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Radiographic evaluation for suspected cerebrospinal fluid shunt obstruction

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Objective: To measure the predictive value of plain radiographs (shunt series) and computed tomography (CT) scans in a group of children undergoing evaluation for suspected shunt obstruction in a tertiary care pediatric emergency department (ED).

Methods: Radiology reports were reviewed for all ED patients who underwent a shunt series over an 18-month period. Two investigators categorized all reports as normal, possibly abnormal (eg, kink in shunt tubing, no prior CT scan for comparison), or abnormal (with definite evidence of shunt dysfunction, such as shunt tubing disconnection and increase in ventricular size since prior CT scan). Studies for which there was disagreement were re-read independently by a pediatric radiologist. Medical records were reviewed to determine outcomes.

Results: A total of 233 patients had shunt series and CT scans ordered. Of these, 60 patients subsequently required surgery for shunt obstruction. The shunt series revealed abnormalities in 12 patients (sensitivity, 20%; negative predictive value, 22%), whereas CT scans showed definite or possible abnormalities in 50 patients (sensitivity, 83%; negative predictive value, 93%). Combined, the two tests detected 53 shunt obstructions (sensitivity, 88%; negative predictive value, 95%). Two obstructed patients had abnormalities on shunt series that would not have been suspected after physical examination or CT scan.

Conclusions: Over one quarter of pediatric ED patients evaluated radiographically for suspected shunt obstruction required surgical management. One in eight obstructed patients had normal radiographic studies. Routine performance of shunt series had a low overall yield but on rare occasions detected abnormalities that were missed by CT. Prospective studies are needed to improve the use of radiographic tests for shunt evaluation and determine clinical indications for further workup when studies are normal.

INTRODUCTION

The evaluation of a child with a suspected cerebrospinal fluid shunt obstruction is a complex and increasingly prevalent clinical

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Address for reprints: Joseph J. Zorc, MD, Division of Emergency Medicine, The Children's Hospital of Philadelphia, 34th Street and Civic Center Boulevard, Philadelphia, PA 19104-4399; e-mail: zorc@email.chop.edu **Key Words:** Cerebrospinal fluid shunt, hydrocephalus, radiography

problem for providers of pediatric emergency care. Since its introduction in the 1950s, cerebrospinal fluid shunting for hydrocephalus has become the most common neurosurgical procedure, with pediatric conditions such as meningomyelocele and intraventricular hemorrhage among the leading indications (1, 2). Although shunting has dramatically improved mortality for these diagnoses, complications are frequent; one longitudinal study found that 80% of shunts required revision within 12 years of placement (3). The symptoms of shunt obstruction, such as headache and vomiting, are nonspecific and difficult to distinguish from common, benign pediatric illnesses. Left untreated, however, a shunt obstruction may progress rapidly to a life-threatening elevation in intracranial pressure. As a result, clinicians typically have a low threshold to perform diagnostic tests to evaluate for shunt malfunction.

Although diagnostic tests to assess shunt function are routinely performed at tertiary care medical centers, little evidence is available to guide clinicians in their interpretation. Physical examination by pumping the shunt bulb reservoir has been shown to be unreliable (4), and directly measuring the pressure by "shunt tap" risks the introduction of infection (5). Radiographic tests that are recommended include computed tomography (CT) scan of the head and plain films of the shunt catheter (shunt series) (6, 7). The CT scan is compared with prior scans to detect an increase in the size of the ventricles or other findings that suggest elevated intracranial pressure. The shunt series is used to detect disruptions in the integrity of the shunt catheter. Several small case series have suggested that CT scan lacks sensitivity in detecting shunt obstruction (8). None of these studies looked at the prevalence of obstruction in emergency department (ED) patients. In addition, we are not aware of any studies that assess the predictive value of routine shunt series or its additive value to CT scan. In this study we sought to measure the predictive value of shunt series and CT scan in a group of children undergoing ED evaluation for suspected shunt obstruction.

METHODS

A computerized radiology department database was used to identify all shunt series ordered for patients at a tertiary care children's hospital over an 18-month period between January 1995 and June 1996. A shunt series is routinely obtained as part of neurosurgical evaluation at this institution. Analysis was limited to patients who were evaluated in the ED on the day the test was ordered. A sample size of at least 225 patients was chosen to provide an estimate of sensitivity with a 95% confidence interval of at most $\pm 10\%$, assuming 25% of patients had obstructions, and a predicted sensitivity of 90%.

Radiographic reports for shunt series and accompanying CT scans were obtained for each patient. Two investigators (J.J.Z. and S.D.K.)

reviewed and categorized each report. Based on a pilot review, shunt series were divided into seven categories: normal, shunt disconnection, shunt tip retracted out of abdomen, proximal discontinuity at shunt bulb, kink in shunt tubing, no tip movement from prior examination (which would suggest a possible loculation within the abdominal cavity), or other abnormality. CT scans were divided into four categories: unchanged from prior CT scan, ventricles increased from prior CT scan, no comparison CT scan available, or possible shunt dysfunction (mild changes in ventricular shape or size). The reliability of categorization was compared between the two investigators using the κ statistic. Studies for which there was disagreement were reviewed independently by a pediatric radiologist (J.B.).

Operative notes and discharge summaries were reviewed to determine clinical outcomes. A shunt obstruction was defined based on the requirement for shunt revision within 2 weeks of ED evaluation. Surgical procedures for reasons other than obstruction, such as infection without obstruction or revision of wound, were excluded.

For each radiographic test and for each combination of tests, a twoby-two table was created based on the result of the test and the clinical outcome. Sensitivity, specificity, the predictive value of negative and positive tests, and 95% confidence intervals were determined. In addition, the likelihood ratio for positive and negative tests was calculated. The study was approved by the Institutional Review Board.

RESULTS

A total of 254 shunt series were ordered for 140 children evaluated in the pediatric ED during the 18 months reviewed. A group of 31 children who had three or more shunt series ordered during the study period accounted for almost half (125) of the studies. The mean age at the time of evaluation was $5.4~(\pm 3.9)$ years, with a range of 0 to 14 years. The majority (64%) of patients were male. Racial distribution was 63% white, 35% African American, and 2% other. For 115 of the studies, the radiology report included one or more clinical indications; the most common indications were vomiting (52%), lethargy (36%), headache (30%), and fever (25%). The types of shunts studied were: ventriculoperitoneal (89%), ventriculopleural (5%), ventriculoatrial (5%), or more than one type (1%). Most patients (74%) had a single shunt; 22% had two shunts, and 4% had three.

When radiographic reports were categorized independently by two investigators, interobserver agreement was high for both shunt series (88% overall agreement, $\kappa=0.6$) and CT scans (89% overall agreement, $\kappa=0.8$). Studies that caused disagreement were

reread and categorized independently by a pediatric radiologist. Three shunt series were excluded because of incomplete examinations or missing films; based on medical record review, none of these patients had abnormalities that we would have expected to see on plain radiographs. Eighteen patients were excluded because a CT scan had not been ordered at the time of the evaluation. Of these, four had documentation in the medical record of an abnormal CT scan from an outside institution and went on to revision of a proximal shunt obstruction. The remaining 14 had no record of a CT scan; of these, only one had an abnormality on the shunt series that was consistent with shunt obstruction (ie, retraction of the distal catheter due to somatic growth of the patient).

Results of radiographic studies and clinical outcomes for the 233 patients with both shunt series and CT scans are summarized in Table 1. One hundred twenty-three (53%) of the patients were admitted to the hospital after the ED evaluation. A total of 60 patients required surgery for shunt obstruction; three of these patients had initially been discharged from the ED but returned within the following 2 days for elective (1) or emergent (2) surgery. Patients who required surgery for shunt infection without obstruction (9) or for revision of a wound (3) were not included in this total. Clinical data at about the time of the most recent shunt revision were available for 45 obstructed patients; 21 (47%) had been revised within the previous 6 months, and 17 (38%) had been revised within the previous 3 months. A history of head trauma was noted in only one obstructed patient.

Of the 233 shunt series analyzed, abnormalities were reported in 38 cases; 12 of these were subsequently found to be related to shunt obstruction. Among the abnormalities reported, six showed a disconnection of the distal catheter that required subsequent revision in the operating room; one other patient had an apparent disconnection radiographically but on physical examination was found to have a long section of non-radio-opaque shunt tubing. Another five shunt series showed evidence of retraction of the distal catheter. Two of these patients were school-aged children with ventriculoperitoneal shunts that were placed in infancy and now required revision because of the insufficient length of the distal catheter. A third patient had a ventriculopleural shunt that had retracted out of the pleural space, producing a subcutaneous fluid collection. The fourth patient was a 5-year-old child with achondroplasia and a ventriculoatrial shunt that terminated high in the chest, proximal to the atrium. Although the position was unchanged from prior films, a high clinical suspicion led to a shunt tap that revealed a distal obstruction. The fifth patient had

TABLE 1
Results of shunt series and head computed tomography scan and clinical outcomes

Radiographic results	Clinical outcome				
	Obstruction $(n = 60)$	No obstruction ($n = 173$)	Sensitivity	Likelihood ratio	
Shunt series					
Findings associated with obstruction					
Disconnection of distal catheter	6	1	10%	17	
Retraction of distal catheter tip	4	1	6%	5.5	
Discontinuity near shunt bulb	2	2	3%	2.8	
Any abnormality	12	4	20%	8.6	
Findings not associated with obstruction					
Kink or coil in shunt tubing	1	7	3%	0.82	
No tip movement from prior exam	2	12	3%	0.48	
Head CT scan					
Increased ventricles since prior CT scan	29	8	48%	10.1	
Possible shunt dysfunction	4	6	6%	1.9	
No prior comparison CT scan	17	27	28%	1.8	
Any abnormality	50	41	83%	3.5	

CT = computed tomography.

undergone a revision from a ventriculoperitoneal to a ventriculopleural shunt; this gave the radiographic appearance of a retracted distal catheter, although no shunt abnormality was present. Another four shunt series showed possible gaps at the beginning of the distal catheter where the tubing is often non–radio-opaque. Of these, one required revision for a disconnection that was also evident on physical examination and was due to a large fluid collection near the shunt bulb. The other three patients had no disconnection, although one required revision for an unrelated obstruction of the proximal catheter. The remaining 22 abnormalities on shunt series were unrelated to shunt malfunction. One patient with a kink in the shunt tubing and two with an absence of movement of the shunt tip required surgery for a proximal shunt obstruction that was unrelated to the radiographic findings.

Of the 233 head CT scans analyzed, 91 had abnormalities reported. Of these, 37 had evidence of increasing ventricular size from a previous examination; 29 of these patients subsequently underwent surgery for shunt obstruction. Another 10 patients had possible abnormalities reported, such as mild changes in the size or shape of the ventricles; four of these patients had shunt obstructions. Finally, 44 patients had no prior CT scan available for comparison; 17 of these patients subsequently required surgery for shunt obstruction.

Sensitivity, specificity, predictive value, and likelihood ratios for shunt series and head CT scan are shown in Table 2. For shunt series, we defined as positive those specific findings that were associated with an increased likelihood of shunt obstruction (eg, disconnection, retraction of the shunt tip, discontinuity near the shunt bulb). For head CT scan, the scan was considered positive if any abnormality was mentioned or if the radiologist noted that the reading was limited by the absence of a prior comparison CT scan. Shunt series had a low overall sensitivity, identifying 12 of the 60 patients with shunt malfunction (20%). If the shunt series was negative, the likelihood of obstruction was not greatly reduced: from 26% prior to the test to 22% afterwards; this corresponds to a likelihood ratio for a negative test of 0.82. Head CT scan identified 50 of the 60 patients with obstruction (sensitivity, 83%). The specificity of head CT scan (76%) was lower than that of shunt series (98%), largely because of the effect of the 44 patients with no prior comparison CT scan, 27 of whom did not have shunt obstruction. If these studies had been categorized as normal, specificity would have increased to 92%, although sensitivity would have decreased to 55%. When the head CT scan was negative, the probability of obstruction was reduced to 7% (likelihood ratio for a negative test, 0.21).

To assess the value of routinely performing the shunt series, we

TABLE 2
Predictive value of shunt series and computed tomography scan of the head

	Shunt series	Head CT scan	Shunt series or CT scan
True positive	12	50	53
False positive	4	41	44
True negative	169	132	129
False negative	48	10	7
Sensitivity (%[95% CI])	20 [11, 32]	83 [71, 92]	88 [77, 95]
Specificity (%)	98 [94, 99]	76 [69, 82]	74 [67, 81]
Predictive value			
Positive test (%)	75 [48, 93]	55 [44, 65]	55 [44, 65]
Negative test (%)	78 [72, 83]	93 [87, 96]	95 [90, 98]
Likelihood ratio			
Positive test	8.6	3.5	3.5
Negative test	0.82	0.21	0.16

compared the results when both tests were combined with the results found by CT scan alone. Fifty-three of the 60 obstructed patients had a radiographic abnormality on one or the other test, resulting in an overall sensitivity of 88%. Three patients with obstruction had an abnormal shunt series in the setting of a normal CT scan; one of these patients had a disconnection near the shunt bulb that was heralded by a large subcutaneous fluid collection. The second patient had a subtle disconnection of a ventriculopleural shunt that was missed when the radiographs were initially reviewed in the ED. This patient was discharged but returned with increasing symptoms several hours later. The correct diagnosis was then made on formal review of the films, and the patient went on to shunt revision without complications. The third patient was the child with achondroplasia and retraction of a ventriculoatrial shunt as described previously.

DISCUSSION

The objective of this study was to determine the predictive value of radiographic studies in a group of children undergoing ED evaluation for suspected shunt obstruction. Overall, several conclusions can be drawn from the results. First, a substantial proportion, over one quarter of those evaluated, required a shunt revision. Current teaching, as reflected in textbooks and review articles, has recommended a high clinical index of suspicion when assessing children with possible shunt malfunction (6, 7). The high prevalence of obstruction observed in our study would support aggressive evaluation when these patients present to a tertiary care ED with suggestive symptoms.

The limitations of radiographic evaluation for suspected shunt obstruction are consistent with the few previously published studies in this area. In one of the original studies using CT scan to assess shunt function, Murtagh et al. (9) noted four patients in whom the ventricles were noncompliant and failed to dilate under conditions of increased intracranial pressure. The estimates of sensitivity of CT scan reported in this and a subsequent study have ranged from 64 to 92% (9, 10). Iskandar et al. (8) recently published a case series of 100 consecutive shunt revisions at Children's Hospital in Birmingham, Alabama. For 24 of these patients, the radiology report made no mention of shunt malfunction on the imaging scan (CT scan or magnetic resonance imaging), for an overall sensitivity of 76%. The investigators argued that a lack of familiarity with shunt malfunction may have contributed to the high rate of misleading radiology reports. For example, in four cases, the neurosurgeon found old scans that showed smaller ventricles that had been missed by the radiologist. In our study, the radiographs were reviewed by neuroradiologists, who were careful to note the absence of a comparison CT scan in their interpretation. Of the 27 scans that were categorized as possibly abnormal because there was no prior comparison film, 17 were eventually diagnosed with shunt obstruction. Clearly, a comparison CT scan is a prerequisite for radiographic evaluation to be considered truly negative. However, even when a comparison scan is available, the overall sensitivity of radiographic studies is not sufficient to rule out obstruction. Because we included nonobstructed as well as obstructed patients in our study, we are able to calculate a negative predictive value of 95% for radiographic studies in this patient population.

None of the previous studies looked systematically at the use of routine plain radiographic shunt series in patients with suspected shunt malfunction. Iskandar et al. (8) reported that three of 100 obstructed patients had abnormalities on shunt series; two of these had normal CT scans. In our study, a shunt series was performed on all 233 patients with suspected obstruction, and 13 had abnormalities.

(Three of these were subsequently found to be unrelated to the cause of the malfunction.) All but two of these abnormalities would have been suspected by a fluid collection around the shunt or increased ventricles on CT scan. Therefore, the yield of routine shunt series in detecting unsuspected abnormalities was low, 0.8% (95% CI, 0.1–3%). Because shunt obstruction has a high potential morbidity, one could argue that this rate is acceptable when compared with other tests with a low yield but a high potential for morbidity (eg, blood cultures for suspected bacteremia, imaging for minor head trauma). However, the benefit of this routine should be weighed against the costs (several hundred dollars per study) and less easily quantified risks, such as the repeated radiation exposure that occurred in a substantial proportion of patients during the time period of this study. In the absence of a more definitive diagnostic test that does not carry a risk of shunt infection, performing both CT scan and shunt series may be appropriate.

Future research efforts to improve the use of radiography for suspected obstruction could include the development of a clinical predictive model. Such a model would incorporate clinical factors from the history and physical examination to provide an estimate of the risk of shunt obstruction. Although it is unlikely that clinical factors alone could adequately rule out shunt obstruction for the majority of patients presenting to an ED, patients at low risk might safely undergo limited radiographic testing (eg, CT scan alone), whereas high risk patients could undergo a full evaluation including shunt tap if radiographic tests were negative. Factors that have previously been associated with a reduced likelihood of obstruction include the presence of fever at the time of evaluation (which would suggest a concurrent infection as the reason for the symptoms) and history of seizure without other symptoms. Preliminary results from a recent study suggest that other clinical factors may also be predictive (11).

There were a number of limitations to this study. Subjects were identified retrospectively by searching for shunt series in a radiology database. Although the shunt series was part of routine evaluation for shunt malfunction, some patients may not have had a shunt series ordered if they required immediate surgical care or had an obvious disconnection on physical examination. Because we were interested in the predictive value for patients needing radiographic studies, we felt that exclusion of such patients was appropriate. The retrospective design limited our analysis to radiographic reports and medical record documentation of outcomes. Although these are objective sources, the actual clinical interpretation at the time of evaluation may have differed from what was documented in the record. For example, the radiologist may suspect a disconnection due to a length of nonopaque tubing, whereas a clinician can rule this out by palpating the shunt in this area. In addition, we did not assess the specific clinical indications or appropriateness of shunt evaluation.

These issues would be more appropriately addressed in a prospective study. Finally, determination of outcomes was limited to medical record review at our institution. Although patients who worsened after discharge would be expected to return, we cannot rule out the possibility that some patients with obstruction sought care at other institutions.

CONCLUSIONS

Although radiographic evaluation by head CT scan and shunt series was able to identify most patients with shunt obstruction, additional evaluation is required in a minority of patients with normal radiographs. Routinely obtaining a shunt series in all patients had a low yield but on rare occasions detected abnormalities that were missed by CT. Future prospective studies should be carried out to identify clinical predictors and to improve the use of radiographic tests in the diagnosis of shunt obstruction.

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