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REVIEW PAPER

Is there an ideal shunt? A panoramic view of 110 years in CSF diversions and shunt systems used for the treatment of hydrocephalus: from historical events to current trends

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Abstract
Object The goal of this study is to evaluate whether an "ideal shunt" exists.
Methods This is a retrospective analysis based on original papers in the field of *Hydrocephalus and Shunts*. Patients of all age groups, who had hydrocephalus, and underwent some form of CSF diversion were included. The study has been divided into four stages: from 1900 to 1949, 1950 to 1974, 1975 to 1999, and from 2000 to 2010.
Results In stage 1 (*historical era*): Saphenous vein grafts, rubber conduits, and other materials were used in CSF diversions. In 1949, the first implantable shunt tube was developed by Nulsen. In stage 2 (*experimental stage*): the Holter valve was developed. Newer innovations were developed in relation to the ventriculo-atrial shunt, which was the preferred CSF diversion. In stage 3 (*developmental stage*), a large number of different design shunt systems were developed, with the aim of reducing complications. The ventriculo-peritoneal shunt had become the preferred CSF diversion. Also, the programmable valve was born. In stage 4 (*era of programmable valve*, there is a preference for the use of programmable shunt systems. However, shunt failure rate at 1 year being around 25 to 40 %, and shunt survival at 1 and 2 years are 50–70 and 47–53 % in most series.
Conclusion Every shunt is an ideal shunt provided the choice of the shunt used should be made by the matching performance of the shunt system to the altered profile of CSF dynamics of a given patient. The most important factor being the opening pressure.

Keywords Hydrocephalus · Shunts · CSF diversion · Complications

Abbreviations

CSF	Cerebrospinal fluid
VP	Ventriculo-peritoneal
VA	Ventriculo-atrial
IVP	Intraventricular pressure
VV	Ventriculo-venous.
FRD	Flow-regulating devise
SCD	Siphon control devise
DSV	Dual switch valve
GAV	Gravitational valve

Introduction

Hydrocephalus has amazed and challenged clinicians throughout the history of medicine, and continues to be one of the most common neurosurgical conditions encountered, both in children and adults. Diversionary shunting for hydrocephalus had its beginning with Ferguson in 1898, who first attempted the lumboperitoneal CSF shunt [42]. Since the first implantation of a cerebrospinal shunt tube in 1949 by Nulsen [89], a variety of shunt systems have been developed in order to provide continuous, failure-free, CSF diversion for patients with hydrocephalus. Despite many years of research and modification to the original shunt valve designs, improvement in treatment results have only been limited [29, 30] and fewer than one third of new shunts survive 10 years without revision [101, 119].

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By far the most frequent cause of shunt failure has been reported to be mechanical complications [28, 46, 86, 146]. Other causes of failure being, selecting the wrong shunt system or surgical technique, or specific patient characteristics. When selecting the type of valve, the most important factor to consider is the opening pressure. When using a valve with a nonadjustable opening pressure, postoperative manipulation of the opening pressure can only be accomplished by exchanging the valve to a different pressure, which entails another operation. In 1973, Hakim proposed the use of a shunt system with an adjustable valve to regulate the opening pressure [49, 50]. The present day trend is more neurosurgeons are in favor of using the programmable valve. This allows the surgeon to adjust the opening pressure setting noninvasively in accordance with clinical or radiological findings in the postoperative period [12, 73, 110].

Materials and methods

This is a retrospective analysis, from 1900 to 2013, based on original papers in the field of "Hydrocephalus and Shunts" research published in major scientific journals and data collected from Pubmed. The study includes patients who had nontumorous hydrocephalus of different etiology and those who underwent some form of CSF diversion. The analysis has been divided into four stages (Table 1): first stage from 1900 to 1949, the second stage from 1950 to 1974, the third stage from 1975 to 1999, and the final stage from 2000 to 2010. In each stage, data has been analyzed in relation to (1) the type of CSF diversions performed, (2) the type of shunt systems and their development, (3) complications in relation to the shunt system and procedure, and (4) advances in the surgical technique to minimize complications.

Table 1 Design of the clinical study

A) Stages
Stage 1: 1900 to 1949
Stage 2: 1950 to 1974
Stage 3: 1975 to 1999
Stage 4: 2000 to 2010
B) Criteria of inclusion
1) Retrospective study
2) All age groups included
3) Patients should have undergone some CSF diversion for nontumorous hydrocephalus
C) Each stage data analyzed
1) Type of CSF diversion
2) Complications encountered
3) Development of shunt systems
4) Advances in surgical technique

Results

Stage I: Historical era

The first stage from 1900 to 1949 can be considered as the era of "Historical Events." In 1908, E. Payr used a saphenous vein graft in an attempt to connect the ventricular system to the superior sagittal sinus, and to the jugular vein, to drain CSF directly from the ventricle, but was unsuccessful [100]. In that same year, W. Kausch was credited for the first ventriculoperitoneal shunt placement, by using a rubber conduit [63]. In 1908, H. Cushing attempted the ventriculo-venous shunt. In addition, he attempted CSF drainage by connecting the lumbar subarachnoid space to the peritoneal cavity or retroperitoneum using silver cannulas passed through apertures in the L4 vertebral body [79]. It was B. Heile who first described the ventriculo-pleural shunt in 1914 and the ventriculo-ureteral shunt with nephrectomy in 1925 [58]. He also attempted to perform spinal CSF drainage into the peritoneum by sewing the serosa of the bowel to the dura matter using a silk suture, or other conduits like veins or latex rubber tubes [57]. In 1920, W. Dandy published his article on cannulation of the aqueduct titled "The diagnosis and treatment of hydrocephalus resulting from strictures of the aqueduct of Sylvius." He proposed reconstructing the obliterated aqueduct using a small rubber catheter left in a newly made channel, which is removed after 2 or 3 weeks [26]. In 1939, Torkildsen described the ventriculo-cisternostomy, where he passed a rubber tube subcutaneously from the lateral ventricle to the cisterna magna, and was credited for the first successful CSF shunting procedure to be internationally accepted [133]. In 1949, W.V. Cone was credited for popularizing the ventriculoperitoneal shunt [105]. In that same year, Matson treated hydrocephalus by performing a ventriculo-ureterostomy, and had a series of 50 cases [76]. The complications encountered were mechanical obstruction of the shunt tube, meningitis, and acute dehydration secondary to intercurrent infection. The breakthrough finally came at the end of this era in 1949 when Nulsen and Spitz from the Children Hospital of Philadelphia developed the first implantable shunt tube. It was constructed of a valve containing two ball valve units with platinum springs and an interposed pumping chamber [89]. They called it the polyethylene ball valve shunt. There was some initial success, but the procedure was beset with problems of blood refluxing into the brain and of blockage of the catheter, and the system was found unsuccessful. They also found the valve was difficult to make, expensive, and thus impractical.

Stage II: Experimental stage

The second stage, "Experimental stage" from 1950 to 1974, is the era of valve-regulated shunt systems for the control of hydrocephalus. The Holter valve representing the first large-

scale implantable device. It was first used in 1956, and the second patient to receive the shunt system was Holter's son, for post-meningitic hydrocephalus. The Spitz Holter valve had an impact on neurosurgery that was immeasurable, and was used for the treatment of hydrocephalus in the late 1950s and 1960. Spitz and Holter never published their surgical work, and had a 96 % success rate, in a series of 400 patients, with 15 deaths occurring due to the technique (E. Spitz, personal communication, 2000) [121]. This period was dominated by the application of these valves in ventriculo-atrial CSF diversions [121]. The valve was used in an estimated 100,000 people during the 1960 [126].

During this period, a wide variety of CSF diversions were performed. The ventriculo-atrial [86, 127, 98, 60, 136], and ventriculo-peritoneal [3, 52, 106] were the preferred CSF diversion's. The other shunts attempted were the ventriculo-venous shunts [8, 97], ventriculo-pleural fenestration shunts [41], subarachnoid/lumboperitoneal shunts [32, 45], and ventriculo-ureteral shunts [75]. The rarer diversions described were the subarachnoid-fallopian shunt [55], subarachnoid-omental bursa shunts [102], ventriculo-gastric shunts [71], and ventriculo-mastoidostomy [14]. Some of these were successful, but most failed. There was a myriad of complications encountered, some related to the procedure, others due the shunt system itself. They were shunt tube migration [84, 108], overdrainage [120], shunt infection [51], disconnection, and abdominal cystic collections [31] with the ventriculo-peritoneal (VP) shunt systems. Nephritis [103], thromboembolic events [88], Cor pulmonale [109], and superior venacaval obstruction [98] with the ventriculo-atrial (VA) shunts have been reported. Shunt infection was a constant problem. Hahn and Raimondi [51] performed 598 VP shunting procedures and encountered 81.7 % complications related to ventriculitis and shunt infection.

The other shunt systems frequently used during this era were the Denver [68], and Hakim shunt [49], both of which are differential pressure valves, and the Pudenz valve [11, 16]. In 1970, Kirsch developed the "Denver shunt." An inexpensive, rugged device, devoid of metallic parts, and capable of passing fluid of high viscosity without obstruction [68]. These valves were used in infants with grossly hemorrhagic ventricular fluid and protein concentration in excess of 5,000 mg. Both the proximal and distal slits in the shunt act as one-way valves, where a positive-pressure gradient in the direction of flow causes the valves to open. He found the number of revisions required in children were less than in adults, and were mostly due to shunt system block, failure of the recipient site to resorb CSF, tube disconnection, and infection. There was no report of subdural hematomas due to overdrainage. In 1973, Portnoy reported his results with the antisiphon and reversible occlusion valves for hydrocephalus, for the prevention of post-shunt subdural hematoma. The system, in contrast to the standard differential pressure valves, closes whenever the pressure within becomes subatmospheric, thus eliminating the siphon effect when the patient stands. Whenever the intraventricular pressure rises above the atmospheric pressure, the valve opens, regulating ventricular pressure within a normal range. However, the antisiphon valve alone did not decrease the incidence of post-shunt subdural hematomas, and this was prevented postoperatively by a percutaneous reversible occlusion valve (PROV) which was connected to the antisiphon valve. The system effectively reduced the hazards of negative intraventricular pressure with the patient in sitting or standing position [104].

Hakim paid much attention to problems of hydraulic mismatching (mismatching of the type of valve to the patient's requirements) and mechanical mismatching (mismatching of CSF pressure and ventricular size). He suggested using a combination of a ball-cone type of valve and a sensor device which adjusts the valve pressure according to the subdural stress, which he thought was the most important parameter in the treatment of hydrocephalus because it indicates the state of compression of the brain [49].

Newer innovations were the development of a new introducer for placement of the Holter ventriculo-caval shunt by Spoerri and Benini [127]. Nishimoto et al. advocate the use of ultrasound for the placement of the cardiac tube in ventriculo-atrial shunts [86]. In 1974, Yamada and Tajima proposed a method for lengthening the distal catheter of a Pudenz ventriculo-atrial shunt [143]. Wise et al. developed a ventriculo-atrial shunt catheter, based on a double telescopic principle, that elongates during longitudinal growth of the child. Radio-opaque markers on the expandable portion of the shunt allow radiographic measurement of the "growth" of the shunt. This shunt system eliminated the need for prophylactic lengthening of VA shunts during growth and also avoided the problem of distal shunt obstruction due to growth [142].

Stage III: Developmental stage

In stage 3, "developmental stage" from 1975 to 1999, a spectrum of different design shunt tubes was introduced into the market. Nearly 190 different shunt systems were available, most of them being prototypes. By this era, the ventriculo-peritoneal shunt had become the most commonly performed cerebrospinal diversion, because of the ease of placement of the distal end of the catheter, and lesser severity of potential complications. Multiple comparative studies between the ventriculo-peritoneal and ventriculo-atrial shunts were conducted [13, 70]. They found no significant difference in the shunt durability between the two. However, the specific complications associated with the ventriculo-atrial shunt method were more severe than in relation to the ventriculo-peritoneal shunt. Keucher and Mealey in their series of 228 patients with infantile nonneoplastic hydrocephalus found mortality and

infection rates were similar for both groups of patients, but children with VP shunts required fewer revisions and late complications occurred more frequently with VA shunts and were more serious [66]. Also, Vernet et al. treated a consecutive series of 120 patients with infantile hydrocephalus with ventriculo-atrial shunting. Two hundred and fifty-three shunt revisions were performed, yielding a revision rate of 2.2 per patient. Of these 253 revisions, 167 (66 %) were elective lengthening of the atrial catheter. The number of reoperations for adjusting the length of the atrial catheter or for revision of the distal end of the shunting system is a major disadvantage of ventriculo-atrial shunting, thereby favoring ventriculo-peritoneal shunting as the primary procedure for the treatment of pediatric hydrocephalus [141]

The other CSF diversions performed were the lumboperitoneal shunts [59, 80], ventriculo-gallbladder shunts [87], ventriculo-subgaleal shunts [129], ventriculo-venous shunts against the direction of blood flow [34], direct cardiac shunts [74, 82], ventriculo-ureteral shunts without nephrectomy [124], and ventriculo-pleural shunts.

Hoffman et al. described a T-tube Silastic shunting device for lumboperitoneal shunts, which was used in patients with communicating hydrocephalus. The shunt was inserted following problems secondary to arachnoiditis created by a polyethylene-type lumboperitoneal shunt. The new shunt system did not lead to arachnoiditis when inserted as a primary shunting procedure [59]. Martin et al. [74] also had good results in their patients managed with direct heart shunts (DHS) for hydrocephalus. The mean shunt survival was 79 months, and distal revision rates were lower than those reported in previous ventriculo-atrial shunt series. They found the DHS an effective self-lengthening cerebrospinal fluid shunt that may be considered as a salvage technique in the challenging shunt-dependent patient. In 1977, Raimondi et al. conducted a comparative study between the three-piece and one-piece shunt systems [107]. The one-piece shunt system was used in 161 patients, and they underwent 302 procedures, at 1.9 % operations per child, with 22 cases of ventriculitis. In the other 196 patients, the three-piece shunt was used, and they underwent 598 procedures, at 3.1 % operations per child, with 106 cases of ventriculitis.

From 1975 onwards, El Shafei popularized ventriculo-venous shunts. He initially shunted CSF to the proximal segment of an occluded neck vein. He postulates that the ligated neck vein is filled with CSF and becomes an extension of the shunt tube, which prevents venous thrombosis [33]. In 1985, he described a new approach, ventriculo-venous shunts against the direction of blood flow, and found shunting CSF in this way prevents the development of siphonage in the erect posture and regulates the intraventricular pressure (IVP) within normal limits. A shunting catheter with a simple unidirectional valve was used to establish the ventriculo-venous (VV) connection [34]. In 1987, he reported ventriculo-jugular shunts against the direction of blood flow, which he performed in 100 hydrocephalic patients, using a specially designed valved catheter and reported no failures or complications related to the dynamics of the shunt [35].

Even with new techniques, complications encountered were many, with shunt infection remaining the foremost problem of shunt implantation after mechanical obstruction and migration. Choux et al. advocated a new protocol for shunt procedures involving modifications in the immediate pre-, intra-, and postoperative management of children undergoing shunt implantation. With the new protocol, their incidence of shunt infection decreased dramatically, with two infections (0.33 %) in 600 patients and a per-procedure rate of 0.17 % [20]. Oi et al. recommended the concepts for an ideal standard shunt system in relation to complications of ventriculo-peritoneal shunts [91]. Migration of shunt tubes intracranially, to the skull base and other locations, was still commonly encountered [6, 62, 122]. Novelli and Reigel reported an unusual occurrence of reflux of bile into the CNS through a lumbar-gallbladder shunt in a patient with communicating hydrocephalus, causing chemical meningitis [87]. McIvor et al. [80] performed 375 lumboperitoneal shunts in children for communicating hydrocephalus and reported a series of orthopedic and neurological complications. Chumas et al. did a retrospective analysis of 143 patients who underwent LP shunt using the T-tube shunt and had complications such as scoliosis in 14 %, back stiffness in 13.7 %, back pain in 10 %, sciatica in 10 %, neurological changes in the lower limbs in 6 %, and symptomatic tonsillar herniation in 4.2 % of the study patients. They have also reported the incidence of hindbrain herniation may be as high as 70 % in asymptomatic patients with LP shunts, suggesting that these unique complications be borne in mind at the time of shunt insertion and at subsequent follow-up [21]. They postulated these deformities to be the result of arachnoiditis involving the conus medullaris and lower lumbar roots.

Post-shunt subdural hematomas continued to be a menace, and were reported in patients even after Portnoy et al. designed a special antisiphon valve [104]. In 1982, Yamada [144] developed a method of regulating CSF flow rate through shunts by the addition of a flow-regulating device (FRD), which consists of a small caliber (0.4 mm) Teflon tube placed in the usual connector. The device has the advantage of converting the original shunt valves to valves for higher resistance flow without replacing the entire distal catheter, and this reduced the CSF flow rate in the shunt system by approximately 30 %. It was found reliable in 32 patients, without causing obstruction.

In 1987, Sante-Rose et al. [118] developed a three-stage valve system, with the aim of providing CSF drainage at or below the CSF secretion rate within a physiological intracranial pressure (ICP) range. The first stage consists of a medium-pressure, low-resistance valve functioning as a

conventional differential pressure valve until the flow through the shunt reaches a mean value of 20 ml/h. A second stage consists of a variable resistance flow regulator that maintains flow between 20 and 30 ml/h at differential pressures of 80 to 350 mm H_2O, and the third stage is a safety device operating at differential pressures above 350 mm H_2O, thereby preventing hyper-elevated ICP.

Czosnyka et al. in 1998 tested different models of valves in vitro using computer-controlled equipment to evaluate their pressure-flow performance curves under various conditions [24, 25]. They found the Medtronic PS Medical CSF-Flow Control Valve, classic differential valves, which tend to stabilize intraventricular pressure. However, the valve allows an excess drainage rate when the body position is upright, as this shunt has low hydrodynamic resistance. Hence, they advocated the use of the Delta Valve, which has a siphon control device (SCD) to prevent CSF overdrainage in the upright position. The SCD was designed to minimize the effect of tissue fibrosis, at the same time remaining sensitive to changes in atmospheric pressure. The Medtronic PS Medical Lumboperitoneal shunt has a high resistance and high opening pressure, which helps to maintain a physiological level of CSF drainage in the vertical position, but may lead to underdrainage in the horizontal position. The overdrainage rate may be decreased due to the very high hydrodynamic resistance of the thin tube of this shunt (internal diameter of 0.8 mm, compared with 1.2 mm diameter for other distal drains). This high resistance may be the cause for underdrainage in the horizontal position. The Heyer-Schulte In-line Valve has an operating pressure range between 5.3 to 8.5 mmHg for medium-pressure valves. The medium- and high-pressure valves have a high hydrodynamic resistance, which may limit an overdrainage in the upright position and thus potentially safer than other differential pressure valves. The Heyer-Schulte Low Profile Valve (with antisiphon device (ASD)) is a classic differential pressure valve to prevent overdrainage related to posture. They found not much difference between this system and the PS Medical Delta Valve. The Heyer-Schulte Pudenz Flushing Valve with ASD is also a classic differential burr-hole valve with an optional mechanism to prevent posture-related overdrainage, with hydrodynamic properties the same as in the Low Profile Valve. The Codman Hakim-Precision Valve is also a classic differential pressure valve, with five fixed performance levels available for treating hydrocephalus. The valve has low hydrodynamic resistance, allowing overdrainage in the vertical position. Hence, not suitable for patients who are likely to develop complications related to overdrainage. The Codman-Medos Programmable Valve shunt, which can be programmed in 18 precise steps from 3 to 20 cm of H_2O. They found the flow through the valve is sensitive to negative hydrostatic outlet pressures, and this may cause overdrainage. As an isolated valve, it has extremely low resistance to fluid flow, thereby its function should be considered not in isolation but in combination with the distal catheter, and implantation of a siphon preventing device in-line with the valve may be considered when necessary. The Sophy Programmable Pressure valve, can be programmed in three precise steps. The flow through the valve is sensitive to a posture-related negative hydrostatic outlet pressure, and may cause increased CSF drainage. They found the hydrodynamic resistance of an isolated Sophy valve is higher than that of a Codman-Medos valve. The flow-regulating valve, the Orbis-Sigma has flow-pressure performance curve, indicating the shunts' ability to stabilize the drainage rate. The high hydrodynamic resistance decreases the rate of drainage in the vertical position making, making the shunt safer than the classic differential pressure shunts when one anticipates complications related to overdrainage.

The antisiphon/siphon control devices used to reduce siphoning often malfunction owing to fibrous scar formation around the valve. In view of these difficulties, Sood et al. designed a valve based on the Starling resistor concept of flow regulation that allows proximal pressure-dependent and distal pressure-independent flow. The valve allows for adjustable negative pressure in the vertical position [125]. Oi et al. also proposed that the excess drainage of CSF via a ventricular shunt system causes morphological changes in the CSF pathways, possibly leading to the isolation of compartments [93–95]. Overdrainage produces slit-like ventricles, which occurs most commonly in young infants and can lead to the slit ventricle syndrome [95, 96]. The presence of slit ventricles can in turn lead to the development of isolated ventricles such as isolated unilateral hydrocephalus (IUH), and isolated fourth ventricle (IFV) [92–95]. The pathogenesis for IUH, being functional occlusion of the foramen of Munro and for IFV they proposed a "functional obstruction," created by a pressure difference established between the supra- and infratentorial compartments [93]. This arises when a shunt overdrains the supratentorial space, causing a low supratentorial intraventricular pressure, pulse pressure, and brain compliance [94, 95]. Occasionally, a trapped fourth ventricle can cause an associated hydromyelia. Oi et al. in 1991 reported their series of such cases and described two other conditions, isolated rhombencephalic ventricle (IRV) and isolated central canal dilatation (ICCD). In IRV, there is involvement of both, the metencephalon and the myelencephalon, and in ICCD, only the central canal of the spinal cord is involved with sparing of the fourth ventricle. They concluded that the pathophysiology of hydromyelia was closely related to hydrocephalus and proposed a new clinical category of hydrocephalus, hydromyelic hydrocephalus [90].

The management of hydrocephalus in premature infants continued to be a problem [96]. Ammar [5] developed the Ammar shunt, to overcome shunt complications in premature and term neonates. The entire shunt is a low-pressure valve, with double distal slit valves. The shunts are made of soft

silicone material, of very small configuration, and no metal has been used, hence MRI compatible. Drake et al. [29] conducted a randomized controlled trial in 344 hydrocephalic children undergoing their first CSF shunt insertion, by receiving one of the three valves: a standard differential-pressure valve; a Delta valve with a siphon control component to reduce siphoning in the upright position; or an Orbis-Sigma valve with a variable resistance. They found the two new valve designs, i.e., the Delta and Orbis-Sigma valves did not significantly affect the shunt failure rates from obstruction and infection (Table 2). Davis et al. [27] also found no significant difference between these newer Delta Valve designs from the standard differential pressure valve.

In 1973, Hakim suggested the valves opening pressure was the most important factor in determining CSF drainage [49, 50]. Based on these studies, Carlos and Solomon Hakim, developed a valve which was noninvasively programmable, adjustable from 30 mm H_2O to 200 mm H_2O in 10 mm H_2O differentials. There are multiple reports with the use of the Codman-Medos Programmable Hakim Valve in the late 1990s [9, 12, 110, 114, 145]. Rohde et al. used the shunt in 60 consecutive children with hydrocephalus. They had an annual complication rate of 24.6 %, with a reoperation rate of 1.7 %. Sixteen children developed clinical and radiological signs of CSF overdrainage [114]. Reinprecht et al. retrospectively analyzed a series of 78 children who underwent a shunting procedure using this programmable valve over a period of 4 years. Treatment was successful with no need for further shunt revision in 29 of the children with primary shunt placements and 27 of the children with shunt revisions. In 10 cases of overdrainage, this was adequately corrected by readjustment of the valve operating pressure alone. They concluded as there are no criteria for prediction of the valve operating pressure needed for any individual patient they consider this valve to be beneficial in various forms of shunt-dependent pediatric hydrocephalus [110]. The other system frequently used was the pressure-adjustable valve SOPHY (PAVS), which could be changed percutaneously with the help of an externally applied magnet in order to select a high-, medium-, or low-valve opening pressure, whenever the need for a change in pressure was necessary. Significant technical complications related to the PAVS did not occur [19, 73].

Clinical trial with implantation of the new dual-switch valve (DSV) was started at the beginning of 1995. The DSV was the first construct available in the market which changes between two different valve chambers in parallel depending on the posture of the patient, and avoided overdrainage-related problems such as subdural hygromas or slit-like ventricles. In 1996, the new DSV avoided overdrainage-related problems such as subdural hygromas or slit-like ventricles, with the high risk of proximal catheter obstruction by the help of two parallel chambers in a titanium casing: one for the horizontal and the other for the vertical position. The control chamber for the horizontal position is closed by a gravity-activated tantalum ball as soon as the patient moves into an upright position [128].

In 1998, Trost et al. [135] reported their experience with the DSV, implanted in 32 adult patients with hydrocephalus. Their clinical results were good, apart from one case with an asymptomatic transient hygroma. They had no valve-related complications like overdrainage, underdrainage, or dysfunction.

During this period, a variety of new techniques were adopted. Direct cardiac shunts for hydrocephalus in infants and childhood in which an adult-size ventriculo-atrial catheter is coiled in an intrathoracic Silastic pouch and implanted in infants with hydrocephalus [82]. The proximal catheter of the shunt system was placed in the third ventricle for hydrocephalus in order to avoid obstruction of the catheter by particles of choroid plexus [48]. The percutaneous placement of VA shunts, for the venous entry, was through the subclavian vein/ transverse sinus, instead of the open cervical approach to the right atrium [15, 72].

Stage IV: Era of the programmable valve

In the final stage, "*era of Programmable valve*" from 2000 to 2010, we see a definite preference for the use of programmable valve shunt systems. Multiple analyses have been done to see if adjustable shunt valves add pressure to the budget. They found that the extra cost of the valve is outweighed by the ability to adjust the opening pressure setting noninvasively, a

Table 2 Comparative complication results of four shunt valve designs

Complications	Standard valve[a] (Differential pressure)	Cordis-Orbis sigma[a] (Flow regulatory)	Delta valve[a] (Standard valve with ASD)	Hakim PV[b] (Programmable valve)
One-year failure rate	40 %	20 %	7.55 %	24.6 %
Shunt obstruction	34.2 %	21 %	33 %	12 %
Overdrainage	2.6 %	0 %	7.8 %	26.7 %
Shunt infection	6.1 %	10.4 %	7.8 %	10 %

[a] Drake et al. [29]

[b] Rhode et al. [114]

reduced hospital stay as well as an increased quality of life for the children [7, 147]. Zemack and Romner did a retrospective study of 583 patients using the Codman Hakim PV. In 42.4 % of the cases, valve pressure adjustments was required at least once (mean number of adjustments 1.2, maximum was 23). Valve malfunction occurred in 2 % of valves implanted, nontraumatic subdural hygroma were seen in 5.1 % of patients, some of which were treated by valve pressure adjustment alone. The 5-year shunt survival was 53.1 % for first-time shunt implantations, and shunt infection rate was 8.5 % of valve implantations (Table 3) [146]. In 2003, Zemack et al. also analyzed the value of making adjustments in the opening pressure of a shunt valve and to determine shunt survival in 158 children and young adults who had received the Codman Hakim programmable valve. The shunt survival was 60.5 % at 1 year, 47.1 % at 2 years, and 43.9 % at 3 years of follow-up review, and concluded adjustment of the valve's opening pressure further improves outcome in pediatric patients [148].

Mauer et al. implanted 585 Medos Hakim programmable valves over a period of 15 years. During this period, reprogramming failure occurred in 12 cases, nine of which, the reprogramming failure required valve explantation. They concluded failure of adjusting the pressure setting is a very rare event in a Medos Hakim programmable valve [78]. Kondageski et al. [69] and Ahn et al. [1] have demonstrated good results with the Strata valve, an externally adjustable valve. Ahn et al. found the valve efficacious in the treatment of shunt-dependent hydrocephalus in children, with a 1-year shunt survival rate of 67.2 %. The necessary adjustments could be made to treat signs of CSF overdrainage and underdrainage [1].

Kondageski et al. treated 24 patients with symptomatic overdrainage, of which 19 patients (79.2 %) had severe symptoms at the time of insertion, and only one patient (4.17 %) still suffered with severe symptoms 1 year after Strata valve insertion. The number of operations was 3.42/patient/year before placement of the Strata valve, and then 0.71 % for the first, 0.56 % for the second, and 0.25 % for the third postoperative years [69]. Meier and Lemcke [81] used the adjustable gravity valve in 30 patients with idiopathic normal pressure hydrocephalus. They had no complications related to infection and shunt dislocation. However, an overdrainage was seen in 3 % of patients. The advantages of this programmable valve with gravitational unit are the absence of unintentional readjustments through external magnets and the ability to control the valve settings using an accessory instrument without the need for X-ray monitoring. Chang et al. [17] managed patients successfully, with normal pressure hydrocephalus using lumboperitoneal shunt system with the Codman Hakim Programmable valve. The valve was incorporated in the lower pressure of 30 mm of H_2O. Takahashi et al. analyzed the possibility of shunt removal using the programmable Medos and Sophy valves. They found that it was possible to remove shunt systems in 50 % or more cases of pediatric hydrocephalus where programmable valves are used [131].

Hanlo et al. [53] conducted a multicenter prospective 5-year shunt survival study to evaluate the long-term results of the flow-regulating shunt the Orbis Sigma Valve II Smart valve system. Five hundred fifty-seven patients (48 % adults and 52 % were children) were selected. Shunt obstruction occurred in 13.5 %, overdrainage in 1.8 %, and infection in 8.2 %. The shunt failure-free interval was 71 % at 1 year, 67 % at 2 years, and 62 % at the 5-year follow-up examination. They demonstrated the effectiveness of flow regulation in the treatment of hydrocephalus both in children and in adults, concluding these shunts limit the incidence of overdrainage and shunt-related complications.

Two different technical principals of gravitational valves are available, the Aesculap-Miethke GA-Valve (Counterbalancer) and the dual-switch valve (Switcher). The *G* valves are posture dependent because they change their total resistance based on the patient's posture, whereas the conventional differential pressure valves are posture independent. In their series of 54 patients with normal pressure hydrocephalus (NPH), Kiefer et al. [64] had 9 patients with clinical or radiological signs of overdrainage. Shunt infection and underdrainage did not occur. Overall, they had a better clinical outcome in 80 % of shunt responders in contrast to earlier

Table 3 Comparative complication results of three shunt valve designs

Author	Zemack	Hanlo et al.	Eymann et al.
Type of valve.	Codman Hakim PV	Orbis Sigma Valve II	Miethke Paedi-Gav
Number of patients	583	575	55
Overdrainage	5.10 %	1.80 %	3.60 %
Infection	8.50 %	8.20 %	N/A
Shunt survival 1 year	N/A	71 %	75 %
Shunt survival 2 years	N/A	67 %	68 %
Shunt survival 5 years.	53 %	62 %	N/A

N/A not available

reports [56, 140]. As a result, they recommended the low-pressure GA-valve as the standard NPH valve. The newer gravitational valves are the adjustable gravity valve by Aesculap, the ProGAV, and a new pediatric gravitational shunt, the Paedi-Gav. Rhode et al. [115] used the ProGAV in 53 children with hydrocephalus. The ProGAV is a differential pressure unit with adjustable opening pressures and a gravitational unit with a fixed opening pressure. They did not observe any valve-related complications. The infection rate was 7.5 % warranting removal of the shunt. In 19 children, the opening pressure was changed at least once during the follow-up period, for underdrainage in 10, overdrainage in 8, and shunt weaning in 1, with substantial clinical improvement in 18 children. They found overall good clinical results in 47 (88.7 %) of the 53 valve placements. The advantage of the ProGAV is the absence of unintentional readjustments through external magnets and the possibility of controlling the valve settings without the need for X-ray monitoring. The most significant disadvantage is adjustment problems after implantation. The results of the study following the usage of the system by Meier et al. [81] showed an overall improvement rate of 72 %, comparable with the range in the literature. Eymann et al. [40] also showed good results with the Miethke Paedi-GAV with a 1- and 2-year survival rates of 75 and 68 %, and overdrainage rate of 3.6 % which falls within the range of recently published overdrainage rates for different modern shunt designs [28].

Bhasin et al. [10] stated, because of the benefits of the ventriculo-peritoneal over the ventriculo-atrial shunts, every effort should be made to preserve the peritoneum as the target for the distal catheter. As an alternative following VP shunt failure, or for other reasons, the other CSF diversions performed are the ventriculo-atrial shunts via transfemoral vein placement [137], ventriculo-subgaleal shunts [138, 139], ventriculo-cholecystic shunts [44, 130], lumboperitoneal shunts [132], ventriculo-pleural shunts [134], ventriculo-venous shunts [36], ventriculo-vesical shunts [4], ventriculo-omental bursa shunts [77], and direct heart shunts [74]. With the increasing use of neuroendoscopy, endoscopic stent placement for the treatment of trapped fourth ventricle [116] and for Dandy-Walker malformation [83] are the other CSF diversions performed.

Shunt complications continue to be a neurosurgeons nightmare. Visceral perforation and migration [18, 47], shunt infection [38], overdrainage, and the slit ventricle syndrome [2, 67, 111] are still commonly encountered. Enger et al. from their study conclude the risk factors for shunt infections appear to relate to epidemiological characteristics rather than to surgical factors [38]. Albright and Tyler-Kabara postulated that long-standing overdrainage of cerebrospinal fluid via shunts dampens the normal cerebral pressure waves; resulting in retardation of calvarial growth, leading to ossification of the sutures, which become unable to expand to allow normal brain growth. They concluded shunt-induced craniostenosis should be considered for children with symptoms of slit ventricle syndrome for whom shunts are functional but intracranial pressure is increased, and cranial expansion surgeries are more appropriate treatments than subtemporal decompressions for such children, given the diffuseness of the suture pathological features [2]. Rare cases of unusual migration of the distal catheter of a ventriculo-peritoneal shunt into the heart have been reported [43, 113].

Ventriculo-subgaleal shunts have shown to be successful in pediatric neurosurgery. It is simple, inexpensive, and especially useful as a temporary CSF diversion in cases where the peritoneal cavity is not suitable for distal shunt implantation [54, 129, 139]. Tubbs et al. [138] found the average length of survival of the primary, secondary, and tertiary subgaleal shunts was 37.4, 32.4, and 19.6 days. Patients obtain therapeutic diversion of their CSF for greater than 2.5 months with intermittent subgaleal shunt revisions, and this length of time is sufficient to manage and resolve issues making ventriculo-peritoneal shunting undesirable. There is also increasing reported literature on the retrograde ventriculo-sinus (RVS) shunts, which is a water-tight connection delivering excess CSF to the superior sagittal sinus against the direction of blood flow [23]. El Shafei et al. had no problems related to improper CSF drainage or sinus thrombosis, and conclude the RVS shunt is a simple, minimally invasive, physiological procedure for treatment of hydrocephalus, suitable for all ages [36, 37].

Shunt infection is still a severe life-threatening complication. A promising alternative is to use antibiotic-impregnated shunt (AIS) catheters, which have been tested extensively in vitro, showing promising sustained antibacterial activity. However, there have been fewer reports of the impact of these catheters on the infection rate in vivo. Multiple studies have been conducted using these shunt systems [112, 123]. Ritz et al. in their study found antibiotic-impregnated shunts did not significantly reduce shunt infections in hydrocephalus patients [112].

The innovations in surgical technique to reduce complications are many. The Seldinger technique is for distal catheter placement in the superior sagittal sinus for reducing the risk of blood loss and air embolism in ventriculo-sagittal sinus shunts [117]. Laparoscopy-assisted implantation of a peritoneal catheter has been found to be safe, minimally invasive, and technically an easy approach, allowing accurate placement of the distal catheter in the peritoneal cavity in children. It also enables retrieval of fractured catheter segments and allows confirmation of the patency of the shunt system [61]. Endoscopic aqueductal stent placement has been effective for treatment of isolated fourth ventricle, and aqueduct stenosis in which endoscopic third ventriculostomy is not feasible [22, 39, 116]. The percutaneous transfemoral placement of ventriculo-atrial shunts has proven useful when all other measures have failed [137]. An alternative site of placement of the

lower end of the shunt in the suprahepatic space has been found advantageous to placing it in the general peritoneal cavity, with rewarding results [99].

Minimally invasive lumboperitoneal shunt placement was conducted after administration of a local anesthetic, the procedure performed placing a guide wire and a peel-away sheath under fluoroscopic and CT guidance. A Codman Hakim programmable valve system is used for the procedure. They found the procedure is less invasive than conventional lumboperitoneal shunt insertion and could be performed as an outpatient surgery for treatment of idiopathic normal pressure hydrocephalus [85]. Since we are faced with 1- and 2-year shunt survival rates of 50–70 and 47–53 % [28, 29, 65, 146], we cannot stop searching for better solutions for more successful CSF diversions and results.

Conclusion

Is there "an ideal shunt"? As we see from the reported literature, there is a wide variety of shunt systems available in the market today, each having its advantages and disadvantages. It will be unfair at this stage to label one shunt system better than the other. From our analysis, it would be correct to conclude that every shunt system is "an ideal shunt" provided the choice of the shunt system used should be made, by matching the performance of the shunt to the disturbed CSF dynamics of a given patient, and not by the manufacturer. The most important factor being the opening pressure, which is the pressure that opens the valve, allowing CSF to drain. Preoperative selection of the most suitable opening pressure for any individual patient is still difficult because the pressure often needs to be adjusted during the postoperative period.

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