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The treatment of infantile hydrocephalus: "differential-pressure" or "flow-control" valves

A pilot study

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Abstract The choice of shunt valve in the treatment of hydrocephalus in children remains controversial. We embarked on a pilot study to determine the differences in outcome between differential-pressure and flow-regulating valves. Prospective data collected on 50 consecutive first-time shunt insertions, performed between June 1993 to June 1996, was analysed. Children with tumour- related hydrocephalus and Dandy-Walker malformations as well as children who had external ventricular drainage prior to definitive shunt insertion were excluded from the study. The defining event was the first complication necessitating surgery, including obstruction, over-drainage and infection. Of the 50 children (31 males), 23 had differential pressure (medium-pressure) and 27 had Delta (performance level 2) valves inserted. The mean age at shunt insertion was 26.4 months. The mean follow-up was 53.8 months. The overall cumulative shunt survival at 5 years was 58.6% for the differential pressure and 58.7% for the Delta valves. The mean shunt life was 37.1 months for the differential pressure group and 34.6 months for the Delta group. This difference was not statistically significant (P=0.72, t-test). Both valves had a similar outcome with respect to obstruction (including proximal, valve, distal). The main differences between the two valves were with respect to the incidence of over-drainage and infection. Amongst the differential pressure valves, there were 4 instances of overdrainage (3 slit-ventricle syndrome, 1 bilateral subdural collection) – all occurring within the first 36 months. The Delta valve group had only one instance of over-drainage (bilateral subdural collection). There were no infections in the differential pressure valve group, whereas 3 of the Delta valve shunts got infected, all within the first month. Whereas both shunt types seemed to have a similar overall survival, there was a relatively higher incidence of over-drainage amongst the differential pressure valves. The Delta valves, on the other hand, had higher rates of infection. Similar studies with larger numbers could suggest whether the choice of shunt type will ultimately have to be a compromise accepting one or the other complication.

Key words Infantile hydrocephalus · Cerebrospinal fluid shunt

Introduction

Introduction of cerebrospinal fluid (CSF) diversion shunts in the late 1950s revolutionised the management of infantile hydrocephalus [15]. However, to this date shunt failure remains the commonest problem in the treatment of hydrocephalus. As many as 40% of shunts fail within the first year [7, 9, 19]. Apart from the economic implications of each episode of shunt malfunction, these complications have significant adverse physical and psychological consequences that affect the patient and family alike.

Shunt malfunctions have been categorised into three groups: "Mechanical" – failure related to improper functioning of the device and include obstructions, fractures, migrations and disconnection, "Infection" – related to colonisation of implanted foreign material into the body and development of clinical infection either of the CSF inside the shunt or the soft tissue around it, and "Functional" – related to the hydrodynamic properties of the shunt. Factors related to shunt malfunction have three potential origins: surgeon, patient and the shunt [7]. The thrust of recent research is aimed at analysing the influence of shunt design as a cause of shunt malfunction, resulting in a proliferation of currently available shunt designs. Newer valves that include flow-regulating or antisiphon devices have been designed to address the problem of over-drainage as a cause of shunt malfunction, and have been reported in uncontrolled series to reduce failure rates [12, 18].

But is this really the case? Is there sufficient evidence of the superiority of these newer designs over the 'standard' differential pressure valves? Do they, by addressing one group of shunt malfunctions, invite complications of a different nature?

In an attempt to address these issues, a pilot cohort study was conducted comparing outcome of two groups of hydrocephalic children who had two different types of valves implanted, differential pressure and Delta valves. The opportunity to carry out this study arose because each of the two surgeons involved at the time favoured the use of one particular type of shunt in all of their patients, regardless of patient age and aetiology of hydrocephalus. The propensity to 'stick to a system that works' is widely recognised as a method of choosing a particular type of shunt and is based on a combination of 'style, comfort, past experience, training, brand loyalty, advertising and scientific evidence' [14].

Materials and methods

Prospective data was collected on 50 children who underwent first-time shunt insertions performed by two surgeons (A.D.H., A.R.W.) between June 1993 and June1996. Follow-up was recorded until February 1999. Children with Dandy-Walker malformations and tumour-related hydrocephalus as well as children who required external ventricular drainage prior to the definitive shunt insertion were excluded from the study. The cases were divided into two groups, those that had differential dressure valves (D.P. group) and those that had Delta valves (Delta group) inserted. The differential pressure shunts were PS Medical Medium Pressure shunt assemblies, which were cylindrical or neonatal, depending on the patient's age. The PS Medical Delta shunts were performance level 2 (PS Medical, Goleta, Calif.). The type of shunt inserted in each patient was decided purely by the personal preference of each of the two surgeons. Age at first shunt insertion, sex, aetiology of hydrocephalus and concurrent illnesses were recorded for each patient. The patients were followed up in the out-patient clinics at 3 months, at 6 months and then annually. Routine interval CT scans were not performed, a practice that has since been modified. All patients that presented with suspected shunt malfunction underwent standardised investigations including CT scan, shunt series X-rays and CSF sampling when infection was suspected. The defining event for each shunt was the first episode of malfunction requiring surgery, and the time to first complication was noted. Based on presenting symptoms, investigations and operative findings, shunt failures were tabulated in the following groups: obstruction (proximal, valve and distal), mechanical failure (fracture, migration), over-drainage (slit ventricles, subdural collections), infection. Following an episode of shunt failure, each patient was followed up according to the preference of the individual surgeon and any further events that occurred up to the end of the study period were noted.

Results

Of the 50 children (32 male), 23 had differential pressure valves and 27 had Delta valves. The mean age at shunt insertion for all patients was 26.4 months (range: 0.25–192 months), and the mean follow-up was 53.8 months (range: 32–70 months). For the D.P. group the mean age at shunt insertion was 31 months (range: 0.25–111 months) and the mean follow-up was 54.7 months (range: 32–70 months). For the Delta group the mean age at shunt insertion was 22.6 months (range: 0.30–192 months) and the mean follow-up was 53.2 months (range: 33–65 range). Figure 1 shows age distribution for both groups. Table 1 shows the various causes of hydrocephalus for the two groups. There were no shunt-related deaths during the course of the study. One child from the Delta group died from congenital heart disease.

Table 1 Etiologies of hydrocephalus (*D.P.* group with differential-pressure valves, *Delta* group with delta valves)

Cause	D.P.	Delta
Aqueduct stenosis	8	9
Spina bifida	5	2
Meningitis	1	3
Intraventricular haemorrhage	3	5
Communicating	4	3
Encephalocele	1	0
Hindbrain hernia	0	1
Head injury	0	1
Other	1	3

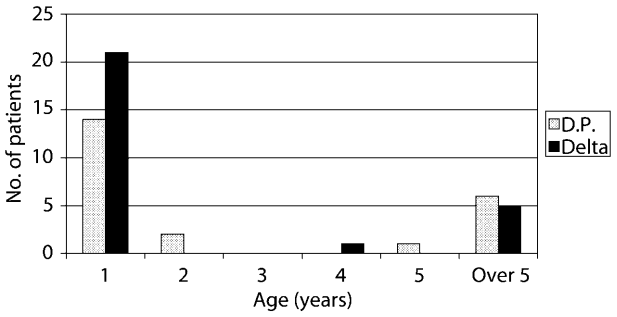

Fig. 1 Age distribution

Table 2 Shunt complications

Complications	D.P.	Delta
Total no. of cases	23	27
Obstruction	5 (21.7%)	5 (18.5%)
Proximal	5	3
Valve	0	1
Distal	0	1
Overdrainage	4 (17.3%)	1 (3.7%)
Slit-ventricle syndrome	3	0
Bilateral subdurals	1	1
Mechanical	0	2 (7.5%)
Fracture	0	1
Disconnection	0	1
Infection	0	3 (11.1%)
Total no. of failed shunts	9 (39.1%)	11(40.7%)

Table 2 shows the complications for the two groups. The mean time to the first shunt complication was 14.3 months for the D.P. group and 7.8 months for the Delta group. The difference was not statistically significant (P=0.26, Student's t-test). From the D.P. group 9 shunts (39.1%) reached the endpoint; shunt obstruction occurred in 5 (21.7%), (all proximal) and overdrainage in 4 (17.3%). From the Delta group 11 shunts (40.7%) reached the endpoint; obstruction occurred in 5 (18.5%), (3 proximal, 1 valve, 1 distal), infection in 3 (11.1%), mechanical complications in 2 (7.4%) and overdrainage in 1 (3.7%).

The mean shunt life was 37.1 months for the D.P. group and 34.6 months for the Delta group. The difference was not statistically significant (P=0.72, Student's t-test). The overall cumulative shunt survival at 5 years was 58.8% for the differential pressure and 58.7% for the Delta valves (Kaplan-Meier survival analysis). Figure 2 shows this in graphical form. There was a difference in the temporal distribution of failures. Whereas in the Delta group most failures occurred within 1 year of implantation, in the D.P. group they occurred over 2 years. After the first 2 years, both groups followed a similar course.

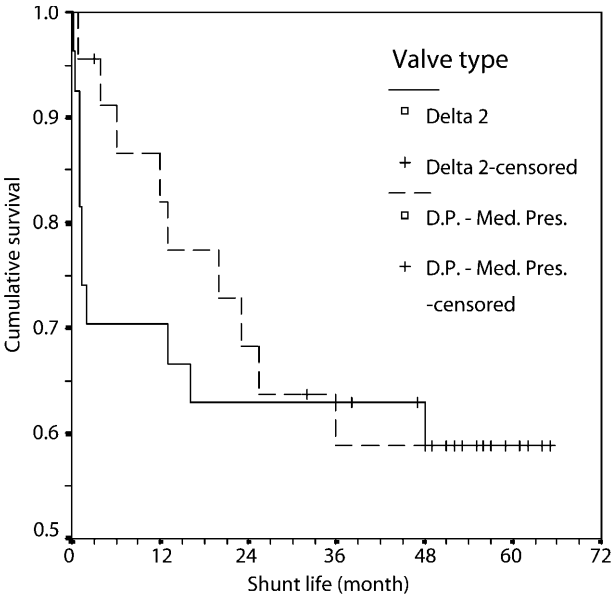

Fig. 2 Cumulative survival of shunt function

The commonest complication was shunt obstruction. There were 5 obstructions in each group, the mean time to obstruction being 14.8 months for the D.P. group and 6.5 months for the Delta group (difference is not statistically significant, P=0.16, Student's t-test). The difference however, was that all the obstructions in the D.P. group were proximal, whereas in the Delta group there were 3 proximal, 1 valve and 1 distal.

The main difference between the two groups was in the incidence of overdrainage and infection. There were 4 instances of overdrainage in the D.P. group (3 slit-ventricle syndrome, 1 bilateral subdural effusions, 17% of all D.P. shunts), as compared to 1 in the Delta group (bilateral subdurals, 3.7% of all Delta shunts). The mean time between shunt insertion and complication was 2.5 months for subdural effusions and 15.3 months for slit-ventricle syndrome (range: 4, 6, 36 months). This early appearance of slit-ventricle syndrome after first shunt insertion was surprising, and contrary to the widely held belief that this type of overdrainage complication occurs after many years of shunting.

There were 3 episodes of infection on the Delta group, all within the first month of shunt insertion. The mean age at shunt insertion was 3 weeks. There were no shunt infections in the D.P. group.

Discussion

The choice of shunt in the treatment of infantile hydrocephalus remains controversial. The plethora of shunt designs currently available bears testimony to the fact that the 'perfect shunt' remains elusive. As commonly

mentioned 'the history of evolution of ventricular shunting for hydrocephalus is largely a history to prevent the complications of shunting' [13]. Evidently, an ideal valve should be flow controlled. Such a valve would have to continuously determine the rate of CSF formation and absorption and regulate the flow so as to remove only the excess [5]. The choice of shunt, however, is very often more subjective than scientific, and is usually a compromise between two or more risk factors. The shunt characteristics, including configuration, hydrodynamic properties, and material, are a compromise between ease of insertion, risk of disconnection, risk of early or late obstruction, expense, and ease of manufacture. From the patient management point of view, one often compromises between the pressure-flow requirements at the time of surgery and those later on in life [7].

The study presented here has two main disadvantages: lack of randomisation in the strict scientific sense and small number of patients. The main advantages, though, include the fact that it represents a single centre experience with only two surgeons involved, which reduces any bias related to surgeon or operating theatre variables. Moreover, there was a consistency in the choice of shunt amongst the two surgeons who performed all operations, thus permitting a direct comparison between two otherwise similar groups. In effect, the patients were randomised into two groups, because the type of shunt each received was a function of when they first presented to the neurosurgical unit.

Of interest is the fact that although the two groups were similar with respect to the number of obstructions from all causes, in the D.P. group they were all proximal. Moreover, 4 of the 5 proximal obstructions in this group occurred at or after 1 year, i.e. were 'delayed' obstructions. It is now recognised that an important factor determining the occurrence of proximal obstructions is the size of the ventricles after shunting, and that proximal obstruction is commoner in patients with slit-like ventricles [7, 17, 19]. A correlation between proximal obstruction and slit ventricles during the postoperative course has been reported in a large series of infantile hydrocephalus treated with differential pressure shunts [19]. In the absence of interval scans in this cohort, the inference that the proximal obstructions were due to overdrainage causing slit ventricles would be purely speculative. However, if this were the case, it makes for a stronger argument in favour of the use of flow-regulating devices to counteract the effects of siphoning in the upright posture causing small ventricles and proximal obstructions. These devices have been shown to significantly reduce the incidence of proximal obstruction caused by slit ventricles, but only at the cost of an increased risk of early valve obstructions [3, 8, 11, 12, 17, 18]. A similar pattern is evident in this study, where 2 of the 5 obstructions in the Delta group occurred at the valve or distally.

There was a notable difference between the two groups in the incidence of overdrainage complications. Although the subgroup analysis did not achieve statistical significance, there was clearly a higher rate of overdrainage complications in the D.P. group. This is consistent with other studies which have elucidated the mechanism of 'siphoning', its association with differential pressure valves and its contributory role in causing the manifestations of the so-called slit-ventricle syndrome [1, 3, 4, 6, 11, 12, 20]. The early occurrence of symptomatic overdrainage was a surprising finding, bringing up the point that the mechanism causing symptoms in the presence of slit ventricles and its temporal evolution is not yet clearly defined.

Another difference between the two groups was in the incidence of infection. There were three cases of shunt infection in the Delta group, all occurring within 1 month of shunt insertion. Notably, in all these cases the shunts had been inserted within 3 weeks of birth. Amongst the various risk factors implicated in the pathogenesis of shunt infection, young age at insertion has represented a higher risk in a number of studies [2, 10, 16]. However, given that both the groups were similar with respect to mean age at shunt insertion, all other factors being equal, one has to speculate that the difference was related to the shunt type. It has been suggested that high-pressure or high-resistance systems may promote CSF leaks around the skin incision, thus predisposing to shunt infection [8].

A discussion of a comparison between two different types of shunts would be incomplete without alluding to the findings of the Randomised Trial of Cerebrospinal Fluid Shunt Valve Design in Paediatric Hydrocephalus, which compared differential pressure, Delta and Orbis Sigma valves [9]. Of the 344 children randomised at 12 centres and followed up for a minimum of 1 year, only 61% were failure free at 1 year. There was no difference in failure-free shunt duration amongst the three valves studied. Comparison between D.P. and Delta groups in this trial showed very similar numbers of complications, but the sites of obstruction were different, more proximal in the D.P. group, more distal in the Delta group, as also shown in our series. The trial concluded that the newer, sophisticated valves that aimed at more 'physiological' control of CSF drainage had not yet made a sufficient impact on failure rates.

In conclusion, the management of paediatric hydrocephalus by CSF shunt procedures involves a complex interaction of patient-, shunt- and surgeon-related factors. Although vast strides have been made in the field of shunt design in the last four decades, shunt failure remains a frustratingly common problem. Choice of shunt type may well turn out to be a preference on the surgeon's part between the lesser of two evils: infection and overdrainage.

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