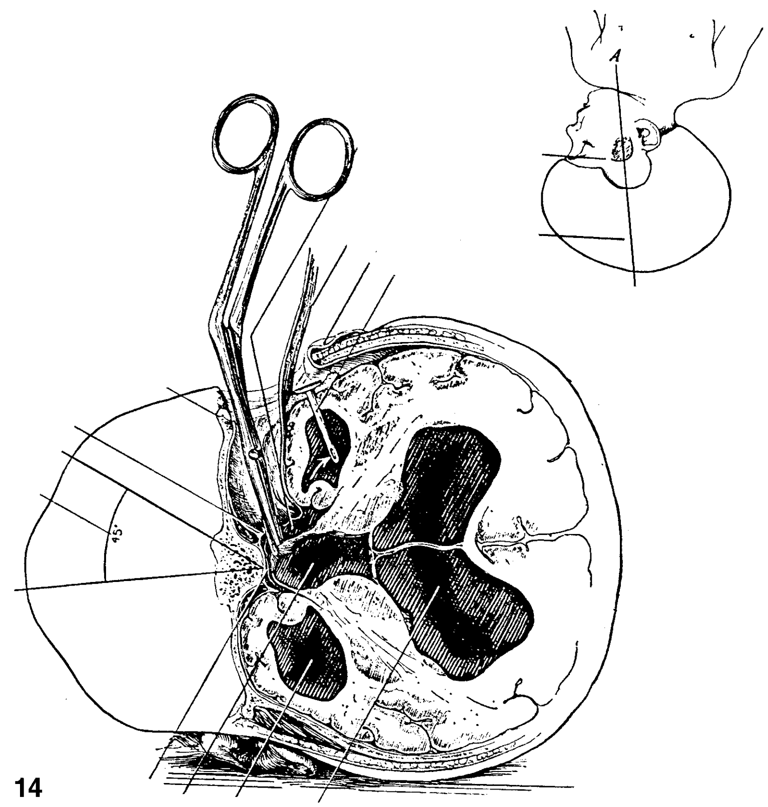
In 1908, Payr implanted a formalin-fixed, paraffin-impregnated, bovine artery into a 16-year-old patient, installing a shunt from the lateral ventricle into the interhemispheric fissure that functioned perfectly for 11 years (Fig. 10) [134]. Other contemporary authors used tubes of glass, copper, silver, metal, guttapercha, and rubber as well as wicks of silk, silver wire, or catgut [74, 75, 100, 138, 183].

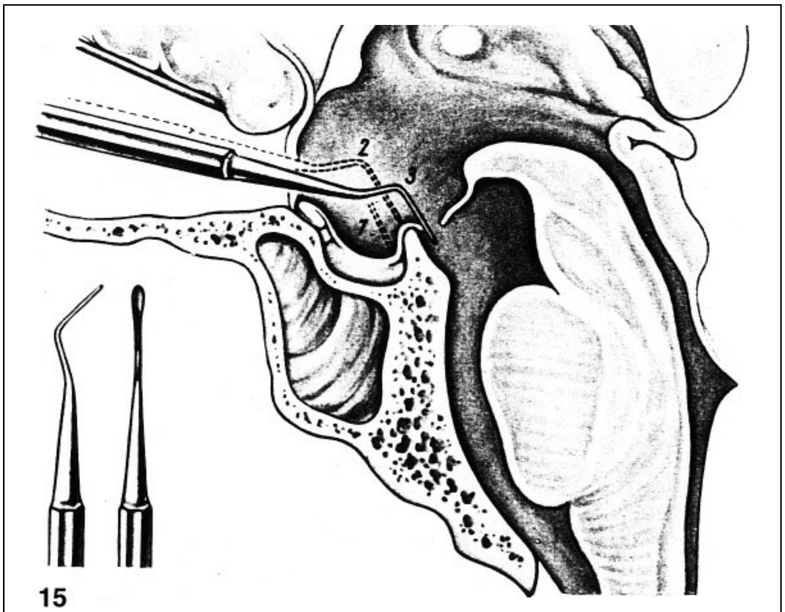
In 1953, Lazorthes [106] used a catheter to create a passage from the ventricle through the anterior corpus callosum into the external subarachnoid space (anterior ventriculocallosal shunt). Kluzer [89] provided a further variation on this theme in 1953 with an approach into the third ventricle through the posterior corpus callosum (posterior ventriculocallosal shunt). Burmeister placed a catheter between the ventricle and the suprachiasmatic cistern in 1959 [18], and Forjaz placed one through the hypothalamus in 1968 [53]. An attempt was also made to shunt CSF into the subdural space [52].

Interventriculostomy III-IV. In 1920, Dandy [27] was the first to perform retrograde recanalization of the aqueduct, using a catheter introduced from the fourth ventricle. Orthograde recanalization of the aqueduct was originated by Leksell in 1949 (Fig. 11) [107]. He passed a stent from the third ventricle distally, using stereotactic technique.

Ventriculocisternostomy (Torkildsen shunt). In 1938, Torkildsen observed a spontaneous cure of hydrocephalus occasioned by ventricular rupture[4], and was thus led

[4] Such occurrences are extremely rare but had been described in numerous published reports [118].





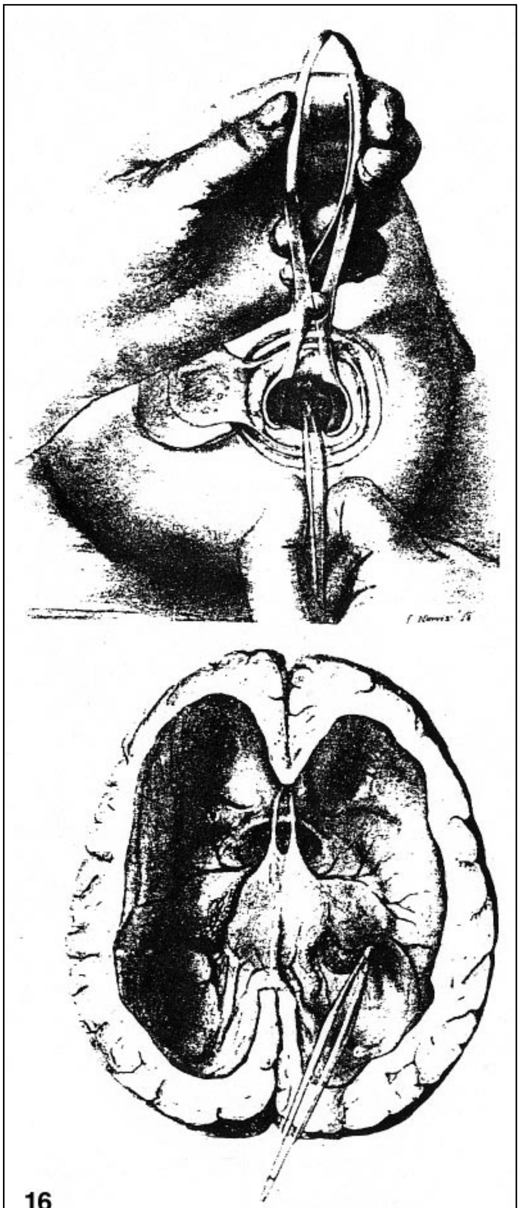


Fig. 14 Dandy 1922: open third ventriculostomy with a temporopolar approach to the third ventricle. Original drawing by Dandy [28]

Fig. 15 Anterior ventriculostomy though the lamina terminalis, combined with a fenestration into the prepontine cistern; according to Scarff and Stookey, 1936. Drawing modified by Riechert in 1960 [153]

Fig. 16 Plexectomy. Original illustration by Dandy, 1918 [24]

to invent a form of bypass drainage from the occipital horn into the cisterna magna using simple, valveless catheters, so-called ventriculocisternostomy [177]. This "Torkildsen shunt" (Fig. 12) was successful in many cases and remained a popular method of treating occlusive hydrocephalus until well into the 1970s [58, 62, 192]. Percutaneous lateral C1-C2 punctures [172] or valveless ventriculolumbar shunts [91] are variants of this method.

Drainage into the external subarachnoid space without implant: third ventriculostomy. In 1908, Anton and Bramann described creating a passage through the corpus callosum into the interhemispheric fissure by means of a special perforator (“Balkenstich”, or callosal puncture) [5]. The results were disappointing (Fig. 13). In Baltimore, Walter Dandy first opened the floor of the third ventricle in 1922 (posterior ventriculostomy). At first, this procedure required the sacrifice of one optic nerve (Fig. 14) [28].

Mixter, in Boston, inspected the ventricles with an endoscope in 1923 and perforated the floor of the third ventricle during this procedure [119]. He performed it in only one patient and did not pursue this type of operation any further. Scarff and Stookey, in 1936, developed a technique of fenestrating the lamina terminalis (anterior ventriculostomy) by a transfrontal approach, combined with perforation of the third ventricle floor (Fig. 15) [156, 157].

Many variations of third ventriculostomy were proposed over the course of time, among them ventriculostomies directed laterally [83] into the ambient cistern [122] or frontally by a transcallosal approach [153]. These procedures were carried out with microsurgical technique starting in the 1970s [11].

These demanding procedures have been replaced in the last few decades by so-called transcutaneous ventriculostomy, first performed by McNickle in 1947, in which the third ventricle is approached through a coronal burr hole and the foramen of Monro and its floor opened into the prepontine cistern [115].

The technique of ventriculostomy has steadily improved with the use of fluoroscopic image intensifiers[64], targeted stereotaxy [135], and especially endoscopy [56], with the result that the procedure experienced a rebirth during the 1990s [42, 77, 185]. An interventriculostomy III-IV without tubes, using balloon dilatations or perforations of septa, “aqueductoplasty” was sometimes described in endoscopic studies in the 1990s [159].

Destruction of the choroid plexus. Victor Darwin Lespinasse, a Chicago surgeon experienced in cystoscopy who mainly performed urological procedures, listed destruction of the choroid plexus for internal hydrocephalus under “Research Interests” in his 1913 application to the American College of Surgeons. Unfortunately, it is no longer possible to reconstruct Lespinasse’s operative experience, and the number and fate of patients he treated in this way are unknown. A lecture to a local medical society mentioned by Davis in 1936 is not further documented [60].[5] Unaware of this possible pioneering effort, Dandy resected the choroid plexuses of four patients with open surgical technique in 1918 (Fig. 16) after successful preliminary experiments in dogs [26, 24]. Three patients died and one was cured.

In the 1930s, Putnam [142] and Scarff [158] achieved better but still harsh results – a mortality rate of 25% – with endoscopic cauterization. In the 1960s, Scarff further reduced mortality after this procedure to 5% [158], and Griffith later brought it to 1% [61]. The method is now seldom practiced, because 67% of patients treated this way go on to require a shunt in the course of time [136].

[5] According to Davis [33], the first cystoscopic plexectomy of Lespinasse was performed in 1910. One child is reported to have died and another to have been helped for 3 years by the procedure.

Shunts into extrathecal, low pressure compartments

Valveless extrathecal drainage. Gärtner, in 1895, proposed shunting the CSF into extrathecal low pressure compartments such as the venous or lymphatic system or the abdominal cavity [57].

Shunts into the abdominal cavity. This idea was first applied in 1898 by Ferguson, who resected a portion of the fifth lumbar arch, drilled a small burr hole through the body of the vertebra into the peritoneal cavity, and established a connection between this space and the spinal canal by a loop of silver wire [49]. This first lumboperitoneal shunt temporarily succeeded in draining the hydrocephalus. The procedure was taken up by several authors, including Cushing, who used a transvertebral silver cannula in 1905, but the results were mostly disappointing [138].

Kausch led a rubber tube from the lateral ventricles into the peritoneal cavity in 1905, thereby originating the ventriculoperitoneal (VP) shunt [94]. The patient survived the procedure only a few hours, and autopsy revealed that the mantle of brain parenchyma around the hydrocephalic ventricles was only 3 mm thick. Hartwell, in 1910, was more successful, employing silver wire as a ventriculoperitoneal wick [74]. The child thus treated lived well for 2 years, then died after a stress fracture of the wire in the neck. Autopsy revealed that a tube of connective tissue had formed around the wire. This was not the first successful VP shunt: Hartwell expressly mentioned other practitioners of this technique and named Robert Abbé as its inventor [74].[6]

VP shunts remained a rarity for some time and experience with them was mostly frustrating [31]. Success rates improved from the 1950s onward with the availability of substantially better implant materials [32, 70, 162]. As early as 1960, Riechert collected some series with 214 VP shunts; the operative mortality still averaged 5% and delayed mortality between 38% and 47% [153].

In 1959, Luyendijk introduced another variation, the lumboperitoneal “petticoat shunt” (Fig. 17), in which a tube placed through the retroperitoneum ended in a funnel-shaped, perforated structure resembling a watering can [108]. This was intended to prevent distal obstruction, which was then still the major problem with abdominal shunting.

Shunts into the venous system

Shunts into intracranial veins. On 19 December 1907, Erwin Payr of Greifswald (Germany) created the first drainage from the ventricular system into the superior sagittal sinus of a 9-year-old girl with hydrocephalus [132]. He used a piece of autologous saphenous vein, with preserved venous flap valves, as the shunt material (Fig. 18).

[6] Abbé was one of the most innovative early neurosurgeons and had previously inaugurated the techniques of osteoplastic laminectomy in 1890 and syringostomy in 1891.

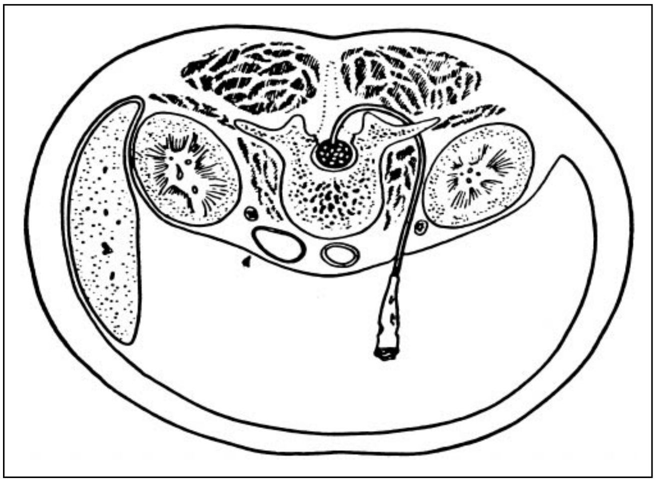


Fig. 17 Lumboperitoneal "petticoat shunt." Luyendijk, 1959 [108]

Thereupon, the patient's symptoms of elevated intracranial pressure improved somewhat. A second shunt implanted on the opposite side 6 weeks later was complicated by poor wound-healing, with egress of CSF. The child nevertheless made a good recovery and regained her eyesight but died of meningitis 7 months after the procedure. Autopsy revealed that both venous shunts had healed well in place with patent lumens containing no old blood clots. Fresh blood in both veins (which had perhaps entered post mortem) indicated an open connection to the superior sagittal sinus; the blood had not reached the ventricles, despite the open passage into them [132].

After two additional lethal cases, Payr changed to ventriculovenous (jugular) and subarachnoid shunts made from formalin-fixed calf veins [133]. His concept of a drainage into the sinus sagittalis was taken up by Sharkey [164] in 1965 with a valved catheter, but this (and similar) trials failed, due to a high quota of thrombotic occlusions.

In 1913, Haynes [74] shunted CSF through rubber catheters from the cisterna magna into the transverse sinus and parietal emissary veins.[7]

Valveless shunts into extracranial veins According to Haynes, [74] Beck tried unsuccessfully in 1904 to shunt CSF into a large caliber scalp vein. Bier did it successfully in 1908, but the shunt functioned for only 5 days. In 1909, Kanavel had longer lasting success with a shunt into a scalp vein. McClure, in 1909, was the first to shunt into the neck veins [74]. Payr, starting in 1911, attempted to create venous transplant shunts into the jugular vein made from stiff, formalin-fixed calf veins, while Enderlen, also in 1911, used a harvested temporal artery as a bioimplant for the same purpose [133]. Despite some therapeutic success, the unsolved problems of thrombotic occlusion and blood going up the shunt delayed the creation of reliable shunts into the venous system for another four decades.

Since the invention of valves, there have been only a few reports of valveless vascular shunting. To avoid overdrainage, El Shafei (1975) implanted valveless ventricular catheters into neck veins against the direction of blood flow, but changed to valves later on [45]. In 1988, Xue described valveless ventriculoatrial catheters 3 mm in diameter without significant reflux problems [193].

Shunts into alternative low pressure compartments Especially the years from 1920 to 1970 saw many attempts to place CSF shunts elsewhere (Table 1) [31, 35, 138, 183]. Recipient structures tested included the retro-or-

[7] Haynes's article of 1913 is an excellent source for the early history of shunt surgery, as is Kausch's study from 1908 [94]. Because of the information from these two sources, a number of first descriptions of operative techniques must be ascribed to different authors and dated earlier than is usually reported.

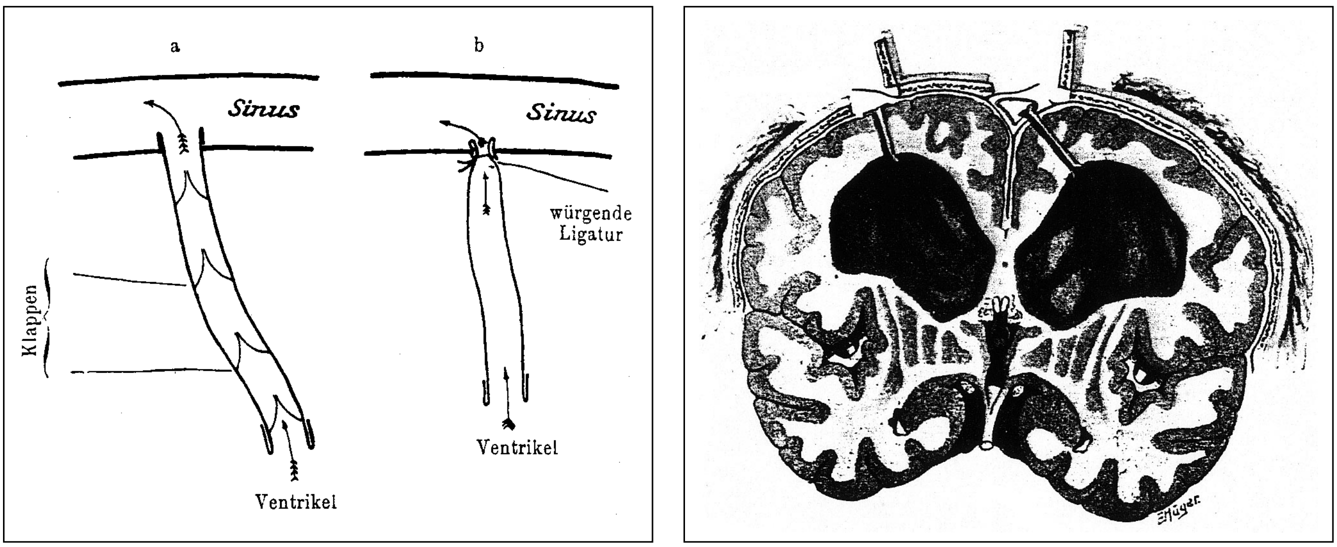


Fig. 18 Bioprosthetic valved shunt (autologous vena saphena with flaps) from ventricle into superior sagittal sinus; Payr, 1908 [132]

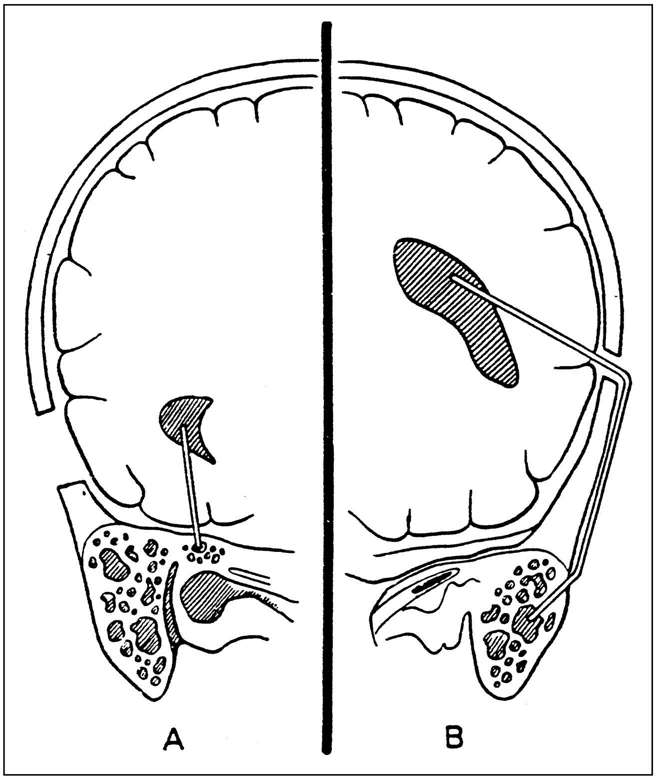


Fig. 19 Ventriculomastoid shunt according to Nosik 1950. Drawing by Brihaye, 1966 [17]

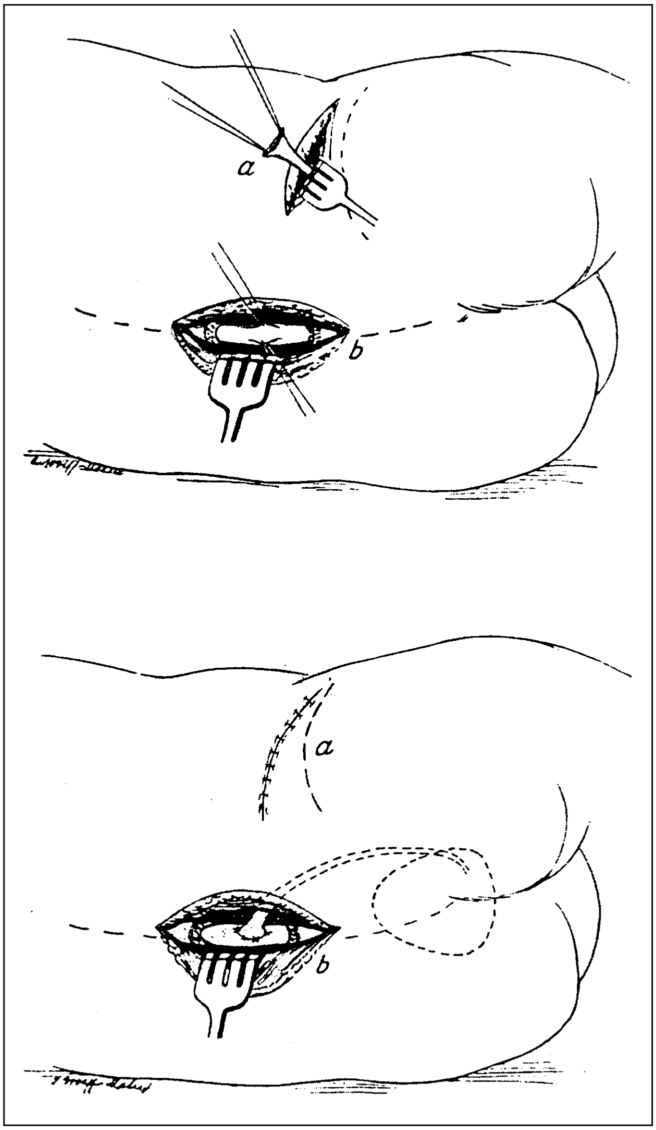


Fig. 20 Lumbo-ureteral shunt requiring a nephrectomy. Heile, 1925 [76]

bital fat [81], the fossa infratemporalis [153], the subgaleal space [94], the mastoid sinuses (Fig. 19) [17, 125], the buccal fat pad of Bichat (between buccinator and masseter) [171], the parotid duct [131], the pleural cavity [75], the thoracic duct [197], the ureter after nephrectomy (Fig. 20) [76, 111][8], the Fallopian tube [70], the gallbladder [123], the spinal epidural space [68, 143], and even the spongiosa of the lumbar vertebral bodies (Fig. 22) (Table 1) [198].

Most of these procedures are of only historical interest today because of severe side effects or limited therapeutic success. Many authors later recommended against pursuing these procedures or restricted their use to special cases. Matson, who favored urethral shunts for many years, eventually made the indication for surgery rest on whether the hydrocephalic child has “two good kidneys, two intelligent parents, an experienced pediatrician who is always available, and a good hospital around the corner” [112].

In 1974, Yasargil and Wennerstand published experiments aimed at improving CSF absorption capacity with omentum [189, 196]. The trials were in part successful, but further papers are not known.

[8] This method, much practiced in the 1950s, was complicated by increased infection and chronic electrolyte loss. Nonetheless, case histories with 26- and 38-year follow-ups have been published [50].

The introduction of valves in the treatment of hydrocephalus

As mentioned above, Payr [132], in 1907, was the first to use a saphenous vein with preserved venous flaps as a bioprosthetic unidirectional valve for a ventriculosinus-sagittalis shunt (Fig. 18).

Ingraham, in 1948, implanted catheters made of the newly developed synthetic, polyethylene, into the venous systems of hydrocephalic dogs. Nearly all of the catheters became obstructed by blood flowing into them. This experience proved the necessity of developing unidirectional valves [86]. Vannevar Bush of the Massachusetts Institute of Technology, collaborating with Donald Matson, was possibly the first to construct a magnetically operated valve [6, 138, 186]. According J. Shillito, (personal communication, July 1999) the valve was a gold

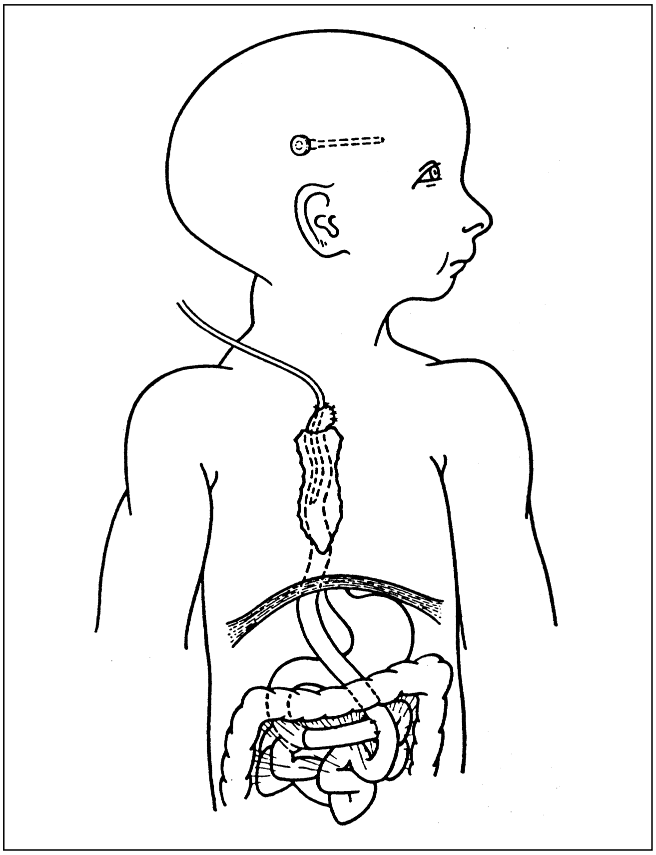


Fig. 21 Ventriculojejunostomy with a retrosternal y-loop; von der Oelsnitz 1971 [180]

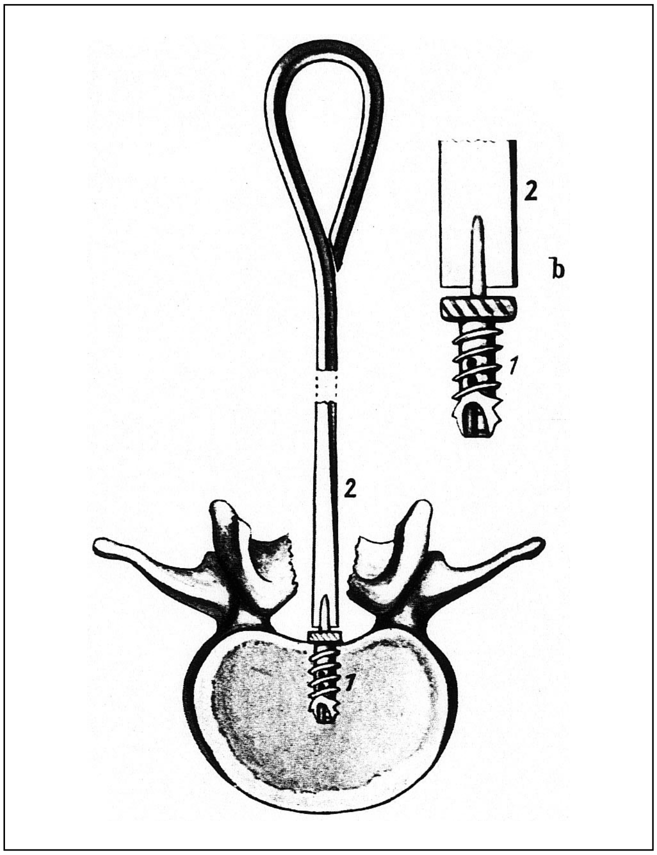
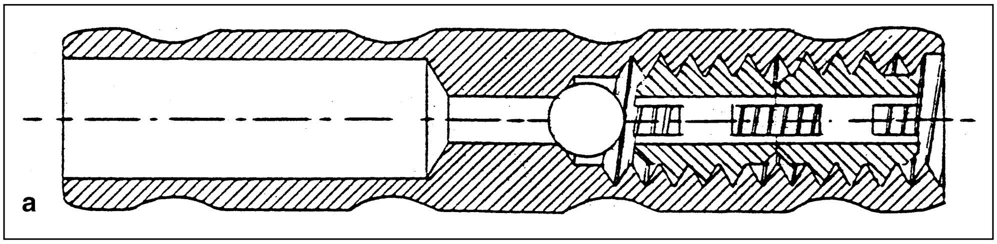


Fig. 22 Shunt from the lumbar subarachnoid space into the spongiosa of a vertebral body. Zimnowicz, 1950 [198]

Table 1. Valveless shunts into extrathecal low pressure compartments

Proximal and distal compartment (craniocaudal order)	Reference, year of publication (year of procedure)
Ventricle, subgaleal tissue with wick of glass wool and gold plated metal tube	Henle 1896 (Mikulicz 1893/1895)
Ventricle, jugular vein	McClure 1909[a]
Cisterna magna, transverse sinus	Haynes 1913
Cisterna magna, parietal emissary veins	Haynes 1913
Ventricle, peripheral neck veins, antidromic orientation	El Shafei 1975
Ventricle, supraorbital fat via orbital roof	Hildebrand 1923
Ventricle, fossa infratemporalis	Vána 1926[b]
Ventricle, buccal fat pad of Bichat	Sokolowski 1929
Ventricle, mastoid sinus	Nosik 1950
Ventricle, parotid duct	Parkinson 1961
Ventricle, buccal dental gap (animal experiment)	Hemmer 1959 [b]
Ventricle, pleura	Heile 1914
Ventricle, thoracic duct	Yokoyama 1959
Ventricle, right atrium (without valve)	Xue 1988
Lumbar, renal pelvis after nephrectomy with ureter	Heile 1925
Ventricle, ureter after nephrectomy with tube	Matson 1949
Lumbar, peritoneum with transvertebral burr hole	Ferguson 1898
Lumbar, peritoneum with catheter	Luyendijk 1959
Ventricle, peritoneum with rubber tube	Kausch 1908
Ventricle, Fallopian tube	Harsh 1954
Ventricle, gallbladder	Smith GW 1959
Lumbar, spongiosa of vertebral body	Zimnowicz 1950
Lumbar, spinal epidural space	Quincke 1891, Hakim 1956
Lumbar, lumbar subcutaneous fat	Fulcher 1958[b]

[a] According to Haynes 1913 [74]; [b] in Riechert 1960 [153]



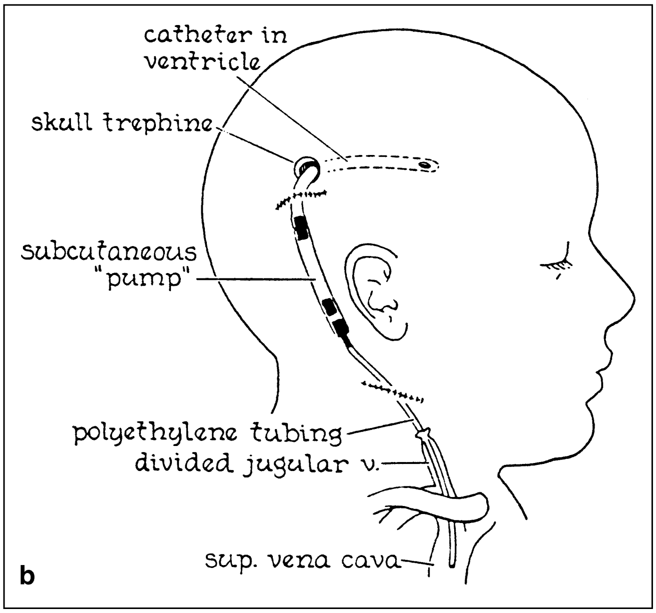
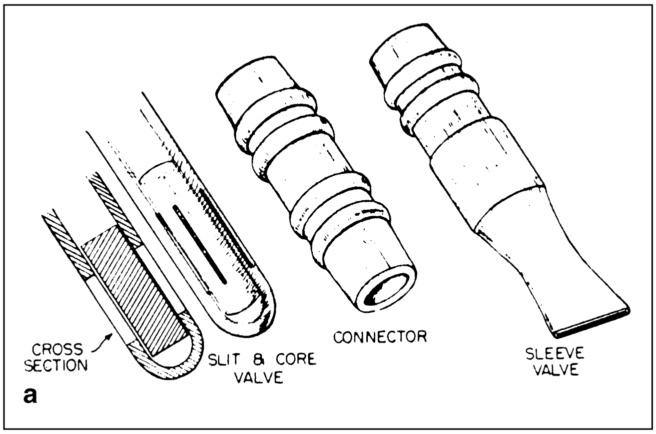


Fig. 23 **a** The first clinically successful valve, introduced by Nulsen and Spitz in 1949. The valve is a direct predecessor of the Hakim valves and contains two ball-valve units with an interposed pumping chamber. **b** The operative site [126]



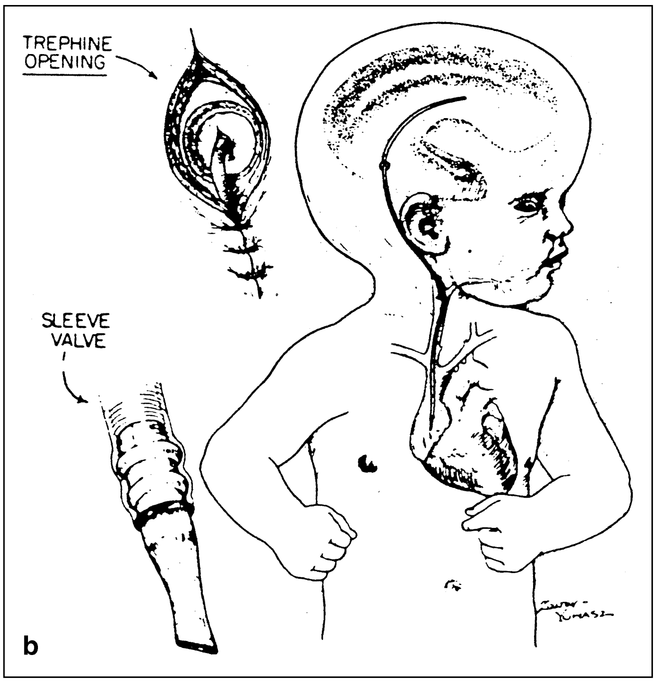


Fig. 24 **a** Distal Teflon valve with transverse slit. **b** Operative site. Pudenz and Heyer, 1955 [140]

plated, magnetically seated ball valve of very small dimensions (a few millimeters), combined with a rubber ventricular catheter and a relatively rigid distal polyethylene tube. The valve was adjusted in the theater by trial and error by passing it quickly through the magnetic field of a large permanent magnet until the desired closing pressure was achieved. These valves were used in the early 1950s; the date of first implantation is not known. Up to June 1957, at least 18 magnetic valves had been implanted in the Harvard Children's Hospital in Boston (R. M. Scott, personal communication, April 1988). They worked well for a time, but because of inadequate follow-up and poor results, the matter was not pursued (Shillito).

In 1949, Frank Nulsen constructed a valve containing two ball-and-cone valves with helix springs in series with a rubber pumping chamber between them (Fig. 23). Eugen Spitz implanted it for the first time in May 1949 at the Children's Hospital (Philadelphia) using a 1.7 mm polyethylene catheter into the superior vena cava. Careful clinical controls over 2.5 years, psychological tests, serial pneumoencephalograms, and a diagnostic shunt occlusion (leading to a recidivism) were able to prove the efficacy of this first successful treatment using a valve [126], while two other cases with short follow-ups were difficult to evaluate. The authors closed their paper with the prophetic statement: "If adequate absorption of cerebrospinal fluid can be secured more uniformly by this procedure in the future, it would seem to be a method which most closely reestablishes normal physiology." Surprisingly, the trials were not continued; the reason remains unclear (reflux problems?).

Fig. 25 Holter with his hydrocephalic son in 1956. The valve is in his hand. Drawing after a photograph. La Fay, 1957 [104]

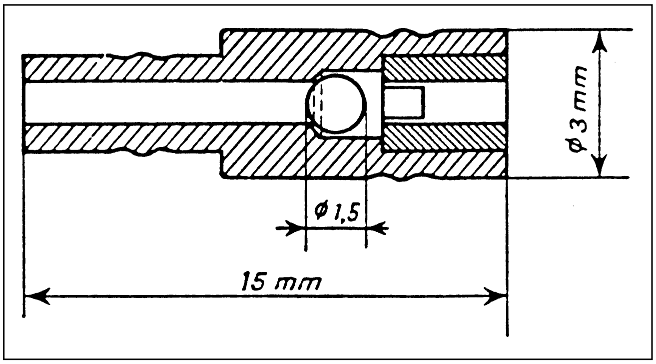


Fig. 26 The first European valve, designed by Engelsman and implanted by Sikkens in Groningen (Netherlands) in 1956 [166]

In 1955, after intensive experimental trials, Robert Pudenz and his engineer Ted Heyer constructed a distal teflon valve with a transverse slit and implanted it into a child (Fig. 24). This atrial shunt functioned for 2 years [140].

John D. Holter, a technician from Philadelphia, was primarily responsible for the spread of valves in neurosurgery. In a dramatic fight for the life of his son, Casey, who suffered from severe hydrocephalus associated with a meningomyelocele, Holter – who did not know Pudenz at the time – took only a few weeks to build a double silicone slit valve mounted on a helix spring that proved its value upon first implantation by Spitz in March, 1956 (Fig. 25) [104]. Serial production of these "Holter" or, more popularly, "Spitz-Holter" valves began in summer 1956 and they are being made in almost unchanged form today.

As often occurs in the history of science, a second pioneer team had the same idea at approximately the same time. In Groningen (Netherlands), W. Engelsman constructed a ball valve combined with a distal slit valve in 1956 (Fig. 26), which was implanted by T. Sikkens in at least six patients [166].

In 1958, Rudi Schulte, a young watchmaker newly emigrated from Germany, joined Pudenz and Heyer, improved their distal slit valve (multiple longitudinal slits), and patented the diaphragm design in 1960 (R. Schulte, personal communication, 1999). In 1958, Ames developed a distal slit VP shunt [3]. In the 1970s, Raimondi improved and simplified this design and set new standards of easy and quick handling [145].

Thus, the four technical principles that have guided the construction of hydrocephalus valves of the first generation until today were already established by 1960 [8, 41].

Silicone, the indispensible material

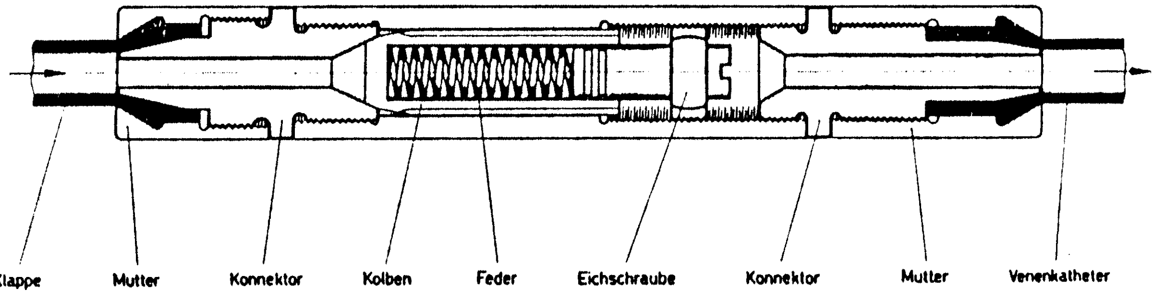
An indispensable prerequisite for durable shunts was the development of biocompatible and fatigue-free materials for use in catheters and valve components that could withstand the severe, long-term mechanical stress they were subject to, particularly in the right atrium.

At the beginning of the century, Kipping, in England, synthesized an oleaginous liquid, polydimethylsiloxan, from silicone and organic components [6, 55]. In the 1930s, glass-like synthetic materials were the next to be invented. During the Second World War, when the search was on for a thermally resistant, electrical isolation material for use in airplane construction, it was discovered that dimethylsiloxan could be transformed with application of silicone foam into any of a group of materials with viscous liquid, rubber-like, or solid glass-like properties, as needed [55]. A high degree of interconnection of the polymer chains is needed for the generation of elastic properties. This can be achieved by heating a mixture of the two components polydimethyl- und vinylmethyl siloxan after addition of silicon dust (so-called vulcanization).

One of the most successful conversions in history occurred in 1946, when this "Silastic" was introduced as a human implant for a bile duct repair in 1946 and in 1956 by Holter and Pudenz as a CSF shunt material [93].

These simultaneous breakthroughs provided working materials that were outstandingly biocompatible[9] and resistant to mechanical stress, an ideal complement to the new valve technology. It is the major reason why shunt operations with valves became the standard procedure for the neurosurgical treatment of hydrocephalus in the years between 1956 and 1961 [6, 35, 41].

[9] Up to now, no case of tumor induction and an extreme low number of allergies [88] related to the silicone formula used in shunts have been reported. Most adverse tissue reactions seem to be caused by latent infections [36].



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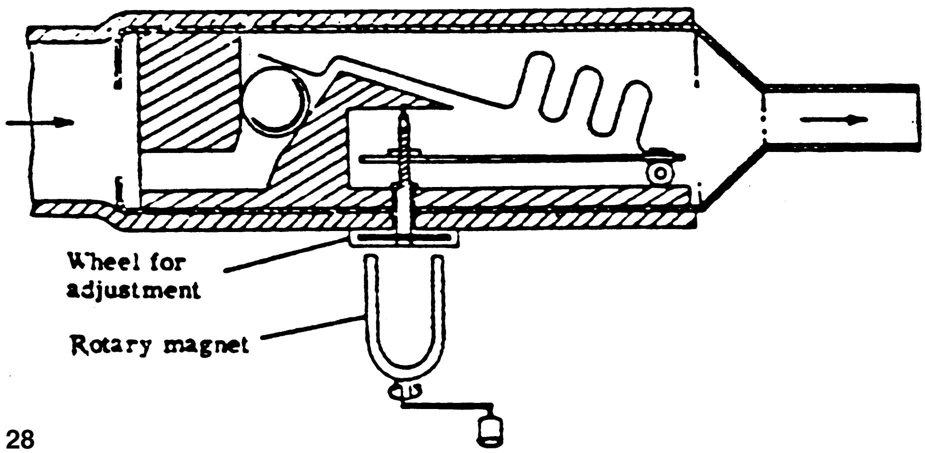


28

Fig. 27 The first adjustable valve by Kuffer and Strub, 1969. Adjustment required a screwdriver [102]

Fig. 28 The initial concept of a percutaneous, magnetically adjustable valve [66]

Development of valve technology since 1960

Approximatively 200 different types of valve have been designed since 1949. As of 1999, at least 127 are commercially available, with more than 450 pressure ranges and 2000 assemblies, while the rest of the 200 are either still in the prototype stage or no longer in production [8].

On closer inspection, most of these valves turn out to function by variations on four, classical principles – ball and cone (13 designs), diaphragm (at least 35 designs but more probably 43), proximal slit (24 designs), and distal slit (30 designs) – and belong technically to the first generation of simple, differential pressure (DP) shunt valves.

The second generation of shunt valves was designed to overcome the problem of possible overdrainage of CSF in the vertical position [48, 139]. The approaches taken to solve this problem include: adjustable valves, autoregulating valves (synonymous with flow-regulated, flow-regulating, and variable resistance valves), and two groups of valves controlled not only by differential pressure but also by body position respective to gravity, "antisiphon" and "gravitational" valves [9].

Adjustable valves

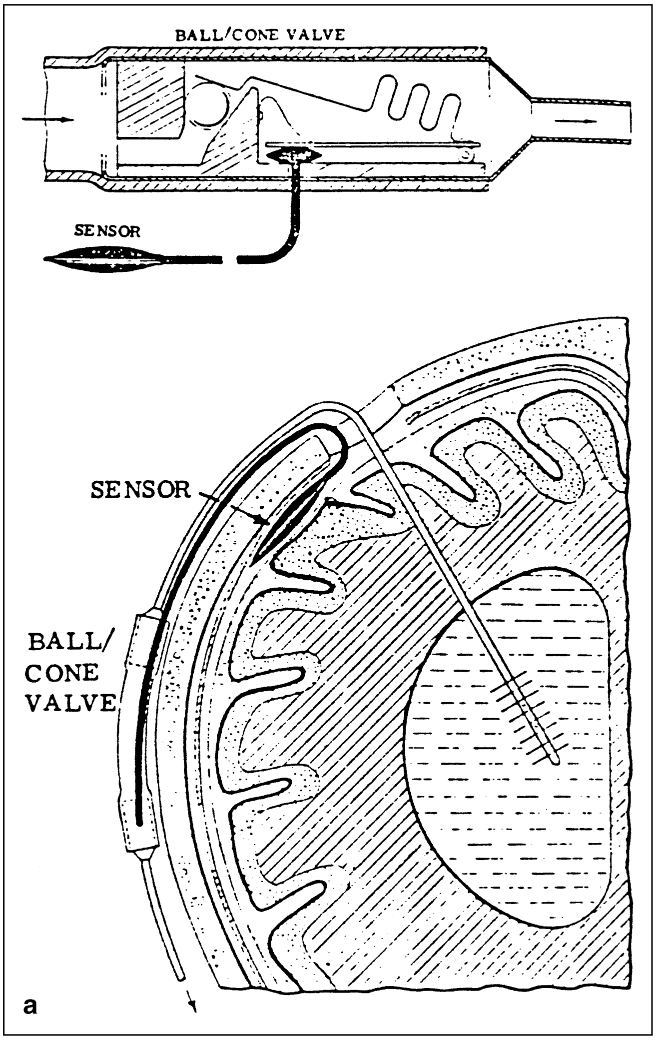
The first step was a piston-type valve made by Kuffer and Strub in 1969 [102] which had a spring that could be adjusted with a screwdriver (Fig. 27). Although this valve was implanted successfully several times, it failed to achieve recognition. A similar construction by Voth [181], designed for EVDs, experienced the same fate.

In 1973, Portnoy devised a percutaneously operable on-off switch for use with the shunt valve [137]. In the same year, S. Hakim [66] published his design for a valve that could be adjusted through the skin with a magnet (Fig. 28).[10]

Marion, a former engineer at the Cordis firm who was acquainted with S. Hakim's projects, anticipated him in 1983 with the three-level Sophy SU3 valve, which was adjustable between 60–160 mm H_2O [8, 41]. More recent variants have eight positions (SU8, Mini-8), an increased range (30–200 mm H_2O), and smaller size.

The "programmable" Medos-Hakim valve with 18 positions between 30 and 200 mm H_2O was designed by C. and S. Hakim, clinically tested in Colombia starting in 1984 (personal communication, S. Hakim, 1992) and

[10] Hildebrand had a similar idea in 1976, but it remained on paper [48].



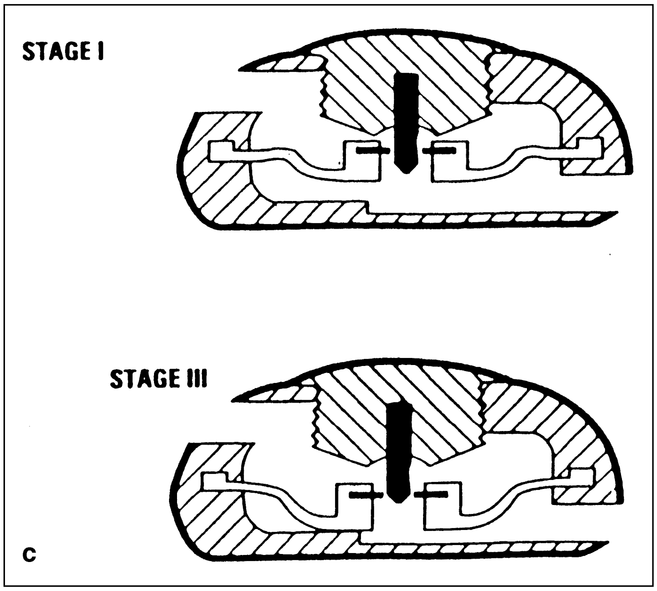
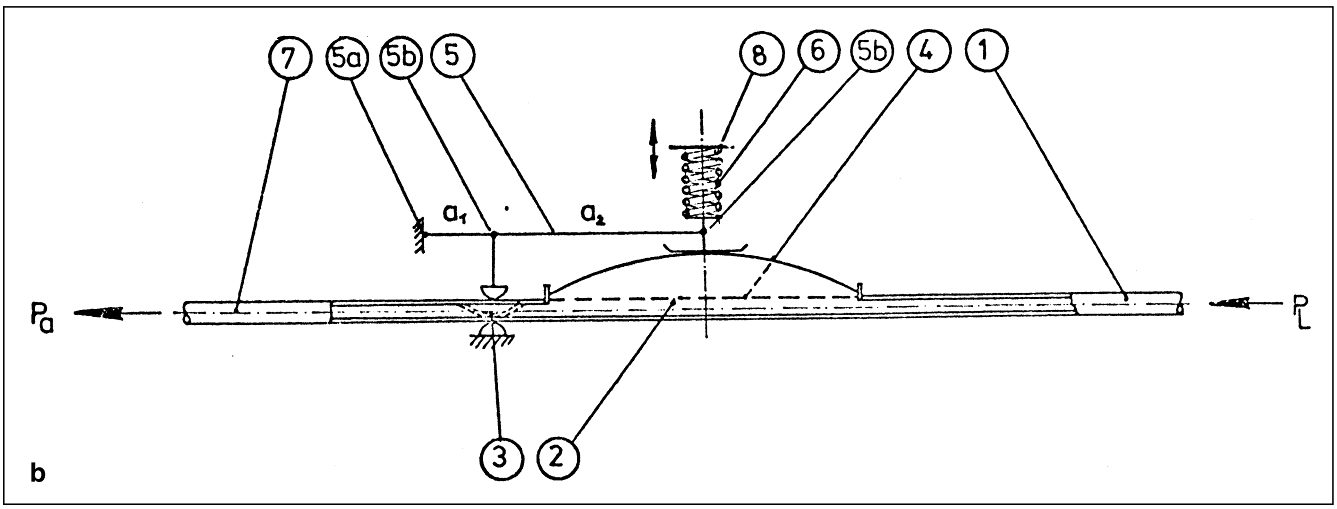


Fig. 29a–c Autoregulating valves, which vary the shunt resistance depending on differential pressure for a more physiological flow regulation: **a** the first design by Hakim 1973 [66]; **b** a patent of Hildebrand, 1976 [80]; **c** Cordis Orbis-Sigma valve



appeared on the market in 1989. After bench tests demonstrating its superior accuracy [8, 9, 23, 178], we introduced it in Europe in 1990. It is now widely used in difficult cases [149]. Out of a total of 14 adjustable designs, eight are now on the market [9, 41, 73].

Autoregulating valves

Valves that change resistance in response to differential pressure and thus limit flow constitute another advanced group of DP valves. The first studies of this concept

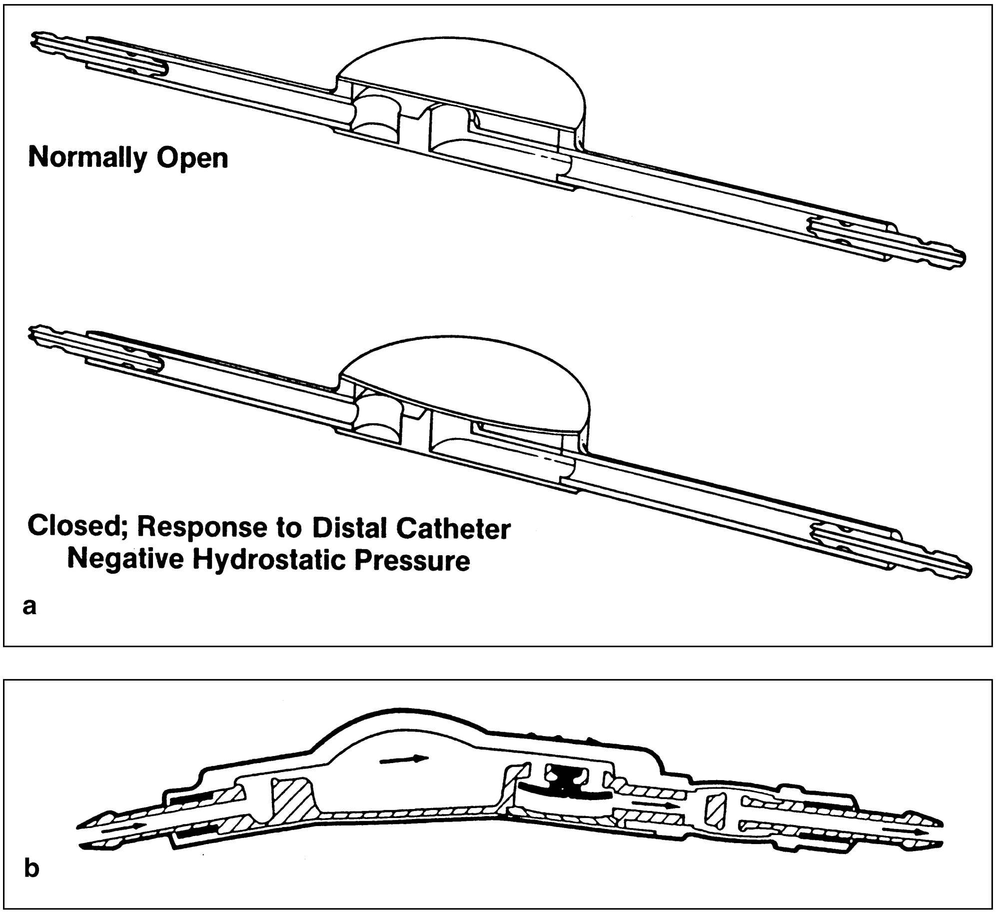


Fig. 30 **a** Antisiphon device, conceived by Portnoy, patent by R. Schulte in 1973. **b** PS Medical delta valve, a combination of conventional diaphragm and modified antisiphon valve in one construction

were carried out by Hakim in 1973 [66] and Hildebrand in 1976 (Fig. 29) [80].

The first practically available, autoregulating device was realized by Sainte-Rose, in 1984, as the Cordis Orbis-Sigma Valve [155] and, after 1996, the Orbis-Sigma II. Despite differences in technology, Paes' Phoenix Diamond [129] and the Codman SiphonGuard also belong to this group.

Antisiphon valves

The first product to exploit gravity for controlling shunt valves was the antisiphon device (Heyer-Schulte ASD) conceived by Portnoy and patented by Schulte in 1973, in which a switching membrane is progressively closed by the weight of the hanging hydrostatic column (negative pressure) in the distal catheter (Fig. 30a) [137]. During verticalization, the negative pressure increases continually to reach a maximum in upright position [9, 41, 63, 101].

Three separate devices are available, e.g., Heyer-Schulte Anti Siphon Device (ASD), Radionics Flow Limiting Device™ PS-Medical Siphon Control Device™ and 31 antisiphon valve combinations, usually with diaphragm valves, e.g., PS-Medical Delta (Fig. 30b), Radionics Equiflow, but also with ball valves (Beverly) or proximal slit valves (Heyer-Schulte Multipurpose).

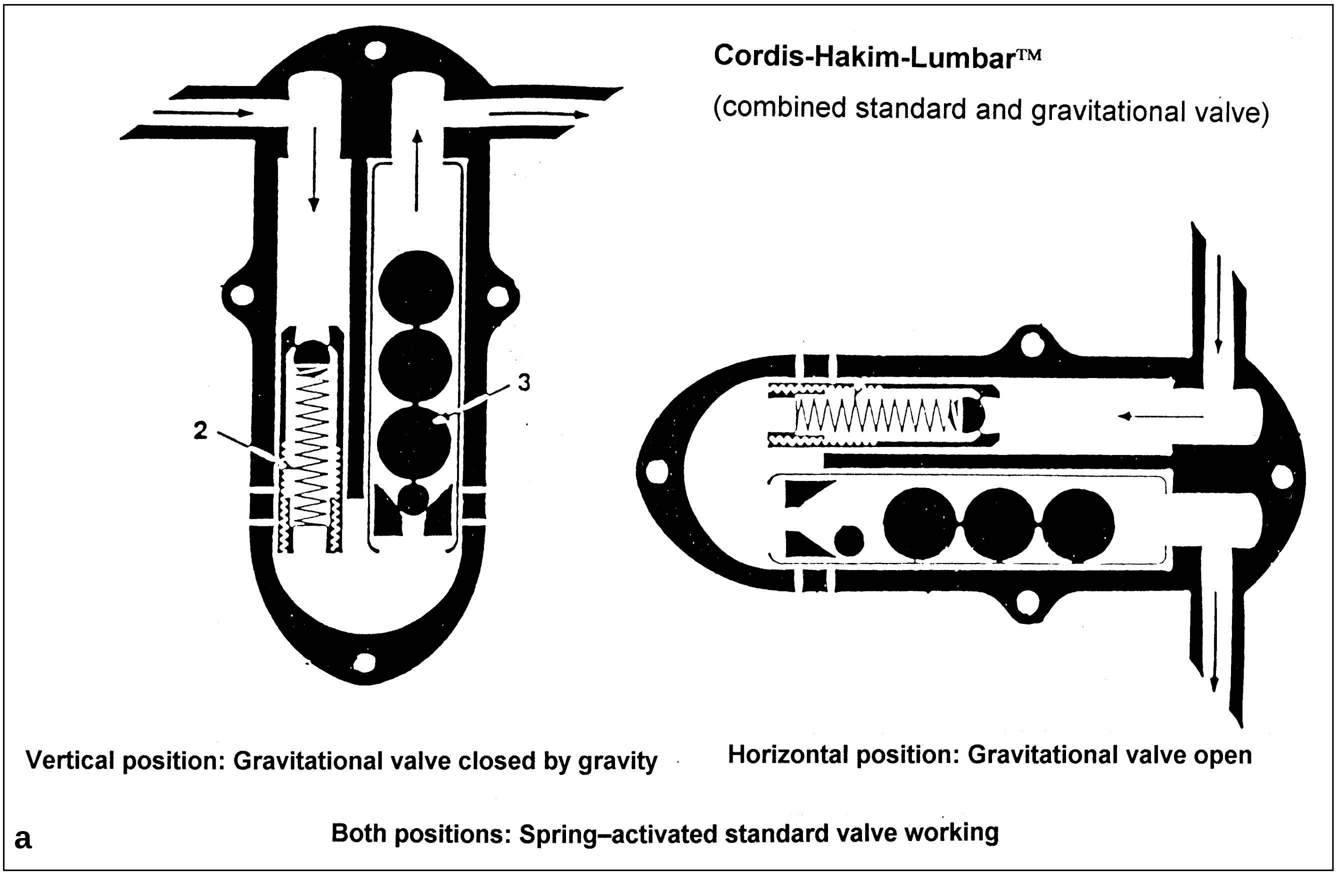
Unfortunately, the external switching diaphragm is highly susceptible to any kind of increased external tissue pressure [101], which can lead to functional obstructions [39]. Most clinical reports showed a decreased rate of overdrainage [63, 90], but often no differences and, in the only randomized study [40], even more overdrainage than the Orbis Sigma and conventional DP-valves. These results may be biased by an unusually high quota (85%) of antisiphon valve combinations with very low pressure (Delta Level I).

Gravitational valves

The same principle is at work in valves that use small metallic balls or similar objects as controlling weights, instead of the hydrostatic column. The first "gravitational" valve was patented by S. Hakim in 1975 (Fig. 31a) and independently published by Yamada in 1979 [66, 195].

Surprisingly, there was minimal interest in these designs for 20 years, but more recently this principle has gained the attention of engineers and laboratory and clinical studies [9, 20, 71, 97, 117, 173, 178]. In 1999, we cataloged 18 gravitational valves, of which six are supplementary devices (Fig. 31b) and 12 are valve-device combinations (Fig. 31a). Eight are on the market.

Conceptual studies have described "active shunts": implanted roller pumps [110], electronic valves [8], and


Cordis-Hakim-Lumbar™
(combined standard and gravitational valve)
2
3
Vertical position: Gravitational valve closed by gravity
Horizontal position: Gravitational valve open
a
Both positions: Spring–activated standard valve working

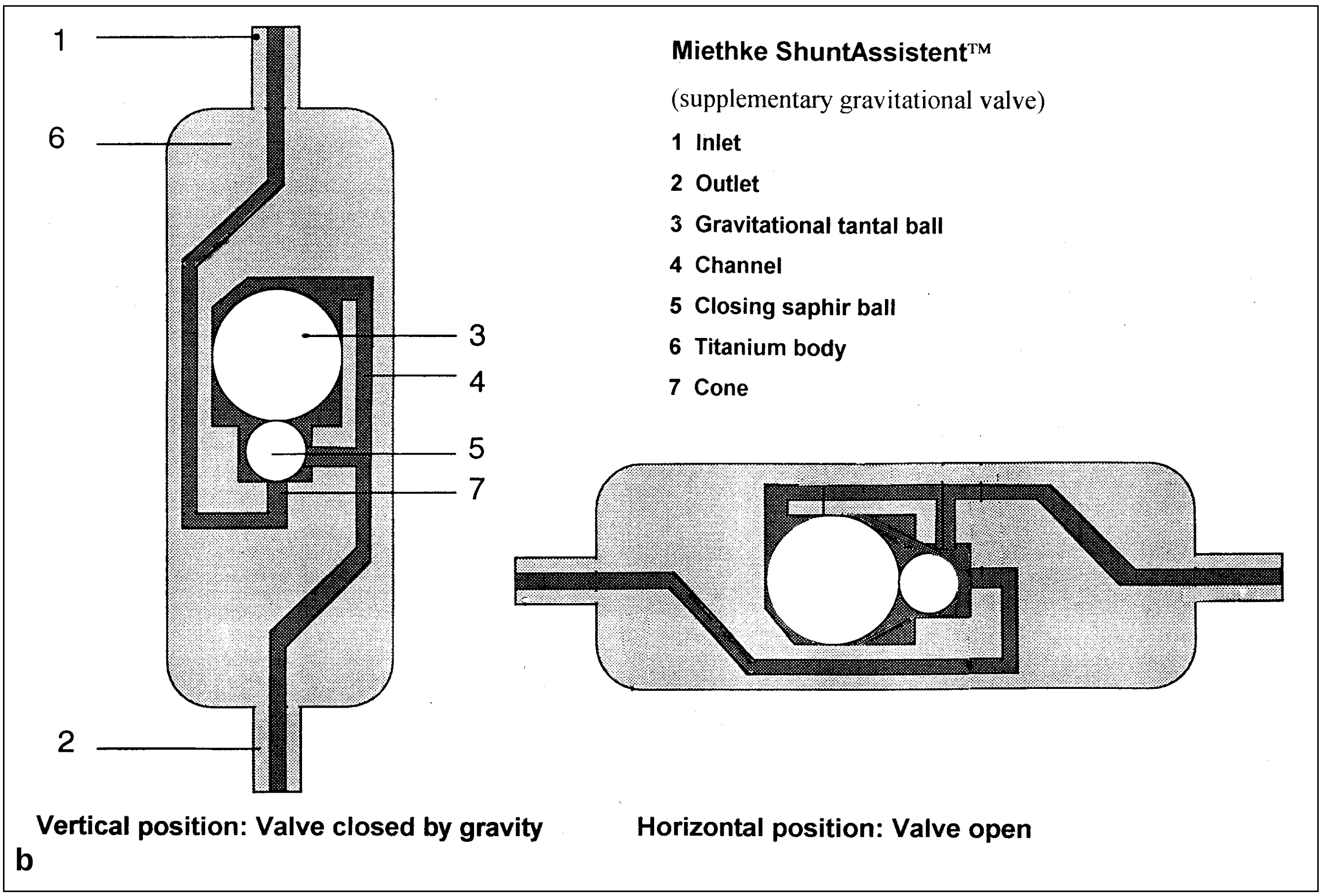

Miethke ShuntAssistent™
(supplementary gravitational valve)
1 Inlet
2 Outlet
3 Gravitational tantal ball
4 Channel
5 Closing saphir ball
6 Titanium body
7 Cone
1
6
3
4
5
7
2
Vertical position: Valve closed by gravity
Horizontal position: Valve open
b

Table 2. Valved shunts into extrathecal low pressure compartments

Proximal and distal compartment (craniocaudal order)	Reference and year of publication (year of procedure)
Ventricle, superior sagittal sinus, with autologous saphenous vein and preserved venous flaps	Payr 1908 (1907)
Ventricle, jugular vein, with autologous saphenous vein and preserved venous flaps, augmented by formalin-fixed calf vein	Payr 1911
Ventricle, vena cava superior	Nulsen 1952 (1949), Matson (1949?)[a]
Ventricle, right atrium	Pudenz 1957 (1955), Spitz 1961 (1956), Sikkens 1957 (1956)
Ventricle, peripheral neck veins, antidromic orientation	El Shafei 1975
Ventricle, pleura	Nixon 1962
Ventricle, azygos vein	Redo 1992
Lumbar, atrial via femoral vein	Aoki 1988
Ventricle, thoracic duct	Kempe 1977
Ventricle, peritoneum	Ames 1967 (1958)
Lumbar, fallopian tube	Jones 1967
Ventricle, gall bladder	Yarzagaray 1987[b]
Ventricle, stomach with special valve	Alther 1965
Ventricle, duodenum	Puri 1977
Ventricle, ileum (isolated segment)	Neumann 1959
Ventricle, jejunum (Y-segment), retrosternal placement	Von der Oelsnitz 1971
Ventricle, ascending colon	Puri 1977
Ventricle, bladder	West 1980
Ventricle, ureter	Smith JA 1980
Lumbar, urethra	Biddle 1966

[a] Implantation of magnetic ball valves, either since the late 1940s (according to Wallmann, 1982) or the early 1950s (J. Shillito, Boston, personal communication, July 99).
[b] In Raimondi 1987 [144], pp 474–476.

components with linear resistance [84, 194]. In three small series, telemetric ICP sensors [19, 73] and flowmeters [92] integrated in shunts have been described.

Since the first independent bench test by Forrest in 1962, 1133 valves have been tested in 65 series. Unfortunately, many papers take the form of internal technical reports, medical theses, and oral transcripts and thus difficult to acquire. There are overviews [8], but no comprehensive meta-analyses of this material.

The early tests can be roughly characterized as simple, usually short (often lasting only minutes!) and without deaerated test fluids, temperature control, or systematic pretest preparation [147, 167]. Most results may be correct, but reliability remains doubtful**.** Improved test rigs and procedures were published by several authors [41, 54, 63, 67, 160, 188], but a really sufficient methodology was first established by Ekstedt in 1980 [43] and later trials [8, 9, 23, 101, 117, 150, 187]. Nevertheless, many problems due to small numbers of probes, insufficient statistical power, and missing long-term and safety tests remain unsolved. Out of 127 valves on the market, more than 60 were never tested in independent laboratories and about 40 tested only one or two specimens, which may lend them only anecdotal value.

Compared to cardiac pacemakers or other implants' high technology, performance, safety, and prices, most hydrocephalus valves today are outdated, inaccurate, and unsafe, but cheap, typically US$ 600 for a standard shunt, as compared to the US$ 29,000 of gross treatment costs per patient [37]. Presuming a 10-year valve life, shunt hardware costs only 17¢ per day. All shunts sold in the USA in 1995 [116] cost a total of US$ 20.8 million, or 8¢ per inhabitant [10].

Although valves were produced almost exclusively in the USA for many years, a considerable shunt valve industry has now developed in France (NMT Cordis, Sophysa, French Neurone, Beverly), followed by Switzerland (Codman-Medos), and Puerto Rico (Heyer-Schulte). It is astonishing that there is so little in the way of shunt production in technologically advanced countries such as Japan (Fuji, MDM 73), England (Nottingham Shunt), and Germany (Schubert, Miethke)[11], while less developed countries such as Russia (Simernitsky and Burdenko valves), India (Upadhyaja [167] and Chhabra valves [20]), the Philippines (Luisa shunt), Colombia (Clunej shunt), Mexico (Sotelo, Dewimed), China (Shanghai-and Jinan valves), and Zimbabwe (Harare shunt [165]) produce functional shunts or at least design them (Ammar, Saudi Arabia).

Development of operative technique

Shunts into the venous system (usually into the right atrium) were preferred in the 1960s [4]. The placement of the

Fig. 31 **a** Cordis Hakim-Lumbar™ valve, which contains a conventional spring- and a gravity-activated ball valve. Patent by S. Hakim 1975. **b** Miethke ShuntAssistent™, a supplementary gravitational valve for integration into conventional shunts

[11] The most important European contribution before the 1980s, Schubert and Zeiner's "Dresden diaphragm valve" [161] was limited to only about 80 valves, due to COMECON trade regulations (Prof. Vogel, Berlin, personal communication, 1992)

atrial tip, not always accurately visible in x-rays, was improved by intracardiac ECG, intra-atrial pressure registration, or transesophageal echography. A time-saving alternative to open venae sectio was puncture and catheterization using the Seldinger technique. Nevertheless, the unsolvable problem of frequent catheter retractions due to children's growth and serious side effects such as thromboembolic complications, pulmonal hypertension, shunt nephritis and sepsis led to a negative trend [35, 78, 144].

Ames introduced valves into peritoneal shunts in 1958 [3]. In the 1970s, the use of VP shunts steadily gained ground, first in children, then in adults, and it became the most commonly used variety of shunt. Today, they are used in 98% of pediatric hydrocephalus patients [38, 92] and in more than 80% of adult patients.

Murtagh, in 1967, introduced valves to lumboperitoneal shunts and was the first to introduce a lumbar catheter through a Touhy needle [121]. This method is used primarily in the treatment of refractory CSF leaks, benign intracranial hypertension, and slit ventricle syndrome, but also in treatment of ordinary hydrocephalus. Variations include shunts from the cervical spinal canal to the peritoneal cavity [15], lumboatrial shunts employing the saphenous vein [7], and lumbourethral [16] and lumbofallopian shunts [89]. More recently published long-term complications such as scoliosis [113] and acquired Chiari malformations [22] limite the application of lumbar shunts.

Meanwhile, attempts have also been made to place the distal end of valved ventricular shunts into stomach [2], duodenum [141], excluded loops of small intestine [123] or jejunum (Fig. 21) [180], ascending colon [141], gallbladder [169], pleural cavity [124], thoracic duct [95], ureter [170], and bladder [191]. Most of these operations are relatively difficult to perform, more likely to cause complications, and rarely successful (Table 2).

Of all these techniques, only the lumboperitoneal and ventriculopleural shunts retain a certain importance [82]. When all other possibilities have failed, one may choose to divert the flow of CSF temporarily into the gallbladder, the ureter (today without nephrectomy, of course), or the bladder.

The three main problems of shunting are (1) infection, (2) improper placement of catheters, and (3) hydraulic mismanagement.

Infections

Infection causes only 10–15% of all shunt revisions, but its impact on mortality, morbidity, quality of life, and economics is enormous [13, 35, 144]. A single shunt infection costs US$30,000, which is equivalent to a typical neurosurgical department's annual expenditures for shunt hardware [10].

Most experts agree that over 98% of infections are caused by intraoperative contamination of implants [13] and that they are therefore primarily a surgical problem. Consequential adherence to classic hygienic rules combined with a high level of experience can diminish infection rates to less than 1% [21], compared to cumulative numbers of 5.8% in the 1990s (3911 procedures in 14 studies) and 8.7% between 1959 and 1992 (24,436 shunts in 108 studies) [8].

The efficacy of systemic antibiotic prophylaxis, which is generally used in over 70% of shunt operations, was shown to be significant in only a few studies, probably due to their using too small samples. Three independent meta-analyses of numerous randomized trials showed a decrease in infections to approx. 5–6% with perioperative antibiotics compared to approx. 10–12% in placebo controls [8, 65, 105]. This is useful to know, but represents no improvement over general results in the 1990s.

The efficacy of antimicrobial surface design with antibiotic impregnations are well-proven in laboratory [14] and animal experiments [69, 99] and have been promising in clinical trials. Hyrogel or antiseptic coatings seem to be useful, too, but the few published data allow no final evaluation.

Improper placement

The most common factor causing shunt failures is misplacement, especially of the proximal catheters [38, 40, 78, 154]. Unfortunately, this topic is not very popular in the literature. These failures are not generally understood as *surgical* failures, due to a misleading terminology and presentation in many studies.

Hydraulic mismanagement

All conventional differential pressure valves present hydraulic mismanagement in either upright or horizontal body position and afford at best a compromise [9, 41].

In the course of time, one of six patients presents evident problems due to overdrainage, such as subdural hygromas, position-dependent headaches, symptomatic slit ventricles [46] or skull deformations. Side effects less often manifested usually include proximal occlusions, trapped ventricles, periventricular hyperemia in upright position, and chronic and in part irreversible loss of brain compliance during ICP/CPP changes [48, 139].

Summary

The scientific history of CSF physiology and the heroic early treatment of hydrocephalus was the subject of many excellent studies by Haynes, Riechert, Scarff, deLange, Pudenz, Torack, Gjerris, Voth, Wallmann, and Aronyk, while a survey of the last decades and particularly of shunt technology remains incomplete.

In 1893, Mikulicz made the first surgical attempt to create a permanent ventriculo-subarachnoid-subgaleal CSF diversion. This was simultaneously the first intrathecal ventriculostomy and the first shunt into an extrathecal low pressure compartment. Thereafter, the results of early lumboperitoneal (Ferguson 1898), ventriculovenous (Payr 1908), and ventriculoperitoneal shunts (Kausch 1908), of plexus coagulation (Dandy 1918, Lespinasse 1910?), third ventriculostomy (Anton 1908, Dandy 1922), and the Torkildsen shunt (1939) were indeed encouraging. But these could often be used only in selected cases and initially had enormous mortality rates.

The year 1949 saw implantation of the first functional hydrocephalus valve and (outside of neurosurgery) also the first silicone prosthesis. The decisive breakthrough in effective treatment came in 1955–1960 with simultaneous construction of several types of valve for clinical use and the introduction of silicone as construction material.

Within a wide variety of operative techniques, the treatment of hydrocephalus by means of a valve is now largely standardized. VP shunts are most widespread, followed by VA shunts. Lumbar and pleural shunts are reserved for special cases.

Despite progress in neuroendoscopy, which can be used successfully to perform ventriculostomy in most adult cases of occlusive hydrocephalus, treatment with valved shunts is expected to remain the method of choice in approx. 80% of patients.

Acknowledgement I thank Mr. Ethan Taub M.D., Zurich, for the translation of most parts of the paper.

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