The Sophy valve and the El-Shafei shunt system for adult hydrocephalus

Gerard O'Reilly, Bernard Williams

Abstract

A selected series of 22 adult patients with hydrocephalus were treated by a shunt system incorporating a variable pressure Sophy valve or by ventriculojugular shunting against the direction of blood flow using the El-Shafei system. One patient had insertion of two Sophy valves and an El-Shafei shunt. Patient selection was reserved to those with hydrocephalus thought to be at high risk when shunted with systems containing a conventional unipressure valve. None of the eight patients who had ventriculojugular shunting by the El-Shafei method demonstrated any notable clinical or radiological improvement subsequent to shunt insertion. Of the 16 Sophy devices inserted only seven produced a satisfactory result.

The current evaluation of shunt malfunction could be improved by support for a national register.

(*J Neurol Neurosurg Psychiatry* 1995;59:621–624)

Keywords: hydrocephalus; Sophy valve; El-Shafei shunt system

There is a subset of hydrocephalic patients whose management remains unsatisfactory. To shunt patients with gross ventriculomegaly with a conventional univalve system is problematic. The risk of developing subdural collections is considerable and may necessitate replacement with an alternative fixed pressure valve.

The use of a valve of a pressure sufficiently high to prevent subdural collection may lead to potentially curable symptoms persisting because the valve pressure cannot be lowered. The incorrectly named "normal pressure" hydrocephalus offers a continued therapeutic challenge. If the patients may be improved by lowering pressure then it seems clear that the pressure has not been "normal" for the patient. We recommend the term "symptomatic hydrocephalus". Contemporary treatment remains occasionally disappointing.

There are other patients with disorders related to CSF pressure such as syringomyelia or intracranial pouches who may require their CSF pressure to be adjusted for optimal clinical improvement to occur. Failure to do so may result in persistence of the syrinx or other cavity. Current practices are often disappointing if they rely on the "hit or miss" characteristics of single pressure valves.

The Midland Centre for Neurosurgery and Neurology, Holly Lane, Warley, West Midlands
G O'Reilly
B Williams

Correspondence to:
Mr Gerard O'Reilly, Midland Centre for Neurosurgery and Neurology, Holly Lane, Warley, West Midlands B67 7JX, UK.

Received 30 January 1995 and in revised form 20 July 1995
Accepted 24 July 1995

In the Midland Centre for Neurosurgery and Neurology either an El-Shafei system or a system incorporating a Sophy valve has been used to treat hydrocephalus in an attempt to find a more controlled drainage method. This has been because of the fear of valve complications due to overdrainage and one of us (BW) has arbitrarily selected such potentially "difficult" cases as being suitable for Sophy or El-Shafei shunting. No special preoperative tests such as long term pressure recordings, repeated lumbar punctures, or resistance to outflow studies were performed. The policy of regarding the insertion of the valve as the test for whether a valved shunt will work seems to us to be rational and cost effective. Such a policy may be criticised[1 2] but in the difficult class of case in which the radiological imaging and the clinical indications may be unclear any further tests proposing to refine the accuracy of selection may also be equivocal.

Patients

Between July 1988 and February 1993, 22 adult patients with symptomatic hydrocephalus had insertion of a Sophy three position variable valve or had ventriculojugular shunting against the direction of blood flow with the El-Shafei system. In addition, one patient had two Sophy valves and an El-Shafei system implanted sequentially. Most hydrocephalic patients presenting during this period were managed with conventional unipressure valves. Insertion of the El-Shafei systems and Sophy valves were restricted to those whose ventriculomegaly was severe. The table summarises the aetiology of the hydrocephalus. Fourteen patients with hydrocephalus (six men and eight women) were treated with Sophy valves. The mean age was 46 with a range of 6 to 75 years.

Seven patients had an El-Shafei system inserted for their hydrocephalus; this group comprised four men and three women. The mean age at shunt insertion was 49 with a range of 13 to 70 years. The additional patient who had multiple insertions of these shunt types was a 65 year old woman who presented with ataxia, dementia, and incontinence.

In total 16 Sophy valves and eight retrograde ventriculojugular shunting devices were inserted. Of the 16 shunt systems incorporating the Sophy valve, eight had their distal catheter located in the peritoneal cavity and the other eight had placement in the cardiac venous system. The lateral ventricle was used for the proximal catheter in 14 patients. The

remaining two patients had their proximal catheters placed in cystic lesions associated with the hydrocephalus. All the Sophy valves were initially set at high and modified according to the clinical situation. Attempts were made in two other instances to insert an El-Shafei system but these were abandoned because of hypoplastic internal jugular veins.

Materials and methods

The Sophy valve is a variable pressure valve which can be adjusted percutaneously to different pressure settings.[3,4] The valves used in these patients had three different settings. The CSF flow characteristics at each pressure are comparable with other devices (fig 1). The pressure is regulated by a semicircular spring, the length of which, crossing over a jewel and pressing it into the inlet port, governs the opening pressure of the valve (fig 2). This can be adjusted by the use of a magnet which rotates the supporting magnetic rotor arm and thus alters the effective length of the spring. The position of the rotor arm can be assessed using a compass over the device or by a skull radiograph. Pressure adjustments can be made without operative valve replacement. Standard surgical techniques for shunt insertion were used, a parieto-occipital burr hole usually being used for the proximal catheter.

The El-Shafei system is a valved ventriculojugular shunt inserted against the direction of blood flow.[5-7] The distal tube was passed retrogradely up the jugular vein via a common facial venotomy (fig 3). It was designed in an attempt to normalise the relation between CSF pressure and the intracranial venous pressure. The outlet of the system is within the skull, thus lessening siphoning effects and approximating to the physiological absorption method. In vivo posture change from recumbent to erect results in collapse of the internal jugular vein and increases the resistance to blood flow. It has been postulated that the internal jugular vein thus acts as a collapsible tube in regulating its upstream pressure.[7] This increase in the resistance to blood flow, according to El-Shafei, acts as a natural self regulating antisyphoning device which counteracts the effect of gravity and regulates the pressure above the vein—that is, the sigmoid sinus—and consequently the intracranial pressure within narrow limits. This happens regardless of the position of the patient.

The technique for insertion was as described by El-Shafei.[6]

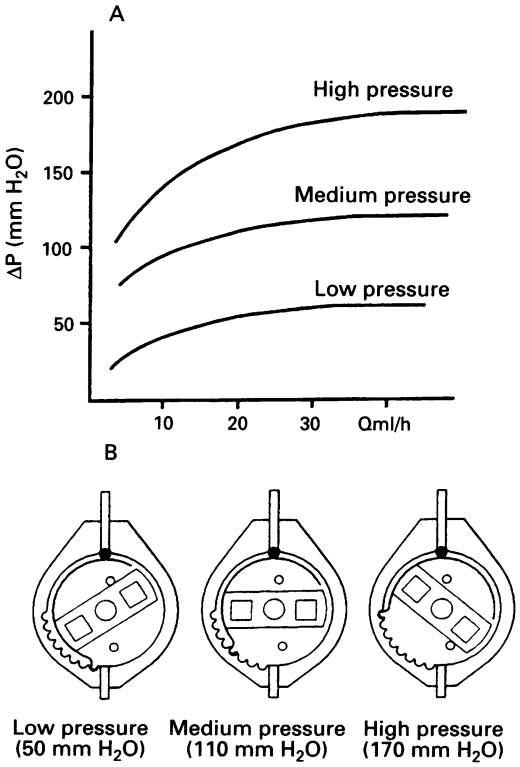

Figure 1 (A) Pressure flow characteristics of the Sophy valve on high, medium, and low settings. (B) Sophy valve on high, medium and low settings, determined by the position of the rotor arm (source: manufacturers literature)

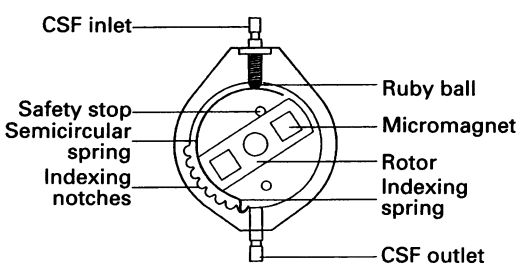

Figure 2 Components of the Sophy valve

Results

There was no operative morbidity or mortality in the series except for one patient who had a Sophy valve inserted and developed a shunt infection. This resulted in shunt removal and external ventricular drainage and it was impossible to assess the effect of the valve. Two other patients with symptomatic hydrocephalus who had Sophy valves inserted failed to improve clinically. It was concluded retrospectively that their presenting cognitive deterioration and ventriculomegaly were secondary to cerebral atrophy. This conclusion is always difficult to validate, particularly if valve placement is used as "the test". Of the remaining 11 patients who had a single Sophy valve inserted, only six patients showed evidence of

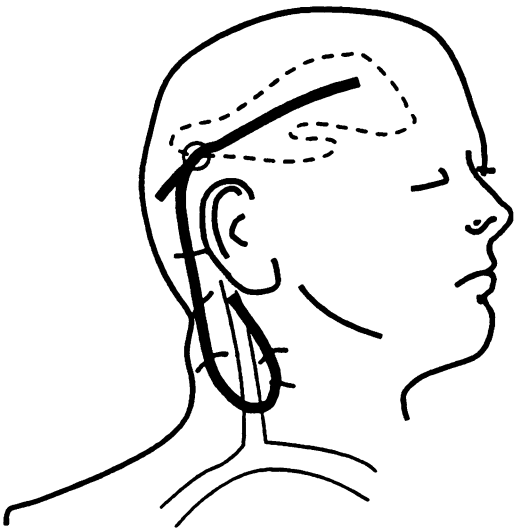

Figure 3 Diagrammatic representation of the El-Shafei system in situ

clinical improvement and radiological diminution of their ventricular size. Two of these patients had required shunt re-explorations and replacement of their proximal catheters, which were found at operation to be blocked. The six patients who had symptomatic resolution after shunting did so when their valve pressure was set to medium on four occasions and to a low pressure setting on two occasions.

The five remaining patients who had a single valve inserted failed to improve symptomatically, although two of these had some diminution in ventricular size. In one of these two patients it proved impossible to change the pressure of the valve down from its original high setting.

Four of the five patients who failed to improve clinically when a Sophy system was inserted subsequently improved when a conventional unipressure system was implanted. Analysis of the five removed valves showed that three were non-functional, and another seemed to drain only at inappropriately high pressures. The fifth device seemed to function normally in vitro.

None of the seven patients who had ventriculojugular shunting against the direction of blood flow improved clinically or had any sustained regression of ventricular size. Five of them continued to deteriorate after insertion of their El-Shafei system. Five of the seven patients later improved clinically when their El-Shafei systems were replaced with conventional valve systems or by alternative adjustable multipressure valvar systems. These "Medos" valves, supplied by Codman, are currently being evaluated.

At reoperation two of the El-Shafei conduits were obviously blocked proximally and three had a distal blockage. After transection of the tubing at reoperation on no occasion could blood be aspirated from the internal jugular vein.

One of our patients who had a ventriculoretrograde jugular shunt inserted had the distal aspect retrieved into the upper cardiac venous system by an overzealous radiologist after a chest radiograph had disclosed the loop in the neck. He had concluded that this was a standard ventriculoatrial shunt the distal end of which had been positioned incorrectly. This shunt system was removed and replaced by an alternate system before relevant overdrainage occurred.

The final patient in our series was a 65 year old woman who presented with incontinence and intellectual decline and had dilated ventricles on CT. A diagnosis of symptomatic hydrocephalus was made and a Sophy valve was inserted. This was set initially at "high". The patient exhibited no clinical or radiological improvement despite serial reduction in the pressure settings. The valve was subsequently removed and found to be non-functional. It was replaced with an El-Shafei system. Again no clinical or radiological improvement resulted, necessitating replacement with another Sophy device. The patient initially exhibited some improvement in intellectual function. This was heightened when the valvar operating pressure was lowered further. At this lower pressure the patient's incontinence resolved.

Discussion

Overdrainage of CSF in hydrocephalic patients treated with conventional single pressure valves is a well recognised and recurrent problem. The complications of subdural and extradural collections, slit ventricle syndromes, secondary synostosis in children, and occlusion of proximal catheters in overshunted ventricles remain familiar. In an attempt to consider these issues an analysis of two shunt systems which have been formulated to give a more controlled CSF drainage mechanism was performed. These were the variable pressure Sophy valve and the ventriculoretrograde jugular shunt of El-Shafei.

The variable pressure concept of the Sophy valve we found to be advantageous. It enabled modification of the valvar opening pressure without the need for further operative intervention. None of the patients in this series treated with a Sophy valve developed a subdural collection. This may be a reflection of the gradual lowering of valvar opening pressure possible with this valve type.

Difficulty was experienced, however, in manipulating pressure settings and in one instance the rotor arm jammed despite attempts to move it. Another drawback of the Sophy valve was that it degraded MRI images and required resetting subsequent to MRI investigation.

The reliability of the Sophy valvar mechanism was questioned. Excluding the two patients who had a retrospective diagnosis of cerebral atrophy and the patient who developed a shunt infection only seven of the 13 Sophy devices resulted in a satisfactory clinical outcome. Better results, however, have been obtained by Battersby and Sutcliffe.[1] There have been subsequent improvements in design but as the device is no longer in use in the Midland Centre for Neurosurgery and Neurology their improved efficiency cannot be attested. Clearly this is a small sample with some errors due to surgery and it is not our intention to dispute the usefulness of this valve.

An adjustable valve not only has advantages in correcting or preventing overdrainage but is also useful in symptomatic hydrocephalus when it is suspected that the initial pressure setting was not low enough. If a non-adjustable valve is inserted and the ventricular size and the clinical condition remains the same then there is a tendency to label the case "non-symptomatic hydrocephalus". It is possible, however, for such patients to be improved by judicious further lowering of pressure. This has been found in this small series and also in the series of adjustable Medos valve cases reported by Kay *et al.*[8]

There is no doubt that the El-Shafei shunt system can work.[6] It quickly and accurately corrects acute hydrocephalus in the short term and has acceptably few complications. The

hope that it might be useful to obtain perfect pressure control in the chronic and "difficult" symptomatic hydrocephalus case, however, has been shown to be unrealistic. It might be thought that the experience with thrombosis around the end of the drainage tube when it is placed in the internal jugular as opposed to the right atrium would be enough evidence to prevent this system being used for long term cases. El-Shafei opined that pointing the end of the drainage tube against the direction of blood flow and placing it in a high flow system such as the jugular bulb might prevent thrombosis. In the present series, however, it is clear that all the distal ends were occluded, presumably by a blood clot, and that therefore this system of drainage is unsatisfactory. Possibly if a non-thrombogenic form of shunt tubing could be developed then this form of hydrocephalus control might be resurrected.

The current appraisal of CSF shunting devices is unsatisfactory and a more comprehensive system of audit is required. Mistakes should not need to be repeated by others, because of failure to publish poor results. A more effective analysis could be cost effective and clinically advantageous. The importance of accruing information cannot be overemphasised. When dealing with the implantation of neurosurgical prostheses, it is essential that not only good results are published but also less than satisfactory results such as these, if a true database is to be acquired.

We think that a national register of shunt systems is overdue and we endorse and commend the efforts of Professor Pickard in this regard[8] and would urge compliancy to the readership. The development of such an information bank would give a quicker indication of the efficiency of the various valvar mechanisms and hopefully speed up improvements in clinical standards.

A more scientific evaluation of shunt malfunction is urgently required. Malfunctioning removed shunt systems need to be assessed to improve our knowledge of shunts and to help correct design faults. Newer shunting devices should ideally undergo neurosurgical pilot studies and only when their efficiency has been assessed should their greater use be encouraged.

We acknowledge the financial assistance from Ann's Neurological Trust Society (ANTS) and the help given by the Medical Illustration Department, Sandwell District Hospital. We thank Ms J Flynn who typed the manuscript.

Mr Bernard Williams died on 9 August 1995.

1 Dauch WA, Zimmermann R. Der Normaldruck—hydrocephalus. Eine Bilanz 25 Jahre nach der Erstbeschreiburg. *Fortschr Neural Psychiatr* 1990;**58**: 178–90.
2 Vanneste JA L. Three decades of normal pressure hydrocephalus: are we wiser now? *J Neurol Neurosurg Psychiatry* 1994;**57**:1021–5.
3 Sutcliffe JC, Battersby RD. Do we need variable pressure shunts? *Br J Neurosurg* 1995;**6**:67–70.
4 Sindou M, Guyotat-Pelissou I, *et al.* Transcutaneous pressure adjustable valve for the treatment of hydrocephalus and arachnoid cysts in adults. *Acta Neurochir (Wien)* 1993;**121**:135–9.
5 El-Shafei IL. Ventriculojugular shunt against the direction of blood flow 1. Role of the internal jugular vein as an antisiphonage device. *Child's Nerv Syst* 1987;**3**:282–4.
6 El-Shafei IL. Ventriculojugular shunt against the direction of blood flow 2. Theoretical and experimental basis for shunting the cerebrospinal fluid against the direction of blood flow. *Child Nerv Syst* 1987;**3**:285–91.
7 El-Shafei IL. Ventriculojugular shunt against the direction of blood flow 3. Operative technique and results. *Child Nerv Syst* 1987;**3**:342–9.
8 UK Shunt Audit Group. Audit of the Medos programmable shunt—a preliminary analysis. Proceedings of the 124th meeting of the Society of British Neurological Surgeons. *J Neurol Neurosurg Psychiatry* 1994;**57**:1150–1.