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Shunt for the Hydrocephalus: the Past and the Present

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ABSTRACT

Shunt is the most popular treatment for hydrocephalus, and of course, it is not a perfect treatment. It was born only in the past century and showed magnificent improvement in these recent decades. The fact that the driving force for these rapid advances was a need to alleviate the complications tells us that we should have deeper understandings about the complications in order to improve our present shunt and come up with a better treatment. It is important that we know well about the mechanisms of shunt: the popular sites, structure of valves and its various types. VP shunt seems to be the most popular choice for drainage location, but choices for valves are more diverse. Various improvements for present shunt are being discussed. We hope to retrospect back on the birth of shunt, see what it is and its problems, and glimpse at the present it would bring on by contributing to the improvements of treatments.

KEYWORDS

Neuropsychology, Cerebral Ventriculomegaly, Shunt, History

INTRODUCTION

The term "Hydrocephalus" is a compound of two Greek words; "hydro" which means "water", indicating CSF, and "cephalus" which means "head". Hydrocephalus is a medical condition in which the CSF is not circulating properly, accumulating in the head, leading to significant increase of intracranial pressure (ICP), convulsion, eye deviation, narrowed vision, and mental disability. Progressive enlargement of the head happens for the infant whose head has not started hardening.

The cause of hydrocephalus is not completely known yet, and there are only some assumptions; inherited genetic abnormalities, developmental disorders, traumatic injury etc. Hydrocephalus is classified congenital or acquired by apriority, and acute or chronic by chronicity. It is also classified as communicating and obstructive [1]. Communicating hydrocephalus does not have CSF-flow obstruction between the ventricles and subarachnoid space. It is thought to be caused by problems in CSF reabsorption. In obstructive hydrocephalus, also called non-communicating hydrocephalus, the passages connecting the ventricles are blocked, preventing the CSF to flow between the ventricles.

Most common surgical treatment for hydrocephalus is the shunt. It drains excessive CSF from head to another place of the body. It is the most commonly used technology to treat hydrocephalus [2]. Shunt system consists of three fundamental parts: proximal catheter, valve and distal catheter. Proximal catheter drains CSF from ventricles and valve gets CSF from the proximal catheter and controls the flow rate of drained CSF. Distal catheter gets CSF from the valve and outlets CSF to appropriate shunt sites, such as peritoneal cavity or heart. To improve accuracy or effectiveness of the system, some shunt systems include ancillary device such as reservoir, anti-siphon, and pumping chamber.

MAIN BODY

History of Shunt

Shunt may now be the most popular treatment for hydrocephalus, but it has not been long since the introduction of this method. Shunt has improved rapidly in these recent decades, after its birth about a-century-ago. Before we go in deeper about the mechanism of shunt, first we would like to talk about the background of how shunt had emerged and the major changes shunt has gone through after its birth.

History of shunt can be divided into 3 periods: (I) before shunt, (II) after the first shunt without valves and (III) after the modern shunt with valves (Figure 1).

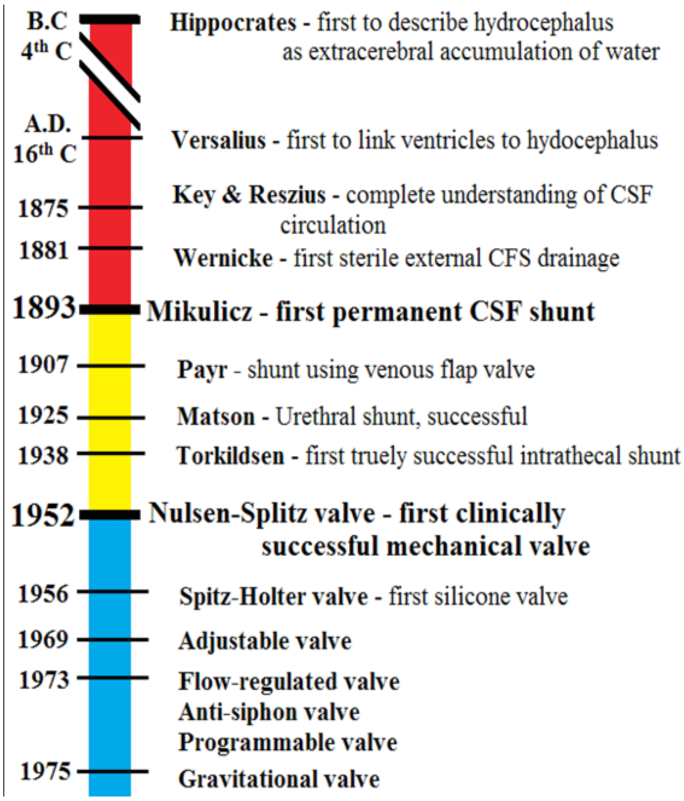

Figure 1. Timeline of major events

I. Before shunt: foundation for development of shunt (~ late 19th C)

Before the late 19th century, shunt was not introduced yet. Instead, inaccurate treatments, some of which being more detrimental, were conducted in vain [3-6].This was mostly due to lack of understanding of the hydrocephalus. In the majority of the time of this period, ventricles were not even linked to hydrocephalus; instead, a false belief prevailed that extracerebral accumulation of water was the cause of hydrocephalus [3-5]. After the ventricles were found responsible for hydrocephalus in the late years, a need for continuous drainage from ventricles emerged, leading to the birth of shunt.

Early efforts to understand Hydrocephalus

Hippocrates first introduced the Hydrocephalus into history in B.C 4th century, he was not only the first to describe and use the term hydrocephalus [3, 5], but also the first to drain extra water of subarachnoid space [4-5].However, when he referred to hydrocephalus, it did not mean the enlargement of the ventricles, but the extracerebral accumulation of water [3-5].

Galen of 2nd century was the first to hypothesize that choroid plexus produced CSF. However he knew only about the fluid collections outside the brain, being unable to link it with ventricles [3-5].

Early understanding of hydrocephalus was limited to outside of the brain. The main reason is that it was mostly based on observations or animal dissections, since human autopsies were highly restricted in the ancient times. Understanding of ventricles was promoted only after the dissection of human cadavers was tolerated after the Renaissance in 16th century [4].

Connection of Ventricles to Hydrocephalus

After centuries of misunderstanding and futile treatments, Andreas Versalius first made the connection of ventricle to hydrocephalus in the 16th century [3]. Through human necropsies, he found that water had collected in the ventricle within the brain, not in between the skull and its outer [3-6]. His finding dismissed the 2000-year-old misunderstanding of hydrocephalus and laid the foundation for more accurate treatments.

Intensified knowledge of CSF circulation was also one of the contributing factors of improved treatments in the years afterwards. In the 17th century, Thomas Willis was the first to suggest that CSF must be drained into the venous system after it was produced in choroid plexus [4-5]. However it was only in the late 19th century that the CSF circulation was proved irrefutably, when Key and Reszius (1875) perspicuously explained the entire CSF circulation from production to absorption [3-5].

In parallel with these enhanced understanding of pathophysiology of ventricles and hydrocephalus, advances in treatments were made, such as attempts to drain extra accumulation of CSF directly from the ventricles. Le Cat (1744) was the first to perform the ventricular puncture [3], and later in 1881, Wernicke conducted the first sterile ventricular puncture and external CFS drainage [5].The low efficiency of these drainage increased the search for permanent procedures of CSF diversionary, which lead to the introduction of shunt [5].

II. Start of shunt: the first shunts with tubes (late 19th C~ mid-20th C)

The first shunt had a simple form, with tubes and mostly valveless. Shunts in this period had a high failure rate mainly due to insufficient implant materials such as rubber, plastic, metal and gold [5, 7]. They were prone to obstruction by collection of particulate matter or adhesions and caused immune reactions for foreign materials [6, 7].Reflux of blood or other body fluids toward the ventricle was also an important factor of the high failure rates [5, 7],which called for unidirectional valves, later invented in the next period.

The First Shunts

The first permanent shunt surgery was conducted by Mikulicz in 1893. He implanted a glass wool wick into the lateral ventricle, which extended to subgaleal area for drainage [3]. Later in 1895, he conducted another shunt surgery using gold metal tubes and cat gut strands [5, 7]. However, the first truly successful intrathecal shunt was the Torkildsen shunt in 1938, which drained CSF to cisterna magna [6]. It even remained as a popular method until 1970s for occlusive hydrocephalus until it was replaced by more current techniques [3, 4].

Extrathecal shunt to low pressure compartments was first proposed by Gärtner, in 1895 [3].Ventricular-Peritoneal shunt, in which CSF is drained to abdominal cavity, was first conducted by Kausch in 1905, using a rubber tube [3, 5, 7]. However, these methods were not largely successful due to many complications, such as distal obstructions [3,17].

Growing needs for valves

In a search for a way to prevent complications by refluxes, the first attempts were to try natural valves; the flaps in our veins. In 1905, Erwin Payr conducted a ventricular shunt, draining to the superior sagittal sinusa [3]. In this surgery, he used autologous saphenous vein, with preserved venous flap valves as tubes. Later autopsy of the patient revealed that despite the open passage, blood had not refluxed into the ventricles [3], suggesting the effectiveness of the valves.

The concept valves and flow regulation was brought into more spotlight after urethral shunts were performed by Matson in 1925. This shunt was a success [4, 7], owing to the natural valve function of the ureter, which has the correct hydrodynamic resistance [5, 7]. These successes in shunts with natural valves in contrast with failures in valveless shunts evidently showed the necessity of unidirectional valves.

III. Start of Modern Shunt: introduction of mechanical valves (~ mid-20th C)

Finally, the need for a unidirectional valve gave birth to the modern shunts with mechanical valves, greatly increasing the safety and effectivity of the shunt surgery. However, it was not the valve alone that contributed to this success; introduction of biocompatible material, the silicone, at about the same time led to the modern era of effective shunts.

Start of Mechanical valves

The first magnetically operated valve was invented in 1948, but it was not very fruitful [3].The first clinically successful valve was the Nulsen-Spitz valve in 1952, a VA shunt using two ball-and-cone valves with springs [3-7].

Introduction of silicone in shunts

Despite the success of the first mechanical valve, the spread of valves in neurosurgery was only after the Spitz-Holter valve in 1956, the first case to introduce silicone as the shunt material [3, 6].This silicone slit valve was very successful, and they are still being made until today in an almost unchanged form [3].

Silicone, introduced during the Second World War, was proved unreactive to the human body, with no adverse effects when in contact with brain tissue, blood, bone and muscles [3, 6]. It did not have the danger of occlusion since nothing stuck to it, and was able to withstand long-term mechanical stress [3, 6].It was only after the advent of silicone as the implant material that the VP shunts gained popularity. After its birth in 1905, the VP shunt had been abandoned due to the fear of frequent complications of occlusion[7]. After this biocompatible silicone was brought into shunt, remarkably rapid progresses were made in the years afterwards.

Development of more valves

After the Spitz-Holter valve, there have been various improvements in the valves and the types have become diverse. Nowdays there are more than hundreds of different valves that are in use. The detailed mechanisms of the valves that would be introduced here would later be explained in the part III of this paper.

Different pressure valves. These valves were the first valves to be invented, including the Nulsen-Spitz, Spitz-Holter valve. 4 types of these valves, diaphragm, proximal slit, distal slit and ball-and-cone valves were all constructed as early as in 1960 [3]. Based on these valves, other types or valves were addressed to overcome the problems it had, such as overdrainage during vertical position [3].

Adjustable valves. The first adjustable valve was introduced in 1969, which had a spring that could be adjusted by a screwdriver. In 1973, Portnoy came up with an on-off switch for valves which can be operated percutaneously. The modern programmable valve has first designed by Hakim and produced in 1984, and owing to its superior accuracy, it is now widely used in difficult cases [3, 18, 19].

Flow-regulated valves. The concept of a valve that changed resistance in response to pressure change and thus regulate flow was first devised by Hakim in 1973. Later in 1984, it was realized as the first practically available CordisOrbis-Sigma Valve, and in 1996 it was amended as the Orbis-Sigma Valve II [3].

Anti-siphon valves & Gravitational valves. Portnoy conceived the Anti-siphon valve, the first use of gravity for controlling the shunt valve, in 1973. It was designed to progressively close as negative pressure increases, which is highest at the upright position [3, 6]. It is usually combined with diaphragm valves. Gravitational valve also use gravity to control the valve, but by using metal balls instead of columns as in the anti-siphon valve. It was first patented by Hakim in 1975, and is mainly used in a valve-device combination or as a supplementary device [3].

As we can see, the history of shunt first started with wrong assumptions and hopeless treatments. However, efforts to ameliorate the complications of the previous treatment gave birth to the first shunts and valves, and worked as the driving force of rapid improvements in the years afterwards. Therefore, understanding the current mechanisms and complications of shunt, the currently most popular treatment, is vital for us to develop more effective treatments in the nearby future.

Mechanism of Shunt

Shunt is a very complicated process, with various drainage sites and hundreds of valves. We will focus on the main shunting sites and then move on to address the main types of valves.

I. Shunt Sites

There are mainly two possibilities of placing shunts to begin cerebrospinal fluid drainage. One called ventricular shunt starts drainage inside the brain, namely ventricles through Ommaya reservoir. The other one called lumbar shunt drains CSF from lumbar thecal sac.

Ventricular shunt is the most common treatment for communicating and obstructive hydrocephalus. Many variations exist in the location for drainage: ventriculo-peritoneal shunt (VP shunt), ventriculo-atrial shunt (VA shunt), ventriculo-pleural shunt (VPL shunt), ventriculo-cisternal shunt (Torkildsen procedure), and ventriculo-superior sagittal sinus shunt (VSSS shunt).

Lumbar shunt is an alternative treatment available for hydrocephalus when ventricular shunts are unavailable to be applied, especially in cases of communicating hydrocephalus, and normal pressure hydrocephalus. [25] The variations are lumbar-peritoneal shunt (LP shunt), and lumbar-subcutaneous shunt (LS shunt).

Ventriculo-peritoneal shunt

Ventriculo-peritoneal shunt, in which the CSF drainage ends at peritoneal cavity located in the abdomen, is the most popular shunt among all the other variation[20]. CSF are routed to peritoneal cavity and absorbed in micro vessels surrounding the cavity. Kausch first successfully performed it at 1905.

VP shunt is more reliable compared to VA shunt, which could lead to serious cardiopulmonary complications [7]. Since the introduction of silicone as the implant material, which significantly reduced peritoneum occlusion [8], VP shunting became the most preferable choice for treating hydrocephalus. VP shunt has some noticeable advantages including low mortality rate, less needed revision with much simpler procedures, and not very critical complications [9, 10, 31, 32].

However, peritoneal shunts are known to have high tendency for obstruction [7], and some peculiar complications such as inguinal hernia, abdominal ascites, perforation of a viscus, and neoplasm spreading or infection to the peritoneal cavity [9]. Also, a hydrocele is reported to be followed [7, 30, 35, 36].

Ventriculo-atrial shunt

The absorption point of this shunt is located in the right or left atrium [11], where CSF diversion flows into the veins. It is well-used, common technique for treating hydrocephalus. However, difficulties exist for accurately placing the catheters, and some major complications are critical: arrhythmias, pulmonary hypertension, bacteremia, renal failure and cardiac tamponade [9, 12, 21, 33].

Ventriculo-pleural shunt

CSF is routed to the pleural cavity where the lungs are located. This is not a mainly used treatment because of pleural effusions [13, 22].

Ventriculo-cisternal shunt

Also known as the Torkildsen shunt, or ventriculocisternostomy, implantation of this shunt began since Torkildsen made the first successful installation in 1938 [7].When a blockage occurs between the ventricles and the subarachnoid cavity, this treatment would be effective for hydrocephalus [23].

Ventriculo-superior sagittal sinus shunt

Effective when long-distance shunts are unavailable, as in growing children or patients on concern for complications [14, 24]. It is considered highly risky, and would be the final option when the other shunts have failed to treat hydrocephalus.

Lumbar-peritoneal shunt

This shunt is an effective treatment for various types of diseases, such as communicating hydrocephalus, idiopathic intracranial hypertension, normal pressure hydrocephalus, spinal and cranial CSF leaks, pseudomeningoceles, slit ventriclesyndrome, and growing skull fractures [15, 25].

Compared to the VP shunt, LP shunt has some advantages since the operations are done extracranially. Revisions are required less often, catheter length is shortened, and infection and malfunction rates are lower [26]. Also it was reported that there was very low occurrence of ACM after LP shunt [15].

Lumbar-subcutaneous shunt

CSF drainage begins from thecal sac and ends at the subcutaneous of surrounding region. It is known to be effective treatment for normal pressure hydrocephalus and benign intracranial hypertension [16, 27, 28].

Other types of shunts

Ventriculo-ureteral shunts or Lumbar-ureteral shunts which had simple, physiological method had been tested by Matson in 1925. However, he had been abandoned this type of shunt later on, because it had repeated complications, such as meningitis or metabolic crises [7, 29].

As we can see, there are too many complicated set of shunts. Each shunt has its own advantages and disadvantages. If not an obvious case, it is highly difficult for the surgeon to determine which shunt would be the best for a patient before conducting the operation. Yet, the researchers are constantly looking for universal solution for treating hydrocephalus and better ways to handle risk factors, which suggests more improvement in shunt in the nearby future.

CONCLUSION

We have followed the trajectory of shunt, from it birth, present usage. Every little success, and every little failure matters since it deepens our understanding about the disease and its treatment and gives us a clue about what we should focus on to improve and innovate for more effective treatment in the future. Since its birth in the previous century, shunt has influenced lived of tremendous amount of people by alleviating them from fears of hydrocephalus. Even though they might disappear in the future, they still would be saving lives, through many other treatments that have learned their ways to improve from our present shunt.

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