Radiographic Evaluation of Pediatric Cerebrospinal Fluid Shunt Malfunction in the Emergency Setting

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Abstract: Children with ventricular cerebrospinal fluid shunts for treatment of hydrocephalus require frequent evaluation for potential shunt malfunction. Current practice relies heavily on neuroimaging, particularly cranial computed tomography, which repeatedly exposes children to ionizing radiation. Rapid cranial magnetic resonance imaging is a new radiation-sparing alternative to CT for evaluation of potential shunt malfunction. We review the diagnostic test performance, radiation exposure, advantages, and limitations of the major neuroimaging modalities available to providers caring for children with possible shunt malfunction in the emergent setting.

Key Words: ventricular shunt, shunt malfunction, CT, MRI, rapid cranial MRI, ultrafast MRI

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TARGET AUDIENCE

This continuing medical education activity is intended for physicians, nurse practitioners, physician assistants, and other emergency personnel who evaluate children with mechanically shunted hydrocephalus for possible ventricular shunt malfunction. Specialists including pediatricians, pediatric, and adult emergency physicians will find this information particularly useful.

LEARNING OBJECTIVES

After completion of this article, the reader should be able to:

1. Review current neuroimaging standards for diagnosing ventricular shunt malfunction in children.
2. Quantify the ionizing radiation exposure associated with current diagnostic standards.
3. Discuss advantages and limitations of rapid cranial magnetic resonance imaging (MRI) as a radiation-sparing alternative for diagnosing shunt malfunction.

Mechanical shunting of cerebrospinal fluid (CSF) is the therapeutic mainstay for children with hydrocephalus and is among the most common pediatric neurosurgical procedures performed.[1,2] A ventriculoperitoneal shunt is the most commonly placed type of CSF shunt because CSF is efficiently absorbed by the peritoneum. Ventriculoatrial, ventriculopleural, and lumboperitoneal shunts are more rarely placed, typically as a second-line device. Most modern ventricular shunts placed in the United States after 2009 consist of a proximal ventriculostomy catheter, a pressure-sensitive valve and reservoir, and a distal catheter.

Ventricular shunts malfunction for numerous reasons, including mechanical obstruction of the shunt lumen, overdrainage, pressure valve or reservoir malfunction, catheter damage or disconnection, catheter migration, or infection.[3,4] Noninfectious causes, particularly proximal obstruction, are the most common etiology of ventricular shunt malfunction in the first 2 years after shunt placement or most recent shunt revision.[3,5] Distal obstruction is more common in long-standing shunts.[3] Shunt infections cause a minority of obstructions by accumulation of debris within the shunt system.

Ventricular shunt failure results in worsening hydrocephalus, which can be life threatening. Unfortunately, ventricular shunt malfunctions are common. Despite advances in design, up to 40% of pediatric shunts require operative revision in the first year after insertion, and an estimated 80% require at least 1 revision in the subsequent decade.[6–8] Introduction of programmable shunt systems has not reduced either shunt failure or revision rates.[8] The financial burden for CSF shunt–related procedures in the United States is estimated to exceed $1 billion per year, with approximately half of these neurosurgical procedures involving shunt removal or replacement.[2,9] Given the high probability of shunt failure, children with ventricular shunts require repeated emergent evaluation when symptomatic with lethargy, headache, vomiting, or other complaints that might arise from shunt malfunction.

The evaluation of a child with possible ventricular shunt failure presents challenges. The clinical presentation of a child with worsening hydrocephalus is often nonspecific with considerable overlap with other childhood illnesses. Retrospective analyses demonstrate that signs and symptoms of shunt malfunction, such as headache and vomiting, lack significant predictive ability to direct decision making alone.[6,10–14] Thus, clinicians rely heavily on emergent neuroimaging and neurosurgical consultation to identify children with ventricular shunt malfunctions.

This review will focus on radiologic adjuncts for diagnosing mechanical causes of ventricular shunt malfunction. Current practice in many centers relies on cranial computed tomography (CT) scans and, to a lesser extent, radiographic shunt series to diagnose shunt malfunction. However, these radiologic studies, particularly CT scans, expose children to ionizing radiation exposure associated with an increase in lifetime malignancy risk.[15–17] We will review the performance characteristics of current neuroimaging standards, as well as a new radiation-sparing diagnostic technique, rapid cranial MRI.

CRANIAL CT

Cranial CT has long been the diagnostic standard for ventricular shunt malfunction. When performed for evaluation of possible ventricular shunt malfunction, cranial CT images are typically acquired axially without intravenous contrast. Cranial

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CT rapidly and reliably assesses interval alterations in ventricle size when compared against a baseline study, thus informing the clinician about the presence of hydrocephalus (Fig. 1). Enlarged ventricles are the radiologic hallmark of shunt obstruction.[18–20] Other CT findings that correlate with increased intracranial pressure include cerebral cortical sulci effacement, loss of the basal cistern, and periventricular edema due to transependymal CSF absorption.[21,22] Shunt overdrainage leading to small or slit-like ventricles and subdural hematomas or hygromas and proximal catheter migration are also easily detected by CT.[22]

Although absolute or relative ventriculomegaly are the expected CT findings in children with a ventricular shunt malfunction, some children never manifest these radiologic changes.[23,24] The inability of CT to detect all ventricular shunt malfunctions is multifactorial. First, shunts require revision for reasons beyond obstruction (eg, infection without obstruction) that may not manifest with ventricle size change, fluid collections, or catheter abnormalities that would be detectable by CT. Second, scarring of the ventricular walls has been hypothesized to prohibit ventricle expansion, leading to signs of increased intracranial pressure in the absence of radiologic findings (ie, stiff ventricles).[22,23] Third, children with partial ventricular shunt obstruction or subacute presentations may manifest more subtle radiographic findings. Finally, patients with complex underlying disease may have multiple baseline ventricular abnormalities that make radiologic interpretation challenging. Thus, the clinical decision to take a child to the operating room for suspected shunt pathology is multifactorial and ultimately rests with the overall assessment of the emergency physicians and neurosurgeons.

In previous largely retrospective studies, cranial CT has a reported sensitivity of 53% to 92% and a specificity of 76% to 93% for detecting ventricular shunt malfunction.[13,25–30] The performance of CT may not be substantially better than an experienced parent's ability to diagnose a shunt malfunction in their child. In 1 study, parental assessment of the likelihood of

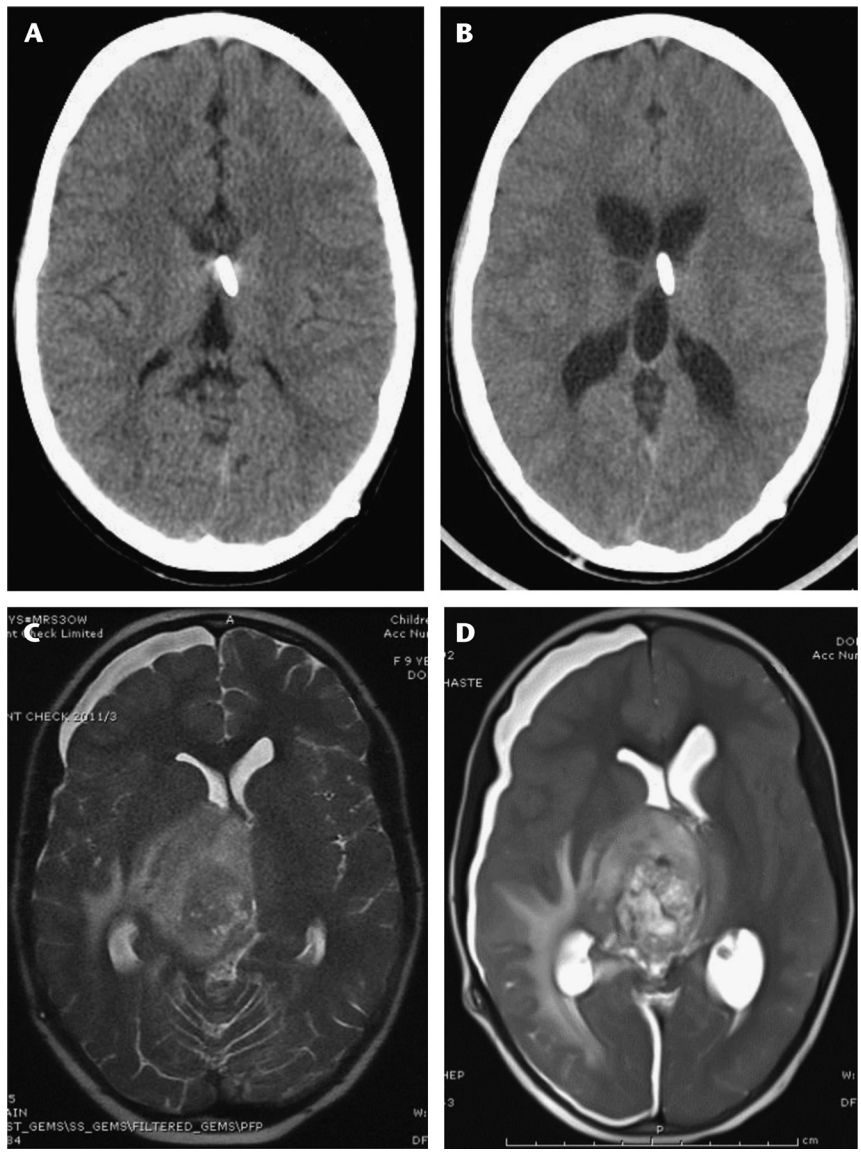

FIGURE 1. Representative axial images from 2 different patients with ventricular shunts at baseline (A and C) and with shunt malfunction (B and D) who underwent CT (A and B) or rapid cranial MRI (C and D).

shunt malfunction using a visual analog scale had 89% sensitivity and 62% specificity for ventricular shunt malfunction.[11]

SHUNT SERIES

A shunt series includes a set of anterior-posterior and lateral plain radiographs to evaluate the entire length of the CSF shunt catheter (eg, skull, chest, and abdominal radiographs for ventriculoperitoneal shunts). These radiographs demonstrate catheter fracture, catheter disconnection, calcification, or migration of the distal tip. Shunt series have poor sensitivity (4%–26%) but high specificity (92%–98%) for diagnosing shunt malfunction.[26,27,29] Furthermore, less than 1% of surgical shunt revisions are prompted by an abnormal shunt series alone.[26] Historically, shunt series have been used concomitantly with CT. However, CT performance is only marginally enhanced when performed with shunt series, improving CT sensitivity from 83% to 88% in 1 study.[26] As radiographic shunt series has low diagnostic utility as a first-line diagnostic test, children with suspected shunt malfunction should not routinely have a shunt series performed.[27,31]

NUCLEAR MEDICINE SHUNTOGRAMS

A technetium-99m (Tc-99m) di-ethylene tri-amine penta-acetic acid (DTPA) shuntogram is a nuclear medicine study that can evaluate shunt flow dynamics and opening pressure using a radiotracer injected into the shunt reservoir.[32] Shunt patency and flow dynamics are evaluated by following contrast progression through the ventricular shunt system. Abnormal tracer clearance can be seen in any of the following scenarios: proximal or distal ventricular catheter obstruction, valve malfunction, or catheter disconnection. Nuclear scintigraphy has a reported sensitivity of 47% to 65% and specificity of 86% to 92% for the diagnosis of ventricular shunt malfunction requiring surgical revision.[27,33–35] However, limited availability, high radiation exposure, and the potential to introduce a shunt infection during injection of radioisotope have made nuclear medicine shuntograms an uncommon imaging modality in the emergent setting.

RADIATION EXPOSURE

Although neuroimaging plays a pivotal role in the diagnostic workup of a child with suspected shunt malfunction, radiographs expose children to ionizing radiation with an increase in the long-term lethal malignancy risk. The highest level of radiation exposure is associated with a nuclear medicine shuntogram followed by a cranial CT (Table 1). Although experts disagree about the precise magnitude of the risk, extrapolations from atomic bomb data as well as long-term follow-up studies of children who underwent CT scans both demonstrate increased lifetime cancer risks attributable to CT radiation exposure.[15,17,37,38] The youngest children are at the highest risk because of more rapidly dividing cells and longer life expectancy.[15–17]

TABLE 1. Effective Radiation Doses in Imaging Modalities Used for Diagnosing Ventricular Shunt Malfunction in Children

Imaging Type	Mean Effective Dose, mSv	Reference
Shunt series	1.6	Shuaib et al[29]
Cranial CT (age ≤21 y)	1.9–2.5	Miglioretti et al,[17] Shuaib et al,[29] Koral et al[36]
Age <5 y	3.5	
Age >5 y	1.1–1.5	
Cranial CT + shunt series (age ≤21 y)	3.2	Shuaib et al[29]

Children with shunted hydrocephalus, who often undergo multiple lifetime CT scans starting at a young age, are a particularly vulnerable population.[36,39] Published estimates of the average number of cranial CT scans performed hover around 2 to 3 per patient-year.[14,36] In a series of children with shunted hydrocephalus, almost 40% underwent more than 5 lifetime head CTs, and 10% underwent more than 15 imaging studies.[40]

The radiation dose delivered by a single cranial CT can vary greatly depending on various technical factors, such as scanner settings and acquisition software, as well as patient factors such as age.[17,38] Estimating the cumulative effective dose in a child additionally depends on the number of lifetime CTs and duration of follow-up. Illustrating this point, the reported cumulative effective dose per patient in 67 patients with shunted hydrocephalus followed over 2 decades varied widely from 2.3 to 64 mSv.[41] Low-dose and limited-slice CT protocols have been implemented in some centers to reduce radiation exposure in shunted children.[42–44] Based on mathematical models with extrapolation from organ-specific effective radiation doses from the International Commission on Radiologic Protection,[45] the average cranial CT is estimated to deliver 2.5 mSv, whereas a low-dose ventricular shunt protocol averages 1.1 mSv.[36]

Evidence linking radiation exposure and lifetime malignancy risk to patient outcomes is growing.[15,46] Determining the ultimate impact of repeated radiation exposure in shunted children is complicated by patient comorbidities and underlying disease processes that may affect life expectancy. That said, assuming a child with shunted hydrocephalus undergoes an average of 2 CT scans annually until the age of 20 years, excess lifetime fatal cancer risk has been estimated to be as high as 1 lifetime cancer per 97 patients for standard dose protocols and 1 per 130 patients for low-dose protocols.[36] Given this risk, radiation sparing alternatives, such as MRI, have gained prominence in many pediatric centers in the last 5 years.[47]

RAPID CRANIAL MRI

Although MRI is radiation sparing, the use of conventional imaging techniques for evaluation of a child with possible ventricular shunt malfunction is limited by lengthy imaging times, potential for motion artifact, and frequent need for sedation. Recent advances in MRI technology have introduced rapid or "ultrafast" sequence protocols (eg, single shot fast spin echo or half-fourier acquisition single shot turbo-spin echo sequences) that dramatically reduce image acquisition time. Single shot fast spin echo sequences use single-section T2-weighted images that are sampled within a fraction of a second, resulting in image acquisition times ranging 1 to 4 minutes that are comparable to the 2-minute acquisition time for CT with minimal motion artifact.[48–50] Half-fourier acquisition single shot turbo-spin echo sequences provide an alternative ultrafast T2-weighted MRI protocol.[49,51,52] Most institutions offering rapid MRI technology use T2-weighted protocols.[47]

Compared with conventional cranial MRI, rapid MRI has 78% sensitivity and 98% specificity for identifying any intracranial abnormality in a child.[50] Ventricle size changes are accurately assessed (Fig. 1), and multicystic loculations are well visualized.[40,52] However, correct identification of congenital malformations, mass lesions, or subtle parenchymal changes that require greater image resolution are less reliable. Similarly, rapid MRI has some advantages over cranial CT. In a prospective study of 114 patients with acute neurological signs and symptoms, rapid MRI provided additional diagnostic information missed by

TABLE 2. Test Performance of Imaging Modalities Used for Diagnosing Ventricular Shunt Malfunction in Children

Imaging Type	Sensitivity	Specificity	Reference
Shunt series	4%–26%	92%–98%	Zorc et al,[26] Lehnert et al,[27] Shuaib et al[29]
Nuclear medicine shuntogram	47%–65%	86%–92%	Lehnert et al,[27] Vernet et al,[33] Ouellette et al,[34] Vassilyadi et al[35]
Cranial CT	53%–92%	76%–93%	Barnes et al,[13] Mater et al,[25] Zorc et al,[26] Lehnert et al,[27] Boyle et al,[28] Shuaib et al[29] Yue et al[30]
Rapid cranial MRI	51%–59%	89%–93%	Boyle et al,[28] Yue et al[30]

cranial CT in a quarter of cases, but generated false-positive results in 3% and false-negative or unreliable findings (unrelated to ventricle size) in approximately 10%.[53]

Driven to spare ionizing radiation exposure while rapidly and reliably assessing ventricle size change, some pediatric centers have begun to offer rapid cranial MRI for assessment of children with shunted hydrocephalus.[40] Sedation needs for children undergoing rapid cranial MRI have generally been limited.[28,52] Less than 1% of children undergoing rapid cranial MRI for evaluation of possible ventricular shunt malfunction at a single center required sedation to obtain the neuroimaging.[28] Rapid cranial MRI, however, has several important limitations to recognize including poor visualization of shunt catheter position, intracranial hemorrhage, and pneumocephaly.[48,49,52–54]

Recently, the test performance of rapid cranial MRI was compared to the current practice standard, cranial CT for diagnosing ventricular shunt malfunction in children.[28,30] In 2 independent, large retrospective cohort studies, rapid cranial MRI demonstrated a sensitivity of 51% to 59%, specificity of 89% to 93%, and accuracy of 82% to 84% for the diagnosis of confirmed ventricular shunt malfunction.[28,30] In one of those studies, the accuracy and specificity of rapid cranial MRI were not inferior to CT (within a priori noninferiority margin of 10%), although the study was underpowered to compare sensitivity.[28] Rapid cranial MRI seems to be a viable diagnostic alternative to CT when evaluating a child with a possible shunt malfunction, particularly if there is concern for obstruction. However, clinical concerns for catheter migration or hemorrhage should prompt consideration of CT as the initial diagnostic modality.

Although the potential benefits of rapid MRI are clear, particularly in children with shunted hydrocephalus, there have been numerous obstacles to widespread implementation of this technology. In a recent survey of 56 of the 101 North American institutions providing pediatric neurosurgical care, 79% reported having a rapid MRI protocol to evaluate ventricle size, with only 64% reporting routine use. Barriers to implementation and routine use included lack of emergency access to MRI facilities, radiologic staffing limitations, and difficulty with reimbursement of rapid MRI protocols.[47] Not unexpectedly, although considerably less than conventional cranial MRI, the charge for rapid cranial MRI compared with CT has institutional variability.[28,49] At the authors' institution, the charge for rapid cranial MRI is marginally greater than CT. However, these cost differences are arguably balanced by the long-term benefits of reducing repeated ionizing radiation exposure. In addition, both MRI technologists and radiologists may need specific training in acquisition and interpretation of ultrafast images.

Furthermore, children with some programmable shunt systems are at risk for unintentional valve resetting in the scanner's magnetic field and may need reprogramming by a trained provider after neuroimaging is obtained.[55–57] In a large series of shunted patients, including children, nearly 27% had shunt valves accidentally reset by MRI.[59] Having an identification system to notify medical personnel of the presence of a programmable valve, and the previous setting of opening pressure is recommended. The presence of a programmable valve may be confirmed by patient or parent knowledge (although potentially less reliable) or by plain skull radiograph. Some newer adjustable shunt valves are resistant to the effects of the MRI magnet and may not require adjustment after imaging. Most centers that routinely use rapid cranial MRI rely on neurosurgical consultations to determine valve type and to readjust settings as needed, as the consequences of incorrect adjustment could be substantial for the patient. Providers at institutions without an available neurosurgeon should either perform a CT (low-dose protocol) or transfer the patient to a center with pediatric neurosurgical coverage and experience with ultrafast MRI protocols to evaluate for possible shunt malfunction.

Finally, potential delays in care related to emergency access to rapid cranial MRI imaging should be examined on an institutional basis. Magnetic resonance imaging scanners are often located outside the ED and require patients to undergo additional checks before exposure to the magnet. After completion of the MRI scan, patients need assessment for possible shunt valve reprogramming. At our institution, we found a median increase of 30 minutes (interquartile range, 18–42 minutes) in the time from ED arrival to completion of neuroimaging and 48 minutes (interquartile range, 24–66 minutes) in the overall ED length of stay for ED visits where rapid cranial MRI rather than CT was performed for possible shunt malfunction.[28] Fortunately, short imaging times with ultrafast protocols make insertion of emergent rapid MRI scans into the existing MRI schedule often feasible within existing work flow.

CONCLUSIONS

In conclusion, diagnosing ventricular shunt malfunction in a child remains challenging. The sensitivity of neuroimaging overall is limited because of the multifactorial mechanisms of shunt malfunction and the complexity of the underlying pathology leading to ventricular shunt malfunction in children (Table 2). Furthermore, the current reliance on repeated CT scans leads to substantial lifetime ionizing radiation exposure in children with ventricular shunts. Rapid cranial MRI is a new radiation sparing alternative that is not inferior to CT for diagnosing shunt malfunction in children. Institutions caring for children with ventricular shunts on a routine and emergent basis should strive to overcome barriers to widespread implementation of rapid MRI protocols. Emergency providers should consider rapid cranial MRI as a first-line diagnostic alternative to CT when caring for a child with a possible shunt malfunction given the long-term benefits of sparing the malignancy risks related to ionizing radiation exposure.

This CME activity has summarized the radiographic alternatives for diagnosing ventricular shunt malfunction in the emergency setting.

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