The treatment of hydrocephalus is a challenging one. The development of shunt devices have greatly improved the survival and quality of life of paediatric patients with hydrocephalus; however, shunt dysfunction is a common problem which represents a significant scope of work for paediatric neurosurgeons with shunt failures occurring in up to 40 to 50% of patients during the first two years after shunt surgery.

Numerous pathologies ranging from congenital to acquired conditions can result in the development of hydrocephalus in the paediatric population. Obstruction of proximal or distal catheter ends, misplacement, infections and over drainage are some of the common problems accounting for shunt failures.

We discussed some of the pertinent problems and nuances involved in treatment of paediatric hydrocephalus with VPS as well as to review the role of endoscopic procedures as an alternative to VPS.

Keywords: Hydrocephalus, Ventriculo-peritoneal shunt, Ventriculomegaly

Introduction

The development of shunts for treatment of hydrocephalus has greatly improved the clinical outcome of neurosurgical patients over the past few decades. This has been made possible through technological advancement as well as the development of new shunt devices and materials through a closer understanding of the pathophysiology of hydrocephalus and complications of shunt insertion. However, in this modern neurosurgical era with years of advancement and development of neurosurgical techniques, instruments and devices; we are still humbled by the fact that many shunt related complications remain a persistent problem in management of patients with paediatric hydrocephalus. Numerous pathologies ranging from congenital to acquired conditions can result in the development of hydrocephalus in the paediatric population. This clinical condition which appears to be easily solved through a simple procedure of creating an alternative pathway of drainage by insertion of a ventriculo-peritoneal shunt (VPS) is often fraught with a multitude of complications arising from the time of insertion to long term problems which may develop many years after the surgical procedure.

It has been reported that shunt failures occur in up to 40 to 50% of patients during the first two years after shunt surgery. (1) Obstruction of proximal or distal catheter ends, misplacement, infections and over drainage are some of the common problems accounting for shunt failures. There are several adjunctive measures used to improve the accuracy of shunt placement ranging from trajectory of insertion, entry point for insertion, ideal location and placement of shunt tip. The use of various types of shunt valve designs as well as the use of antibiotic impregnated shunts are some of the current measures used to reduce shunt infection and shunt failures.

This review article serves to discuss some of the pertinent problems and nuances involved in treatment of paediatric hydrocephalus with VPS as well as to review the role of endoscopic procedures as an alternative to VPS.
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Types of CSF Shunts

Overdrainage of shunts can result in the development of extra-axial fluid collections, subdural hematomas or cause the problem of slit ventricles. The change in design of shunt valves with programmable functions and addition of anti-siphon devices have been advocated to minimise or prevent these complications. The development of shunt valves began with the simple ball-in-spring, miter, slit and diaphragm valves. These standard shunt valves control CSF flow unidirectionally by opening at a particular pressure differential and allow for a reduced resistance to CSF flow. Gravity on CSF from a vertical posture leads to an increase in flow of CSF within the shunt tubing downwards and creates a siphoning effect which ultimately results in the problem of over drainage. Standard valves were subsequently augmented with anti-siphon devices to deal with the problems of over drainage however; there has been no randomised trials to validate the perceived benefits in an in-vivo setting.

Further technological advancements lead to modifications in shunt valve design in an attempt to reduce shunt failure rates from over drainage. The development of horizontal-vertical valves, flow controlled valves and more recently various programmable valves were designed to adapt to physiological flow of CSF. Programmable valves also allow adjustments to resistance of flow in order to tailor to variations in different patients where physiological changes may result in changes of CSF drainage rates. The benefit of such valves were tested in two major prospective randomised trials. Pollack et. al, investigated the programmable Medos valve (Codman, Johnson & Johnson) with other valve designs and results also showed no difference in failure rates. (2) An investigation was also carried out comparing the Delta valve (Medtronic PS Medical) which contains a siphon control component and the Orbis-Sigma (Cordis) valve with a variable resistance flow-limiting component as compared with a standard differential-pressure valve. (1) The results did not reveal any significant difference in shunt failure rates which included shunt obstruction, over drainage, loculated ventricles and infection. A further post hoc analysis was performed in the above mentioned trial to rule out the possibility of catheter position as an influencing factor of shunt failure. (3) This further revealed that ventricular volume decreased in an exponential fashion, forming a plateau at 14 months and was not affected by the different shunt valve designs. Hence implying the possibility that the physiological and mechanical principles applied to the design of these valves were inaccurate.

Further recent retrospective reviews carried out between programmable valves and standard valves have also shown varying results. A review of 253 patients with insertion of programmable shunts in about 30% of patients showed no statistical significance in the ability of programmable valves to reduce shunt failure rates (4). Another retrospective review comparing 100 patients with programmable shunts versus 89 patients with non-programmable shunts showed programmable valve malfunction of 11.1% per year while non-programmable shunts had no malfunctions for the same period for patients with similar causes of hydrocephalus (5). In contrast, another review of 279 cases comprising implantation of programmable valves in 27% of patients versus set pressure valves showed a statistically significant reduction in risk of proximal shunt obstruction and shunt revision in patients with programmable valves. (6)

The mixed findings in these studies lead us to question the validity of usage of the more expensive and sophisticated valves which may ultimately serve no significant benefit to our patients in reduction of complications. Alternatively, our failure to fully appreciate the pathophysiology and dynamics of hydrocephalus in various disease conditions as well as the variability in confounding factors from these current studies may prevent us from identifying certain select groups of patients where there is a potential benefit of utilising programmable shunt devices.

One argument for the usage of programmable valves applies to patients with over drainage after shunting and development of
subdural collections. These patients may benefit from a programmable valve to reduce CSF drainage in order to facilitate brain expansion after evacuation of the subdural collections. Patients with bony craniotomy defects and subsequent development of post-traumatic hydrocephalus may also benefit from the ability to perform adjustment in valve resistance to temporarily manage the problem of a severely sunken skin flap by increasing valve resistance prior to cranioplasty.

Shunt Infections

Shunt infection in hydrocephalus is a serious complication with the potential to be life-threatening. Current reported infection rates in large trials are approximately 8-10% during the first few months after shunt insertion (7). A recent study reported infections developing in greater than 11% of children who underwent uncomplicated initial CSF shunt placements within 24 months (8). A multitude of factors have been attributed to the development of shunt infections and optimization of operating theatre and shunt procedures (9), double gloving (10) and the use of systemic prophylactic antibiotics (11) have been shown to reduce the incidence of infections.

In recent years, antibiotic-impregnated shunts (AIS) with rifampicin and clindamycin have been developed in an attempt to reduce rate of shunt infection in the critical early post-operative period when most shunt infections occur. Several major studies have been conducted to determine the benefits of AIS. A prospective study on adult and pediatric patients evaluating 243 shunt procedures in 178 patients and retrospective comparison with preceding shunt procedures showed a statistically significant reduction in infection rates (12). Parker et al carried out a retrospective review of patients shunted over a ten year period comprising insertion of 502 AIS shunts vs 570 non-AIS shunts (13). A reduction in infection was found in patients with AIS shunts with the most significant reductions in high risk patients such as premature neonates, post-meningitis patients and patients with a prolonged hospital stay. Another retrospective study on paediatric shunt procedures found that AIS had a significant impact on infection rates in the neonatal hydrocephalic population (14). In an analysis of patients in the UK Shunt Registry, a search was carried out to match 994 procedures in which AIS was used as compared to a similar number of controls. The results suggested AIS to have the potential to significantly reduce shunt infections (15).

Cost-benefit ratio analysis on cost of AIS was analysed where additional cost of AIS was balanced with economic consequences for additional surgeries and prolonged hospital stay from clinical and economic perspectives. This study showed AIS to have a potential as a valuable addition in hydrocephalus treatment (16). In comparison, a review of 160 shunt procedures with 80 patients in each group did not show any significant reduction in the pediatric CSF shunt infection rate with the use of AIS (17). Similar conclusions were also reported in another observational study involving 258 patients (18).

The use of silver impregnated catheters containing silver nano-particles and an insoluble silver salt have also been introduced in clinical practice to reduce the risk of infections. In-vitro and early clinical studies have shown a potential benefit in the ability of these shunts in reducing as well as eradicating the presence of infections. An in-vitro study to analyze the antimicrobial activities of two antimicrobial-agent-impregnated ventricular catheters and to compare their efficacies on the bacterial cultures showed that the antibiotic-impregnated catheters had greater inhibition zones when tested on Staphylococcus aureus, Staphylococcus epidermidis, and Pseudomonas aeruginosa while the silver-impregnated catheters showed the lowest bacterial colonization in agar (19). Another in-vitro study showed that the commercial silver-impregnated catheter was not able to eradicate MRSA or E. coli while it showed activity against S. epidermidis (20).

In the clinical setting, a study carrying out retrospective analysis on silver-impregnated external ventricular drains showed a decrease in incidence of positive cultures and bacterial colonization of silver catheters when compared to conventional catheters (21). The clinical efficacy regarding use of silver-impregnated polyurethane ventricular catheter for shunting of
CSF in patients with infected hydrocephalus was recently investigated in a small study. Seven patients who had hydrocephalus with high protein level and positive CSF culture and prior shunt failure from infection underwent implantation of VPS with silver-impregnated polyurethane ventricular catheter. Follow-up was carried out for 14 months and preliminary results suggested a complete improvement of infection (22).

These current studies performed have been based on prospective or retrospective analysis in comparison with historical control groups. These results though promising are conflicting and cannot confirm the benefit of AIS or silver-impregnated catheters. Further prospective randomized control trials will need to be carried out in order to determine its real benefit if any.

**Accuracy in placement of Ventricular Catheters**

One of the most common causes of shunt failure is malpositioning of the ventricular catheter which leads to obstruction at the proximal intra-ventricular segment. Free-hand techniques of insertion can be prone to error and we have undoubtedly seen alarming locations of catheters on follow-up imaging. The position of the ventricular shunt tip is one of the important factors in determining long term shunt patency. Long term patency of a the proximal ventricular catheter tip has been shown to be dependent on the positioning of the hole-bearing segment of the catheter. A properly functioning shunt over the long term must have a patent catheter with all the hole bearing segments within the CSF fluid space. Occlusion over the hole bearing segment of the proximal catheter may develop due to its proximity to adjacent choroid plexus, ependymal lining of ventricles or damaged ependymal surfaces from multiple attempts at ventricular catheterisation. It is critical that during ventricular insertion, minimal ependymal damage as well as accurate placement of the proximal catheter in the most ideal position is needed to minimise the rate of occurrence of occlusion.

The placement of a ventricular catheter has been based on the time honoured art of appreciation of known anatomical landmarks to decide on the trajectory of placement. Experienced neurosurgeons through years of experience have an almost “stereotactic” vision which enables them to target a ventricular catheter accommodating for anatomical variations in different head shapes with fairly good accuracy. Pang et al (23), described a free-hand insertion technique of ventricular catheters based on simple stereotactic coordinates based on visible and palpable surface anatomy with an insertion trajectory aligned to the coronal obliquity of the lateral ventricle. Their review of 160 patients showed an obstruction rate of 1.9%. Radiographic evaluation in 112 patients showed they achieved satisfactory placement of catheters in over 90% of patients.

Surface anatomical landmarks have been the traditional method used to get a sense of spatial orientation in order to target a particular location within the ventricles. This is unfortunately not consistently accurate and the trajectory and depth of placement is subject to significant variation depending on the entry points as well as surgical experience. The common entry points for ventricular shunt placement into the lateral ventricles include the frontal, parietal and occipital burr holes. Lind et al, performed a morphometric study and geometrically analysed the possible approaches based on reconstructed MRI scans in patients with normal and pathologically dilated ventricles (24). They found that the range of possible angles for successful catheter insertion was smallest for the occipital approach and greatest for the frontal approach. It was also found that there was no single landmark for aiming occipital or parietal catheters that achieved accurate placement in every case. A more recent retrospective review of freehand initial shunt surgery with post-operative radiological imaging showed parietal and frontal catheters are more likely to be placed successfully in the target ventricle (25).

In view of the increase in risk of misplacement of catheters from posterior approaches, a virtual reality simulation study was carried out to study the insertion of ventricular catheters from Dandy’s point (3cm above and 2cm lateral to the inion) and Frazier’s point (6cm ? and 4 cm lateral to the inion)
based on imaging data from ten patients. Satisfactory placement of catheter tip was achieved using target points of 4cm above the contralateral medial canthus for Frazier’s point and target points of 2cm above the glabella for Dandy’s point (26). The shortening a ventricular shunt catheter associated with growth of the cranium can also account for displacement of catheter tip which affects long patency of the shunt. One study found the insertion of a ventricular catheter via the frontal route in children resulted in a higher incidence of shortening due to greater displacement of the burr hole adjacent to the coronal suture, hence arguing that the parietal route is recommended in order to maintain long-term shunt function (27).

The findings from the above studies and the great variability and potential risk of error from identification of critical landmarks intra-operatively suggests that patient specific stereotaxy rather than the use of anatomical landmarks will probably allow a more reliable first-pass to achieve accurate placement of catheters and this is especially so for occipital entry points.

**Use of Navigational Systems**

The presence of small ventricles in neonates and young paediatric patients or patients with slit ventricles make free-hand insertion of ventricular catheters a formidable challenge. The use of navigational systems to assist in ventricular cannulation are becoming increasingly widespread. A major benefit of navigation in comparison to traditional methods of insertion is in cases where ventricles are small or there is the presence of distortion of the ventricular system. Frameless neuronavigation has been used by various authors who report it to be a safe and beneficial option for optimal positioning of ventricular catheters (28, 29). Image guidance utilizing CT or MRI images allows a three dimensional reconstruction of the ventricular system and real time visualisation based on imaging using pre-operative images during catheter insertion in tri-planar views.

Pre-operative planning can be carried out to define the exact target points thereby allowing the most ideal trajectory, entry point and length of catheter to be selected and tailored for each individual patient. The ventricular tip can be accurately placed in free cerebrospinal fluid space away from the choroid plexus or too close to ependymal surfaces. Trajectory planning also ensures that the selected trajectory avoids any vascular structures during insertion which may result in unnecessary complications. In addition, the use of navigation also means that the entry point of insertion is flexible and is not reliant on the standard anatomical entry points; this may be come useful in cases where there is a need to avoid the re-use of a prior scalp incision or burr hole and in the absence of the cranium after craniectomy.

One major disadvantage regarding the use of navigational systems is longer set-up and operative time when compared to free-hand insertion techniques. In addition, the accuracy of utilising navigation is assuming the radiological images used for localisation are close to the actual situation during surgery. This requires that pre-operative scans are performed as close to the surgical date as possible as time may result in evolution of size and shape of ventricles causing further antomical distortion. During surgical insertion, any excess loss of CSF can also result in brain shift and change in ventricular anatomy; hence rendering navigation inaccurate.

Other disadvantages include the need for rigid head fixation in order to ensure accuracy in navigation and this has its limitations in neonates and younger paediatric patients. This limitation has however been over come with the development of newer electromagnetic strap-on devices to the head which allow for reasonable accuracy. Such devices may however be cumbersome and may not be ideal as they can interfere with the surgical process of shunt placement. The cost to develop facilities for stereotactic navigation is easily affordable by many major neurosurgical institutions worldwide; however the lack of financial resources in many developing nations where the problem of hydrocephalus in many children limits its application utility.

**Ultrasound Guidance**

Intra-operative ultrasound is an alternative
tool to stereotactic navigation which can be used to identify the target location and determine the best location for the burr hole based on trajectory and distance to the target point. Real-time ultrasound was found to be a feasible, safe and effective technique to aid in the insertion and accurate placement of ventricular shunts (30). This method utilises a small footprint ultrasound probe to provide continuous real-time monitoring while the catheter is being inserted hence allows for insertion with a single pass. The only disadvantage to this elegant procedure however is the need to enlarge the burr hole up to 2cm in diameter in order to accommodate the ultrasound probe.

**Use of Robotics**

Image guided robotic targeting of a specific location is a potential tool to allow accurate placement of ventricular catheters. Such robotic systems have been utilized for safe and accurate placement of catheters to an accuracy of within 1.5mm from a targeted point with a single pass (31). The use of robotics has the advantage of a high degree of precision and accuracy in catheter placement and careful pre-operative planning can help minimise the risk of injury to vascular structures and the ability to select the most ideal trajectory. The major disadvantages of such a setup is no doubt the cost of such a system as well as longer operative times involved in the peri-operative set-up. The field of robotic surgery is in its infancy and further advances in technology and micro engineering holds the promise of further refinements in allowing better feasibility of robotics for these procedures.

**Endoscopic Procedures**

There have been several applications of endoscopic procedures used to assist in the implantation of VPS. One major advancement is the use of endoscopic techniques specifically for the use of endoscopic third ventriculostomy (ETV) as an alternative to treat paediatric hydrocephalus.

ETV is a procedure involving perforating the floor of the third ventricle between the infundibular recess and the mamillary bodies. The use of ETV as a first line procedure in management of conditions resulting in obstructive hydrocephalus is gaining favour as it eliminates the need for shunt insertion and its complications. Relative contraindications to ETV include patients with infections resulting in basal arachnoiditis, patients with a small pre-pontine cistern, small ventricles and communicating hydrocephalus. The increase in number of cases performed and the surgical experience gained has allowed many neurosurgeons to push the limits and successfully perform the procedure.

ETV is however not without its complications. A serious complication is that of rapid deterioration and sudden death from closure of the ventriculostomy and rapid development of hydrocephalus (32). Other potential complications include closure of stoma, CSF leak, meningitis, hypothlamic injury, vascular injury and injury to the optic chiasma. The failure rate of ETV has been reported to be as high as 13.6% based on the Canadian ETV series (33). However, in spite of these factors, there is a school of thought that avoidance of shunt placement in view of the cumulative life time risk of complications is worth attempting ETV as a first line.

Endoscopes have also been utilised to assist in placement of both proximal and distal catheters in VPS. Miniature scopes passed through the lumen of the catheter have been performed in order to allow direct visualisation while inserting proximal catheters to achieve optimal positioning. The multicenter randomised Endoscopic Shunt Insertion Trial unfortunately did not reveal any significant benefit when compared to free-hand insertion techniques (34). Patients with distal shunt problems such as recurrent obstruction or pseudo cyst formation secondary to loculations from dense peritoneal adhesions have also benefitted from the use of direct visualisation to break down adhesions or to guide the distal peritoneal catheter into a suitable region of the abdominal cavity to allow adequate reabsorption of CSF.

The improvement in endoscopic instruments and advancement in optical technology have rapidly progressed and is now being used for endoscopic resection of intraventricular as well
as intra-axial tumours causing obstructive hydrocephalus. Endoscopic procedures such as fenestration of arachnoid cysts, tumour cysts and loculated ventricles to relieve the problem of hydrocephalus are other examples of treatment options as an alternative to VPS. The use of endoscopic biopsy procedures for third ventricular, pineal region or midbrain tumours can also be coupled with ETV to manage the problem of obstructive hydrocephalus which minimises the need for multiple surgical procedures.

**Conclusion**

CSF shunt failure is a time-related event. We have made significant progress in the treatment of paediatric hydrocephalus over the decades. However, much still needs to be investigated and learned about CSF flow dynamics, absorption and how this interacts with ventricular compliance in various pathophysiological conditions in the development of hydrocephalus. This will hopefully allow for the further development of newer biomaterials, sophisticated valve control systems which are dynamic in regulation of CSF flow to better manage the life long problem of hydrocephalus in the paediatric population. The need for well designed prospective randomised trials investigating various treatment modalities will be crucial in order to validate the benefits of new technology and treatment techniques.

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