An overview of hydrocephalus and shunts used in the clinical management of hydrocephalus.

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Abstract

Hydrocephalus is a complex neurological condition characterized by an abnormal accumulation of cerebrospinal fluid (CSF) within the ventricles of the brain, leading to increased intracranial pressure. The clinical management of hydrocephalus often involves the surgical placement of a shunt system to divert CSF and restore normal fluid dynamics. This article provides a comprehensive overview of hydrocephalus, its etiology, clinical presentation, and various types of shunts used in its management. The outline of this report encompasses the principles of shunt surgery, indications for shunt placement and primary types of shunts used in the clinical management of hydrocephalus, including ventriculoperitoneal (VP), ventriculoatrial (VA), ventriculopleural (VPL), lumboperitoneal (LP) and ventriculo-subgaleal (VS) shunts. Additionally some non-conventional shunts such as ventriculo-osseous, ventriculo renal, ventriculo gall bladder, intraventricular (Aqueductal stents) and long-tunnelled external ventricular drains are also discussed. The distinct features of each shunt type, its associated advantages, limitations and complications are also outlined together with a comparison between pressure vs flow regulated valves. An understanding of the different shunt types and their specific characteristics is essential for clinicians to make informed decisions in tailoring treatment to individual patients. This review offers a comprehensive understanding of shunt characteristics which we believe will empower clinicians to make informed decisions tailored to individual patients, ensuring effective CSF diversion and long-term management of hydrocephalus.

Key words: Shunts, hydrocephalus, pressure valves, flow valves, neurosurgery.

Background

Hydrocephalus is a condition in which there is an abnormal accumulation of cerebrospinal fluid (CSF) in the ventricles of the brain, leading to increased intracranial pressure and potential brain damage.[1, 2] The incidence rate of hydrocephalus varies among different geographical regions. In general, the reported incidence rate is around 0.5 to 1.5 per 1,000 live births worldwide. However, the incidence rate can be higher in certain regions due to various factors such as genetic predisposition, environmental factors, and healthcare access.[3] The reported incidence rate of hydrocephalus in Europe is around 1.3 per 1,000 live births. A study conducted in Italy reported an incidence rate of 0.9 per 1,000 live births, while in Denmark an incidence rate of 1.5 per 1,000 live births was reported. In the United States, the reported incidence rate of hydrocephalus is around 1 in 1,000 live births, with an estimated 700,000 people living with the condition.[4] In developing countries, the incidence rate can be higher (1.5 to 6 per 1,000 live births) due to factors such as limited access to healthcare, poor maternal nutrition, and higher rates of infections such as meningitis.[5] The incidence rate of hydrocephalus in Asia is reported as 0.5 to 2.6 per 1,000 live births. Highest incidence of hydrocephalus is reported in sub-Saharan Africa, where incidence range from 3.0 to 6.6 per 1,000 live births. Other regions that have reported higher incidence rates include Latin America (1.7 to 4.5 per 1,000 live births).[2]

Factors which can contribute to incidence of hydrocephalus

Several factor can contribute to the incidence of hydrocephalus (Figure 1). In many cases, the cause of hydrocephalus can be multifactorial, involving a combination of genetic, environmental, and developmental factors.[2, 6, 7] Some commonly associated causes of hydrocephalus are: 1) Congenital abnormalities (neural tube defects, spina bifida, and other genetic or chromosomal disorders). 2) Infections (meningitis or encephalitis, can cause inflammation and scarring that disrupts the flow of cerebrospinal fluid and leads to hydrocephalus). 3) Trauma (Head injuries sustained in car accidents or falls, can cause bleeding or swelling in the brain that interferes with the normal circulation of cerebrospinal fluid). 4) Tumours (Brain tumours or other growths can block the flow of cerebrospinal fluid and cause hydrocephalus). 5) Haemorrhage (Intracranial haemorrhages caused by an aneurysm or arteriovenous malformation, can increase pressure on the brain and lead to hydrocephalus). 6)

Idiopathic: In some cases, the cause of hydrocephalus is unknown and referred to as idiopathic. 7) Premature birth (Premature infants are at increased risk for developing hydrocephalus due to the immaturity of their brains and the potential for bleeding and associated complications).8) Family history: Some types of hydrocephalus may be inherited or associated with genetic conditions (L1CAM gene mutations, defects with transthyretin or the SPG11 protein). 9) Environmental factors: Exposure to certain toxins (lead, mercury, or pesticides) or environmental factors may increase the risk of developing hydrocephalus.

Clinical management of hydrocephalus

Treatment for hydrocephalus typically involves among others the surgical placement of a shunt or other device to divert the excess CSF away from the brain (Figure 2). In some cases, medication or other non-surgical interventions may be used to manage symptoms or underlying causes of hydrocephalus.[1, 8, 9] Shunts are medical devices used to treat hydrocephalus by diverting excess CSF from the brain to another part of the body, such as the abdominal cavity, where it can be reabsorbed. Besides the use of shunts, endoscopic third ventriculostomy (ETV) which is a minimally invasive surgical procedure that involves making a fenestration in the floor of the third ventricle of the brain to allow CSF to flow out of the ventricles and into the subarachnoid space is also considered.[10] ETV is typically used in cases of non-communicating hydrocephalus, in which there is a blockage preventing CSF from flowing freely between the ventricles. Ventriculocisternostomy (VC) is a similar procedure to ETV, but involves creating a hole in the floor of the fourth ventricle to allow CSF to flow into the cisterna magna (reservoir of CSF located at the base of the brain and is used for spinal tapping). VC is typically used in cases of non-communicating hydrocephalus in which the blockage is located near the fourth ventricle.[11, 12] Alternatively choroid plexus coagulation (CPC) which is a surgical procedure that involves using heat or other methods to destroy part of the choroid plexus, which is responsible for producing CSF is also performed.[13, 14] By reducing the amount of CSF produced, CPC can help alleviate the symptoms of hydrocephalus. However, CPC is less commonly used than shunt surgery or ETV/VC, as it is a more invasive procedure with a higher risk of complications. Additionally medication such as diuretics may be used to reduce the production of CSF together with antibiotics to treat infections that may be contributing to hydrocephalus.

Shunts in the management of hydrocephalus

Shunt surgery is considered a relatively safe and effective treatment for hydrocephalus.[1, 15] The success rate of shunt surgery varies depending on the specific type of shunt used and the individual patient's response to the procedure. Some studies have reported success rates of up to 80-90% for shunt surgery in the short term, with long-term success rates ranging from 50-70%.[16, 17] The prognosis for shunt surgery to treat hydrocephalus depends on several factors, including the underlying cause of the hydrocephalus, the age of the patient, the presence of any other medical conditions, and the effectiveness of the shunt placement. Shunt surgery is associated with certain risks and complications, including infection, shunt malfunction or blockage, over drainage or underdrainage of cerebrospinal fluid, and other neurological complications. The overall prognosis for shunt surgery depends on the individual patient's response to treatment, as well as ongoing management and monitoring of the shunt system. In principle the excess fluid in the ventricles can be drained to any feasible outlet in the body restricted only by the neurosurgeons competence and creativity. As a consequence several types of shunts have evolved over the years.

Here is a list of various types of shunts (Figure 3) which are available for the treatment of hydrocephalus:

1. Ventriculoperitoneal (VP) shunt: This is the most common type of shunt used for hydrocephalus. It involves placing a catheter from the ventricle of the brain to the peritoneal cavity in the abdomen.
2. Ventriculoatrial (VA) shunt: This type of shunt involves placing a catheter from the ventricle of the brain to the right atrium of the heart.
3. Ventriculopleural (VPL) shunt: This type of shunt involves placing a catheter from the ventricle of the brain to the pleural cavity surrounding the lungs.
4. Lumboperitoneal (LP) shunt: This type of shunt involves placing a catheter from the lumbar region of the spine to the peritoneal cavity in the abdomen.
5. Ventriculosubgaleal(VS) shunt: This type of shunt involves placing a catheter under the scalp and skin, into a pocket or space that has been created between the scalp and skull.
6. Miscellaneous shunts: Ventriculorenal (ureteral &vesical), Ventriculo-osseous (sternal, iliac, humeral, mastoid, diploic) and Ventriculo-gallbladder shunt, Aqueductal stents, Long-tunnelled external ventricular drains are less

commonly used types of shunts to treat hydrocephalus. These approaches are considered when the above options are not feasible due to medical complications.

The choice of shunt type will depend on factors such as the patient's age, the underlying cause of hydrocephalus, the location of the blockage in the CSF flow, and the presence of any other medical conditions. We further elaborate on the characteristics of each of the shunt types which we believe is essential for clinicians to make informed decisions in tailoring treatment to individual patients.

Ventriculoperitoneal (VP) shunt is the most common type of shunt used for the treatment of hydrocephalus. The VP shunt is a medical device that diverts excess CSF from the ventricles of the brain to the peritoneal cavity in the abdomen, where it can be reabsorbed by the body.[18, 19] There are several types of VP shunts available, each with its own features and benefits.

Fixed-pressure valve VP shunt*:* This type of shunt has a fixed pressure valve that allows the CSF to drain from the ventricles of the brain to the peritoneal cavity at a predetermined pressure. These shunts are typically used in patients with stable CSF pressure levels.[20, 21] This type of shunt has a valve that is designed to maintain a constant pressure differential between the ventricles of the brain and the peritoneal cavity in the abdomen. The fixed-pressure valve is set at a predetermined pressure, usually between 70-200 mmH_2O, which determines the rate at which CSF is drained from the brain. When the pressure in the ventricles of the brain exceeds the pressure set by the valve, the valve opens to allow the excess CSF to drain into the peritoneal cavity, where it can be reabsorbed by the body. The fixed-pressure VP shunt consists of three main components: a catheter, a valve, and a distal catheter. The catheter is a flexible tube that is inserted into the ventricle of the brain to drain excess CSF. The valve is located along the catheter and regulates the flow of CSF from the brain to the peritoneal cavity. The distal catheter is a second flexible tube that carries the excess CSF from the valve to the peritoneal cavity. The fixed-pressure VP shunt is typically used in patients with stable CSF pressure levels. However, this type of shunt may not be appropriate for patients who experience fluctuations in their CSF pressure, as the valve cannot adjust to these changes. In addition, the fixed-pressure VP shunt may require revision if the patient's pressure requirements change over time. Like all VP

shunts, the fixed-pressure VP shunt carries a risk of complications such as infection, blockage, or failure of the valve. Patients with a VP shunt should be monitored closely for signs of complications and undergo regular imaging studies to ensure the shunt is functioning properly. Some of the commercially available fixed-pressure VP shunts are: 1) *Codman Hakim Fixed Pressure Valve (FPV) System*: This shunt is designed to provide continuous cerebrospinal fluid drainage at a fixed pressure between 30 and 200 mmH_2O. The valve is adjustable prior to implantation, allowing for customization based on the patient's needs. 2) *Medtronic Strata II Valve:* This shunt has a fixed pressure range between 35 and 200 mmH_2O and is designed to prevent over-drainage and under-drainage of cerebrospinal fluid. The valve is MRI compatible and can be adjusted non-invasively using a magnetic field. 3) *Sophysa Polaris Fixed Pressure Valve*: This shunt has a fixed pressure range between 30 and 200 mmH2O and is designed to provide a stable flow of cerebrospinal fluid. The valve can be adjusted using a magnetic field and is MRI compatible. 4) *Integra Lifesciences Certas Plus Fixed Pressure Valve System:* This shunt has a fixed pressure range between 30 and 200 mmH_2O and is designed to provide consistent cerebrospinal fluid drainage. The valve is adjustable prior to implantation and can be removed or repositioned if necessary. 5) Braun Hydrocephalus Fixed Pressure Valve System: This shunt has a fixed pressure range between 50 and 200 mmH_2O and is designed to provide a constant flow of cerebrospinal fluid. The valve is adjustable prior to implantation and can be removed or repositioned if necessary. 6) *The Medtronic PS Medical CSF-Flow Control valve* is a pressure differential valve built to open at a set pressure. In vivo, the pressure that the valve responds to is the difference between the intracranial pressure and the pressure of the space into which the distal end of the system is draining into (e.g., the intraperitoneal pressure).[22-24]

Programmable valve VP shunt: This type of shunt has a programmable valve that can be adjusted externally to change the pressure at which the CSF drains from the brain to the peritoneal cavity. Unlike a fixed-pressure VP shunt, a programmable VP shunt allows for adjustment of the pressure setting after implantation, without requiring surgery.[21, 23] These shunts are typically used in patients who require frequent adjustments to their CSF drainage rate. The programmable VP shunt consists of three main components: a catheter, a valve, and a programmable unit. The catheter is a flexible tube that is inserted into the ventricle of the brain to drain excess CSF. The

valve is located along the catheter and regulates the flow of CSF from the brain to the peritoneal cavity. The programmable unit is a small device located outside the body that allows the pressure setting of the valve to be adjusted as required. The programmable VP shunt valve has a small, battery-operated motor that controls the opening and closing of the valve. The motor is connected to a magnet that can be activated by an external programmer. The programmer is used by a trained healthcare provider to adjust the pressure setting of the valve. The programmer sends a signal to the motor, which moves the magnet to open or close the valve to achieve the desired pressure setting. The ability to adjust the pressure setting of the valve without surgery is a significant advantage of the programmable VP shunt. This allows the pressure to be tailored to the individual needs of the patient and can reduce the need for revision surgery. However, there are also some disadvantages to the programmable VP shunt. The device requires a battery, which may need to be replaced periodically. The device also carries a risk of malfunction or failure, which can lead to complications such as over-drainage or under-drainage of CSF. Some of the commercially available programmable VP shunts are: 1) *Medtronic Strata NSC*: This shunt has a programmable valve with a range of 36 to 200 mmH_2O. The valve is MRI compatible and can be programmed non-invasively using an external programming device. 2) *Codman Certas Plus*: This shunt has a programmable valve with a range of 30 to 200 mmH2O. The valve is MRI compatible and can be programmed non-invasively using an external programming device. 3) *Sophysa Polaris SPV*: This shunt has a programmable valve with a range of 20 to 200 mmH_2O. The valve is MRI compatible and can be programmed non-invasively using an external programming device. 4) *Integra Lifesciences Codman EDS III*: This shunt has a programmable valve with a range of 20 to 200 mmH_2O. The valve is MRI compatible and can be programmed non-invasively using an external programming device. 5) *Aesculap-Miethke ProGAV 2.0*: This shunt has a programmable valve with a range of 0 to 30 cmH_2O. The valve is MRI compatible and can be programmed non-invasively using an external programming device.

Anti-siphon VP shunt: This type of shunt has an anti-siphon device that prevents the siphoning effect of CSF drainage, which can cause a decrease in the pressure in the ventricles of the brain.[20, 22] These shunts are typically used in patients who have a tendency to over drain CSF, resulting in low-pressure headaches. An anti-siphon valve

is a specialized type of valve used in VP shunts for the treatment of hydrocephalus. The valve is designed to prevent over drainage of CSF when a patient is in an upright position. Over drainage can occur when the pressure in the shunt system is greater than the pressure in the brain, leading to complications such as headaches, vomiting, and seizures. The anti-siphon valve consists of a small chamber with a ball bearing that moves up and down in response to changes in pressure. When the patient is upright, the pressure in the shunt system decreases due to the effect of gravity. This causes the ball bearing to move down, which restricts the flow of CSF through the valve. This restriction helps to maintain a higher pressure in the brain, reducing the risk of over drainage. In addition to preventing over drainage, the anti-siphon valve can also improve the flow of CSF when the patient is lying down. When the patient is in a supine position, the pressure in the shunt system can increase due to the lack of gravity. This can cause the ball bearing to move up, increasing the flow of CSF through the valve. This increased flow helps to maintain a stable pressure in the brain. These shunts are typically recommended for patients who are at high risk for over drainage, such as those with normal-pressure hydrocephalus or patients who have undergone previous shunt surgeries. While the anti-siphon valve can help to prevent over drainage and improve the flow of CSF, it is important to note that it is not suitable for all patients with hydrocephalus. The choice of shunt should be made in consultation with a neurosurgeon and based on the individual needs of the patient. Some of the commercially available Anti-siphon VP shunts are: 1) *Medtronic Strata NSC and Strata II*: These shunts have an anti-siphon device called the SiphonGuard that is designed to prevent over drainage. 2) *Codman Hakim Precision and Codman Certas Plus*: The anti-siphon device is called the SiphonGuard MPV. 3) *Sophysa Polaris and Polaris II* with anti-siphon device called the SiphonGuard IVP. 4) *Aesculap-Miethke proSA and proGAV*: with anti-siphon device called the gravitational unit (GAV).

Gravity-assisted VP shunt: This type of shunt relies on gravity to allow the CSF to drain from the brain to the peritoneal cavity. These shunts are typically used in patients who have low CSF flow rates and require a higher pressure to drain the excess fluid. It is designed to improve the flow of CSF by taking advantage of the force of gravity.[25, 26] Unlike traditional VP shunts, which rely on a pressure valve to regulate the flow of CSF, the gravity-assisted shunt uses a simple design that allows CSF to flow freely from the brain to the abdomen. The gravity-assisted shunt consists of two

components: a long catheter that is inserted into the brain ventricles and a short catheter that is inserted into the abdominal cavity. The two catheters are connected by a flexible tube that allows CSF to flow freely from the brain to the abdomen. Unlike traditional VP shunts, the gravity-assisted shunt does not have a pressure valve. Instead, it relies on the force of gravity to regulate the flow of CSF. When the patient is upright, the force of gravity pulls the CSF downward, creating a pressure gradient that allows the CSF to flow freely through the shunt and into the abdominal cavity. When the patient is lying down, the pressure gradient decreases, which can lead to a temporary increase in the flow of CSF. However, this increase is usually short-lived, and the shunt quickly returns to its normal flow rate. Because the gravity-assisted shunt does not have a pressure valve, it is less likely to malfunction or become blocked compared to traditional VP shunts. It is also less expensive and easier to implant, as it does not require any specialized equipment or programming devices. However, the gravity-assisted shunt is not suitable for all patients with hydrocephalus. It is typically recommended for patients with normal-pressure hydrocephalus or those who are not at high risk for over drainage. One commercially available gravity-assisted shunt is the Delta valve from Codman Neuro, a Johnson & Johnson company. The Delta valve features a simple design with a long catheter that is inserted into the brain ventricles and a short catheter that is inserted into the abdominal cavity. The two catheters are connected by a flexible tube that allows CSF to flow freely from the brain to the abdomen, regulated by gravity. The Delta valve does not have a pressure valve, relying instead on the force of gravity to regulate the flow of CSF. It is typically used for the treatment of normal pressure hydrocephalus and is available in different configurations to meet the specific needs of individual patients.

Magnetic programmable VP shunt: This type of shunt uses a magnetic field to adjust the pressure of the valve, allowing for non-invasive adjustments. These shunts are typically used in patients who require frequent adjustments to their CSF drainage rate, but who are unable to tolerate external pressure adjustments due to skin sensitivity or other medical issues. It is designed to allow the flow of CSF to be adjusted non-invasively using a magnetic field.[27, 28] The shunt consists of two main components: the valve and the magnet. The valve is a small device that is inserted into the catheter of the VP shunt. It regulates the flow of CSF and can be adjusted using a magnet. The magnet is an external device that is used to change the setting of the valve. It is held

over the site of the valve and used to adjust the strength of the magnetic field. This changes the pressure setting of the valve, allowing the flow of CSF to be increased or decreased as needed. The advantage of the magnetic programmable VP shunt is that it allows the flow of CSF to be adjusted without the need for invasive procedures. This can reduce the risk of complications associated with surgery and allow for more precise control of the flow of CSF. The magnetic programming can be done in a doctor's office, and the procedure is typically quick and painless. The shunt can be adjusted to a variety of settings based on the individual needs of the patient. The magnetic programmable VP shunt is typically used for patients with normal-pressure hydrocephalus or those at high risk for over drainage. The shunt can also be used for patients who require frequent adjustments or who have difficulty with traditional VP shunts. While magnetic programmable shunts offer several advantages over traditional fixed-pressure or programmable VP shunts, they do have some disadvantages such as: 1) Magnetic programmable VP shunts are more expensive than traditional VP shunts. This can be a significant barrier to access for patients who do not have adequate insurance coverage. 2) The use of magnetic fields can interfere with the function of the shunt valve. This can result in unintended changes to the pressure setting of the shunt, which can lead to over drainage or underdrainage of cerebrospinal fluid. 3) While the range of pressure settings that can be programmed using a magnetic field is broader than that of fixed-pressure shunts, it is still limited. This means that some patients may not be able to achieve the optimal pressure setting with a magnetic programmable shunt. 4) The use of magnetic fields can pose safety concerns for patients who need to undergo MRI scans. While most magnetic programmable VP shunts are considered MRI safe, there is still a risk of complications and patients may require careful monitoring during MRI scans and 5) Magnetic programmable VP shunts require periodic adjustments to ensure that the pressure setting is optimal for the patient's needs. While this can be done non-invasively using a magnet, it still requires regular follow-up appointments with a neurosurgeon. Some commercially available magnetic programmable VP shunts are: 1) Codman Certas Plus, 2) Medtronic Strata II, 3) Sophysa Polaris, 4) Miethke ProGAV, 5) Aesculap Prosa, 6) Spiegelberg Magnetom, and 6) Integra RadiaFlow.

Low-profile VP shunt: This type of shunt has a smaller profile than traditional VP shunts, making it less visible and more comfortable for patients. These shunts are

typically used in paediatric patients and patients who require a lower flow rate of CSF.[27-29] It is designed to have a smaller profile than traditional VP shunts, making it less visible and more comfortable for the patient. The shunt consists of several components, including the catheter, the valve, and the connector. The catheter is a flexible tube that is inserted into the ventricles of the brain to drain excess CSF. The valve is a small device that regulates the flow of CSF and prevents over drainage or underdrainage. The connector is used to attach the catheter to the valve. Low-profile VP shunts are typically made from materials that are biocompatible and resistant to corrosion. They are available in a variety of configurations to suit the needs of different patients. Some low-profile VP shunts have a fixed pressure setting, while others are programmable. The main advantage of low-profile VP shunts is that they have a smaller profile than traditional VP shunts, which can make them more comfortable for the patient and less visible under the skin. This can improve patient satisfaction and reduce the risk of complications such as infections and dislodgement. Low-profile VP shunts are also available in a variety of lengths and diameters, which can allow for more precise placement and improved drainage of CSF. This can be particularly beneficial for patients with smaller body frames or those who require a shunt in a challenging location. Some of the potential disadvantages of these shunts are: 1) Limited availability, Low-profile VP shunts may not be available in all regions or countries. This can limit the options for patients who require this type of shunt. 2) Higher cost: Low-profile VP shunts may be more expensive than traditional VP shunts, which can be a barrier for some patients who do not have insurance coverage or who cannot afford the cost. 3) Limited pressure settings: Some low-profile VP shunts have a fixed pressure setting, which may not be appropriate for all patients. Patients who require a programmable or magnetic programmable shunt may not be able to use a low-profile shunt. 4) Increased risk of complications: While low-profile VP shunts may be less visible and more comfortable for the patient, they still carry a risk of complications such as infection, malfunction, and over drainage or underdrainage of cerebrospinal fluid. 5) Limited research: Low-profile VP shunts are a relatively new type of shunt, and there is limited research available on their long-term safety and efficacy. As such, the long-term risks and benefits of these shunts are not fully understood. Some examples of commercially available low-profile VP shunts include the Medtronic Strata II LP, the Codman Certas LP, and the Sophysa Polaris LP.

Ventriculoatrial (VA) shunt works by draining excess CSF from the ventricles of the brain into the right atrium of the heart, where it is then absorbed by the bloodstream.[30, 31] The VA shunt consists of several components, including a catheter, a valve, and a connector. The catheter is inserted into the ventricles of the brain and then tunnelled under the skin to the neck. From there, it is directed down to the chest and threaded through a vein to the right atrium of the heart. The valve is located along the length of the catheter and controls the flow of CSF, preventing over drainage or underdrainage. The connector is used to attach the catheter to the valve and secure it in place. VA shunts are typically recommended for patients who have difficulty tolerating a VP shunt or who have experienced complications with a VP shunt, such as peritonitis or bowel obstruction. However, VA shunts are less commonly used than VP shunts due to a higher risk of complications, such as infection and thrombosis. Major disadvantages of VA shunt are: 1) Higher risk of infection: Because the catheter of a VA shunt is threaded through a vein and into the heart, there is a higher risk of infection compared to other types of shunts. Infection can lead to serious complications and may require removal of the shunt. 2) Increased risk of thrombosis: The catheter of a VA shunt can also cause blood clots to form in the vein or the heart. This can lead to a blockage of the catheter or a pulmonary embolism, which is a potentially life-threatening condition. 3) Potential cardiac complications: VA shunts may cause cardiac complications such as arrhythmias or cardiac valve dysfunction. 4) Limited valve options: VA shunts typically use a fixed-pressure valve, which may not be appropriate for all patients. Patients who require a programmable or magnetic programmable valve may not be able to use a VA shunt. 5) Difficulty adjusting pressure: Adjusting the pressure settings of a VA shunt can be more challenging than adjusting the pressure settings of a VP shunt. This may lead to over drainage or underdrainage of cerebrospinal fluid. 6) Limited availability: VA shunts are less commonly used than VP shunts and may not be available in all regions or countries. This can limit the options for patients who require this type of shunt. Some examples of commercially available VA shunts are: 1) Medtronic Strata VA, 2) Codman Hakim VA, 3) Sophysa Polaris VA and 4) Integra Pudenz VA

Ventriculopleural (VPL) shunt works by draining excess CSF from the ventricles of the brain into the pleural cavity around the lungs, where it is then absorbed by the lymphatic system.[32-34] The VPL shunt consists of several components, including a

catheter, a valve, and a connector. The catheter is inserted into the ventricles of the brain and then tunnelled under the skin to the chest. From there, it is directed down to the pleural cavity around the lungs. The valve is located along the length of the catheter and controls the flow of CSF, preventing over drainage or underdrainage. The connector is used to attach the catheter to the valve and secure it in place. VPL shunts are typically recommended for patients who have difficulty tolerating a VP shunt or who have experienced complications with a VP shunt, such as peritonitis or bowel obstruction. However, VPL shunts are less commonly used than VP shunts due to a higher risk of complications, such as pneumothorax or haemothorax. Potential disadvantages and risks associated with VPL shunts include: 1) Risk of pneumothorax: VPL shunts have a higher risk of pneumothorax (air leaking into the pleural cavity around the lungs) compared to other types of shunts. This can cause difficulty breathing and may require additional medical intervention. 2) Risk of haemothorax: VPL shunts can also cause bleeding into the pleural cavity, a condition known as haemothorax. This can be a serious complication that may require surgery to address. 3) Risk of infection: As with all types of shunts, there is a risk of infection with VPL shunts. Infection can occur at the site of the incision, along the length of the catheter, or at the valve. Symptoms of shunt infection may include fever, headache, and changes in mental status. 4) Risk of over drainage or underdrainage: VPL shunts must be carefully adjusted to prevent over drainage or underdrainage of CSF. Over drainage can cause headaches, nausea, vomiting, and other symptoms, while underdrainage can lead to a build-up of CSF in the brain and potentially life-threatening complications. 5) Although not common, coughing CSF due to erosion of the pleural catheter into the bronchus creating a fistula is also reported. Some examples of commercially available VPL shunts are: Medtronic Strata VPL, Codman Hakim VPL, and Sophysa Polaris VPL.

Lumboperitoneal (LP) shunt drain CSF from the lumbar region of the spine into the peritoneal cavity. This type of shunt is typically used in cases where VP shunts are not suitable or have failed, such as in patients with blocked or scarred ventricles or those with previous abdominal surgery.[35, 36] The LP shunt consists of a catheter that is inserted into the lumbar subarachnoid space, usually at the L4-L5 or L5-S1 level, and a valve that controls the flow of CSF. The catheter is tunnelled under the skin and connected to a reservoir or valve that is implanted in the abdomen. From there, the

CSF drains passively into the peritoneal cavity, where it is absorbed and eliminated by the body. LP shunts catheter is inserted into the lumbar subarachnoid space under fluoroscopic guidance, and the valve or reservoir is placed in the abdomen. The catheter is then connected to the valve or reservoir. LP shunts have several potential advantages over VP shunts. Because the CSF is drained from the lumbar region rather than the brain's ventricles, there is a lower risk of complications such as infection, haemorrhage, or damage to brain tissue. LP shunts may also be easier to adjust or revise than VP shunts, as the catheter can be repositioned or replaced without accessing the brain. However, LP shunts also have some potential disadvantages. They may be associated with a higher risk of complications such as CSF leaks, infections, or mechanical failure compared to VP shunts. They may also be less effective at controlling intracranial pressure in some patients, particularly those with communicating hydrocephalus or other complex medical conditions. Additionally, LP shunt placement may be more technically challenging than VP shunt placement, and requires specialized expertise and training. Some examples of commercially available LP shunts are the Strata II valve (Medtronic), Codman Hakim Programmable valve (Integra LifeSciences), and Delta valve (Miethke).

Ventriculo-subgaleal shunt is a relatively uncommon type of cerebrospinal fluid (CSF) diversion surgery used to treat hydrocephalus. It is a simple surgical procedure that offers a passage between the dilated ventricle and the subgaleal pouch developed in the opposite side of the scalp through a small silicone tube as the conduit.[37, 38] Subgaleal shunts are typically reserved for patients who are not candidates for traditional shunting procedures due to medical comorbidities, previous surgical interventions, or other factors (post-infective hydrocephalus). This form of diversion of infected CSF into an avascular subgaleal pocket has not given rise to any increased rate of shunt infection as compared to VP shunt. Furthermore, this avoids iatrogenic infection risk associated with external ventricular drain (EVD) or the risk of developing porencephalic cysts associated with repeated anterior fontanelle ventricular taps and avoids the risk of infection with insertion of a needle into the ventricular access device (VAD). They may also be used in emergency situations, such as in patients with acute hydrocephalus who require immediate intervention. It is also most commonly used in treating neonates with germinal matrix haemorrhage, as these children have CSF with high RBC and protein content and also very low body weight, and are considered

unsuitable for VP shunt.[39, 40] It is presumed that in a recumbent child, raised intracranial pressure will force CSF flow from the ventricle to the tube and then to the avascular pocket from where it will be absorbed back through the walls of the pouch kept distended by the incoming CSF. The procedure to place a subgaleal shunt typically involves making a small incision in the scalp and inserting the catheter into the subgaleal space. The catheter is then tunnelled under the scalp and connected to an extracranial reservoir or valve, usually placed in the neck or chest. The reservoir or valve is typically buried under the skin to minimize the risk of infection or damage. One potential advantage of subgaleal shunts is that they may be associated with a lower risk of complications compared to traditional shunting procedures. Because the catheter is placed subcutaneously rather than directly into the brain or abdomen, there is a lower risk of infection, CSF leakage, or mechanical failure. Additionally, subgaleal shunts may be easier to adjust or remove than traditional shunts, as the catheter can be accessed relatively easily under the scalp. However, subgaleal shunts also have some potential disadvantages. They may be less effective at controlling intracranial pressure in some patients, particularly those with complex medical conditions or severe hydrocephalus. Additionally, they require specialized expertise and training to place and may be associated with unique complications such as skin erosion or displacement of the catheter. Subgaleal shunts are not commonly used in clinical practice, and there are currently no commercially available subgaleal shunt systems. The procedure to place a subgaleal shunt is typically performed using components from traditional shunt systems, including catheters, valves, and reservoirs, that are modified or adapted for subgaleal placement. As such, subgaleal shunts may be considered an off-label use of existing shunt components. Majority of these shunts may require replacement to other standard shunts like VP shunt after a preterm neonate gain adequate weight, scalp development and the protein/blood load in the CSF improves or removal after a period of time determined by the treating team of neonatologist, paediatrician or paediatric neurosurgeon.

Miscellaneous shunts: Ventriculoureteral, ventriculovesical, ventriculosternal, ventriculodiploic, ventriculohumeral, ventriculoiliac, ventriculomastoid, Vertebral and Ventriculo-gallbladder shunt[41, 42] are examples of some non-conventional shunts used for the clinical management of hydrocephalus. In general these non-conventional approaches are used when the classical approaches cannot be performed due to

patient specific technical/medical limitations.[43, 44] Aqueductal stents can be placed for aqueductal stenosis and trapped 4th ventricle requires shunting of 4th ventricle to peritoneum, pleura or atrium. Rarely the shunt is connected outside the body in the form of long-tunnelled external ventricular drain to drain proteinaceous CSF secondary to inoperable brain tumours. This defines the concept that CSF can be diverted to various organs in the body including bone. The treating physician and the team of surgeons lead by neurosurgeons must be aware of the pros and cons of the individual shunts, awareness of its indications & the rationale for its placement.

Ventricular access device (VAD) and External ventricular drain(EVD)

Ventricular access device is used to treat hydrocephalus both in paediatric and adults. This is often used instead of external ventricular drain(EVD).[45-47] This forms a safe conduit for CSF drainage when CSF diversion can be performed on a regular basis particularly avoiding repeated lumbar punctures or trans fontanelle tap in neonates.[45-47] VAD help to temporize situation with post haemorrhagic hydrocephalus to clear the blood load in the ventricles and to optimise the protein content in the CSF in neonates before they can have a VP shunt. This comprises of a standard ventricular catheter connected to an Ommaya reservoir that sits under the scalp to aid repeated CSF tap. This can be used to drain CSF in a continuous fashion until the underlying cause is treated or intermittently as and when required.[45, 47, 48] There is no increased risk of VAD over the external ventricular drain(ventricular catheter that is tunnelled outside the scalp to drain CSF) from our own institutional experience (RHCYP and RIE). EVD needs to be changed on a regular basis if it were to remain longer. The recommended duration of changing the catheter is 7-10 days. In exceptional circumstance the EVD can be used on a long term basis.[43, 44]

Variations in shunt valves (Pressure vs Flow regulated valves)

Shunts typically consist of a series of valves that regulate the flow of fluid between the brain and the abdomen. The valve plays a critical role in regulating the flow of CSF and maintaining the appropriate pressure within the shunt system. Pressure and flow regulated valves are the two primary types of valves used in shunts, each with its advantages and limitations.[25, 49, 50]

Pressure Regulated Valves: Pressure-regulated valves are designed to maintain a constant pressure differential between the brain and the abdomen. These valves have

a spring-loaded mechanism that opens or closes depending on the pressure difference.[25, 49-51] When the pressure in the brain exceeds a certain threshold, the valve opens and allows the fluid to drain into the abdomen. Once the pressure has equalized, the valve closes and prevents excessive drainage of CSF. Some key features of pressure regulated valves include: 1) Constant Pressure Regulation: Pressure regulated valves ensure a consistent CSF pressure within the shunt system, irrespective of patient posture or activity level. This helps prevent over drainage or underdrainage of CSF. 2) Passive Mechanism: Pressure regulated valves operate based on the principles of hydrostatic pressure. They do not require any external power source or complex mechanisms, simplifying their design and reducing the risk of malfunction. 3) Fixed Opening Pressure: Pressure regulated valves have a fixed opening pressure, which is determined during the manufacturing process. This value cannot be adjusted after implantation, making it crucial to choose the appropriate valve based on individual patient requirements. 4) Limited Control over Flow Rate: Pressure regulated valves offer limited control over the flow rate of CSF. They rely on the body's posture and hydrostatic pressure to regulate CSF drainage, which may not be sufficient in all circumstances.

Flow Regulated Valves: Flow-regulated valves are designed to maintain a constant flow rate of CSF, irrespective of the ICP. The valve opens when the flow of CSF exceeds the pre-set flow rate, allowing the excess fluid to drain.[25, 49-51] As the flow rate drops, the valve closes, reducing the drainage of CSF. Flow-regulated valves can be further classified into fixed or variable orifice valves. Fixed orifice valves have a fixed flow rate setting, while variable orifice valves have a variable flow rate setting, which can be externally adjusted using a magnet. Hence flow regulated valves, also referred to as adjustable valves or programmable valves, allow for active control of the CSF flow rate within the shunt system. These valves include an adjustable mechanism that can be externally programmed to achieve the desired flow rate. Some key features of flow regulated valves include: 1) Customizable Flow Rate: Flow regulated valves offer the advantage of customizable flow rates. This allows healthcare professionals to tailor the shunt system's performance to the specific needs of each patient, ensuring optimal drainage and pressure regulation. 2) Active Mechanism: Flow regulated valves utilize a complex mechanism that can be adjusted using external magnets or programming devices. This mechanism provides greater control over CSF drainage and can be fine-

tuned as needed. 3) Adaptability: Flow regulated valves are designed to adapt to the patient's changing needs. They can adjust the flow rate in response to variations in CSF pressure, ensuring appropriate drainage during different activities or postures. 4) Complex Design: Flow regulated valves are more complex than pressure regulated valves, incorporating additional components and mechanisms. This complexity can increase the risk of malfunctions, requiring regular monitoring and potential adjustments.

When considering pressure vs flow regulated valves for shunts, several factors should be taken into account: 1) Patient-Specific Needs: The choice between pressure and flow regulated valves depends on the patient's condition, age, activity level, and other individual factors. Flow regulated valves offer greater customization options, making them suitable for patients with complex requirements, while pressure regulated valves may suffice for others. 2) Risk of over drainage and Underdrainage: Pressure regulated valves tend to provide a more stable pressure within the shunt system, minimizing the risk of over drainage or underdrainage. Flow regulated valves allow for responding to changes in CSF dynamics, such as changes in posture, activity level, or intracranial compliance. While flow-regulated valves are easy to use, require minimal adjustments after implantation and can accommodate a wide range of flow rates, making them suitable for patients with varying CSF dynamics, they have a higher risk of over-drainage, which can lead to complications such as subdural hematomas, brain herniation, and slit ventricle syndrome.

Conclusions

In summary, the management of hydrocephalus relies heavily on the surgical placement of shunt systems, and selecting the appropriate shunt type is crucial for optimizing patient outcomes. This comprehensive overview provides insights into the various types of shunts available for clinical management, their indications, complications, and emerging advancements. A comprehensive understanding of shunt characteristics empowers clinicians to make informed decisions tailored to individual patients, ensuring effective CSF diversion and long-term management of hydrocephalus. Future research and technological advancements hold the potential for further improving shunt therapies and enhancing the quality of life for patients with hydrocephalus.

Declaration of interest statement: none

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Figure legends

Figure 1: The factors reported to be responsible for development of hydrocephalus.

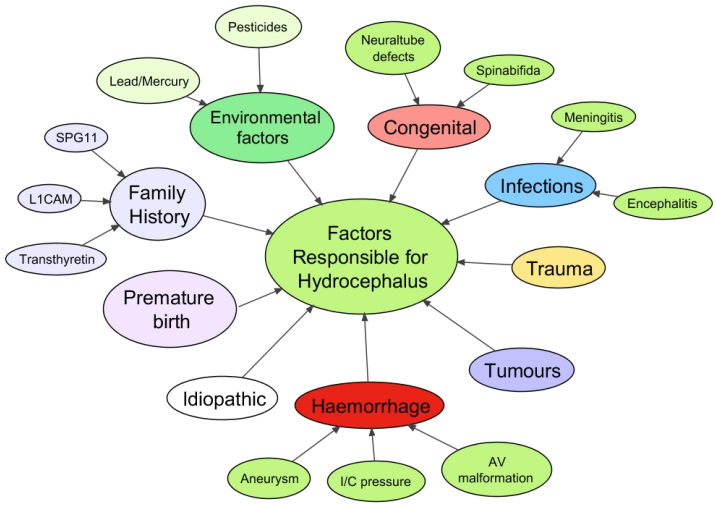

Figure 2: The various approaches to clinical management of hydrocephalus.

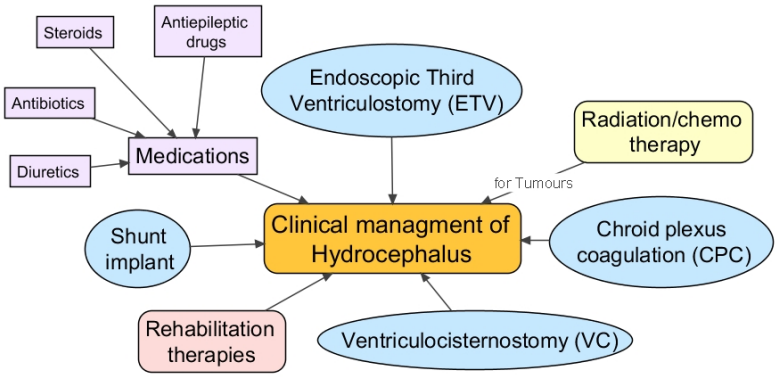

Figure 3: Various types of shunts used in the clinical management of hydrocephalus.