ORIGINAL ARTICLE

Ventriculoperitoneal shunt block: what are the best predictive clinical indicators?

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Aims: To evaluate the predictive value of symptoms, signs, and radiographic findings accompanying presumed ventriculoperitoneal (VP) shunt malfunction, by comparing presentation with operative findings and subsequent clinical course.

Methods: Prospective study of all 53 patient referrals to a paediatric neurosurgical centre between April and November 1999 with a diagnosis of presumed shunt malfunction. Referral pattern, presenting symptoms and signs, results of computed tomography (CT) scanning, operative findings, and clinical outcome were recorded. Two patient groups were defined, one with proven shunt block, the other with presumed normal shunt function. Symptomatology, CT scan findings, and the subsequent clinical course for each group were then compared.

Results: Common presenting features were headache, drowsiness, and vomiting. CT scans were performed in all patients. Thirty seven had operatively proven shunt malfunction, of whom 34 had shunt block and three shunt infection; 84% with shunt block had increased ventricle size when compared with previous imaging. For the two patient groups (with and without shunt block), odds ratios with 95% confidence intervals on their presenting symptoms were headache 1.5 (0.27 to 10.9), vomiting 0.9 (0.25 to 3.65), drowsiness 10 (0.69 to 10.7), and fever 0.19 (0.03 to 6.95). Every patient with ventricular enlargement greater than their known baseline had a proven blocked shunt.

Conclusions: Drowsiness is by far the best clinical predictor of VP shunt block. Headache and vomiting were less predictive of acute shunt block in this study. Wherever possible CT scan findings should be interpreted in the context of previous imaging. We would caution that not all cases of proven shunt blockage present with an increase in ventricle size.

Cerebrospinal fluid (CSF) shunts remain the mainstay of treatment for most cases of hydrocephalus in the paediatric population. All are prone to malfunction, with block being the commonest reported complication in most series. In the largest reported cohort of 1719 patients, 56% experienced at least one episode of shunt block in the 12 years following insertion.[1] Similarly, Lazareff and colleagues[2] recently reported a 44% prevalence of shunt malfunction, including block, in 244 children with CSF shunts followed up over a period of up to six years post initial insertion. The peak "danger" period for blockage is in the first year after insertion, with rates as high as 20% recorded in some series.[3] Annual rates of shunt blockage have been estimated by Rekate to be approximately 5%.[4] These findings are in keeping with our published unit experience in which we noted a 28% incidence of shunt block over a 10 year period, and that 55% of patients experienced at least one episode of shunt malfunction during this time.[5]

The presentation of shunt block may undoubtedly be with what might be termed "classical" symptoms, namely headache, vomiting, and drowsiness,[6] or may on occasion be more atypical and misleading.[7] Documented more atypical presentations include seizures, abdominal pseudocyst, syringomyelia, cranial nerve palsies, and hemiparesis.[6] Parkinson like rigidity,[6] visual failure,[8] and developmental standstill have also been documented.

The importance of prompt diagnosis and operative treatment of blocked ventriculoperitoneal (VP) shunts cannot be overemphasised. Death or major neurological sequelae, including blindness, are well described sequelae of delayed treatment.[5] Difficulty in making the diagnosis may stem from a considerable symptom overlap with other common childhood illnesses. This is compounded further by the fact that an accurate first hand history may not prove forthcoming, as a result either of patient age, or that up to 60% attend special school and so a first hand history may not be possible.[5 9 10]

We present a blinded prospective study in which we evaluate which symptoms commonly prompting an urgent neurosurgical opinion are truly predictive of acute shunt block. This is further correlated with the results of computed tomography (CT) scanning in each patient and ultimate clinical outcome.

METHODS

Setting

The department of neurosurgery at Great Ormond Street Hospital provides tertiary level paediatric neurosurgical care to a large part of London and southeast England. The unit carries out approximately 70 operative procedures per year on patients with newly diagnosed hydrocephalus or related conditions, with the workload divided equally between three consultant neurosurgeons. Once shunted, patients are followed up in a paediatric neurosurgical clinic, and an attempt made to acquire a baseline CT scan postoperatively on all patients. They are additionally offered "open door" access to the unit should they develop symptoms suggestive of a shunt block. These symptoms are detailed in an information booklet given to every parent once a shunt has been inserted.

Patients

The study period was from April to November 1999. All children referred to the Department of Neurosurgery at Great Ormond Street Hospital with a tentative diagnosis of shunt

Abbreviations: CSF, cerebrospinal fluid; CT, computed tomography; GCS, Glasgow coma scale; VP, ventriculoperitoneal

Table 1 Data recorded on each patient admission referred with possible VP shunt malfunction over the study period

Age
Aetiology of hydrocephalus
Mode of referral (self, district general hospital, clinic, other)
History of previous shunt block
Presenting symptoms
Presence or absence of fever
Results of CT scanning of brain
Operative findings
Clinical outcome

malfunction were entered into the study. Fifty three such admissions were recorded over this period. These comprised 34 patients, 11 of whom were seen on more than one occasion.

Definitions

The diagnosis of shunt blockage was considered confirmed when peroperative examination revealed no CSF flow from the ventricular catheter, or when manometric evaluation revealed abnormal or no flow through the valve or distal catheter. The diagnosis of "no shunt blockage", which resulted in no surgical intervention, was considered confirmed if the patient made the recovery anticipated from their alternative diagnosis and did not re-present with further symptoms and signs suggesting shunt malfunction.

Data collection

This was a prospective study. None of the surgeons were aware the data was being collected during the time period it was carried out. Table 1 illustrates the data recorded on each of the 53 occasions patients were admitted during the study period.

Statistical analysis

Four patient admission groups were initially defined:

(A) All admissions referred with suspected shunt block/malfunction (n = 53)

(B) Those admissions we diagnosed preoperatively as having shunt block/malfunction, group B1 (n = 37); and those we diagnosed as not having shunt block/malfunction, group B2 (n = 16)

(C) Admissions with confirmed shunt block (n = 34) or shunt infection (n = 3)

(D) Admissions with an initially incorrect clinical diagnosis of "normal shunt function" who later re-presented during the study period with proven block (n = 4).

Surgical proof of shunt blockage was required for inclusion in group C. Odds ratios were then calculated to compare the symptomatology between groups C and B2. In this way we compared the presenting symptomatology in those referred with proven shunt malfunction (group C) with those in whom we considered shunt function to be normal on admission (group B2). Ratios were calculated for headache, vomiting, drowsiness, and fever. The low incidence of other presenting symptoms negated against meaningful statistical analysis as to their significance. Odds ratios were then recalculated with the admissions from group D transferred to group C. By this process symptomatology was compared between two groups; those with proven shunt malfunction, and those with proven normal shunt function.

RESULTS

Patient age ranged from 6 weeks to 17.7 years of age, median 7.2 years. Table 2 summarises the aetiology of hydrocephalus in this cohort.

Table 2 Aetiology of hydrocephalus

Aetiology	%
Tumour	26
Prematurity with intraventricular haemorrhage	26
Congenital	23
Meningitis	8
Non-accidental injury	8
Craniosynostosis	3
Posterior fossa cyst	3
Arnold–Chiari malformation	3

Referral pattern

Forty of the 53 admissions were referred directly by their parents as per the unit "open door" policy. A further 10 came from outlying district general hospitals. Two were admitted directly from clinic at Great Ormond Street Hospital. One was transferred in by air ambulance from abroad. In 39 of the 53 admissions during this study period, the patients involved had suffered a previous episode of shunt malfunction necessitating revision. One patient was admitted on seven occasions during the study period, necessitating five shunt revisions in total. Thirty four individual patients were seen during the study period. Table 3 illustrates the presenting symptoms and the results of CT scanning in each patient admission (53 admissions of 34 individual patients).

On 11 occasions patients were unable to vocalise their symptomatology on admission. Reasons for such included young age, obtundation on admission, and longstanding learning disability.

Surgery was undertaken for 37 of the 53 admissions for a preoperative diagnosis of shunt blockage in 34 and for shunt infection in three. This resulted in resolution of the presenting symptoms in all 34 children with blocked shunts, 84% of whom had increased ventricle size noted on their preoperative CT scanning when admission images were compared with those on file. In the other three operations an infected shunt was removed and replaced with an external ventricular drain. All three had a further ventriculoperitoneal shunt reinserted following intrathecal and intravenous antibiotic therapy.

Of the remaining 16 admissions, alternative diagnoses were proven in five cases. These comprised viral illness (two admissions) and CSF overdrainage (three). For the other 11 admissions, no identifiable unifying diagnosis was apparent. In four of these cases (group D), the patients re-presented within the study period and were subsequently found to have shunt block. In all four shunt revision resulted in symptom resolution. On first presentation none of these four patients had a Glasgow coma scale (GCS) less than 15 or evidence of a recent increase in ventricle size on CT scanning. Symptoms on their first admission comprised headache or vomiting, or both. On re-presentation, symptomatology was different in each case. Three of the four had a Glasgow coma scale less than 15, and two had increased ventricle size on CT scanning. The patients who accounted for the other seven admissions for which no specific diagnosis was made did not re-present during the study period with shunt malfunction.

Statistical analysis

For admissions with and without shunt malfunction, groups C and B2 respectively, the odds ratios with 95% confidence intervals for comparison of the symptomatology were headache 1.5 (0.27 to 10.9), vomiting 0.9 (0.25 to 3.65), drowsiness 10 (0.69 to 10.7), and fever 0.2 (0.03 to 6.95). The equivalent odds ratio for enlarged ventricle size on CT scanning was infinite, as every admission with ventricular enlargement greater than their previous baseline had a proven

Table 3 Presenting symptoms in each case, with the results of CT scanning

Admission	Symptoms			Ventricle size on CT scan	Surgical findings
1		Vomiting		Unchanged	–
2	Headache	Vomiting	Drowsiness	Larger	Block
3		Lethargy		Unchanged	–
4	Headache	Vomiting	Drowsiness	Larger	Block
5		Vomiting	Drowsiness	Unchanged	†
6	Headache		Drowsiness	Larger	Block
7	Headache	Vomiting		Unchanged	–
8	Headache		Drowsiness	Larger	Block
9	Headache	Vomiting	Drowsiness	Larger	Block
10	Headache			Unchanged	–
11		Vomiting	Drowsiness	Larger	Block
12	Headache	Vomiting	Drowsiness	Larger	Block
13	Headache			Unchanged	–
14		Vomiting	Drowsiness	Unchanged	–
15		Vomiting	Drowsiness	Larger	Block
16	Headache	Vomiting	Drowsiness	Unchanged	–
17	Headache	Vomiting		Unchanged	–
18	Headache	Vomiting	Drowsiness	Larger	Block
19	Headache			Larger	Block
20	Headache	Vomiting	Drowsiness	Larger	Block
21		Vomiting	Drowsiness	Unchanged	–
22		Vomiting	Drowsiness	Unchanged	Block
23	Headache	Vomiting		Unchanged	–
24	Headache	Vomiting		Larger	Block
25	Headache	Vomiting	Drowsiness	Larger	Block
26	Headache	Vomiting		Larger*	Block
27	Headache	Vomiting		Larger	Block
28	Headache	Vomiting		Unchanged	–
29	Headache	Vomiting	Drowsiness	Unchanged	Block
30		Vomiting		Unchanged	–
31			Drowsiness	Larger	Block
32	Headache	Vomiting	Drowsiness	Larger	Block
33	Headache	Vomiting	Drowsiness	Larger	Block
34	Headache	Vomiting	Drowsiness	Unchanged	–
35		Lethargy		Larger	‡
36	Tonic-clonic fit			Larger	Block
37	Headache	Vomiting	Drowsiness	Larger	Block
38	Headache	Vomiting	Drowsiness	Larger	Block
39	Headache	Vomiting	Drowsiness	Smaller	–
40	Headache	Vomiting	Drowsiness	Larger	Block
41		Vomiting		Smaller	–
42	Headache			Unchanged	Block
43	Headache	Vomiting		Unchanged	–
44			Drowsiness	Unchanged	–
45	Headache		Drowsiness	Larger	Block
46	Headache			Larger	Block
47	Headache	Vomiting	Drowsiness	Larger	†
48	CSF leak			Larger	†
49		Vomiting	Drowsiness	Larger	Block
50	Headache	Vomiting		Unchanged	‡
51	Headache	Vomiting	Drowsiness	Larger	Block
52	Headache		Drowsiness	Larger	Block
53		Vomiting	Drowsiness	Unchanged	Block

*Radiographic "shunt series" indicated the distal catheter to have broken.
†In admissions 5, 47, and 48, a preceding CSF tap indicated infection; in all three cases the VP shunt was replaced with an external ventricular drain. Only admission 5 presented with fever.
‡At surgery it was found that the distal end of the catheter was in an extraperitoneal position.

blocked shunt, in contrast to the admission group "without shunt block" (group B2), in which ventricular size was uniformly unchanged. In the presence of headache, vomiting, and drowsiness, but no fever, 82% of admissions were subsequently proven to have acute shunt block. A further 6% (three cases) had an infected VP shunt (admissions 5, 47, and 48 in table 3). In only one case was fever an initial presenting sign. All three presented with vomiting and drowsiness; one presented with headache.

If the four admissions (group D) originally considered to have normal shunt function are instead included in the "shunt malfunction group" (group C) (as their initial presentation likely represented shunt block), and the odds ratios for symptomatology in each recalculated, the results are as follows: drowsiness 4.8, headache 2.5, vomiting 1.3, fever 0.1.

Eight of the 10 admissions referred via their district general hospital subsequently had proven shunt block, compared with 24 of 40 (60%) seen as a result of the "open door policy". The four admissions initially incorrectly considered to have normal shunt function had all presented directly to the unit via the open door policy.

DISCUSSION

This cohort illustrates the broad aetiology of hydrocephalus. Many affected have other medical problems and most are looked after by a multidisciplinary team. As a result, the paediatrician working in the district general hospital setting is not infrequently faced with the question "Is this shunt blocked?".

The results of this study indicate that the presence of headache, vomiting, and drowsiness together make it very likely that an affected patient has shunt dysfunction; in most cases this will mean acute shunt block. A minority will have an infected shunt, though in the absence of fever this study

shows there is little to easily clinically distinguish between shunt infection and block. The calculated odds ratios for symptomatology illustrate the striking positive relation between drowsiness and acute shunt block. It is this sign in particular that should prompt urgent neurosurgical referral. Headache or vomiting in isolation is less predictive of acute block, and though one should adopt a low threshold for seeking a neurosurgical opinion, a careful search for an alternative diagnosis is warranted. Recent reported figures[6 11] for symptomatology in proven shunt malfunction (which includes infection in some cases) are headache (47–55% of cases), vomiting (40–90%), and drowsiness (30–60%). Relatively comparable figures from this study, which focuses on shunt block as opposed to all causes of malfunction, are headache 74%, vomiting 73%, and drowsiness 73%.

We are realistic that eliciting a history in this setting may sometimes prove difficult[5 9 10] and justify early neurosurgical advice, but the results of this study are encouraging, with a low false positive rate (20%) of referral from district general hospitals. Furthermore, they are in keeping with previous published studies.[11] This includes the recorded 5% incidence of overdrainage symptoms,[5] a condition probably underestimated by hospital admission rates.

During the time period of this study, in a minority of cases (n = 4), an incorrect diagnosis of presumed normal shunt function was made when each initially presented. In none of the four on first presentation was drowsiness a presenting feature. Following discharge each later re-presented during the study period with a different symptom profile, which included drowsiness in three cases. All four were found to have operatively proven shunt block. In each no discernible neurological morbidity was apparent as a consequence. The "recalculated" odds ratios show a near identical trend in symptom significance, and again highlight drowsiness as an important positive predictive clinical sign of shunt block.

The limitations of performing CT scanning without the facility to compare with previous images are well illustrated, as are the pitfalls of over reliance on scan findings. However, the observation that in 16% of admissions the CT scans were unhelpful should not detract from the fact that in 84% the diagnosis was effectively confirmed by an increase in ventricle size when compared with the most recent previous CT examination. Given the importance of arriving swiftly at an accurate diagnosis, we would recommend that all children have a baseline CT study performed a few weeks after either their initial shunt insertion or a shunt revision. Paediatric units with the facility to carry out scans on children with suspected shunt blockage should hold copies of these images, as should, of course, the regional neurosurgical unit. In some situations, particularly when a family is moving from place to place, it may be sensible for copies to also be held by the child's carers. This may prove an invaluable arrangement for those (often nocturnal) situations when the radiology department is unable to locate a patient's previous studies.

The importance of paying careful attention to the observations of the child's parents and other carers, particularly if they have had experience of shunt block in the past, cannot be overemphasised. In a previous study[11] (which excluded children who had had recent shunt problems) we showed that families were at least as accurate as paediatricians in diagnosing shunt block. Indeed, it is the neurosurgeons who may be reviewing a child perhaps only once a year (or who may have delegated follow up completely to a local paediatric department[12]), who may have the least knowledge of the child's "regular" state of health, complicated by a variety of disabilities.

Key messages

- Always suspect shunt malfunction in a child with a shunt and no alternative convincing explanation for their symptoms
- Be particularly suspicious if drowsiness is a prominent symptom in shunted patients
- In trying to interpret a child's presenting symptoms, always defer to the experience of the family if the child has had previous shunt block
- When in doubt always discuss with a tertiary centre
- All patients should have a CT scan performed as a routine at some stage after a primary shunt insertion or revision procedure, and a copy held at not only the tertiary centre but also at the local district general hospital if it has a CT scanning facility
- An increase in ventricle size is highly suggestive of shunt blockage, but no change, particularly if the ventricles are slit like, does not rule it out

Conclusions

It is well documented that the presentation of acute shunt block is heterogeneous. Furthermore, there is a significant morbidity and mortality in late diagnosis. In conclusion, this study illustrates the importance of considering shunt malfunction in a child with a shunt and no alternative convincing explanation for their symptoms, particularly if drowsiness is a prominent feature. In trying to interpret a child's presenting symptoms, always defer to the experience of the family if the child has had previous shunt block, and if in doubt, always discuss with a tertiary centre. An increase in ventricle size is highly suggestive of shunt blockage, but no change in size, particularly if the ventricles are slit like, does not rule it out.

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