Clinical evaluation and surveillance imaging in children with spina bifida aperta and shunt-treated hydrocephalus

Clinical article

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Object. Most children with spina bifida aperta have implanted CSF shunts. However, the efficacy of adding surveillance imaging to clinical evaluation during routine follow-up as a means to minimize the hazard of shunt failure has not been thoroughly studied.

Methods. A total of 396 clinic visits were made by patients with spina bifida aperta and shunt-treated hydrocephalus in a spina bifida specialty clinic during the calendar years 2008 and 2009 (initial clinic visit). All visits were preceded by a 6-month period during which no shunt evaluation of any kind was performed and were followed by a subsequent visit in the same clinic. At the initial clinic visit, 230 patients were evaluated by a neurosurgeon (clinical evaluation group), and 166 patients underwent previously scheduled surveillance CT scans in addition to clinical evaluation (surveillance imaging group). Subsequent unexpected events, defined as emergency department (ED) visits and caregiver-requested clinic visits, were reviewed. The time to an unexpected event and the likelihood of event occurrence in each of the 2 groups were compared using Cox proportional hazards survival analysis. The outcome and complications of shunt surgeries were also reviewed.

Results. The clinical characteristics of the 2 groups were similar. In the clinical evaluation group, 2 patients underwent shunt revision based on clinical findings in the initial visit. In the subsequent follow-up period, there were 27 visits to the ED and 25 requested clinic visits that resulted in 12 shunt revisions. In the surveillance imaging group, 11 patients underwent shunt revision based on clinical and imaging findings in the initial visit. In the subsequent follow-up period, there were 15 visits to the ED and 9 requested clinic visits that resulted in 8 shunt revisions. Patients who underwent surveillance imaging on the day of initial clinic visit were less likely to have an unexpected event in the subsequent follow-up period (relative risk 0.579, p = 0.026). The likelihood of needing shunt revision and the morbidity of shunt malfunction was not significantly different between the 2 groups.

Conclusions. Surveillance imaging in children with spina bifida aperta and shunted hydrocephalus decreases the likelihood of ED visits and caregiver-requested clinic visits in the follow-up period, but based on this study, its effect on mortality and morbidity related to shunt malfunction was less clear.

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Key Words • myelomeningocele • shunt-treated hydrocephalus • surveillance imaging • spine

Early detection and treatment of CSF shunt malfunction is perhaps the most important strategy for decreasing rates of mortality and morbidity associated with this problem. Several factors may influence the timeliness of care in these patients. Comprehensive patient and caregiver education is necessary to raise awareness of the symptoms of shunt malfunction. For early detection, clinical evaluations at regular intervals may identify signs and subtle symptoms in a mildly symptomatic patient. Last, routine surveillance imaging enables detection of shunt malfunction (that is, disconnected catheter and/or enlarged ventricles) in an asymptomatic child; however, shunt revision in this clinical scenario remains controversial.1,3,5

Abbreviations used in this paper: ED = emergency department; EVD = external ventricular drain; LOS = length of stay.
Although the aforementioned factors are applicable to all patients with CSF shunts, patients with spina bifida aperta and CSF shunts are unique in some respects. In general, these patients and their families are more familiar with the medical system because a good relationship was often established before the child was born. Because of their complex medical and social needs, these children are often cared for in multidisciplinary clinics and have regular follow-up visits. In terms of their treated hydrocephalus, it is generally believed that shunt independence is rare in this group of patients. The majority of these patients and their caregivers are familiar with the symptoms of shunt malfunctions because of multiple past failures. Last, and peculiar to this population, is the fact that a significant number will have no change in ventricular size at presentation for shunt malfunction.

In our multidisciplinary spina bifida specialty clinic, every patient with a CSF shunt is evaluated by a neurosurgeon, but not every patient has a surveillance CT scan at every visit. We first selected a consecutive series of these patients who were deemed to have a functioning shunt by clinical or clinical and radiological examinations, and then we asked whether the addition of a surveillance imaging study at a particular clinic visit correlated with event occurrences in the follow-up period. Our hypothesis is that use of surveillance imaging may alter the number of emergency shunt revisions, the number of ED visits or caregiver-requested visits, and the morbidity and mortality related to shunt malfunction.

**Methods**

*The Care of Patients With Spina Bifida Aperta*

At Children's Hospital, Birmingham, Alabama, patients with spina bifida aperta receive care from a multidisciplinary team of neurosurgeons, orthopedic surgeons, urologists, pediatricians, rehabilitation physicians, physical therapists, and a dedicated full-time clinical coordinator in a specialty clinic setting. Approximately 450 children were followed in this specialty clinic during the study period. A team of 4 board-certified pediatric neurosurgeons provided neurological care to these patients during the same period. Each patient was assigned to 1 of the 4 neurosurgeons to maintain continuity of care. The general consensus among the 4 neurosurgeons was to perform clinical evaluation of the patients at 3-month intervals when the child was less than 12 months old, at 6-month intervals between 1 and 3 years of age, and annually thereafter. All neurosurgeons adopted the practice of routine surveillance CT scanning at this institution. In general, surveillance CT scans were obtained at 6-month intervals between 1 and 3 years of age, annually between 3 and 7 years of age, and biennially after that. All 4 neurosurgeons adopted the practice of revising asymptomatic enlargement of ventricles in these patients.

**Patient Selection**

Patients who had a scheduled visit to the spina bifida specialty clinic in the calendar years 2008 and 2009 (“initial” visit) were considered for this study. The included patients had a documented diagnosis of spina bifida aperta and shunt-treated hydrocephalus. Only those patients who had already attended the follow-up visit were included in the analysis. The period between the initial clinic visit and the subsequent scheduled visit was defined as the “follow-up period.”

Eighty patients had only one visit between January 1, 2008, and the present, and these patients were excluded. Twenty-three of these 80 patients were transitioned to the care of neurosurgeons who treat adult patients. Patients with spina bifida aperta but without a CSF shunt were excluded (36 patients). One hundred six visits were excluded because the patient had been evaluated by the neurosurgery service in the previous 6 months. This included any clinic visits, ED visits, or 23-hour hospital admissions for observation. Patients were excluded if they were hospitalized by other services and the neurosurgery service was consulted to participate in their care. Patients were excluded regardless of whether the clinical evaluation led to a shunt exploration. Infants younger than 6 months of age were excluded because they did not have a 6-month interval free of neurosurgical evaluation (7 patients).

A total of 396 clinical visits made by 396 individual patients fulfilled the above inclusion criteria. These were visits made by patients with spina bifida aperta and shunted hydrocephalus who had a documented follow-up visit and an unremarkable clinical course in the 6-month period prior to the initial visit.

**Statistical Analysis**

Statistical analysis was performed using SPSS software (SPSS, Inc.). The event-free survival of the CSF shunt was defined as the length of time between the “start” and the “end” events. The “start” event was a scheduled visit to the spina bifida specialty clinic during 2008–2009. At the “start” event, a surveillance CT scan may or may not have been previously scheduled. The “end” events were an ED visit or a request from a caregiver to have the patient examined in the clinic prior to the previously scheduled follow-up visit. If neither of the “end” events occurred before the patient attended the follow-up visit, the CSF shunt was considered to function adequately in the event-free survival analysis. In the analysis, the use of surveillance CT was defined as a categorical predictor (patients who did or did not undergo surveillance CT). Cox proportional hazard analysis was then used to examine whether the CSF shunt event-free survival was predicted by the use of surveillance CT. The occurrence of surgical complications from shunt exploration in the 2 groups was compared using a contingency table. Probability values < 0.05 were considered statistically significant.

**Results**

*Events at the Initial Clinic Visit*

The mean ages of the patients in Group 1 (clinical evaluation only) and Group 2 (clinical and radiological evaluations) were 10.1 ± 6.1 years and 9.9 ± 6.1 years, respectively. The mean length of follow-up was 10 ± 5.4
Surveillance imaging for shunt-treated hydrocephalus

months and 10.8 ± 4.7 months in the two groups. The mean values are expressed ± SD. Consistent with our patient care strategy, children who were younger were seen more often in the spina bifida clinic than those who were older (Fig. 1).

For patients who were evaluated by a neurosurgeon without a surveillance CT scan, 9 CT scans were obtained on the day of the clinic visit because of various clinical findings. Two of these 9 patients underwent shunt exploration (Table 1). One asymptomatic patient had a known shunt disconnection that was previously followed at another institution (Case 12, Table 2). One patient underwent a CT scan because “the patient has not had a surveillance scan for more than 5 years” per the physician requesting the scan, and the ventricles were found to be enlarged compared with baseline (Case 13, Table 2). The remaining 7 patients did not require neurosurgical intervention.

For patients who had a surveillance CT scan and clinical evaluation, 11 underwent surgical intervention as a result of imaging and clinical findings (Table 1). Nine patients were asymptomatic, with enlarged ventricles seen on CT scans. One patient reported paresthesias in the upper extremities, and a syrinx was found on the CT scan (Case 1, Table 2). The shunt was explored to check its patency. One patient had enlarged ventricles and an accelerated head growth (Case 2, Table 2).

Events in the Follow-Up Period

Patients who underwent shunt exploration due to findings at the initial clinic visit were excluded from this part of the analysis. Therefore, the event-free survival analysis included only those patients who were deemed to have a functioning shunt based on clinical (228 patients) or clinical and radiological (155 patients) findings. The relevant clinical events (ED visits and caregiver-requested visits for shunt-related complaints) in the follow-up periods of the 2 groups are summarized in Table 1. The 2 CSF shunt event-free survival curves were then plotted and compared using the Cox proportional hazards method (Fig. 2). The addition of a radiological evaluation to clinical evaluation at the time of the initial clinic visit was associated with a 42% decrease in the likelihood of ED visits and caregiver-requested clinic visits in the follow-up period (p = 0.026; risk ratio 0.579, CI 0.352–0.952).

Number of Shunt Surgeries

The number of shunt surgeries for the 2 groups of patients is summarized in Table 1. Patients were much more likely to undergo shunt exploration if a surveillance CT study was performed at the time of the initial clinic visit (11 vs 2). In the follow-up period, despite a significantly higher number of visits to the ED and the clinic by patients in the clinical evaluation group, the rate of shunt revision was similar in the 2 groups (12 [5.3%] of 228 vs 8 [5.2%] of 155).

Surgical Complications

The details of clinical presentation, surgical findings, and complications of shunt explorations are summarized in Tables 2 and 3. In the 13 patients whose shunts were revised because of findings at the initial clinic visit (Table 2), complications occurred in 4 patients within the first 6 months. Proximal obstruction and a fractured catheter were the most common findings. Eleven of these patients had no signs or symptoms suggestive of shunt malfunction. The mean LOS (the day of surgery counted as Day 1) was 2.2 days.

Twenty patients underwent shunt revision in the follow-up period (Table 3). Three intraoperative complications occurred. The patient in Case 3 suffered scalp injury due to electrocautery use. Intraventricular hemorrhage occurred with the proximal catheter removal in the patient in Case 7. With the patient in Case 10, brisk backflow from the distal tubing was observed on disconnecting the distal catheter from the valve, and the shunt was removed and an EVD placed due to suspicion of an abdominal pseudocyst. One dialysis-dependent patient presented with peritonitis, and his shunt failed due to poor abdominal absorption (Case 4). Excluding this last patient, complications occurred in 8 of 19 patients, and the average LOS was 3.6 days. The differences in complication rates (31% vs 42%) and LOS between the 2 groups were not statistically significant.

TABLE 1: Summary of clinical events after the initial clinic visit and in the follow-up period in patients with shunt-treated hydrocephalus

<table>
<thead>
<tr>
<th>Clinical Events</th>
<th>Clinical Evaluation Only</th>
<th>Clinical &amp; Radiological Evaluation</th>
</tr>
</thead>
<tbody>
<tr>
<td>no. of patients</td>
<td>230</td>
<td>166</td>
</tr>
<tr>
<td>ops due to findings at initial clinic visit</td>
<td>2</td>
<td>11</td>
</tr>
<tr>
<td>shunts deemed to be functional at initial clinic visit</td>
<td>228</td>
<td>155</td>
</tr>
<tr>
<td>events in FU period</td>
<td></td>
<td></td>
</tr>
<tr>
<td>ED visits (no. of ops)</td>
<td>27 (7)</td>
<td>15 (7)</td>
</tr>
<tr>
<td>caregiver-requested visits (no. of ops)</td>
<td>25 (5)</td>
<td>9 (1)</td>
</tr>
<tr>
<td>total no. of shunt ops during FU period</td>
<td>12</td>
<td>8</td>
</tr>
</tbody>
</table>

* FU = follow-up.

Fig. 1. Graph showing the mean time interval in months between clinic visits for children of various ages.
Discussion

Study Design

Many factors affect the care of children with shunted hydrocephalus; thus the utility of surveillance imaging to detect early shunt malfunction is not easily assessed. Although cases of hydrocephalus secondary to intraventricular hemorrhage of prematurity or third-ventricular neoplasm may both be treated with a CSF shunt, the differences in underlying pathological features and patient characteristics do not allow for direct comparison of the shunt management strategy in the 2 groups. To provide a more homogeneous background, children with spina bifida and shunted hydrocephalus were chosen for this study. In general, these children and their families regularly attend follow-up appointments and have an ongoing relationship with the medical community. Because of the establishment of a spina bifida specialty clinic, they tend to receive all of their medical care at a single institution. They are often not strangers to shunt malfunctions, which implies a certain degree of vigilance and familiarity with the presentation of shunt malfunction. The choice of this patient population decreases the generalizability of the results of this study, but it may help to direct future studies on hydrocephalus from other causes.

All of our patients had a well-defined follow-up period that was flanked by 2 scheduled clinic visits. This is important because it implies that in all likelihood, the patient continued to receive care from our institution. In addition, a defined follow-up period is necessary for the construction of a Cox proportional hazards regression model, which analyzed not only whether an event had occurred but also the timing of the event. The stepwise occurrence of events throughout the follow-up period as shown in Fig. 1 further adds to the validity of the results.

Surgeries for Asymptomatic Shunt Malfunction

The first and foremost reason to perform imaging routinely in asymptomatic patients with CSF shunts is to identify those who are at an increased risk of shunt malfunction, in the hope that the mortality and morbidity related to shunt malfunction can be decreased by preemptive intervention. However, to revise the CSF shunt in this context is to assume that these patients are shunt dependent. In our study, 11 asymptomatic patients underwent shunt revisions for this reason (Table 2), and not surprisingly, the majority of these patients were identified through the use of routine surveillance imaging. If one assumes that all children with spina bifida aperta become shunt dependent once they undergo shunt placement, one would then expect that the patient cohort that underwent clinical and radiological evaluation should have a lower rate of shunt revision in the subsequent follow-up period. However, our data suggested otherwise. The rate of shunt exploration surgery did not differ between the 2 groups

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs)</th>
<th>Surveillance CT</th>
<th>Reason for Revision</th>
<th>LOS (days)</th>
<th>Shunt Exploration Findings</th>
<th>Complications w/in 6 Mos</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>5.1</td>
<td>yes</td>
<td>syrinx, arm paresthesia</td>
<td>3</td>
<td>proximal obstruction</td>
<td>proximal obstruction</td>
</tr>
<tr>
<td>2</td>
<td>1.0</td>
<td>yes</td>
<td>increased head circumference, enlargement of ventricles</td>
<td>2</td>
<td>negative</td>
<td>proximal obstruction</td>
</tr>
<tr>
<td>3</td>
<td>2.8</td>
<td>yes</td>
<td>asymptomatic enlargement of ventricles</td>
<td>2</td>
<td>proximal obstruction</td>
<td>none</td>
</tr>
<tr>
<td>4</td>
<td>20.8</td>
<td>yes</td>
<td>asymptomatic enlargement of ventricles; broken catheter</td>
<td>2</td>
<td>fractured catheter</td>
<td>none</td>
</tr>
<tr>
<td>5</td>
<td>3.8</td>
<td>yes</td>
<td>asymptomatic enlargement of ventricles</td>
<td>2</td>
<td>proximal obstruction</td>
<td>proximal obstruction</td>
</tr>
<tr>
<td>6</td>
<td>2.4</td>
<td>yes</td>
<td>asymptomatic enlargement of ventricles</td>
<td>2</td>
<td>proximal obstruction</td>
<td>none</td>
</tr>
<tr>
<td>7</td>
<td>23.7</td>
<td>yes</td>
<td>asymptomatic disconnection found on CT</td>
<td>2</td>
<td>fractured catheter</td>
<td>none</td>
</tr>
<tr>
<td>8</td>
<td>8.9</td>
<td>yes</td>
<td>asymptomatic enlargement of ventricles</td>
<td>2</td>
<td>fractured catheter; distal obstruction</td>
<td>none</td>
</tr>
<tr>
<td>9</td>
<td>14.9</td>
<td>yes</td>
<td>asymptomatic enlargement of ventricles</td>
<td>2</td>
<td>fractured catheter</td>
<td>none</td>
</tr>
<tr>
<td>10</td>
<td>1.6</td>
<td>yes</td>
<td>asymptomatic enlargement of ventricles</td>
<td>2</td>
<td>proximal obstruction</td>
<td>none</td>
</tr>
<tr>
<td>11</td>
<td>12.7</td>
<td>yes</td>
<td>asymptomatic enlargement of ventricles</td>
<td>2</td>
<td>proximal obstruction</td>
<td>none</td>
</tr>
<tr>
<td>12</td>
<td>14.7</td>
<td>no</td>
<td>asymptomatic disconnection that was previously followed</td>
<td>3</td>
<td>distal catheter disconnected from valve</td>
<td>none</td>
</tr>
<tr>
<td>13</td>
<td>20.5</td>
<td>no</td>
<td>asymptomatic enlargement of ventricles</td>
<td>2</td>
<td>proximal &amp; distal obstruction</td>
<td>shunt infection</td>
</tr>
</tbody>
</table>

Fig. 2. Graph showing event-free survival of a CSF shunt after the initial clinic visit. Solid line, clinical evaluation group; dotted line, surveillance imaging group.
(Table 1) despite the fact that all asymptomatic patients with radiological evidence of shunt malfunction were preemptively treated.²

It is important to point out that this study does not argue for or against revisions for asymptomatic ventriculomegaly. The utility of shunt revision in this context is not only to decrease morbidity and mortality associated with shunt malfunction but also to protect against subtle neurocognitive deterioration. A prospective randomized clinical trial with serial neurocognitive testing would be necessary to provide the answer. However, before randomizing patients to imaging and no-imaging groups, there needs to be a study that shows that the severity and likelihood of harm in the no-imaging group are not greater than in the imaging group; otherwise randomization would not be ethically acceptable.

Our study cannot address whether a malfunctioning shunt in a patient who is asymptomatic should be revised because intellectual function outcome was not assessed. However, it does address the question of safety. Our study shows that it is likely to be safe to randomize patients into imaging and no-imaging groups because the rates of subsequent shunt revision and the associated complications are not different between the 2 groups. Without this assurance, a randomized trial as advocated above would not have been possible.

**Effect of Surveillance Imaging**

Patients who underwent surveillance imaging were less likely to visit the ED or to request a clinic visit with shunt-related complaints after the initial clinic visit. The most plausible reason is that surveillance imaging provided the caregiver and the patient a degree of confidence in adequate shunt function. This psychological effect was significant and was not expected by the authors. We do not advocate performing surveillance imaging on every patient at every visit to decrease ED visits or requested clinic visits. It is possible that a follow-up clinic visit at a shorter interval may provide the necessary assurance. The potentially harmful effects of radiation and the cost of imaging and ED and clinic visits are factors that need to be considered.

**Conclusions**

Surveillance imaging in children with spina bifida aperta and shunted hydrocephalus has a significant effect on the number of unexpected hospital and clinic visits, but based on our study, its effect on the mortality and morbidity related to shunt malfunction was less apparent.

**Disclosure**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Chern. Acquisition of data: Muhleman, Miller. Analysis and interpretation of data: Chern.
References


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