

ACR Appropriateness Criteria[®]

Hydrocephalus-Child

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Abstract

Hydrocephalus is a disorder of cerebrospinal fluid circulation that leads to ventricular enlargement and increased intracranial pressure. Presentation and imaging choice vary with age: infants often develop progressive macrocephaly because of skull compliance, whereas older children more commonly present with headache, vomiting, and lethargy. In newly suspected hydrocephalus, imaging confirms ventricular dilation, helps assess acuity, may identify an underlying cause, and informs management. In patients with cerebrospinal fluid diversion, imaging primarily evaluates suspected shunt malfunction, with modality selection driven by clinical urgency, patient cooperation, and age. In infants with open fontanelles, cranial ultrasound provides rapid assessment of ventricular size. MRI is generally preferred to define ventricular caliber and sites of obstruction; abbreviated/rapid MRI can shorten examinations and may avoid sedation. Noncontrast CT is useful when rapid evaluation is required or MRI is not feasible.

The American College of Radiology Appropriateness Criteria are evidence-based guidelines for specific clinical conditions that are reviewed annually by a multidisciplinary expert panel. The guideline development and revision process support the systematic analysis of the medical literature from peer reviewed journals. Established methodology principles such as Grading of Recommendations Assessment, Development, and Evaluation or GRADE are adapted to evaluate the evidence. The RAND/UCLA Appropriateness Method User Manual provides the methodology to determine the appropriateness of imaging and treatment procedures for specific clinical scenarios. In those instances where peer reviewed literature is lacking or equivocal, experts may be the primary evidentiary source available to formulate a recommendation.

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Disclaimer: The ACR Committee on Appropriateness Criteria and its expert panels have developed criteria for determining appropriate imaging examinations for diagnosis and treatment of specified medical condition(s). These criteria are intended to guide radiologists, radiation oncologists and referring physicians in making decisions regarding radiologic imaging and treatment. Generally, the complexity and severity of a patient's clinical condition should dictate the selection of appropriate imaging procedures or treatments. Only those examinations generally used for evaluation of the patient's condition are ranked. Other imaging studies necessary to evaluate other co-existent diseases or other medical consequences of this condition are not considered in this document. The availability of equipment or personnel may influence the selection of appropriate imaging procedures or treatments. Imaging techniques classified as investigational by the FDA have not been considered in developing these criteria; however, study of new equipment and applications should be encouraged. The ultimate decision regarding the appropriateness of any specific radiologic examination or treatment must be made by the referring physician and radiologist in light of all the circumstances presented in an individual examination.

Key Words: Appropriateness Criteria, Appropriate Use Criteria, AUC, endoscopic third ventriculostomy, hydrocephalus, shunt failure, ventriculoperitoneal shunt

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ACR Appropriateness Criteria® Hydrocephalus-Child. [Variants 1 to 6](#) and [Tables 1 and 2](#).

Variant 1. Infant. Suspected hydrocephalus. Abnormal prenatal imaging or signs or symptoms of elevated intracranial pressure. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
US head	Usually Appropriate	○
MRI head without IV contrast	Usually Appropriate	○
MRI head without IV contrast abbreviated	Usually Appropriate	○
MRI head without and with IV contrast	May Be Appropriate	○
CT head without IV contrast	May Be Appropriate	⚠⚠⚠
MRI head with IV contrast	Usually Not Appropriate	○
MRV head with IV contrast	Usually Not Appropriate	○
MRV head without IV contrast	Usually Not Appropriate	○
CT head with IV contrast	Usually Not Appropriate	⚠⚠⚠
CT head without and with IV contrast	Usually Not Appropriate	⚠⚠⚠⚠

Variant 2. Infant. Known hydrocephalus. Follow-up imaging.

Procedure	Appropriateness Category	Relative Radiation Level
MRI head without IV contrast	Usually Appropriate	○
MRI head without IV contrast abbreviated	Usually Appropriate	○
Radiography head neck chest abdomen	May Be Appropriate	⚠⚠
US head	May Be Appropriate	○
CT head without IV contrast	May Be Appropriate	⚠⚠⚠
MRI head with IV contrast	Usually Not Appropriate	○
MRI head without and with IV contrast	Usually Not Appropriate	○
MRV head with IV contrast	Usually Not Appropriate	○
MRV head without IV contrast	Usually Not Appropriate	○
CT head with IV contrast	Usually Not Appropriate	⚠⚠⚠
CT head without and with IV contrast	Usually Not Appropriate	⚠⚠⚠⚠

Variant 3. Infant. Macrocephaly. No signs or symptoms of elevated intracranial pressure. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
US head	Usually Appropriate	○
MRI head without IV contrast	Usually Appropriate	○
MRI head without IV contrast abbreviated	Usually Appropriate	○
CT head without IV contrast	May Be Appropriate	⚠⚠⚠
Radiography head neck chest abdomen	Usually Not Appropriate	⚠⚠
MRI head with IV contrast	Usually Not Appropriate	○
MRI head without and with IV contrast	Usually Not Appropriate	○
MRV head with IV contrast	Usually Not Appropriate	○
MRV head without IV contrast	Usually Not Appropriate	○
CT head with IV contrast	Usually Not Appropriate	⚠⚠⚠
CT head without and with IV contrast	Usually Not Appropriate	⚠⚠⚠⚠

Variant 4. Child greater than 1 year of age. Suspected hydrocephalus. Signs or symptoms of elevated intracranial pressure. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
MRI head without IV contrast	Usually Appropriate	○
MRI head without IV contrast abbreviated	Usually Appropriate	○
CT head without IV contrast	Usually Appropriate	⚠⚠⚠
MRI head without and with IV contrast	May Be Appropriate	○
US head	Usually Not Appropriate	○
MRI head with IV contrast	Usually Not Appropriate	○
MRV head with IV contrast	Usually Not Appropriate	○
MRV head without IV contrast	Usually Not Appropriate	○
CT head with IV contrast	Usually Not Appropriate	⚠⚠⚠
CT head without and with IV contrast	Usually Not Appropriate	⚠⚠⚠⚠

Variant 5. Child greater than 1 year of age. Known hydrocephalus. No signs or symptoms of shunt malfunction. Follow-up imaging.

Procedure	Appropriateness Category	Relative Radiation Level
MRI head without IV contrast	Usually Appropriate	○
MRI head without IV contrast abbreviated	Usually Appropriate	○
CT head without IV contrast	May Be Appropriate	⚠⚠⚠
Radiography head neck chest abdomen	Usually Not Appropriate	⚠⚠
US head	Usually Not Appropriate	○
MRI head with IV contrast	Usually Not Appropriate	○
MRI head without and with IV contrast	Usually Not Appropriate	○
MRV head with IV contrast	Usually Not Appropriate	○
MRV head without IV contrast	Usually Not Appropriate	○
CT head with IV contrast	Usually Not Appropriate	⚠⚠⚠
CT head without and with IV contrast	Usually Not Appropriate	⚠⚠⚠⚠

Variation 6. Child. Known hydrocephalus with cerebral spinal fluid diversion. Signs or symptoms of shunt malfunction. Follow-up imaging.

Procedure	Appropriateness Category	Relative Radiation Level
Radiography head neck chest abdomen	Usually Appropriate	⦿⦿
MRI head without IV contrast	Usually Appropriate	○
MRI head without IV contrast abbreviated	Usually Appropriate	○
CT head without IV contrast	Usually Appropriate	⦿⦿⦿
US abdomen	May Be Appropriate	○
MRI head without and with IV contrast	May Be Appropriate	○
Nuclear medicine ventriculography	May Be Appropriate	⦿⦿⦿⦿
US head	Usually Not Appropriate	○
MRI head with IV contrast	Usually Not Appropriate	○
MRV head with IV contrast	Usually Not Appropriate	○
MRV head without IV contrast	Usually Not Appropriate	○
CT head with IV contrast	Usually Not Appropriate	⦿⦿⦿
CT head without and with IV contrast	Usually Not Appropriate	⦿⦿⦿⦿

Table 1. Appropriateness category names and definitions

Appropriateness Category Name	Appropriateness Rating	Appropriateness Category Definition
Usually Appropriate	7, 8, or 9	The imaging procedure or treatment is indicated in the specified clinical scenarios at a favorable risk-benefit ratio for patients.
May Be Appropriate	4, 5, or 6	The imaging procedure or treatment may be indicated in the specified clinical scenarios as an alternative to imaging procedures or treatments with a more favorable risk-benefit ratio, or the risk-benefit ratio for patients is equivocal.
May Be Appropriate (Disagreement)	5	The individual ratings are too dispersed from the panel median. The different label provides transparency regarding the panel's recommendation. "May be appropriate" is the rating category and a rating of 5 is assigned.
Usually Not Appropriate	1, 2, or 3	The imaging procedure or treatment is unlikely to be indicated in the specified clinical scenarios, or the risk-benefit ratio for patients is likely to be unfavorable.

Table 2. Relative radiation level designations

RRL	Adult Effective Dose Estimate Range (mSv)	Pediatric Effective Dose Estimate Range (mSv)
○	0	0
⦿	<0.1	<0.03
⦿⦿	0.1-1	0.03-0.3
⦿⦿⦿	1-10	0.3-3
⦿⦿⦿⦿	10-30	3-10
⦿⦿⦿⦿⦿	30-100	10-30

Note: Relative radiation level (RRL) assignments for some of the examinations cannot be made, because the actual patient doses in these procedures vary as a function of a number of factors (eg, region of the body exposed to ionizing radiation, the imaging guidance that is used). The RRLs for these examinations are designated as "varies."

SUMMARY OF LITERATURE REVIEW

Introduction/Background

Hydrocephalus is a disorder of altered cerebrospinal fluid (CSF) physiology and circulation, resulting in abnormal ventricular enlargement associated with increased intracranial pressure [1]. Infants with hydrocephalus typically present with progressive macrocephaly due to the high compliance of the cranial vault, whereas older children more commonly display symptoms of increased intracranial pressure, such as headaches, vomiting, and lethargy [1]. The classic understanding of hydrocephalus as the direct result of obstruction to bulk CSF flow, either in the ventricles or subarachnoid space, is evolving toward models that incorporate various hemodynamic contributions, including arterial pulsation, parenchymal compliance, venous pressure, and cellular water/ion transport mechanisms [2].

The epidemiology of hydrocephalus varies by geography and socioeconomic status. Approximately 400,000 new cases of pediatric hydrocephalus occur annually, with more than 80% arising in low- and middle-income countries [3]. Congenital hydrocephalus frequently results from mutations affecting genes that regulate brain development, particularly those involved in cerebral aqueduct formation, or is associated with open neural tube defects. In high-income countries, posthemorrhagic hydrocephalus associated with preterm birth is a leading cause of hydrocephalus, accounting for nearly 6,000 new cases every year [4]. Hydrocephalus can also be acquired due to other pathological processes that affect CSF pathways in the ventricular system or subarachnoid space, such as infection, hemorrhage from other causes (such as trauma or vascular malformations), or direct mass effect from tumors.

The primary goal of hydrocephalus treatment is to restore CSF homeostasis through surgical CSF diversion. Ventriculoperitoneal shunting is a widely used approach, providing a reliable method for CSF diversion in the short term. Although effective, shunts have high complication rates, with up to a 40% failure rate in the first 2 years due to obstruction, infection, or over-shunting [3,5], and an independent 5% risk of infection with every procedure [6]. Endoscopic third ventriculostomy (ETV), often performed with choroid plexus cauterization, has gained adoption as an alternative approach, particularly for noncommunicating hydrocephalus [1,7]. Those who remain symptom free beyond the initial postoperative months tend to have more durable long-term outcomes compared to shunted patients. Additionally, infection rates are lower than with shunts and overall morbidity is reduced [7].

Special Imaging Considerations

Factors unique to children influence both the choice and appropriateness of imaging. The developmental neuroanatomy

of the skull plays a crucial role, as in newborns and infants, the open fontanelles serve as acoustic windows for sonographic evaluation of the brain parenchyma, ventricles, and extra-axial CSF spaces. Second, the heightened sensitivity of younger children to ionizing radiation necessitates careful selection of imaging modalities and optimized protocols for radiation-based techniques. Third, younger children may be unable to follow commands or remain still during image acquisition, which complicates MRI scans and may necessitate sedation.

In certain cases of congenital hydrocephalus, abnormalities may be detectable via obstetric ultrasound (US) and fetal MRI; however, these modalities fall outside the scope of this document.

Initial Imaging Definition

Initial imaging is defined as imaging at the beginning of the care episode for the medical condition defined by the variant. More than one procedure can be considered usually appropriate in the initial imaging evaluation when:

- There are procedures that are equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care)

OR

- There are complementary procedures (ie, more than one procedure is ordered as a set or simultaneously wherein each procedure provides unique clinical information to effectively manage the patient's care).

DISCUSSION OF PROCEDURES BY VARIANT

Variant 1: Infant. Suspected hydrocephalus. Abnormal prenatal imaging or signs or symptoms of elevated intracranial pressure. Initial imaging

Infants presenting with hydrocephalus are a heterogeneous group of patients, including those with known prenatal abnormalities, those with undetected anomalies on prenatal screening US, and a subgroup who experience a perinatal insult, such as hemorrhage or infection, leading to a derangement in CSF dynamics.

Genetic conditions can manifest prenatally or in the early postnatal period. Several mutations with Mendelian inheritance are known to cause congenital hydrocephalus, most of which follow an X-linked pattern. These include *LICAM*, which accounts for approximately 10% of cases in boys, as well as *OTUD5*, *AP1S2*, and *Xp22.33* duplications. Other patterns of inheritance have also been described, including autosomal recessive mutations in *MPDZ*, *CCDC88C*, and genes associated with dystroglycanopathies [8]. In addition to ventriculomegaly,

many of these conditions are associated with aqueductal stenosis.

Hydrocephalus may also be associated with complex malformation patterns, such as open neural tube defects. Normal brain and CSF pathway development is significantly impaired by the presence of an open defect, which allows CSF leakage and permits amniotic fluid entry into the neural tube. The resulting complex abnormalities include Chiari II malformations, gray matter heterotopia, callosal dysgenesis, and ventriculomegaly. Up to 80% of patients with postnatally repaired myelomeningoceles develop hydrocephalus and require CSF diversion, compared to only 40% in those who underwent prenatal repair [9].

The epidemiology of acquired hydrocephalus in the perinatal period varies significantly by geography and socioeconomic conditions [3]. In high-income countries, most cases are related to posthemorrhagic hydrocephalus in the setting of preterm birth. The risk of hemorrhage with intraventricular extension is particularly high in very-low-birth-weight infants (<1,500 g), affecting approximately 25%. In turn, an estimated 20% to 25% of patients with high-grade intraventricular hemorrhage of prematurity will develop hydrocephalus [4]. Globally, the burden of disease is much higher due to postinfectious hydrocephalus, which is most often acquired during or immediately after birth [3]. The severe inflammation associated with neonatal meningitis damages cilia, causes scarring, and obstructs drainage pathways and cisternal spaces. Approximately 35% of newborns with neonatal meningitis develop postinfectious hydrocephalus and require CSF diversion [10].

US Head. US is a safe and portable imaging modality that can aid in evaluating infants with suspected hydrocephalus, who have an open anterior fontanelle, particularly those 6 months of age or younger. The use of the fontanelles as acoustic windows allows reliable assessment of the size and configuration of the lateral and third ventricles, as well as the frontal and parietal extra-axial CSF spaces [11]. Assessment of the fourth ventricle and posterior fossa cisterns is more challenging, although these can be visualized via a transmastoid approach. US measurements for evaluating hydrocephalus have been shown to correlate well with MRI findings [12]. US may be useful as an initial assessment of suspected hydrocephalus, when the primary interest is in supratentorial ventricular caliber, for example with history of repaired myelomeningocele, fetal ventriculomegaly, or new concern for hydrocephalus in setting of prior germinal matrix hemorrhage.

MRI Head Without IV Contrast. Noncontrast MRI is the usual imaging modality for evaluating infants with suspected hydrocephalus. Its high soft tissue contrast is a critical

advantage in this population. MRI demonstrates both supra- and infratentorial ventricular caliber, is sensitive for intracranial hematoma, and characterizes tumors resulting in obstruction. High spatial resolution T2-weighted or flow sensitive sequences may characterize the level of CSF bulk flow obstruction, if any, distinguishing obstructive versus communicating hydrocephalus [13,14]. Detailed anatomy of the third ventricular floor and adjacent adhesions may inform surgical planning and decision making regarding ETV [15,16]. The primary disadvantage is the longer acquisition time, which can lead to motion artifacts or require sedation for a diagnostic examination. However, many young infants can tolerate the examination without sedation during natural sleep, with feed and swaddle techniques efficacious [17].

MRI Head Without IV Contrast Abbreviated. Abbreviated MRI (AB-MRI) protocols address one of the limitations of comprehensive multisequence noncontrast MRI examinations, long acquisition time and consequent motion artifact, by using motion-insensitive and fast sequences. These protocols usually employ single-shot T2-weighted sequences, which allow single-slice acquisition in about a second and whole brain coverage is under a minute. No sedation is required. Supra- and infratentorial ventricles as well as extra-axial CSF spaces are well delineated, with parenchymal assessment less robust than on conventional MRI [18,19]. An AB-MRI protocol for initial assessment of hydrocephalus may be useful when the primary clinical interest is ventricular caliber, for example with a history of repaired myelomeningocele or fetal ventriculomegaly.

MRI Head Without and With IV Contrast. If the clinical findings are suspicious for a meningitis or an infectious etiology, MRI head without and with intravenous (IV) contrast may be considered. It readily demonstrates both supra- and infratentorial ventricular caliber. The addition of gadolinium-based contrast in the initial evaluation may reveal active leptomeningeal inflammation, as well as relevant complications including ventriculitis, cerebritis, parenchymal abscess, extra-axial empyema, and infarction [20].

CT Head Without IV Contrast. CT head without IV contrast may be appropriate in the assessment of an infant with suspected hydrocephalus. It demonstrates both supra- and infratentorial ventricular caliber, is sensitive for intra- and extra-axial hemorrhage, and detects mass lesions resulting in ventricular obstruction. Basal cistern caliber and herniation are readily depicted on accompanying multiplanar reformats. When an infant's clinical or neurologic status dictates urgent imaging, multidetector CT acquisition of the head takes only seconds, mitigating motion

artifacts and abrogating the need for sedation. Furthermore, in settings where ventricular caliber is the primary concern, for example with a history of repaired myelomeningocele or fetal ventriculomegaly, lower dose CT techniques allow for the targeted assessment of ventricular size and extra-axial CSF space caliber, with only cursory evaluation of the parenchyma [21,22].

MRI Head With IV Contrast. There is no relevant literature to support the use of MRI head with IV contrast in the initial evaluation of an infant with suspected hydrocephalus.

MRV Head With IV Contrast. There is no relevant literature to support the use of MR venography (MRV) head with IV contrast in the initial evaluation of an infant with suspected hydrocephalus.

MRV Head Without IV Contrast. There is no relevant literature to support the use of MRV head without IV contrast in the initial evaluation of an infant with suspected hydrocephalus.

CT Head With IV Contrast. There is no relevant literature to support the use of CT of the head with IV contrast in an infant with suspected hydrocephalus. If the clinical findings are suspicious for meningitis or an infectious etiology as the source of the symptomatology, this examination could be considered.

CT Head Without and With IV Contrast. There is no relevant literature to support the use of CT head without and with IV contrast in the initial evaluation of an infant with suspected hydrocephalus.

Variant 2: Infant. Known hydrocephalus. Follow-up imaging

Management of hydrocephalus often requires follow-up imaging to assess for stability of ventricle size, shunt complications, or shunt failure. Complications of ventriculoperitoneal shunting can include malfunctions of the catheter or reservoir, infection, or overdrainage. In general, about a third of shunts fail by 1 year and up to 80% by 10 years [23]. Reported revision rates, however, are as high as 72% for patients with posthemorrhagic hydrocephalus and 67% for those with congenital defects, with a higher shunt revision rate for patients <6 months of age [24].

As an alternative to ventriculoperitoneal shunting placement, ETV with or without choroid plexus cauterization can be performed. Long-term success rates of ETV are variable but in selected patients approach 60%, with success rates correlating with the ETV success score. In this score, older age, absence of previous shunting, and obstructive etiologies about aqueduct/tectum favorably

affect outcome [16]. Imaging may influence patient selection by characterizing etiology and the morphology of aqueduct, third ventricular floor, and adjacent cisterns, as well as in follow-up to assure stability of ventricular size and ventriculostomy patency [25,26].

MRI Head Without IV Contrast. Infants with known hydrocephalus and CSF diversion may be followed using noncontrast MRI of the head, which facilitates assessment of supra- and infratentorial ventricular caliber as well as shunt continuity. In those infants with previous third ventriculostomy, high-resolution T2-weighted sequences or flow sequences may assess patency [26,27]. The favorable safety profile of nonsedated MRI is especially important in this patient population, who often require repeat imaging studies. However, as infants grow, the likelihood of requiring sedation for the acquisition of a comprehensive MRI examination increases, which may necessitate consideration of alternate imaging strategies.

MRI Head Without IV Contrast Abbreviated. AB-MRI protocols address one of the primary limitations of comprehensive multisequence noncontrast MRI examinations, long acquisition time and consequent motion artifact, by using motion-insensitive and fast sequences. These protocols usually employ single-shot T2-weighted sequences, which allow single-slice acquisition in about a second and whole brain coverage is under a minute. As such, no sedation is required. An AB-MRI protocol for assessment of hydrocephalus in the setting of CSF diversion is usually appropriate for surveillance, as it allows assessment of supra- and infratentorial ventricular caliber, as well as accompanying size of extra-axial CSF spaces [18,19]. Direct shunt visualization may be more challenging with single-shot sequences, with the author's institution adding a fast gradient echo T1 or T2 sequence to assist in this regard.

Radiography Head Neck Chest Abdomen. Radiography of the neck, chest, and abdomen can be useful in evaluating suspected shunt malfunction, particularly to assess for shunt discontinuity, disconnection, fracture, or migration. Shunt series radiographs provide a rapid method to evaluate the entire shunt tract from the skull to the peritoneum. In symptomatic patients, radiographs are typically used in conjunction with cranial imaging to help localize potential shunt failure. In asymptomatic patients undergoing routine surveillance, radiography is generally not indicated unless there is clinical concern for hardware complications.

US Head. Cranial US may be a useful imaging modality for infants with known hydrocephalus who have open fontanelles, particularly those 6 months of age or younger. It may be considered for follow-up in infants who are

asymptomatic or require frequent monitoring. The use of the anterior fontanelle as an acoustic window allows reliable assessment of the size and configuration of the lateral and third ventricles, as well as the frontal and parietal extra-axial CSF spaces [11]. Assessment of the fourth ventricle and posterior fossa cisterns is more challenging, although these can be visualized via a transmastoid approach. US measurements for evaluating hydrocephalus have been shown to correlate well with MRI [12].

CT Head Without IV Contrast. Noncontrast head CT may be a useful modality for longitudinal evaluation of patients with known hydrocephalus and CSF diversion. Lower dose CT techniques allow targeted assessment of supra- and infratentorial ventricular caliber, accompanying size of sulci and cisterns, and continuity of shunt if present [21,22].

MRI Head With IV Contrast. There is no relevant literature to support the use of MRI head with IV contrast as follow-up imaging in infants with known hydrocephalus.

MRI Head Without and With IV Contrast. There is no relevant literature to support the use of MRI head without and with IV contrast as follow-up imaging in infants with known hydrocephalus. This may be considered, however, when clinical suspicion for intracranial infection is high.

MRV Head With IV Contrast. There is no relevant literature to support the use of MRV head with IV contrast as follow-up imaging in infants with known hydrocephalus.

MRV Head Without IV Contrast. There is no relevant literature to support the use of MRV head without IV contrast as follow-up imaging in infants with known hydrocephalus.

CT Head With IV Contrast. There is no relevant literature to support the use of CT head with IV contrast as follow-up imaging in infants with known hydrocephalus.

CT Head Without and With IV Contrast. There is no relevant literature to support the use of CT head without and with IV contrast as follow-up imaging in infants with known hydrocephalus.

Variant 3: Infant. Macrocephaly. No signs or symptoms of elevated intracranial pressure. Initial imaging

Macrocephaly is defined as head circumference exceeding two standard deviations above normative data, placing it above the 97th percentile of the population [28]. In asymptomatic and otherwise healthy infants with macrocephaly, imaging is usually normal or reveals benign enlargement of the subarachnoid spaces (BESS) [28,29].

BESS typically presents clinically with a rapid increase in head circumference around 6 months of postnatal age, without signs of acute neurological illness. Localizing symptoms, such as a downward gaze, are absent. Findings on physical examination are generally mild and variably present, including mild frontal bossing, prominence of the fontanelle, irritability, or hypotonia [30]. The pathophysiology of BESS is not fully understood, but is frequently attributed to immature arachnoid villi, which temporarily impair CSF resorption [31,32]. Neuroimaging in BESS typically demonstrates prominence of the subarachnoid spaces, particularly in the frontal regions, with only mild to moderate enlargement of the ventricular system [33]. BESS does not typically require imaging follow-up, as the progressive maturation of the arachnoid villi leads to normalization of head growth by approximately 18 months of age. Occasionally, infants with BESS develop small subdural collections, which are usually inconsequential [34].

A large series of asymptomatic infants with imaging for macrocephaly have demonstrated a 1% to 2% incidence of clinically significant findings. This includes instances of hydrocephalus secondary to aqueductal stenosis or intracranial tumor, as well as subdural hemorrhages attributed to nonaccidental trauma [28,29,35].

US Head. Cranial US offers a rapid and portable method to assess supratentorial ventricular size and extra-axial CSF spaces. In cases of BESS, it demonstrates abundant anechoic CSF along the convexities of the frontal and parietal lobes. Although most asymptomatic infants with macrocephaly will demonstrate normal imaging or BESS, 8 of 440 (1.8%) such infants screened by US in a series by Thomas et al [29] demonstrated clinically relevant findings including aqueductal stenosis with hydrocephalus, intracranial tumors, and subdural hematomas. In a series by Naffaa et al [35], 4 out of 318 such infants (1.2%), 3 to 6 months of age, screened with US needed neurosurgical intervention, again for hydrocephalus, intracranial tumor, and subdural hematoma associated with nonaccidental trauma. Haws et al [33] found that no additional imaging was required following a US diagnosis of benign macrocrania/BESS in a series of 466 infants without neurologic findings.

MRI Head Without IV Contrast. Noncontrast MRI provides high-resolution assessment of the ventricular system, extra-axial CSF spaces, and brain parenchyma. MRI reliably detects the characteristic findings of BESS, one of the more common etiologies of asymptomatic macrocephaly, while simultaneously evaluating for alternative causes. However, it may require sedation in older infants and is time intensive [28].

MRI Head Without IV Contrast Abbreviated. AB-MRI protocols, consisting primarily of single-shot T2-

weighted sequences, can be used to assess the ventricular system and extra-axial CSF spaces, reliably detecting characteristic findings of BESS. Missios et al [36], found that 97.5% of 102 asymptomatic macrocephaly patients screened with quick (abbreviated) brain MRI required no additional imaging.

CT Head Without IV Contrast. Noncontrast head CT can assess ventricular size, evaluate extra-axial CSF space enlargement and/or fluid collections, and identify space occupying lesions including masses. Sedation is usually not necessary, and CT can be used in older infants with a closing fontanelle when US may have a limited acoustic window [28].

Radiography Head Neck Chest Abdomen. There is no relevant literature to support the use of radiography in the initial evaluation of an asymptomatic infant with macrocephaly.

MRI Head With IV Contrast. There is no relevant literature supporting the use of MRI head with IV contrast in the initial evaluation of an asymptomatic infant with macrocephaly.

MRI Head Without and With IV Contrast. There is no relevant literature supporting the use of MRI head without and with IV contrast in the initial evaluation of an asymptomatic infant with macrocephaly.

MRV Head With IV Contrast. There is no relevant literature to support the use of MRV head with IV contrast in the initial evaluation of an asymptomatic infant with macrocephaly.

MRV Head Without IV Contrast. There is no relevant literature to support the use of MRV head without IV contrast in the initial evaluation of an asymptomatic infant with macrocephaly.

CT Head With IV Contrast. There is no relevant literature to support the use of contrast-enhanced head CT in the initial evaluation of an asymptomatic infant with macrocephaly.

CT Head Without and With IV Contrast. There is no relevant literature to support the use of CT head without and with IV contrast in the initial evaluation of an asymptomatic infant with macrocephaly.

Variant 4: Child greater than 1-year of age. Suspected hydrocephalus. Signs or symptoms of elevated intracranial pressure. Initial imaging

In older children whose skulls are less compliant, hydrocephalus more frequently manifests with signs and symptoms of elevated intracranial pressure. It is important to

recognize that even in this group of patients, the presentation is influenced by the acuity with which the inciting cause develops. Acute hydrocephalus is more frequently obstructive and can present with signs and symptoms including headache, nausea, vomiting, altered mental status, papilledema, and vision changes. If not recognized in a timely manner, acute hydrocephalus may be lethal.

The role of imaging at initial presentation is to confirm the diagnosis of acute hydrocephalus by demonstrating abnormal dilation of the ventricular system, documenting the presence of transependymal edema as a surrogate marker for acuity, and potentially outlining the etiology. This facilitates triage to an appropriate intervention that relieves the obstruction and potentially addresses the underlying cause.

In childhood, brain tumors frequently present with obstructive hydrocephalus. Approximately half of pediatric brain tumors arise in the posterior fossa, where the confined space and intricate anatomy predispose lesions to obstruct the cerebral aqueduct, fourth ventricle, or its outlets. Both high-grade tumors, such as medulloblastoma, ependymoma, and atypical teratoid/rhabdoid tumor, as well as low-grade tumors, such as pilocytic astrocytoma, can present with equally florid symptoms. Lesions in the supratentorial space can also result in hydrocephalus, including those at the foramen of Monro (eg, subependymal giant cell astrocytoma, colloid cyst).

Nonneoplastic lesions can also result in acute obstruction leading to symptomatic elevation of intracranial pressure. Rarely, infectious agents such as intraventricular neurocysticercosis can obstruct CSF drainage either due to associated inflammation or mass effect from cystic lesions. Congenital lesions such as aqueductal stenosis and webs have a more insidious onset and are therefore less likely to present with a florid case of intracranial pressure elevation.

MRI Head Without IV Contrast. Noncontrast MRI is the usual imaging modality for evaluating children with suspected hydrocephalus. Its high soft tissue contrast is a critical advantage in this population. MRI demonstrates both supra- and infratentorial ventricular caliber, is sensitive for intracranial hematoma, and characterizes tumors resulting in obstruction. High spatial resolution T2-weighted or flow sensitive sequences may characterize the level of CSF bulk flow obstruction, if any, distinguishing obstructive versus communicating hydrocephalus [13,14]. Detailed anatomy of the third ventricular floor and adjacent adhesions may inform surgical planning and decision making regarding ETV [15,16]. The primary disadvantage is the longer acquisition time, which can lead to motion artifacts or require sedation for a diagnostic examination.

MRI Head Without IV Contrast Abbreviated. AB-MRI protocols address one of the limitations of comprehensive multisequence noncontrast MRI examinations, long acquisition time and consequent motion artifact, by using motion-insensitive and fast sequences. These protocols usually employ single-shot T2-weighted sequences, which allow single-slice acquisition in about a second and whole brain coverage is under a minute. No sedation is required. Supra- and infratentorial ventricles, as well as extra-axial CSF spaces are well delineated, with parenchymal assessment less robust than on conventional MRI [18,19]. As such, in children with signs or symptoms of elevated intracranial pressure, AB-MRI may be useful as the initial assessment of ventricular caliber, but may be limited in characterizing underlying hydrocephalus etiology.

CT Head Without IV Contrast. Noncontrast head CT is a ubiquitous modality for the rapid assessment of suspected hydrocephalus [21]. It demonstrates both supra- and infratentorial ventricular caliber, is sensitive for intra- and extra-axial hemorrhage, and detects mass lesions resulting in ventricular obstruction. Basal cistern caliber and herniation are readily depicted on accompanying multiplanar reformats. When a child's clinical or neurologic status dictates urgent imaging, multidetector CT acquisition of the head takes only seconds, mitigating motion artifacts and abrogating the need for sedation.

MRI Head Without and With IV Contrast. If clinical findings are suspicious for meningitis or an infectious etiology as a cause of hydrocephalus, initial MRI head without and with IV contrast may be considered. It readily demonstrates both supra- and infratentorial ventricular caliber. The addition of gadolinium-based contrast in the initial evaluation may reveal active leptomeningeal inflammation, as well as relevant complications including ventriculitis, cerebritis, parenchymal abscess, extra-axial empyema, and infarction [20]. Contrast-enhanced imaging may also be useful if there is a high clinical suspicion of findings or noncontrast MRI findings suggesting a vascular lesion, tumor, or other abnormality with disruption of the blood brain barrier.

US Head. In older children with closed fontanelles, cranial US has no significant role in the initial evaluation of suspected hydrocephalus in children. Its usefulness is limited to infants.

MRI Head With IV Contrast. There is no relevant literature to support the use of MRI head with IV contrast in the initial evaluation of suspected hydrocephalus in children.

MRV Head With IV Contrast. There is no relevant literature supporting the use of contrast-enhanced MRV in

the initial evaluation of suspected hydrocephalus in children. MRV may be considered if there is clinical concern for venous sinus thrombosis or anomalies contributing to elevated intracranial pressure.

MRV Head Without IV Contrast. There is no relevant literature supporting the use of noncontrast MRV in the initial evaluation of suspected hydrocephalus in children. MRV may be considered if there is clinical concern for venous sinus thrombosis or anomalies contributing to elevated intracranial pressure.

CT Head With IV Contrast. There is no relevant literature supporting the use of contrast-enhanced head CT in the initial evaluation of suspected hydrocephalus in children. A contrast-enhanced head CT may be considered if there is clinical suspicion for neoplasm, vascular malformation, or infection.

CT Head Without and With IV Contrast. There is no relevant literature to support the use of CT head without and with IV contrast in the initial evaluation of suspected hydrocephalus in children.

Variant 5: Child greater than 1-year of age. Known hydrocephalus. No signs or symptoms of shunt malfunction. Follow-up imaging

Surveillance of asymptomatic children with CSF diversion, beyond stabilization of the hydrocephalus, aims to identify individuals with late-onset shunt or ETV failure. The radiological surveillance of these children lacks standardization and is highly dependent on the practitioner. Imaging surveillance carries the theoretical potential of identifying occult shunt malfunction or ETV failure before catastrophic consequences ensue [37]. Insufficient CSF drainage may go unrecognized if signs and symptoms are subtle, if the patient's baseline neurological status is severely compromised, or if personnel involved in care are unfamiliar with its manifestations. Evidence suggests that <4% of shunt revisions occur as a result of a routine visit in an asymptomatic child [38].

In a recent survey of pediatric neurosurgeons, 75% of respondents reported obtaining imaging in asymptomatic patients with a ventricular shunt. Among these, 33% obtained annual imaging, 26% every 2 to 3 years, and 15% every 5 to 10 years. A slightly lower percentage of respondents (67%) performed imaging for asymptomatic patients with an ETV. Of these, 41% conducted imaging annually, 18% every 2- to 3-years, and 12% every 5- to 10-years [37]. Independent of the modality of CSF diversion used, respondents of this survey favor imaging.

MRI Head Without IV Contrast. Noncontrast MRI is a useful modality for following asymptomatic children with

known hydrocephalus. It readily demonstrates both supra- and infratentorial ventricular caliber. In those children with previous third ventriculostomy, high-resolution T2-weighted sequences or flow sequences may assess patency [26,27].

MRI Head Without IV Contrast Abbreviated. AB-MRI is the usual modality for routine surveillance in asymptomatic children with known or treated hydrocephalus. Abbreviated protocols address one of the primary limitations of comprehensive multisequence noncontrast MRI examinations, long acquisition time and consequent motion artifact, by using motion-robust and fast sequences. These protocols usually employ single-shot T2-weighted sequences, which allow single-slice acquisition in about a second and whole brain coverage is under a minute. As such, no sedation is required. An AB-MRI protocol for assessment of hydrocephalus allows assessment of supra- and infratentorial ventricular caliber, as well as accompanying size of extra-axial CSF spaces [14,18]. Direct shunt visualization may be more challenging with single-shot sequences, with the author's institution adding a fast gradient echo T1 or T2 sequence to assist in this regard.

CT Head Without IV Contrast. Noncontrast head CT may be a useful modality for longitudinal evaluation of patients with known hydrocephalus and CSF diversion. Lower dose CT techniques allow targeted assessment of supra- and infratentorial ventricular caliber, accompanying size of sulci and cisterns, and continuity of visualized shunt if present [21,22].

Radiography Head Neck Chest Abdomen. Radiography is usually unwarranted in the routine follow-up of asymptomatic children with hydrocephalus. Shunt series radiographs may be considered if there is specific clinical concern for hardware complications, such as fracture, disconnection, or migration. However, in asymptomatic patients without suspicion for shunt malfunction, routine radiographic surveillance is not indicated.

US Head. In older children with closed fontanelles, cranial US has no significant role in the follow-up imaging of asymptomatic children with known hydrocephalus. Its usefulness is limited to infants.

MRI Head With IV Contrast. There is no relevant literature supporting the use of MRI head with IV contrast in the follow-up imaging of asymptomatic children with known hydrocephalus.

MRI Head Without and With IV Contrast. There is no relevant literature supporting the use of MRI head without and with IV contrast in the follow-up imaging of asymptomatic children with known hydrocephalus.

MRV Head With IV Contrast. There is no relevant literature supporting the use of MRV head with IV contrast in the follow-up imaging of asymptomatic children with known hydrocephalus.

MRV Head Without IV Contrast. There is no relevant literature supporting the use of noncontrast MRV head in the follow-up imaging of asymptomatic children with known hydrocephalus.

CT Head With IV Contrast. There is no relevant literature to support the use of contrast-enhanced head CT in the follow-up imaging of asymptomatic children with known hydrocephalus.

CT Head Without and With IV Contrast. There is no relevant literature to support the use of CT head without and with IV contrast in the follow-up imaging of asymptomatic children with known hydrocephalus.

Variant 6: Child. Known hydrocephalus with cerebral spinal fluid diversion. Signs or symptoms of shunt malfunction. Follow-up imaging

The two most common forms of CSF diversion in pediatric hydrocephalus are ventricular shunt catheters and ETV. The approach to a child with CSF diversion who presents with symptoms concerning for perturbed or worsening CSF dynamics varies depending on the type of diversion in place.

Ventricular shunt catheters most commonly consist of a proximal intracranial catheter, a subcutaneous reservoir, a one-way valve, and a distal catheter. The proximal catheter resides in the CSF, exits through a burr hole, and connects to the distal catheter via the reservoir and valve. The distal catheter typically terminates in the peritoneum, with less common placements in the pleural space or right heart atrium. Early complications of ventricular shunt placement may result from incorrect positioning, segment disconnection, or infection. Late-stage complications can include catheter breakage or migration, obstruction, over-drainage, or infection [39].

Children with shunt failure generally present with signs of elevated intracranial pressure, such as nausea, vomiting, seizures, irritability, headaches, and altered mental status. To identify abnormalities in catheter integrity, such as kinks, disconnections, or fractures, imaging of the entire catheter course is necessary, often using conventional radiography. To assess catheter obstruction, infection, CSF accumulation, or over-drainage, imaging of the brain—and occasionally the abdomen to evaluate the distal tip—may be required. In equivocal cases, catheter patency and flow can be assessed by injecting a radionuclide or iodinated contrast agent into the reservoir and documenting its transit using scintigraphy or fluoroscopy/CT.

ETV has gained popularity as a CSF diversion option in suitable patients. Compared to ventricular shunts, ETV may carry a higher risk of failure within the first 3 months. However, beyond this period, the risk of failure is lower for ETV and continues to decrease over time [7]. Direct evaluation of ETV patency can only be performed using MRI due to its soft tissue resolution and sensitivity to flow. In the absence of MRI, surrogate markers of failure such as enlarging ventricles may be used.

Radiography Head Neck Chest Abdomen. Shunt series, frontal and lateral radiographs of the head, neck, chest, and abdomen, are used to detect mechanical disruption of the shunt catheter, including fracture (usually in the neck), disconnection at the valve, and kinking. Radiographs may also suggest adhesions or pseudocysts at the abdominal level. However, shunt failure in children is more commonly due to intrinsic catheter obstruction, which is occult radiographically. Sensitivity for shunt failure is therefore low, below 20% [40,41]. As such, radiographs are usually used in conjunction with cranial cross-sectional imaging.

MRI Head Without IV Contrast. Noncontrast head MRI is useful for evaluating children with suspected shunt malfunction. Its high soft tissue contrast is a critical advantage in this population. MRI demonstrates both supra- and infratentorial ventricular caliber, cranial shunt catheter components, and allows visualization of transpendymal edema. In those children with previous third ventriculostomy, high-resolution T2-weighted sequences or flow sequences may assess patency [26,27].

MRI Head Without IV Contrast Abbreviated. In the acutely symptomatic child with shunted hydrocephalus, AB-MRI protocols are useful and frequently considered a first-line strategy. AB-MRI address one of the primary limitations of comprehensive multisequence noncontrast MRI examinations, long acquisition time and consequent motion artifact, by using motion-insensitive and fast sequences. These usually employ single-shot T2-weighted sequences, which allow single-slice acquisition in about a second and whole brain coverage is under a minute. No sedation is required. AB-MRI protocols reliably assess supra- and infratentorial ventricular caliber, as well as accompanying size of extra-axial CSF spaces [18,19]. Direct shunt catheter visualization may be more challenging with single-shot sequences, with the author's institution adding fast gradient echo T1 or T2 sequences to assist in this regard.

CT Head Without IV Contrast. Noncontrast head CT is a widely used and effective imaging modality for evaluating suspected shunt malfunction in symptomatic children [42,43]. It allows rapid assessment of ventricular size and the position of cranial shunt components. Lower dose CT

techniques allow for the targeted assessment of ventricular size and extra-axial CSF space caliber, with only cursory evaluation of the parenchyma [21,22].

US Abdomen. Abdominal US is not usually indicated in the initial imaging of suspected shunt malfunction. However, it may be useful when there is clinical concern for abdominal complications related to the distal shunt catheter, such as pseudocyst formation, ascites, bowel perforation, or infection. US offers a noninvasive method to evaluate for these complications.

MRI Head Without and With IV Contrast. There is no relevant literature to support the use of MRI head without and with IV contrast in the evaluation of suspected shunt malfunction. It might be considered if there is a high suspicion of intracranial infection accompanying clinically suspected shunt malfunction.

Nuclear Medicine Ventriculography. Nuclear medicine shuntography is not routinely used for initial imaging in suspected shunt malfunction. It may be employed secondarily, for example, when clinical concern for shunt dysfunction persists in the setting of stable ventricular caliber, or when a shunt is radiographically fractured but there are no clinical symptoms (to assess for functioning fibrous tract) [44]. With ventricular reflux of radionuclide and prompt distal drainage and dispersion following injection into shunt reservoir, Thompson et al [45] reported a negative predictive value exceeding 90%.

US Head. In older children with closed fontanelles, cranial US has no significant role in the evaluation of suspected shunt malfunction. Its usefulness is limited to infants with an open anterior fontanelle.

MRI Head With IV Contrast. There is no relevant literature to support the use of MRI head with IV contrast in the evaluation of suspected shunt malfunction.

MRV Head With IV Contrast. There is no relevant literature to support the use of MRV head with IV contrast in the evaluation of suspected shunt malfunction.

MRV Head Without IV Contrast. There is no relevant literature to support the use of noncontrast MRV head in the evaluation of suspected shunt malfunction.

CT Head With IV Contrast. There is no relevant literature to support the use of contrast-enhanced head CT in the evaluation of suspected shunt malfunction.

CT Head Without and With IV Contrast. There is no relevant literature to support the use of CT head without and with IV contrast in the evaluation of suspected shunt malfunction.

SUMMARY OF HIGHLIGHTS

This is a summary of the key recommendations from the variant tables. Refer to the complete narrative document for more information.

■ **Variants 1 and 2:** For infants with suspected hydrocephalus due to abnormal prenatal imaging (fetal ventriculomegaly), for those presenting with signs and symptoms that suggest increased intracranial pressure, and for follow-up imaging, MRI head without IV contrast or alternatively AB-MRI head without IV contrast is recommended. These examinations accurately assess ventricular size, characterize level of obstruction if any, and assess for underlying causes including tumors or hemorrhage. AB-MRI is particularly apropos when the main objective is assessment of current ventricular caliber. Alternatively, US of the head is appropriate, initially or in follow-up, when ventricular caliber is the primary concern and the anterior fontanelle is open. CT head without IV contrast may be considered when an infant's clinical or neurologic status dictates urgent and rapid imaging. If infection is a concern at initial assessment, MRI head without and with IV contrast may assess for accompanying intracranial complications, including abscess or empyema. Radiography of the head, neck, chest, and abdomen (shunt series) may be used in follow-up of symptomatic infants or when there is clinical concern for shunt disconnection.

■ **Variant 3:** In the evaluation of infants with macrocephaly, but no accompanying neurologic signs or symptoms, US head, MRI head without IV contrast, or AB-MRI is recommended. These usually demonstrate normal findings or reveal benign external enlargement of the subarachnoid spaces. When abnormal, underlying obstructive hydrocephalus or subdural collections associated with nonaccidental trauma may be detected.

■ **Variants 4, 5, and 6:** For children with suspected hydrocephalus due to signs and symptoms that suggest increased intracranial pressure as well as for both asymptomatic or symptomatic follow-up imaging, MRI head without IV contrast or alternatively AB-MRI is recommended to accurately assess ventricular size, characterize level of obstruction if any, and assess for underlying causes. AB-MRI is particularly apropos when the main objective is assessment of current ventricular caliber. CT head without IV contrast is an alternative recommendation at initial presentation or during symptomatic follow-up when clinical or neurologic status dictates urgent and rapid

imaging. If infection is a concern at initial assessment, MRI head without and with IV contrast may assess for accompanying intracranial complications, including abscess or empyema. Radiography of the head, neck, chest, and abdomen (shunt series) is recommended in follow-up of symptomatic children but is insensitive in isolation and complements cross-sectional MRI or CT imaging.

SUPPORTING DOCUMENTS

The evidence table, literature search, and appendix for this topic are available at <https://acsearch.acr.org/list>. The appendix includes the strength of evidence assessment and the final rating round tabulations for each recommendation.

For additional information on the Appropriateness Criteria methodology and other supporting documents, please go to the ACR website at <https://www.acr.org/Clinical-Resources/Clinical-Tools-and-Reference/Appropriateness-Criteria>.

GENDER EQUALITY AND INCLUSIVITY CLAUSE

The ACR acknowledges the limitations in applying inclusive language when citing research studies that predates the use of the current understanding of language inclusive of diversity in sex, intersex, gender, and gender-diverse people. The data variables regarding sex and gender used in the cited literature will not be changed. However, this guideline will use the terminology and definitions as proposed by the National Institutes of Health [46].

RELATIVE RADIATION LEVEL INFORMATION

Potential adverse health effects associated with radiation exposure are an important factor to consider when selecting the appropriate imaging procedure. Because there is a wide range of radiation exposures associated with different diagnostic procedures, a relative radiation level (RRL) indication has been included for each imaging examination. The RRLs are based on effective dose, which is a radiation dose quantity that is used to estimate population total radiation risk associated with an imaging procedure. Patients in the pediatric age group are at inherently higher risk from exposure, because of both organ sensitivity and longer life expectancy (relevant to the long latency that appears to accompany radiation exposure). For these reasons, the RRL dose estimate ranges for pediatric examinations are lower as compared with those specified for adults (see [Table](#) below). Additional information regarding radiation dose assessment for imaging examinations can be found in the ACR

Appropriateness Criteria® Radiation Dose Assessment
Introduction document [47].

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