4 Introduction: The Use of Extrathecal CSF Shunts, Optional vs Mandatory, Unavoidable Complications

Rizwan A. Khan

Contents

R.A. Khan, MD, MS, MCh
Department of Pediatric Surgery, JNMC, AMU, Aligarh, India
e-mail: drrizwanahmadkhan@yahoo.co.in

4.1 Types of Extrathecal Shunts

Since 1895 when Gartner proposed shunting the CSF into extrathecal low-pressure compartments such as the venous or lymphatic system or the abdominal cavity, the treatment of hydrocephalus has come a long way [1]. The idea was first applied by Fergusson in 1898 and later was taken up by several authors, including Cushing, but the results were mostly disappointing. The modern shunting era began with Nulsen and Spitz, creating a one-way pressure-regulated valve which they placed in the atrium via the jugular vein (Figs. 4.1 and 4.2). There are various types of extrathecal shunts, some of which are only of historical importance. Extrathecal shunts still in vogue are lumboperitoneal, ventriculovenous, ventriculoatrial, ventriculopleural, ventriculoureteric, ventriculo-gallbladder, and ventriculoperitoneal shunts [2, 3]:

1. Lumboperitoneal (LP) shunts are used in patients with communicating hydrocephalus or small slit ventricles (due to overdrainage caused by other extrathecal shunts like ventriculoperitoneal and ventriculoatrial shunts) and patients who have had multiple ventricular shunt malfunctions. These shunts have the advantage of avoiding injury by the ventricular catheter, maintaining the patency of ventricles (unlike ventricular shunts where the ventricles tend to collapse leading to shunt malfunction), and have lower risk of obstruction and infection. However, LP shunts are more prone to malfunction from mechanical

C. Di Rocco et al. (eds.), *Complications of CSF Shunting in Hydrocephalus: Prevention, Identification, and Management*, DOI 10.1007/978-3-319-09961-3_4,

© Springer International Publishing Switzerland 2015

failures like migration out of the spine or peritoneum and carry a definite risk of development of hindbrain herniation over a period of time especially in children. The assessment of function of a lumbar shunt is more difficult as compared to ventricular shunt [4].

2. Ventriculoatrial (VA) shunts are provided to divert cerebrospinal fluid from the cerebral ventricle into the right atrium. These are not chosen as the first-line method to redirect the CSF rather when there have been multiple failure of the VP shunt (Table 4.1). The intraoperative appropriate vein selection and exact shunt placement are important to reduce complications such as obstruction [5].

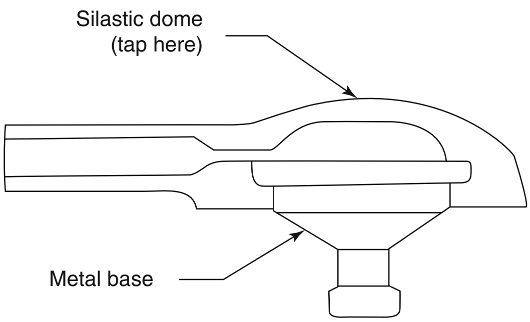

Fig. 4.1 Conventional ventriculoperitoneal shunt

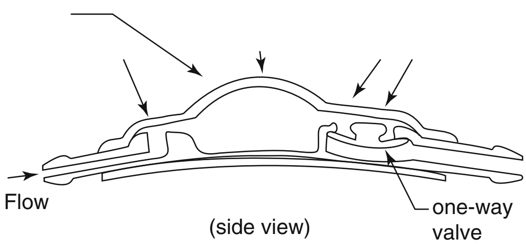

Fig. 4.2 Ventriculoperitoneal shunt with antisiphon device

3. Ventriculopleural shunts were introduced as a management option for hydrocephalus by Ransohoff in 1954. Ventriculopleural shunts have been used infrequently in the management of hydrocephalus and have become the "next preferred procedure" in case if the ventriculoperitoneal shunt fails. It has low complication rates and is easy to perform, but involvement and collaboration of the thoracic surgery team is required. The risk of pleural effusion is highest among the infants which seemed to be reduced with the introduction of antisiphon device [6].
4. Ventriculoureteric shunt (VUS) is rarely used nowadays in neurosurgical practice owing to much better techniques that are available. It is technically demanding – exposure of the renal pelvis and ureter requires the help of an urologist. As previously thought, it no more requires nephrectomy and reimplantation of the ureter, but to prevent expulsion, flanged at the distal catheter must be used. Hyponatremia is a known metabolic complication immediately following the VUS [7].
5. Ventriculo-gallbladder shunts (VGS) are used because of the ability of the biliary tree to adequately control intracranial pressure, low risk of infection due to the sterility of bile, and also the prospect for electrolyte reabsorption from the small intestine (Table 4.1). But it is not the first line of treatment. Indications described are multiple shunt revisions, abdominal pseudocysts, and ascites secondary to shunts in optic chiasmal hypothalamic astrocytomas. Bile calculus causing a distal

Table 4.1 Showing features of the extrathecal shunts

Type of extrathecal shunt	Drainage into	Practicality	Common complication
Ventriculoperitoneal	Peritoneal cavity	Most commonly performed	Mechanical, fracture, infection
Ventriculopleural	Pleural cavity	Second most commonly performed procedure	Pleural effusion
Ventriculoatrial	Right atrium	Multiple failure of VP shunt	Obstruction, embolic episodes
Ventriculoureteric	Ureter/renal pelvis	Rarely used	Hyponatremia
Ventriculo-gallbladder	Gallbladder	Multiple failures	Bile calculus with distal obstruction
Lumboperitoneal	Lumbar space to peritoneum	Overdrainage syndrome due to other extrathecal shunts	Mechanical complications, hindbrain herniation

obstruction in the VG shunt catheter is a frequently reported complication. There are also chances of cholecystitis causing retrograde and descending shunt infections [8].

6. It is widely accepted that ventriculoperitoneal shunts represent the most common extracranial CSF diversion choice, given that the peritoneal cavity is the most efficient and reliable location for CSF absorption. The peritoneal cavity is the preferred drainage site in children because it enables the implantation of drainage catheter of sufficient length to allow for the growth and predisposes to less severe complications than does the right atrium and ease of shunt revision. The first ventriculoperitoneal shunt is probably attributed to Kaush. In 1908 he reported a patient in whom he connected a lateral ventricle to the peritoneum using a rubber tube, but the patient died because of overdrainage.

4.2 Indications

It is easy to identify patients who would benefit from CSF diversion if the presentation includes clear evidence of increased intracranial pressure (ICP), manifested as severe headaches with projectile vomiting, diplopia, upward gaze paresis, and a dilated ventricular system on radiologic imaging [2, 9] (Table 4.1).

1. In adults with occlusive hydrocephalus with raised ICP, CSF diversion is indicated to prevent permanent neurologic deficits or neurologic deficits progress. To supplement clinical findings, radiological evidence of ventricular enlargement may be assessed with Evan's index which is a ratio of greatest width of the frontal horns of the lateral ventricles to the maximal internal diameter of the skull. An index exceeding 0.3 is indicative of a hydrocephalus. T2-weighted MR images can show transependymal flow of CSF and subependymal white matter damage.
2. Assessing patients of NPH for CSF diversion is difficult. Contrary to its name, there are periodical fluctuations of intracranial pressure in the early stages of the disease. Presence of this intermittent elevation of ICP provides a justification for treatment by shunting, which cuts off the peaks of increased ICP damaging the brain. On radiology widened temporal horns and flattened cortical sulci at the top of the brain are also found in NPH. A twenty-four-h ICP monitoring showing B-waves is regarded to be a good indicator of NPH likely to benefit from shunting. Patients with NPH who are considered for shunting should have gait disturbances and at least one of the two other elements of the triad: disturbances of urination and cognition. Patterns of concentration of biomarkers like NFL (neurofilament protein light) may also help in reaching the diagnosis [10].
3. The most important point to be considered in decision making in premature infants is the rapid head growth with bulging and tense anterior fontanelle. There may be prominent scalp vein and episodes of apnea and bradycardia. Term infants may also show irritability, vomiting, drowsiness, axial hypotonia, and setting-sun sign. Retinal hemorrhages may be present, but papilledema is uncommon. Older children may show headaches, projectile vomiting, diplopia (abducens nerve palsy), blurred vision, loss of visual acuity, and papilledema. Transfontanellar craniosonography may help in showing the progress of ventricular enlargement, and CT and MRI can be used to assess the severity by measuring Evan's ratio or by the presence of transependymal CSF absorption [11]. The most difficult is the evaluation of a child with macrocephaly and normal development. Computed tomography scanning shows increased extra-axial collections, with a normal or mildly enlarged ventricle. This entity, sometimes referred to as benign extra-axial collections of infancy, may be a stage of transient communicating hydrocephalus that may resolve in a period of 12–18 months with a normal outcome. It is a clinical and not an imaging diagnosis, and it must be distinguished from true communicating hydrocephalus. If the child is

developing normally, close observation with neurodevelopmental examination and head circumference measurements are adequate. If the child is neurologically abnormal, then a radionuclide cisternogram plays a role in evaluating the contribution of abnormal fluid dynamics. It has been used to identify patients with communicating hydrocephalus. A high lumbar pressure, poor 4-h urinary excretion (normal 50 %, borderline 30–40 %, definitely abnormal <30 %), persistence of ventricular filling at 24 h, and poor flow over convexity are highly suggestive of communicating hydrocephalus and an indication for CSF diversion [12].

4.3 Unavoidable Complications

Shunt system is a foreign body placed for relieving the intracranial pressure and will have some complications inherent to its very presence inside the body. There are many complications associated with the shunt like infection, obstruction, overdrainage, underdrainage, loculation, etc. These complications can be avoided or best minimized with proper precautions and use of appropriate shunting device. But there are certain complications that are going to be present, and the neurosurgeon has no control over these complications.

4.3.1 Complications Due to Material

Holter developed the first shunt to employ silicone with multiple slit valve design. Medical grade silicone (dimethyl polysiloxane), the indispensible material for the shunt systems of current generation, meets all the criteria of biocompatibility, nonimmunogenic, noncarcinogenic, fatigue-free (which could withstand the severe, long-term mechanical stress they were subject to particularly in the right atrium), thermally resistant (ideal for heat sterilization), and electrical stability. Although chemically inert, silicones are not necessarily biologically inert, and there are several evidences of silicone allergy in some patients with silicone shunts. At the time of insertion of a ventricular catheter, the blood–brain barrier is not healed for 2–3 weeks. This leads to adherence of the platelets and serum proteins including immunoglobulins, albumin, fibronectin, and fibrinogen which in turn potentiate the inflammatory response and also adhere to silicone rubber and alter the function of more distal shunt components (e.g., the valve mechanism). In the peritoneal cavity, shunt catheters can be obstructed by ingrowth of mesothelial cells and fibroblasts, and adsorption of protein is associated with greater adhesion of inflammatory cells.

The body's reaction to silicone can be classified into three types: local reaction and granuloma formation, silicone migration, and autoimmune disease. In all cases following the implantation of silicone, there is the development of a chronic type of reaction to the implant, which includes the formation of a fibrous tissue pseudocapsule with minimal inflammatory reaction. This is best represented by the very familiar shunt tract. Another form of reaction that has been recognized is the formation of granulomas, focal collections of macrophages, histiocytes, epithelioid cells, giant cells, lymphocytes, and plasma cells often termed "siliconomas." These are seen along the tract in patient with multiple revisions. Silicone allergy is known to affect an implant in a patient by the phenomenon of migration. This spread can occur by lymphatic, hematogenous, and local route. This is seen more commonly in patients with breast implants and cardiac valve prosthesis, in those undergoing cardiopulmonary bypass, and in patients on frequent hemodialysis. Autoimmune disease can occur following implantation of silicone polymers. This is also seen more commonly in women who have undergone injection or implantation of silicone for breast augmentation. Symptoms include arthritis, arthralgia, and local and regional lymphadenopathy.

In patients with suspected or documented silicone allergy, the use of polyurethane or CO2-extracted silicone catheters has been postulated but not proven to offer some advantage in reducing the risk of recurrent malfunctions [13, 14].

4.3.2 Complications Due to Design

A physical design modification, i.e., addition of soft radial flanges to push the catheter away from the brain tissue and protect the catheter lumen from the problem of tissue ingrowth, has not been successful. It has since become clear that the simple addition of flanges does not prevent tissue ingrowth, and in fact some neurosurgeons report that tissue ingrowths on flanged catheters are even more tenacious than those associated with standard design catheters.

The technological advancements made to valve systems (like the staircase mechanism of the Codman Medos valve) have provided additional CSF pathways that are turbulent and susceptible to areas of buildup of debris that may be a reason for the higher failure rate. Diaphragm-based antisiphon devices are prone to obstruction from encapsulation. There is no evidence that using an open system has any advantage over using a closed system, but theoretically any malfunction in an open system would result only in loss of antisiphon function, without obstruction to CSF flow as against the closed system, in which the flow stops once the pressure reaches zero. In flow control devices, patients develop nighttime or early morning headaches. This is because these patients have limited pressure volume compensatory reserve and there can be an excessive increase in ICP during cardiovascular fluctuations, especially at night [15]. Silicone rubber components of CSF shunts, especially in the subcutaneous compartment, degrade with time, becoming fragile and likely to fracture. This occurs because of deposition of calcium phosphate and aluminum in the external layers of shunt tubing. Evidence suggests that the barium used in the silicone catheters is probably not an important factor in promoting calcification and degradation. But using barium-free catheters makes it difficult to assess a shunt system on radiologic imaging [16].

4.3.3 Complication Due to Age

Age at which the shunt is performed is also a predictor of shunt survival. Patients older than 2 years have a longer shunt survival period than those who were younger. It has been suggested that this observation is related to both immunological deficiency and particular bacterial flora in infants. Prematurity at first shunt insertion also predisposes to subsequent shunt revisions. This point to some fundamental tissue reaction occurs in response to shunt insertion [17].

4.3.4 Complications Due to Miscellaneous Causes

Concurrent surgical procedure is noted to increase the risk of shunt failure. Shunt surgery is generally performed as a single procedure but, sometimes, like in patients with tumors requiring a biopsy, it is done as simultaneous procedure. But in such a setting the procedure becomes long and the risk of infection and malfunction increases [18].

It has been seen that whites have significantly longer shunt survival times than nonwhites. Multiple factors especially increased incidence of prematurity and general immunocompromised state among nonwhites may be responsible for this [19].

As a matter of fact there is no ideal or flawless shunt up to this time. An ideal shunt still needs to be the goal in the future of treating hydrocephalus. The ideal shunt would allow for a flow-regulated control to drain a specific amount of fluid, which could be tailored to an individual patient's needs. All the CSF shunt systems that are currently available show a given failure rate, and therefore search for the perfect shunt continues.

References

1. Greenberg MS (1997) Handbook of neurosurgery, 4th edn. Greenberg Graphics, Lakeland
2. Milhorat TH (1995) Hydrocephalus: pathophysiology and clinical features. In: Wilkins RH, Rengachary SS (eds) Neurosurgery, vol 3, 2nd edn. McGraw Hill, New York, pp 3625–3632
3. Nulsen FE, Spitz EB (1952) Treatment of hydrocephalus by direct shunt from ventricle to jugular vein. Surg Forum 2:399–403

4. Aoki N (1990) Lumboperitoneal shunt: clinical applications, complications, and comparison with ventriculoperitoneal shunt. Neurosurgery 26:998–1003
5. Nash DL, Schmidt K (2010) Complications and management of ventriculoatrial (VA) shunts, a case report. Cerebrospinal Fluid Res 7(Suppl 1):S42
6. Piatt JH Jr (1994) How effective are ventriculopleural shunts? Pediatr Neurosurg 21:66–70
7. Pittman T, Steinhardt G, Weber T (1992) Ventriculoureteral shunt without nephrectomy. Br J Neurosurg 6:261–263
8. Aldana PR, James HE, Postlethwait RA (2008) Ventriculogallbladder shunts in pediatric patients. J Neurosurg Pediatr 1:284–287
9. Kausch W (1908) Die Behandlung des Hydrocephalus der Kleinen Kinder. Arch Klin Chir 87:709–796
10. Gallia GL, Rigamonti D, Williams MA (2006) The diagnosis and treatment of normal pressure hydrocephalus. Nat Clin Pract Neurol 2:375–381
11. Robinson S, Kaufman BA, Park TS (2002) Outcome analysis of initial neonatal shunts: does the valves make the difference? Pediatr Neurosurg 37:287–294
12. Fouyas IP, Casey AH, Thompson D, Harkness WF, Hayward RD, Rutka JT et al (1996) Use of intracranial pressure monitoring in the management of childhood hydrocephalus and shunt-related problems. Neurosurgery 38(4):726–732
13. Jimenez DF, Keating R, Goodrich JT (1994) Silicone allergy in ventriculoperitoneal shunts. Childs Nerv Syst 10(1):59–63
14. Snow RB, Kossovsky N (1989) Hypersensitivity reaction associated with sterile ventriculoperitoneal shunt malfunction. Surg Neurol 31:209–214
15. Drake JM, Kestle J, Milner R et al (1998) Randomized trial of CSF shunt value design in paediatric hydrocephalus. Neurosurgery 43:294–305
16. Echizenya K, Satoh M, Murai H et al (1987) Mineralization and biodegradation of CSF shunting systems. J Neurosurg 67:584–619
17. Serlo W, Fernell E, Heikkinen E et al (1990) Functions and complications of shunts in different etiologies of childhood hydrocephalus. Childs Nerv Syst 6:92–94
18. Shurtleff DB, Stuntz JT, Hayden PW (1985/1986) Experience with 1201 cerebrospinal fluid shunt procedures. Pediatr Neurosci 12:49–57
19. Vinchon M, Dhellemmes P (2006) Cerebrospinal fluid shunt infection: risk factors and long-term follow-up. Childs Nerv Syst 22:692–697