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Pitfalls in the Diagnosis of Ventricular Shunt Dysfunction: Radiology Reports and Ventricular Size

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ABSTRACT. ***Introduction.*** **The diagnosis of shunt malfunction can be difficult even for the experienced clinician and may lead to disastrous circumstances when misinterpreted. Less experienced physicians may rely more on radiographic reports as a primary diagnostic modality. In this study, we evaluated the reliability of using these reports without accurate clinical assessment.**

Methods. **All shunt revisions seen at Children's Hospital (Birmingham, AL) between January 1996 and August 1996 were reviewed, excluding patients with brain tumors, supratentorial extraaxial fluid collections, and infections. Sixty-eight patients underwent 100 operations for shunt malfunction. All patients had evidence of shunt blockage, disconnection, catheter malposition, or valve pressure incompatibility. The prospective radiographic interpretation of preoperative computed tomography and magnetic resonance imaging scans was reviewed in each case.**

Results. **Twenty-four percent of the reports made no mention of shunt malfunction. In this group, the ventricular system was described as "unchanged," "stable," "normal," "unremarkable," "small," "smaller," "slit," "negative," and "no hydrocephalus," with no other comment to support a diagnosis of shunt malfunction. An additional 9% of reports contained the same terms, while also hinting at some other clinical or radiographic data that suggest the possibility of shunt failure (eg, a shunt disconnection seen on plain radiographs), despite the scan findings. In all patients in this group, symptoms improved after surgery.**

Conclusion. **We conclude that as many as one third of patients presenting with shunt malfunction will not have the diagnosis of shunt malfunction supported by a prospective radiologic interpretation of brain imaging. Although the neurosurgical community can assess the clinical situation to determine the need for surgery, other clinicians can be easily reassured by a radiographic report that does not mention or diagnose shunt malfunction. Today, more than ever, nonneurosurgeons are being called on to evaluate complex clinical situations and may rely on radiographic reports.** ***Pediatrics*** **1998;101:1031–1036;** ***hydrocephalus, ventricle size, radiology reports, ventricular shunt.***

ABBREVIATIONS. CT, computed tomography; MRI, magnetic resonance imaging; CSF, cerebrospinal fluid; ICP, intracranial pressure.

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Shunt malfunction is one of the most common clinical problems in pediatric neurosurgery. The diagnosis can be both difficult and perplexing even for the experienced clinician. With the advent of managed care, primary care physicians are being asked with more frequency to evaluate patients with possible shunt malfunction, a task typically reserved for the neurosurgeon. Because of their relative inexperience in reading cranial imaging and managing shunt patients, generalists tend to rely heavily on radiology reports in their assessments. In this study, we objectively evaluate the reliability of these reports in diagnosing shunt dysfunction.

METHODS

One hundred consecutive shunt revisions at the Children's Hospital in Birmingham, AL, were reviewed retrospectively. These occurred in 68 patients between January 1996 and August 1996. In each case, the medical records and radiographic studies including computed tomography (CT) and magnetic resonance imaging (MRI) scans were examined. The prospective (preoperative) interpretation of the CT and MRI studies in the radiology reports were then noted. These reports were generated primarily by pediatric radiologists in a busy children's hospital. Patients with associated disorders that may have caused symptoms independently from the hydrocephalus were excluded from the study in order not to confuse our diagnostic criteria. These included patients with brain tumors, shunt infections, and supratentorial extraaxial fluid collections. One other patient was excluded because of a negative surgical finding (no shunt malfunction was demonstrated). In all patients, therefore, a diagnosis of shunt failure was determined intraoperatively. The preoperative imaging reports were evaluated for the presence of information that may have inferred incorrectly that the shunt had not failed.

RESULTS

Age and Etiology of Hydrocephalus

The patients ranged from 1 month to 26 years of age (mean, 6 years; median, 4 years). There were minimal differences in the distributions of age and etiology of hydrocephalus between the subset of patients with misleading reports and the rest of the patients (patients whose diagnosis of shunt failure was obvious from the outset) (Figs 1, 2). An important observation, however, is that the Dandy Walker and miscellaneous groups are somewhat overrepresented in the category of misleading reports. This may be explained partially by the fact that multiple compartment hydrocephalus is more difficult to evaluate than simple hydrocephalus. In the latter all ventricles tend to dilate proportionally, whereas in the former the ventricles do not communicate freely, and more than one shunt catheter is frequently needed. A typical example is that of the Dandy

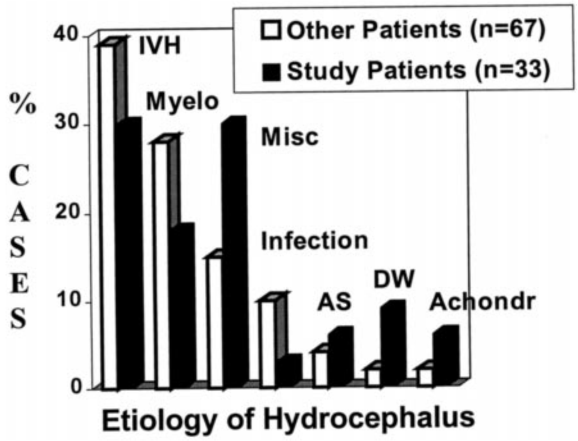

Fig 1. Graph comparing the etiology of hydrocephalus in the study group and the rest of the patients.

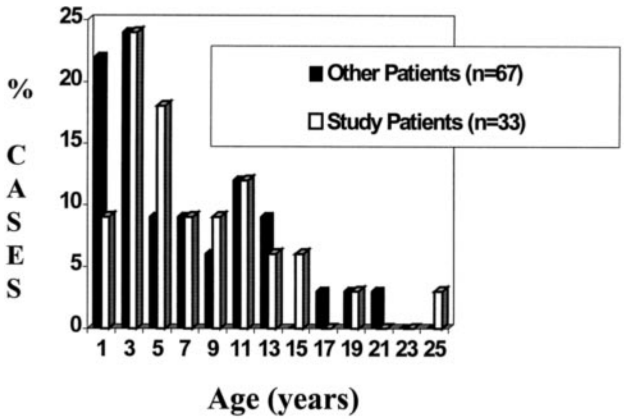

Fig 2. Graph comparing the age distributions of the study group (the 33 shunt revisions with misleading reports) and the rest of the patients (67 revisions).

Fig 3. T1-weighted sagittal MRI of the cervical spine showing a syrinx. The head CT scan revealed "small" ventricles. The syrinx and symptoms improved after the ventriculoperitoneal shunt was revised.

Walker patient whose posterior fossa shunt fails and the supratentorial shunt remains functional. In any case, hydrocephalus of different etiologies may have different hydrodynamic considerations, thus the added complexity in diagnosing shunt failure.

Incidence of Misleading Radiology Reports

In 24 of 100 cases, the imaging reports included terms describing the scan or the size of the ventricular system, which implied that the shunt functioned properly: "unchanged," "stable," "normal," "unremarkable," "small," "smaller," "slit," "negative," and "no hydrocephalus." In an additional 9 cases, the same terms were used along with other statements indicating that the possibility of shunt failure still existed despite the unremarkable ventricles. This often was attributable to two factors: the radiologists at this institution are aware that small ventricles still could be consistent with shunt failure, and they discuss most cases with the neurosurgeons for clinical correlation. These statements included the following: 1) in 3 cases, an enlarged syrinx (Fig 3) was noted on a cervical MRI scan (one of these extended into a syringobulbia seen on the initial brain MRI scan). The radiologist concluded that the syrinx may be a manifestation of shunt failure despite the stable or unremarkable ventricles; 2) 1 patient had a shunt disconnection (Fig 4) seen on plain radiographs (shunt series); 3) subgaleal fluid was noted along the shunt tract on the CT scan of 2 patients, indicating shunt failure (Fig 5); 4) in 1 other case, the radiologist noted that the comparison scan (in which the ventricles were "unchanged") was obtained at a time when there was shunt malfunction; 5) the radiologist noted in the report that after discussion with the neurosurgeon, shunt malfunction could not be ruled out despite the "stability" of the scan; and 6) another report noted that although the ventricles were small, one cannot rule out slit ventricle syndrome. This patient was seen to have a shunt disconnection on plain radiographs that were not seen initially by the radiologist. Therefore, of 100 cases with surgically or radiographically proven shunt failure, 33 had radiology reports that were potentially misleading.

Finally, in at least 11 of the 100 cases, or one third of the 33 cases with misleading reports, the ventricles were noted to be small by both the radiologist and the neurosurgeon. Therefore, the diagnosis of shunt malfunction in these cases could not be made from the CT scan findings.

Fig 4. Head CT scan showing small ventricles. Note the subgaleal fluid collection (arrow), which indicates in this case that the shunt was not working properly.

Diagnosis

Despite the misleading reports, the clinical suspicion of shunt failure in these 33 cases (30 patients) was enough to necessitate operative exploration. These diagnoses were made as follows: most patients (21 cases) had symptoms reminiscent of previous shunt dysfunction. In addition, there were 4 cases of subgaleal fluid accumulation, 2 cases of increasing head circumference, 1 case of cerebrospinal fluid (CSF) leak, 3 cases of shunt disconnection seen on plain radiographs, and 3 cases of an enlarging syrinx seen on MRI scan. In instances in which the diagnosis was uncertain, a shunt patency (nuclear medicine) study (3 cases) or Camino continuous intracranial pressure (ICP) monitoring (2 cases) demonstrated evidence of shunt failure or raised intracranial pressure. Finally, in another 7 cases, the neurosurgeon added additional information to the radiologist's report, showing shunt failure: 1) in 2 patients, a shunt disconnection was seen on plain radiographs read by a different radiologist; 2) the neurosurgeon noted increased mass effect from a suprasellar cyst in a shunted patient; once the shunt was revised, the mass effect improved and the symptoms resolved; 3) in 4 cases, the neurosurgeon found other previous scans with smaller ventricles; the radiologist had reviewed only the previous scans with larger or same-size ventricles.

Intraoperative Findings

Intraoperative findings helped confirm the diagnosis of shunt malfunction in 28 of the 33 patients with misleading reports. There were 19 obstructions (8 proximal, 6 distal, and 5 combined proximal and distal obstructions); five shunt disconnections or fractures, two of which were not evident on plain radiographs, even retrospectively; and one case of lack of abdominal absorption of CSF, one overdrainage, and two catheter migrations (one catheter migrated backward into the subcutaneous tissues, and the other migrated back into the pleural space, demonstrating that the tubing originally had been tunneled under a rib). There were two cases of valve pressure incompatibility.

Postoperative Improvement

In 25 of the 29 symptomatic cases (patients with subgaleal collections and CSF leaks were considered symptomatic), there was complete resolution of symptoms after shunt revision; in the other 4 cases, the symptoms and signs improved; all 4 had shown evidence of shunt dysfunction intraoperatively.

The Table summarizes the findings in all 33 cases, including the terms and conclusions used in the imaging reports, the interpretation of the scan by the neurosurgeon, the presenting clinical findings, and how a final diagnosis was made.

DISCUSSION AND RECOMMENDATIONS

Missing the diagnosis of shunt malfunction may lead to permanent neurologic injury or death. In this study, we have attempted to evaluate the re-

Fig 5. Skull radiograph in a patient with "unchanged" ventricles on head CT scan. This patient has lateral as well as fourth ventricular catheters. Note the shunt disconnection distal to the three-way connector.

TABLE. Summary of Presentations and Results Among the Shunt Failure Patients Who Had Potentially Misleading Radiology Reports (33 Revisions in 30 Patients)

Patient	Disease	Reason for Evaluation	Diagnosis Made By	Cause of Failure	CT: Ventricle Size, by Radiologist	by Surgeon
1	Intraventricular hemorrhage	Fractured shunt	Plain radiographs	Fractured shunt	Unchanged	Large
2	Other	Ventricular enlargement	Comparison with old CT	Unknown	Unchanged from failure: large	Larger
3	Myelomeningocele	Nausea/vomiting and subgaleal collection	Subgaleal fluid	Distal malfunction	Smaller; subgaleal fluid	Large
4	Encephalocele	Enlarging head circumference/ bulging fontanel	CT; surgical exploration	Proximal obstruction	Unchanged; large; cannot rule out dysfunction	Unchanged; large
5	Intraventricular hemorrhage	Headache, nausea/ vomiting, lethargy	Surgical exploration	Proximal/distal obstruction	Unchanged	smaller; tiny
6	Intraventricular hemorrhage	Headache	Surgical exploration	Proximal obstruction	Unchanged	Small
7	Intraventricular hemorrhage	Headache	Continuous ICP monitoring	Proximal obstruction	Unchanged; small	Unchanged; small
8	Intraventricular hemorrhage	Subgaleal collection	Subgaleal collection	Proximal obstruction	Smaller; mild dilatation	Smaller
9	Myelomeningocele	Fatigue, speech problems, neck pain	CT; surgical exploration	Proximal/distal obstruction	Stable; large; no malfunction	Large
10	Other	Enlarged ventricles	CT; surgical exploration	Proximal obstruction	Smaller, but still large	Large
11	Intraventricular hemorrhage	Headache, nausea/ vomiting	Continuous ICP monitoring	Overdrainage	Unchanged; large occipital horns	Large
12	Meningitis	Headache, nausea/ vomiting	CT; surgical exploration	Proximal/distal obstruction	Normal; no malfunction	Large
13	Cyst	Increasing head circumference	Plain x-rays; compared with old CT	Shunt fracture	Stable; large	Larger
14	Aqueductal stenosis	Moderate ventricular dilatation	CT; surgical exploration	Distal malfunction	Unchanged	Large
15	Myelomeningocele	Nausea/vomiting, respiratory distress	Plain radiographs	Displaced catheter	Unchanged; large; unlikely failed	Large
16	Intraventricular hemorrhage	Headache	Surgical exploration	Proximal obstruction	Slit; stable; previously large	Slit
17	Other	Nausea/vomiting, seizures	CT; surgical exploration	Unknown	Smaller	Large
18	Dandy Walker	Subgaleal collection	Subgaleal collection	Fractured shunt	Smaller; mild dilatation	Large
19	Dandy Walker	Subgaleal collection	Subgaleal collection	Displaced catheter	Small; subgaleal fluid	Larger
20	Dandy Walker	CSF leak	CSF leak	Valve malfunction	Slit	Slit
21	Intraventricular hemorrhage	Headache, nausea/ vomiting, lethargy	Plain radiographs	Shunt disconnection	Stable; can't r/o slit vent syndrome	Small
22	Myelomeningocele	Headache, lethargy, parinaud's	MRI	Shunt disconnection	Small; syrinx	Small
23	Achondroplasia	Headache, nausea/ vomiting	Surgical exploration	Unknown	Unchanged; small	Small
24	Achondroplasia	Headache, nausea/ vomiting, lethargy	Comparison with old CT	Proximal/valve obstruction	Normal	Larger
25	Cyst	Headache, ataxia	MRI	Valve malfunction	Unchanged; stable; large	Large
26	Trauma	Headache, papilledema	Papilledema	Distal malfunction	Normal	Normal
27	Other	Headache, nausea/ vomiting, lethargy	Shunt tap	Proximal obstruction	Slit	Slit
28	Intraventricular hemorrhage	Headache, nausea/ vomiting, papilledema	Papilledema	Proximal/distal obstruction	Small	Small
29	Intraventricular hemorrhage	Headache, nausea/ vomiting	Nuclear medicine study	Valve incompatibility	Smaller; no recent films	Large
30	Myelomeningocele	Headache, back pain, clumsiness	Nuclear medicine study	Valve incompatibility	Unchanged; large; syrinx larger	Unknown
31	Trauma	Worsening esotropia	Surgical exploration	Valve/distal obstruction	Stable	Large
32	Cyst	Cyst mass effect on MRI	MRI scan	Distal obstruction; cyst	Stable; large	Large; suprasellar cyst
33	Myelomeningocele	Syrinx	MRI; nuclear medicine study	Proximal obstruction	Unremarkable; large syrinx	Small; syrinx

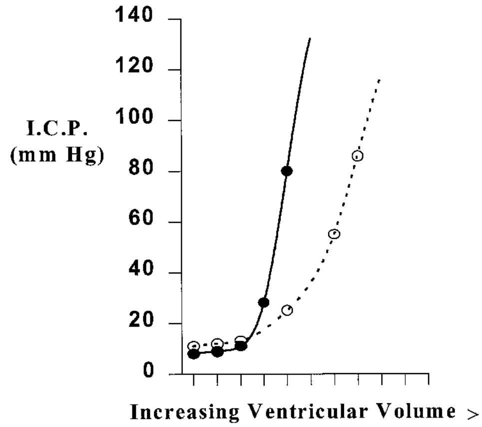

Fig 6. Schematic diagram of a hypothetic relationship between ICP and ventricular volume. The ICP changes minimally as the ventricle enlarges, until the ventricular volume reaches a certain point, after which the ICP shows a rapid increase. The dotted curve represents a patient with increased compliance, causing a shifting of the curve to the right.

liability of radiology reports in the assessment of shunt function to minimize the chance of such misdiagnoses. The results support several important conclusions:

1. Approximately 11% of shunt failures present with brain imaging studies showing small ventricles, therefore, CT and MRI scans should not be used as the only or definitive diagnostic modality when evaluating shunt function. Causes of small ventricles despite shunt failure include poor compliance, overdrainage, slit ventricle syndrome, intermittent shunt malfunction, and the possibility that the ventricles may have been even smaller in the past (therefore, the current ventricles actually are enlarged compared with the previous study). The presence of shunt dysfunction with normal ventricular size is well documented in the literature.[1–4] The phenomenon of poor compliance of the ventricles may occur after chronic shunting and is probably, in part, a reflection of changes in the biomechanical and biological properties of the brain and ependyma.[3,5] In at least one study, patients who tended to suffer from acute deteriorations in neurologic function had noncompliant pressure-volume curves, as opposed to patients with subtle (usually intellectual, social, or developmental) deteriorations, who had compliant (normal) curves and ventricles that enlarged routinely after shunt failure. In the latter group, deteriorations occurred only after the intracranial pressure rose to the steep portion of the normal curve (Fig 6).[3,6]
2. When a scan shows large ventricles, an effort should be made to find out whether the ventricles have ever been smaller in size, thus usually implying that the current shunt has failed. (This is not always the case, however. For instance, in cases of slit ventricle syndrome, larger ventricles are preferable.) All previous scans should be reviewed and compared with the current scan. Furthermore, it would be important to figure out which of these scans was obtained at a time when the shunt was not functional. Finally, even if the ventricles had never changed in size, the presence of large ventricles still should raise the suspicion for shunt failure. In the general radiology literature, the need for comparisons with previous radiologic studies and the extent of descriptive detail in the radiology reports have been debated.[7,8] The seriousness with which such issues should be taken in evaluating shunts cannot be overstated.
3. The presence of disorders that shunt the CSF away from the ventricular system, thus keeping the ventricle size small, should be recognized. These disorders include CSF leak from one of the skin incisions, subgaleal collections of CSF, and the formation or enlargement of a syringohydromyelia.
4. In all of the cases in this study, the appropriate diagnosis of shunt dysfunction was made, despite a "stable" scan finding, because the neurosurgeon had sufficient clinical suspicion to pursue other diagnostic avenues. In addition, the radiologists at our institution often discuss their cases with the neurosurgeons, recognizing that scans may not always be reliable in diagnosing malfunction. Excluding shunt malfunction can be difficult even for experienced neurosurgeons. Additional studies often are obtained by the neurosurgeon to solidify a diagnosis. Such studies include shunt taps, intracranial pressure monitoring, shunt patency studies, long periods of observation in the hospital, and even an occasional surgical exploration. When the shunt evaluation is performed by pediatricians and emergency room physicians, the rate of misdiagnosis has the potential of being even greater. A primary care physician without specialized training is less able to read brain scans and less experienced in managing children with shunts. Nonspecialists will tend to rely more on the radiologist's report for their assessment, and thus risk missing the diagnosis and sending the patient home with a tenuous situation. The consequences in this case may be disastrous.
5. Finally, it is important to note that the final determination of shunt function in our patient population has been the surgeon's intraoperative evaluation. The design of this study is open to criticism because there was no independent confirmation of the findings by nonneurosurgeons. Although this fault is inherent to the design, there was no obvious practical alternative that could have been pursued in this retrospective evaluation. In support of the neurosurgeon's assessment was the improved clinical course of the patients after the vast majority of revisions, and the fact that intraoperative evaluation of shunt function often is relatively straightforward.

Physicians who may be called on to evaluate shunts are required to appreciate the complexity and difficulties of managing shunt patients and seek early neurosurgical assistance. Currently, even under the best circumstances (ie, in a children's hospital with a busy and competent radiology department), radiology reports may be misleading.

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