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ORIGINAL PAPER

Hydrocephalus shunt practice of experienced pediatric neurosurgeons

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Abstract
Objective The objective of this study is to evaluate the ways experienced pediatric neurosurgeons insert ventriculoperitoneal shunts and manage the shunted children afterward.
Methods Seven pediatric neurosurgeons with extensive experience in hydrocephalus were surveyed about their choice of shunts, methods of shunt insertion, shunt follow-ups, management of incidental ventriculomegaly, and prevention of slit-ventricle syndrome. The author completed the survey also.
Results No particular shunt was used by a majority of respondents, although differential pressure valves were used most often. Adjuncts to insert the ventricular catheter were used by half. Shunt catheters were inserted frontally in half and posteriorly in half. No one obtained annual follow-up scans after 5 years of age, and no one operated on asymptomatic children with ventriculomegaly except perhaps in spina bifida cases. No techniques were identified to prevent slit-ventricle syndrome, but respondents emphasized the need for reticence in initial shunt insertions and in shunt revisions.
Conclusions There are substantial variations among extremely experienced pediatric neurosurgeons in their choice of shunts and their techniques of shunt insertion but reasonable uniformity in their frequency of follow-up, in not-obtaining routine scans after age five, and in rarely revising asymptomatic children. Methods to accurately position ventricular catheters and to prevent slit-ventricle syndrome need to be evaluated in multicenter studies.

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Introduction

Randomized clinical trials conducted in the past decade have demonstrated that no commercially available shunt functions significantly longer than any other [4, 10], that programmable shunts function no better than nonprogrammable shunts [8, 12], that endoscopic shunt placement is associated with no better outcome than freehand placement [9], and that positioning of the ventricular catheter away from the choroid plexus is associated with significantly longer shunt function [9]. A large case series indicated that antibiotic-impregnated shunts do not significantly reduce the frequency of shunt infections in children [6].

No prospective study has demonstrated that either frontal or posterior shunt insertions are better or how to prevent slit-ventricle syndrome. In day-to-day pediatric neurosurgery, there are other practical questions, such as how often to see patients in follow-up, when to get follow-up scans, and what to do if scans demonstrate ventricular enlargement in asymptomatic patients.

This survey was carried out to determine if pediatric neurosurgeons with extensive experience caring for children with hydrocephalus had similar shunt practices. The survey was intended to evaluate the practice patterns of a particular small group of pediatric neurosurgeons, not to evaluate shunt outcomes or obtain generalizable data.

Methods

James Drake, John Kestle, Jerry Oakes, Harold Rekate, Michael Scott, Paul Steinbok, and Marion Walker were

asked to participate in the survey. They had practiced pediatric neurosurgery from 18 to 33 years, with a mean of 25 years, and all had published manuscripts about hydrocephalus research. They were asked the following questions: (1) What shunt do you use most frequently? (2) Do you routinely use any adjunct to insert the ventricular catheter? (3) Do you insert ventricular catheters frontally or posteriorly? (4) How frequently do you see shunted patients in follow-up after infancy? (5) Do you get annual computed tomography (CT) or magnetic resonance (MR) scans in follow-up? (6) What would you do if you see an asymptomatic shunted child with normal fundi whose ventricles have increased (a) from slits to normal, (b) from normal to mildly enlarged, and (c) from mildly enlarged to moderately enlarged? (7) How do you prevent slit-ventricle syndrome? The author's answers to the questions were included with the seven respondent answers.

Results

Replies to the questions are shown in Tables 1, 2, 3, 4, 5, 6, and 7. The most commonly used shunt was a medium pressure, differential pressure valve. None of the respondents used antibiotic-impregnated shunts routinely, and only one used a programmable shunt routinely. Most neurosurgeons did not use adjuncts to position the intraventricular catheter; of those who did use adjuncts, ultrasound was the modality used most often. Ventricular catheters were inserted frontally and posteriorly in approximately equal numbers. Shunted patients were seen in follow-up at widely differing intervals but were most commonly seen every year until age 5 then every 2 to 3 years thereafter. Routine follow-up scans were generally not obtained; no one obtained annual scans after age 5. If scans were obtained in shunted, asymptomatic children who had normal fundi, most respondents would follow the child, although if the child had spina bifida, a shunt revision would be considered more seriously; if moderate ventriculomegaly was detected, then ICP monitoring was considered. Respondents thought the frequency of slit-ventricle syndrome could be decreased primarily by judicious shunting and revising and potentially by the use of Orbis–Sigma valves (OSV).

Table 1 Which shunt do you use most frequently?

Orbis–Sigma II integral shunt
Differential pressure valve—low pressure in babies, medium in older children, Orbis–Sigma if subdural hematomas are a concern
Differential pressure valve—usually medium pressure to start with
Delta 1.5 in 95%; Orbis–Sigma occasionally. Have never placed a programmable shunt.
Medium pressure PS Medical valve with a Rickham reservoir
PS Medical medium pressure but recently tending to OSVII to decrease slit ventricles
PS Medical 65%, Codman Medos 25%, Orbis–Sigma 10%
Codman Hakim Programmable valve with siphonguard except with high protein or particulate matter

Table 2 Do you routinely use any adjunct in ventricular catheter insertion?

No
Yes, ultrasound in every case
Yes, ultrasound to place the tip in the frontal horn, anterior to the foramen of Monro
Pen endoscope for ~30% of new shunts and >90% of revisions; ultrasound rare
No, but we are getting intra-op CT to decrease our rate of malpositioned catheters
No
Endoscope for 25% of revisions, ultrasound rarely, Stealth if guidance is needed
Infants: no adjuncts; older patients and revisions: Stealth system with PCI probe

Discussion

The pediatric neurosurgeons who were surveyed are some of the most experienced in the USA and Canada. Several have written authoritatively about hydrocephalus and shunting. All but two were from different medical centers. There are few ways for pediatric neurosurgeons to know if their practice is similar to that in other centers. The results of this survey indicate that there is considerable variability in shunt practice, except that no one obtained routine annual scans after age 5, and these neurosurgeons were loathe to revise shunts in asymptomatic children whose ventricles had enlarged.

When questioned about the shunt of one's preference, most of the respondents used differential pressure valves, but there was no preference for a particular shunt. Their practice is in keeping with data from Kestle et al., who

Table 3 Do you insert shunts frontally or posteriorly?

Frontally
Posteriorly
Frontally
Frontally in 90%
Posteriorly (the frontal ones are guaranteed to be too short in a few years)
Usually posteriorly but frontally if considering an ETV later
Frontal unless contraindicated
Posteriorly (parieto-occipital) with frameless stereotaxy for new shunt insertion

Table 4 How frequently do you see shunted patients in follow-up after age 2?

Annually until age 5 then every 2 years until age 13, then every 3 years
Every 2–3 years
Annually until ages 5 to 6 then every 2 years
Every 6 months until age 5 then annually
Annually until adolescence then every 2 years
Every 2 years for maybe two visits then every 5 years
Every 6 months until 4 to 5 years old then annually
Annually until age 5 then every 2 years

demonstrated no significant difference in long-term function rates between differential pressure, Delta, and Orbis–Sigma valves inserted in infancy, although function of the OSV shunt was 10% greater than the other two valves at 5 years [10].

Seven of the eight neurosurgeons preferred nonprogrammable shunts. Data support that practice also: randomized, prospective studies demonstrate significant difference in function neither of programmable and nonprogrammable Codman shunts [12] nor of programmable (Strata) and nonprogrammable Medtronic shunts [8]. Yet, in spite of Class I data confirming nondifference, two of the top five shunts inserted in the USA (Medtronic Delta, Medtronic Strata, Codman programmable, Orbis–Sigma II, Codman Hakim) are programmable (personal communication, Cathy Dorangrichia, Medtronic).

The respondent who used programmable shunts used models with an antisiphon device and adjusted the valve to its highest setting after ventricles had decreased to their smallest size to minimize the development of slit-ventricle syndrome. Whether programmable shunts are associated with significantly fewer chronic headaches that nonprogrammable shunts has not been determined. In one study, 63% of symptoms—mainly headaches—improved after shunt valve reprogramming [7].

Table 5 Do you get yearly CT or MR scans in follow-up?

No, prn
No, prn
Scan at 3 months and 1 year, then prn
CT scan annually until age 5, then every 2 years
Baseline CT or MR 1 year after shunting, then every 3–5 years after age 5
Scan 1 year post-shunt, then at about 5 years, then before transitioning to adult NS
Obtain baseline CT/MR but do not obtain them routinely
No routine imaging of asymptomatic patients except quick MRIs every 3 years in children with spina bifida

Table 6 What would you do if you see an asymptomatic shunted child with normal fundi whose ventricles have increased (a) from slits to normal, (b) from normal to mildly enlarged, or (c) from mildly enlarged to moderately?

(a) observe, (b) observe, (c) consider ICP monitoring
Get shunt series to exclude disconnection or fracture, follow more closely, make sure they really are asymptomatic, revise for any new symptoms
If the shunt series is normal, I would follow the patient in all three scenarios and have ophthalmology evaluate and follow
(a) Nothing, (b) take a detailed history and consider monitoring, (c) revise
Tend to just watch. The exception is the myelo patient where if there is ventricular dilatation, I almost always revise to avoid syringomyelia or Chiari symptoms later
Observe in all cases
Observe in all cases. Repeat the scan in a few months and be more concerned if the ventriculomegaly progressed
Very loathe to intervene on asymptomatic non-spina bifida patients; low threshold to intervene for apparently asymptomatic enlargement in SB patients

Although half of the respondents did not use adjuncts to position ventricular catheters, Kestle et al. demonstrated that catheters positioned away from choroid plexus function significantly longer than those adjacent to it [9]. Endoscopic catheter insertion was postulated to position catheters away from choroid plexus, but the endoscope trial demonstrated that catheters inserted endoscopically function no better (and almost-significantly worse) than freehand inserted catheters [9]. Techniques are available to position catheters in more desirable locations. In the series of Drake et al., 10/10 catheters, inserted with real-time ultrasonagraphhy, were properly positioned [13]. Stealth-guided catheter insertions, used routinely by one of the respondents in this survey, can be done and are particularly helpful in children with slit ventricles.

Table 7 How do you prevent slit-ventricle syndrome?

Unsure. OSV shunts are associated with a lower risk of slit ventricles
Not sure, maybe Orbis–Sigma
Try to avoid a shunt; be conservative in shunting and revising
Never use low pressure valves, only shunt patients who really need it, no distal slit valves, use high pressure valves when the ventricles are initially large
Have seen it rarely. Put the catheter anteriorly with the scope and the tip through the foramen of Monro
Not a significant problem in our practice. Accept intermittent headaches. Do not reoperate unless symptoms are affecting lifestyle
I have no idea
Have a high tolerance for ventriculomegaly and try not to shunt in the first place in the absence of overt problems. Attempt to dial the programmable valve to its highest setting when the ventricles are the smallest.

The debate continues as to whether shunts function better if inserted frontally or posteriorly. In 1988, we reported a retrospective study in which the 10-year shunt function rate was significantly better in frontally inserted shunts (55%) than in posteriorly inserted shunts (25%) [1]. Three years later, Bierbrauer et al. reported a randomized prospective study that observed no significant difference in shunt function between frontal and posterior shunts, but the study duration was only 14 months [2]. Unpublished data from Kestle's shunt design trial demonstrate 45% shunt function at 5 years for frontal shunts vs a 30% rate for posterior shunts, but the difference is not statistically significant because of large confidence intervals at 5 years (John Kestle, personal communication, 2009).

Questions 4–7 on the survey asked about individual practice and cannot be compared to published standards yet are important issues in the management of children with shunted hydrocephalus. The frequency of shunt patient follow-up probably varies according to the purpose of the visit: practitioners who schedule visits to identify occult shunt malfunction follow patients more frequently than those who schedule visits primarily to reeducate patients and parents about the symptoms and signs of shunt malfunction. Routine clinic visits rarely identify patients with shunt malfunction. In 1997, we reported 100 children who were seen in routine follow-up after shunting to determine how many were diagnosed with shunt malfunction; four had symptoms that were suspicious for malfunction; yet, none were confirmed to have malfunction [3]. We also evaluated the last 100 children who had undergone shunt revisions to determine how many were diagnosed during routine follow-up; only one child who had been shunted for more than a year was diagnosed during routine follow-up.

All respondents indicated that they obtained a "baseline" CT or MR scan approximately 1 year after shunt insertion, but the frequency of scans thereafter differed widely. The rationale for obtaining annual scans is questionable. CT scans do not demonstrate ventricular enlargement in one third of children with shunt malfunction [5]. The question about management of a shunted, asymptomatic child with normal fundi, whose scan demonstrated some ventricular enlargement from the baseline, indicated a strong preference of this cohort of pediatric neurosurgeons for the maxim, "treat the child, not the scans," and reflects extensive experience. There is probably no justification for revising a shunt in an asymptomatic child with normal fundi whose ventricles have enlarged unless monitoring confirms increased intracranial pressure.

Few publications address the question of whether to obtain CT or MR scans during follow-up visits of asymptomatic shunted patients. Liptak et al. retrospectively reviewed the charts of 112 children with shunted hydrocephalus secondary to myelomeningoceles who were monitored with CT scans, usually one scan per year after age 1, for an average of 12.2 years [11]. Shunt malfunction was diagnosed by routine CT scans in 8%, and in children with shunts that functioned for more than a year, 14% were diagnosed with shunt failure by routine CT scans; 76% were diagnosed by clinical symptoms and 15% by clinical signs. However, the authors did not specify the criteria by which shunt failure was diagnosed.

In this survey, there was no consensus about the treatment of slit-ventricle syndrome. Several respondents emphasized the need for its prevention, particularly by judicious shunting and revising. Those opinions presumably reflect the difficulties encountered in treating shunt malfunctions in children with slit ventricles and the paucity of good therapeutic options for that syndrome. The frequency of slit ventricles appears to be lower with Orbis–Sigma valves than with other types. Kan et al. reported that slit ventricles occurred in 10.8% of children with differential pressure valves, 10.5% of those with Medtronic Delta valves, and 3.6% of those with Orbis–Sigma valves (p=.0007) [7].

Conclusions

Experienced pediatric neurosurgeons manage hydrocephalus similarly in judicious shunting and shunt revision—even in the face of ventricular enlargement—and in not obtaining annual scans after age 5. Their practice differed in terms of shunts used and in the use of adjuncts for shunt insertion. Better data are needed to evaluate methods to prevent slit-ventricle syndrome.

Conflict of interest The author has no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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