American College of Radiology ACR Appropriateness Criteria® Imaging of Suspected Intracranial Hypotension

<u>Variant 1:</u>
Adult. Orthostatic headache from suspected intracranial hypotension, without recent spinal intervention that could cause CSF leakage. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level	
MRI complete spine without and with IV contrast	Usually Appropriate	0	
MRI complete spine without IV contrast	Usually Appropriate	0	
MRI head without and with IV contrast	Usually Appropriate	0	
MRI head without IV contrast	Usually Appropriate	0	
CT myelography complete spine	May Be Appropriate	❖❖❖❖❖	
Radiographic myelography digital subtraction complete spine	Usually Not Appropriate	❖❖❖❖	
MR myelography complete spine	Usually Not Appropriate	0	
MRI complete spine with IV contrast	Usually Not Appropriate	0	
MRI head with IV contrast	Usually Not Appropriate	0	
CT head cisternography	Usually Not Appropriate	❖❖❖	
CT head with IV contrast	Usually Not Appropriate	❖❖❖	
CT head without and with IV contrast	Usually Not Appropriate	૽	
CT head without IV contrast	Usually Not Appropriate	❖❖❖	
CT complete spine with IV contrast	Usually Not Appropriate	❖❖❖❖	
DTPA cisternography	Usually Not Appropriate	**	
CT complete spine without and with IV contrast	Usually Not Appropriate	❖❖❖❖	
CT complete spine without IV contrast	Usually Not Appropriate	❖❖❖❖	
CT myelography dynamic complete spine	Usually Not Appropriate	***	

<u>Variant 2:</u>
Adult. Orthostatic headache from suspected intracranial hypotension within 72 hours of dural puncture or other spinal intervention that could cause CSF leakage. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
Radiographic myelography digital subtraction complete spine	Usually Not Appropriate	❖❖❖❖
MR myelography complete spine	Usually Not Appropriate	0
MRI complete spine with IV contrast	Usually Not Appropriate	0
MRI complete spine without and with IV contrast	Usually Not Appropriate	0
MRI complete spine without IV contrast	Usually Not Appropriate	0
MRI head with IV contrast	Usually Not Appropriate	0
MRI head without and with IV contrast	Usually Not Appropriate	0
MRI head without IV contrast	Usually Not Appropriate	0
CT head cisternography	Usually Not Appropriate	���
CT head with IV contrast	Usually Not Appropriate	❖❖❖
CT head without and with IV contrast	Usually Not Appropriate	❖❖❖
CT head without IV contrast	Usually Not Appropriate	❖❖❖
DTPA cisternography	Usually Not Appropriate	⋧ ��
CT complete spine with IV contrast	Usually Not Appropriate	**
CT complete spine without and with IV contrast	Usually Not Appropriate	❖❖❖❖
CT complete spine without IV contrast	Usually Not Appropriate	❖❖❖❖
CT myelography complete spine	Usually Not Appropriate	***
CT myelography dynamic complete spine	Usually Not Appropriate	***

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<u>Variant 3:</u>

Adult. Orthostatic headache from suspected intracranial hypotension without improvement post 72 hours of dural puncture or other spinal intervention that could cause CSF leakage. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
Radiographic myelography digital subtraction complete spine	Usually Not Appropriate	❖❖❖❖
MR myelography complete spine	Usually Not Appropriate	0
MRI complete spine with IV contrast	Usually Not Appropriate	0
MRI complete spine without and with IV contrast	Usually Not Appropriate	0
MRI complete spine without IV contrast	Usually Not Appropriate	0
MRI head with IV contrast	Usually Not Appropriate	0
MRI head without and with IV contrast	Usually Not Appropriate	0
MRI head without IV contrast	Usually Not Appropriate	0
CT head cisternography	Usually Not Appropriate	**
CT head with IV contrast	Usually Not Appropriate	**
CT head without and with IV contrast	Usually Not Appropriate	♦
CT head without IV contrast	Usually Not Appropriate	**
DTPA cisternography	Usually Not Appropriate	**
CT complete spine with IV contrast	Usually Not Appropriate	***
CT complete spine without and with IV contrast	Usually Not Appropriate	❖❖❖❖
CT complete spine without IV contrast	Usually Not Appropriate	❖❖❖❖
CT myelography complete spine	Usually Not Appropriate ♀�����	
CT myelography dynamic complete spine	Usually Not Appropriate ������	

Variant 4: Adult. Obtundation with initial brain imaging features of suspected intracranial hypotension. Next imaging study.

Procedure	Appropriateness Category	Relative Radiation Level
MRI complete spine without and with IV contrast	Usually Appropriate	0
MRI complete spine without IV contrast	Usually Appropriate	0
CT myelography complete spine	May Be Appropriate	❖❖❖❖❖
Radiographic myelography digital subtraction complete spine	Usually Not Appropriate	❖❖❖❖
MR myelography complete spine	Usually Not Appropriate	0
MRI complete spine with IV contrast	Usually Not Appropriate	0
DTPA cisternography	Usually Not Appropriate	���
CT complete spine with IV contrast	Usually Not Appropriate	***
CT complete spine without and with IV contrast	Usually Not Appropriate	⊕ ���
CT complete spine without IV contrast	Usually Not Appropriate	❖❖❖❖
CT myelography dynamic complete spine	Usually Not Appropriate	***

<u>Variant 5:</u>

Adult. Chronic daily headache from suspected intracranial hypotension with negative initial brain and spine imaging, but with history and clinical examination suggesting CSF leakage. Next imaging study.

Procedure	Appropriateness Category	Relative Radiation Level
Radiographic myelography digital subtraction complete spine	Usually Appropriate	❖❖❖❖
CT myelography dynamic complete spine	Usually Appropriate	***
MR myelography complete spine	May Be Appropriate	0
DTPA cisternography	May Be Appropriate	♦
CT head cisternography	Usually Not Appropriate	**

<u>Variant 6:</u> Adult. Rebound headache following epidural blood patch or fibrin glue patch treatment for suspected intracranial hypotension. Initial imaging.

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Procedure	Appropriateness Category	Relative Radiation Level
Radiographic myelography digital subtraction complete spine	Usually Not Appropriate	ዏዏዏ
MR myelography complete spine	Usually Not Appropriate	0
MRI complete spine with IV contrast	Usually Not Appropriate	0
MRI complete spine without and with IV contrast	Usually Not Appropriate	0
MRI complete spine without IV contrast	Usually Not Appropriate	0
MRI head with IV contrast	Usually Not Appropriate	0
MRI head without and with IV contrast	Usually Not Appropriate	0
MRI head without IV contrast	Usually Not Appropriate	0
CT head cisternography	Usually Not Appropriate	⊕⊕⊕
CT head with IV contrast	Usually Not Appropriate	⊕⊕
CT head without and with IV contrast	Usually Not Appropriate	€€
CT head without IV contrast	Usually Not Appropriate	⊕⊕
DTPA cisternography	Usually Not Appropriate	⊕ ⊕ ⊕
CT complete spine with IV contrast	Usually Not Appropriate	₩₩
CT complete spine without and with IV contrast	Usually Not Appropriate	♦♦♦
CT complete spine without IV contrast	Usually Not Appropriate	❖❖❖❖
CT myelography complete spine	Usually Not Appropriate	♥♥♥♥
CT myelography dynamic complete spine	Usually Not Appropriate	***

IMAGING OF SUSPECTED INTRACRANIAL HYPOTENSION

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Summary of Literature Review

Introduction/Background

The clinical syndrome of intracranial hypotension refers to the symptoms caused by cerebrospinal fluid (CSF) hypovolemia and is primarily characterized by postural headaches [1,2]. According to the *International Classification of Headache Disorders, third edition*, the low CSF pressure headache is typically orthostatic in nature, is caused by low CSF pressure (<6 cm H₂O) or CSF leakage, and remits after normalization of CSF pressure or a successful sealing of the CSF leak [3]. The onset of symptoms may be spontaneous or secondary depending on a temporal relation to a presumed cause, such as a recent procedural dural puncture. Headache symptoms may be accompanied by additional symptomatology to include nausea, vomiting, neck pain, tinnitus, changes in hearing, and photophobia [4].

It is estimated that spontaneous intracranial hypotension (SIH) occurs with an incidence of approximately 5 per 100,000 individuals annually [5]. The true incidence of this condition may be higher, because SIH is thought to be both highly underdiagnosed and a misdiagnosed disorder [4,6-8]. Clinical risk factors for the development of SIH include spinal osteophytes that can perforate the dura, weakened ectatic dura/meningeal cysts as can be seen with collagen vascular disease, and a history of bariatric surgery in which it is postulated that rapid loss of epidural fat may weaken the dura and predispose to CSF leakage [4,9-12]. Secondary intracranial hypotension symptoms related to dural puncture are thought to occur with a frequency of approximately 2% to 8% of cases, with risk factors in part related to needle type and gauge used [13-15].

The pathophysiologic mechanism of headache symptoms and various neurological deficits in patients with intracranial hypotension is not well understood but likely multifactorial and may be attributed to 1) compensatory venodilitation, blood volume expansion, and dural sinus stretching as the body attempts to maintain a stable intracranial volume in response to decreased CSF volume; and 2) downward traction on the meninges, nerves, and brain parenchyma as the brain loses buoyancy and begins to sag in response to decreased CSF volume. The findings of venodilitation and brain sagging have well-defined imaging findings and have been shown to resolve along with clinical improvement of headache symptoms [16,17].

The 3 main causes of intracranial hypotension are CSF leakage through a dural defect, leaking meningeal diverticulum, and CSF-venous fistula [18-20]. It is important to note that in the upright position, intracranial hydrostatic pressure is slightly negative relative to atmosphere, whereas hydrostatic pressure in the spine is positive relative to atmosphere [21]. Consequently, the spine has been shown to represent the anatomical source of most symptomatic CSF leaks and venous fistulas, such that the imaging investigation of leak source should be directed primarily toward the spine and not intracranially [22].

Imaging plays a critical role in the diagnostic evaluation of intracranial hypotension. The goals of imaging are 2-fold: 1) to confirm the diagnosis of intracranial hypotension and 2) to localize the source of leak for targeted therapy such as epidural blood patch, percutaneous fibrin glue treatment, endovascular venous fistula embolization, and surgical dural repair or venous fistula ligation [23,24]. Intracranial imaging features suggestive of intracranial

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hypotension include qualitative signs (engorgement of venous sinuses, pachymeningeal enhancement, midbrain descent, superficial siderosis, subdural hygroma or hematoma, and convex superior surface of the pituitary) and quantitative signs (pituitary height, pontomesencephalic angle, suprasellar cistern, prepontine cistern, midbrain descent, venous-hinge angle, mamillopontine angle, tonsillar descent, and area cavum veli interpositi) [4,16,25-27]. The cumulative presence of these intracranial findings has been shown to correlate with a likelihood of finding a spinal leak source [16]. The spinal imaging findings associated with SIH include direct evidence of CSF leakage via epidural fluid collections and CSF-venous fistula as well as secondary indirect signs of CSF leakage such as dilated epidural venous plexus, subdural hygromas, and dural enhancement [24,28-30]. It should be noted that CSF pressure can be normal in patients with SIH, and the absence of a low CSF pressure should not exclude this condition [31,32]. It also bears mentioning that the diagnosis of SIH is challenging, and in some cases SIH cannot be definitely diagnosed or excluded until a full diagnostic workup with invasive imaging (myelography) has been performed. In cases in which symptoms persist after a negative full imaging workup, the possibility of SIH mimicking pathologies such as positional orthostatic tachycardia syndrome, cervicogenic headaches, migraines, or new daily persistent headache syndromes should be considered [33].

Using the best available evidence, this document provides diagnostic imaging recommendations for intracranial hypotension across various clinical presentations. In cases in which conservative/medical management or a therapeutic procedural intervention may be a more appropriate first step over imaging, the evidence to support such practices is discussed in the variant narrative.

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 where each procedure provides unique clinical information to effectively manage the patient's care).

Discussion of Procedures by Variant

Variant 1: Adult. Orthostatic headache from suspected intracranial hypotension, without recent spinal intervention that could cause CSF leakage. Initial imaging.

This clinical scenario refers to a patient who demonstrates postural orthostatic headaches with or without additional secondary SIH symptoms. The absence of a recent spinal intervention such as a dural puncture is relevant because orthostatic headaches are known, usually self-limiting sequelae of such procedures that do not typically require an imaging workup [4,13]. Orthostatic headaches without a temporal relation to dural puncture usually require 2 initial imaging examinations: brain imaging to help confirm a suspected SIH diagnosis and spine imaging to assist in localizing a potential source of CSF leak [4].

CT Complete Spine With IV Contrast

There is no relevant literature to support the use of CT complete spine with intravenous (IV) contrast in the initial imaging of suspected SIH.

CT Complete Spine Without and With IV Contrast

There is no relevant literature to support the use of CT complete spine without and with IV contrast in the initial imaging of suspected SIH.

CT Complete Spine Without IV Contrast

There is no relevant literature to support the use of CT complete spine without IV contrast in the initial imaging of suspected SIH.

CT Head Cisternography

There is no relevant literature to support the use of CT head cisternography in the initial imaging of suspected SIH. The spine has been shown to represent the anatomical source of most symptomatic CSF leaks and venous fistulas,

such that the imaging investigation of leak source should be directed primarily toward the spine and not intracranially [22].

CT Head With IV Contrast

There is no relevant literature to support the use of CT head with IV contrast in the initial imaging of suspected SIH.

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 imaging of suspected SIH.

CT Head Without IV Contrast

There is no relevant literature to support the use of CT head without IV contrast in the initial imaging of suspected SIH.

CT Myelography Complete Spine

Although CT myelography complete spine can detect epidural contrast collections suggestive of dural defect or leaking meningeal diverticulum, MRI complete spine optimized with fluid sensitive sequences has been shown to detect epidural collections with equal sensitivity and is a preferred initial imaging option over CT myelography complete spine because it avoids the need for lumbar puncture [34-36].

CT Myelography Dynamic Complete Spine

There is no relevant literature to support the use of dynamic CT myelography complete spine in the initial imaging of suspected SIH. Dynamic CT myelography plays an important role in the subsequent imaging workup of SIH after the initial brain and spine imaging [37,38]. Results from an initial spine MRI or conventional CT myelogram provide useful information on how a subsequent dynamic CT myelogram will be performed, such as prone positioning for suspected ventral dural defect or decubitus positioning for suspected leaking meningeal diverticulum or CSF-venous fistula [24,37-39]. Dynamic CT myelography involves an initial scan followed by delayed phase scans in immediate succession and, when performed in the decubitus position, may require 2 separate contrast injections due to the transient temporal characteristics associated with CSF-venous fistula visualization as well as limitations in contrast dosing [40-44].

DTPA Cisternography

Although diethylenetriamine pentaacetate (DTPA) cisternography can detect epidural collections suggestive of dural defect or leaking meningeal diverticulum, MRI complete spine optimized with fluid sensitive sequences is typically the preferred initial imaging option over DTPA cisternography complete spine because it avoids the need for lumbar puncture and has superior spatial resolution for lesion localization [45,46].

MR Myelography Complete Spine

There is no relevant literature to support the use of MR myelography complete spine in the initial imaging of suspected SIH. An MR myelogram with intrathecal gadolinium administration has been used in several studies as a subsequent follow-up imaging examination after initial spine imaging to increase sensitivity for the detection of slow leaking dural and meningeal diverticular defects [47-49]. Some limited evidence suggests a potential role for MR myelography in the detection of CSF-venous fistulas [50]. It should be noted that intrathecal use of gadolinium is currently off label. Although studies have suggested that intrathecal gadolinium can be administered safely in small doses, the safety profile of such imaging is not formally approved for such use, and special dosing caution is required to avoid potential of gadolinium induced neurotoxicity [51].

MRI Complete Spine With IV Contrast

There is no relevant literature to support the use of MRI complete spine with IV contrast in the initial imaging of suspected SIH.

MRI Complete Spine Without and With IV Contrast

MRI complete spine without and with IV contrast can be useful in the initial evaluation of suspected SIH. The noncontrast component of this examination optimized with fluid sensitive sequences is most useful, particularly when performed with 3-D T2-weighted fat saturated sequences, which increases sensitivity for detecting fluid collections outside of the thecal sac [52]. It can detect with a high degree of accuracy the presence of epidural fluid collections and meningeal diverticula that can inform positioning and regions of interest for subsequent CSF leak localization imaging examinations, such as dynamic CT myelogram complete spine and digital subtraction myelography complete spine [25,34-36]. The contrast component of this examination may demonstrate dural

enhancement and engorged epidural venous plexus, which are also imaging features that support a diagnosis of SIH [28,29].

MRI Complete Spine Without IV Contrast

MRI complete spine without IV contrast optimized with fluid sensitive sequences is most useful in the initial evaluation of suspected SIH, particularly when performed with 3-D T2-weighted fat saturated sequences, which increases sensitivity for detecting fluid collections outside of the thecal sac [52]. This examination can detect with a high degree of accuracy the presence of epidural fluid collections and meningeal diverticula that can inform positioning and regions of interest for subsequent CSF leak localization imaging examinations, such as dynamic CT myelogram complete spine and digital subtraction myelography complete spine [25,34-36].

MRI Head With IV Contrast

There is no relevant literature to support the use of MRI head with IV contrast in the initial imaging of suspected SIH.

MRI Head Without and With IV Contrast

MRI head without and with IV contrast is most useful in the initial evaluation of suspected SIH. Suggestive imaging features of SIH are best visualized on MRI head without and with IV contrast and include qualitative signs (engorgement of venous sinuses, pachymeningeal enhancement, midbrain descent, superficial siderosis, subdural hygroma or hematoma, and convex superior surface of the pituitary) and quantitative signs (pituitary height, pontomesencephalic angle, suprasellar cistern, prepontine cistern, midbrain descent, venous-hinge angle, mamillopontine angle, tonsillar descent, and area cavum veli interpositi) [16,25,26]. The cumulative presence of these intracranial findings has been shown to correlate with the likelihood of finding a spinal leak source [16].

MRI Head Without IV Contrast

MRI head without IV contrast can be useful in the initial evaluation of suspected SIH. Although imaging features of suspected SIH are best visualized on MRI head without and with IV contrast, some imaging features such as midbrain descent, superficial siderosis, subdural hygroma or hematoma, convex superior surface of the pituitary, pituitary height, pontomesencephalic angle, suprasellar cistern, prepontine cistern, midbrain descent, venous-hinge angle, mamillopontine angle, tonsillar descent, and area cavum veli interpositi may be evaluated on noncontrast MRI sequences [16,25,26]. The cumulative presence of these intracranial findings has been shown to correlate with likelihood of finding a spinal leak source [16].

Radiographic Myelography Digital Subtraction Complete Spine

There is no relevant literature to support the use of dynamic digital subtraction myelography in the initial imaging of suspected SIH. Dynamic digital subtraction myelography plays an important role in the subsequent imaging workup of SIH after initial brain and spine imaging [53]. Results from an initial spine MRI or conventional CT myelogram provide useful information on how a subsequent dynamic digital subtraction myelogram will be performed, such as prone positioning for suspected ventral dural defect or decubitus positioning for suspected leaking meningeal diverticulum or CSF-venous fistula [24,53,54]. Dynamic digital subtraction myelography involves continuous real-time fluoroscopic x-ray imaging of the entire spine or of a focused region of the spine where there is suspicion for CSF leak. When performed in the decubitus position, 2 separate contrast injections may be required due to the transient temporal characteristics associated with CSF-venous fistula visualization as well as limitations in contrast dosing [40-44].

Variant 2: Adult. Orthostatic headache from suspected intracranial hypotension within 72 hours of dural puncture or other spinal intervention that could cause CSF leakage. Initial imaging.

Leakage of CSF following a dural puncture may be of sufficient volume to elicit symptoms of intracranial hypotension. Postdural puncture headaches occur with an estimated frequency of 2% to 8% [13-15]. Risk factors for postdural puncture headaches include the use of a larger gauge needle, multiple attempts at dural puncture, the use of a cutting needle versus pencil point tip needle, needle orientation perpendicular rather than parallel to spine longitudinal axis (when using a cutting needle), and dural puncture in the sitting position as opposed to lateral decubitus positioning [55-58]. Imaging is not typically indicated in this clinical setting because postdural puncture headaches are typically self-limited, with most symptoms fully resolving within 1 week without any treatment [13]. The initial management of postdural puncture headaches is conservative medical management, with consideration of epidural blood patch if symptoms are severe or not beginning to resolve by 2 to 3 days postdural puncture [59-61].

CT Complete Spine With IV Contrast

There is no relevant literature to support the use of CT complete spine with IV contrast in the initial imaging of suspected intracranial hypotension within 72 hours of dural puncture.

CT Complete Spine Without and With IV Contrast

There is no relevant literature to support the use of CT complete spine without and with IV contrast in the initial imaging of suspected intracranial hypotension within 72 hours of dural puncture.

CT Complete Spine Without IV Contrast

There is no relevant literature to support the use of CT complete spine without IV contrast in the initial imaging of suspected intracranial hypotension within 72 hours of dural puncture.

CT Head Cisternography

There is no relevant literature to support the use of CT head cisternography in the initial imaging of suspected intracranial hypotension within 72 hours of dural puncture.

CT Head With IV Contrast

There is no relevant literature to support the use of CT head with IV contrast in the initial imaging of suspected intracranial hypotension within 72 hours of dural puncture.

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 imaging of suspected intracranial hypotension within 72 hours of dural puncture.

CT Head Without IV Contrast

There is no relevant literature to support the use of CT head without IV contrast in the initial imaging of suspected intracranial hypotension within 72 hours of dural puncture.

CT Myelography Complete Spine

There is no relevant literature to support the use of CT myelography complete spine in the initial imaging of suspected intracranial hypotension within 72 hours of dural puncture.

CT Myelography Dynamic Complete Spine

There is no relevant literature to support the use of dynamic CT myelography complete spine in the initial imaging of suspected intracranial hypotension within 72 hours of dural puncture.

DTPA Cisternography

There is no relevant literature to support the use of DTPA cisternography in the initial imaging of suspected intracranial hypotension within 72 hours of dural puncture.

MR Myelography Complete Spine

There is no relevant literature to support the use of MR myelography complete spine in the initial imaging of suspected intracranial hypotension within 72 hours of dural puncture.

MRI Complete Spine With IV Contrast

There is no relevant literature to support the use of MRI complete spine with IV contrast in the initial imaging of suspected intracranial hypotension within 72 hours of dural puncture.

MRI Complete Spine Without and With IV Contrast

There is no relevant literature to support the use of MRI complete spine without and with IV contrast in the initial imaging of suspected intracranial hypotension within 72 hours of dural puncture.

MRI Complete Spine Without IV Contrast

There is no relevant literature to support the use of MRI complete spine without IV contrast in the initial imaging of suspected intracranial hypotension within 72 hours of dural puncture.

MRI Head With IV Contrast

There is no relevant literature to support the use of MRI head with IV contrast in the initial imaging of suspected intracranial hypotension within 72 hours of dural puncture.

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 initial imaging of suspected intracranial hypotension within 72 hours of dural puncture.

MRI Head Without IV Contrast

There is no relevant literature to support the use of MRI head without IV contrast in the initial imaging of suspected intracranial hypotension within 72 hours of dural puncture.

Radiographic Myelography Digital Subtraction Complete Spine

There is no relevant literature to support the use of dynamic digital subtraction myelography complete spine in the initial imaging of suspected intracranial hypotension within 72 hours of dural puncture.

Variant 3: Adult. Orthostatic headache from suspected intracranial hypotension without improvement post 72 hours of dural puncture or other spinal intervention that could cause CSF leakage. Initial imaging.

This clinical variant refers to a patient with orthostatic headache in temporal relation to a dural puncture that is not improving following a trial of conservative management. Imaging is not usually warranted in this clinical scenario, because the next management step for such patients typically involves an epidural blood patch procedure directed at the level of dural puncture [59-61].

CT Complete Spine With IV Contrast

There is no relevant literature to support the use of CT complete spine with IV contrast in the initial imaging of suspected intracranial hypotension without improvement after 72 hours of dural puncture or other spinal intervention that could cause CSF leakage.

CT Complete Spine Without and With IV Contrast

There is no relevant literature to support the use of CT complete spine without and with IV contrast in the initial imaging of suspected intracranial hypotension without improvement after 72 hours of dural puncture or other spinal intervention that could cause CSF leakage.

CT Complete Spine Without IV Contrast

There is no relevant literature to support the use of CT complete spine without IV contrast in the initial imaging of suspected intracranial hypotension without improvement after 72 hours of dural puncture or other spinal intervention that could cause CSF leakage.

CT Head Cisternography

There is no relevant literature to support the use of CT head cisternography in the initial imaging of suspected intracranial hypotension without improvement after 72 hours of dural puncture or other spinal intervention that could cause CSF leakage.

CT Head With IV Contrast

There is no relevant literature to support the use of CT head with IV contrast in the initial imaging of suspected intracranial hypotension without improvement after 72 hours of dural puncture or other spinal intervention that could cause CSF leakage.

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 imaging of suspected intracranial hypotension without improvement after 72 hours of dural puncture or other spinal intervention that could cause CSF leakage.

CT Head Without IV Contrast

There is no relevant literature to support the use of CT head without IV contrast in the initial imaging of suspected intracranial hypotension without improvement after 72 hours of dural puncture or other spinal intervention that could cause CSF leakage.

CT Myelography Complete Spine

There is no relevant literature to support the use of CT myelography complete spine in the initial imaging of suspected intracranial hypotension without improvement after 72 hours of dural puncture or other spinal intervention that could cause CSF leakage.

CT Myelography Dynamic Complete Spine

There is no relevant literature to support the use of dynamic CT myelography complete spine in the initial imaging of suspected intracranial hypotension without improvement after 72 hours of dural puncture or other spinal intervention that could cause CSF leakage.

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DTPA Cisternography

There is no relevant literature to support the use of DTPA cisternography in the initial imaging of suspected intracranial hypotension without improvement after 72 hours of dural puncture or other spinal intervention that could cause CSF leakage.

MR Myelography complete spine

There is no relevant literature to support the use of MR myelography complete spine in the initial imaging of suspected intracranial hypotension without improvement after 72 hours of dural puncture or other spinal intervention that could cause CSF leakage.

MRI Complete Spine With IV Contrast

There is no relevant literature to support the use of MRI complete spine with IV contrast in the initial imaging of suspected intracranial hypotension without improvement after 72 hours of dural puncture or other spinal intervention that could cause CSF leakage.

MRI Complete Spine Without and With IV Contrast

There is no relevant literature to support the use of MRI complete spine without and with IV contrast in the initial imaging of suspected intracranial hypotension without improvement after 72 hours of dural puncture or other spinal intervention that could cause CSF leakage.

MRI Complete Spine Without IV Contrast

There is no relevant literature to support the use of MRI complete spine without IV contrast in the initial imaging of suspected intracranial hypotension without improvement after 72 hours of dural puncture or other spinal intervention that could cause CSF leakage.

MRI Head With IV Contrast

There is no relevant literature to support the use of MRI head with IV contrast in the initial imaging of suspected intracranial hypotension without improvement after 72 hours of dural puncture or other spinal intervention that could cause CSF leakage.

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 initial imaging of suspected intracranial hypotension without improvement after 72 hours of dural puncture or other spinal intervention that could cause CSF leakage.

MRI Head Without IV Contrast

There is no relevant literature to support the use of MRI head without IV contrast in the initial imaging of suspected intracranial hypotension without improvement after 72 hours of dural puncture or other spinal intervention that could cause CSF leakage.

Radiographic Myelography Digital Subtraction Complete Spine

There is no relevant literature to support the use of dynamic digital subtraction myelography complete spine in the initial imaging of suspected intracranial hypotension without improvement after 72 hours of dural puncture or other spinal intervention that could cause CSF leakage.

Variant 4: Adult. Obtundation with initial brain imaging features of suspected intracranial hypotension. Next imaging study.

This clinical scenario represents a severe clinical manifestation of SIH, which may represent sequela of more severe mechanical traction forces from brain sagging in the setting of CSF hypovolemia [62,63]. There are some limited reports of successful acute management of these critically ill patients with intrathecal saline infusions via lumbar drains, which may serve as a temporizing measure to raise CSF pressure and reverse obtundation [64-67]. The subsequent management of such patients is otherwise similar to patients with SIH without impaired consciousness and focuses on spine imaging to localize a source of leak that may be targeted for definitive treatment [24].

CT Complete Spine With IV Contrast

There is no relevant literature to support CT complete spine with IV contrast as a next imaging study in the clinical setting of obtundation with initial brain imaging features of suspected intracranial hypotension.

CT Complete Spine Without and With IV Contrast

There is no relevant literature to support CT complete spine without and with IV contrast as a next imaging study in the clinical setting of obtundation with initial brain imaging features of suspected intracranial hypotension.

CT Complete Spine Without IV Contrast

There is no relevant literature to support CT complete spine without IV contrast as a next imaging study in the clinical setting of obtundation with initial brain imaging features of suspected intracranial hypotension.

CT Myelography Complete Spine

Although CT myelography complete spine can detect epidural contrast collections suggestive of dural defect or leaking meningeal diverticulum, MRI complete spine optimized with fluid sensitive sequences has been shown to detect epidural collections with equal sensitivity and is a preferred initial imaging option over CT myelography complete spine because it avoids the need for lumbar puncture [34,35].

CT Myelography Dynamic Complete Spine

There is no relevant literature to support the use of dynamic CT myelography complete spine as a next imaging study in the clinical setting of obtundation with initial brain imaging features of suspected intracranial hypotension. Dynamic CT myelography plays an important role in the subsequent imaging workup of SIH after initial brain and spine imaging [37,38]. Results from an initial spine MRI or conventional CT myelogram provide useful information on how a subsequent dynamic CT myelogram will be performed, such as prone positioning for suspected ventral dural defect or decubitus positioning for suspected leaking meningeal diverticulum or CSF-venous fistula [24,37-39]. Dynamic CT myelography involves an initial scan followed by delayed phase scans in immediate succession and, when performed in the decubitus position, may require 2 separate contrast injections due to the transient temporal characteristics associated with CSF-venous fistula visualization as well as limitations in contrast dosing [40-44].

DTPA Cisternography

Although DTPA cisternography can detect epidural collections suggestive of dural defect or leaking meningeal diverticulum, MRI complete spine optimized with fluid sensitive sequences is typically the preferred initial imaging option over DTPA cisternography complete spine because it avoids the need for lumbar puncture and has superior spatial resolution for lesion localization [45,46].

MR Myelography Complete Spine

There is no relevant literature to support the use of MR myelography complete spine as a next imaging study in the clinical setting of obtundation with initial brain imaging features of suspected intracranial hypotension. An MR myelogram with intrathecal gadolinium administration has been used in several studies as a subsequent follow-up imaging examination after initial spine imaging to increase the sensitivity for detection of slow leaking dural and meningeal diverticular defects [47-49]. Some limited evidence suggests a potential role for MR myelography in the detection of CSF-venous fistulas [50]. It should be noted that intrathecal use of gadolinium is currently off label. Although studies have suggested that intrathecal gadolinium can be administered safely in small doses, the safety profile of such imaging is not formally approved for such use, and special dosing caution is required to avoid potential of gadolinium induced neurotoxicity [51].

MRI Complete Spine With IV Contrast

There is no relevant literature to support the use of MRI complete spine with IV contrast as a next imaging study in the clinical setting of obtundation with initial brain imaging features of suspected intracranial hypotension.

MRI Complete Spine Without and With IV Contrast

MRI complete spine without and with IV contrast can be useful as a next imaging study in the clinical setting of obtundation with initial brain imaging features of suspected intracranial hypotension. The noncontrast component of this examination optimized with fluid sensitive sequences is most useful, particularly when performed with 3-D T2-weighted fat saturated sequences, which increases the sensitivity for detecting fluid collections outside of the thecal sac [52]. It can detect with a high degree of accuracy the presence of epidural fluid collections and meningeal diverticula that can inform positioning and regions of interest for subsequent CSF leak localization imaging examinations, such as dynamic CT myelogram complete spine and digital subtraction myelography complete spine [25,34-36]. The contrast component of this examination may demonstrate dural enhancement and engorged epidural venous plexus, which are also imaging features that support a diagnosis of SIH [28,29].

MRI Complete Spine Without IV Contrast

MRI complete spine without IV contrast optimized with fluid sensitive sequences is most useful as a next imaging study in the clinical setting of obtundation with initial brain imaging features of suspected intracranial hypotension, particularly when performed with 3-D T2-weighted fat saturated sequences, which increases sensitivity for detecting fluid collections outside of the thecal sac [52]. This examination can detect with a high degree of accuracy the presence of epidural fluid collections and meningeal diverticula that can inform positioning and regions of interest for subsequent CSF leak localization imaging examinations, such as dynamic CT myelogram complete spine and digital subtraction myelography complete spine [25,34-36].

Radiographic Myelography Digital Subtraction Complete Spine

There is no relevant literature to support the use of dynamic digital subtraction myelogram complete spine as a next imaging study in the clinical setting of obtundation with initial brain imaging features of suspected intracranial hypotension. Dynamic digital subtraction myelography plays an important role in the subsequent imaging workup of SIH after initial brain and spine imaging [53]. Results from an initial spine MRI or conventional CT myelogram provide useful information on how a subsequent dynamic digital subtraction myelogram will be performed, such as prone positioning for suspected ventral dural defect or decubitus positioning for suspected leaking meningeal diverticulum or CSF-venous fistula [24,53,54]. Dynamic digital subtraction myelography involves continuous real-time fluoroscopic x-ray imaging of the entire spine or of a focused region of the spine where there is suspicion for CSF leak. When performed in the decubitus position, 2 separate contrast injections may be required due to the transient temporal characteristics associated with CSF-venous fistula visualization as well as limitations in contrast dosing [40-44].

Variant 5: Adult. Chronic daily headache from suspected intracranial hypotension with negative initial brain and spine imaging, but with history and clinical examination suggesting CSF leakage. Next imaging study.

Negative initial imaging should not preclude continued diagnostic workup in a patient with clinically suspected SIH [4,53]. Approximately 20% of initial brain MRIs and 46% to 67% of initial spine imaging in patients with clinically suspected SIH may have normal results [4]. Of note, CSF-venous fistulas and slow meningeal diverticular leaks are often subtle imaging findings that may not be readily detectable using conventional imaging techniques with limited temporal resolution and may require follow-up imaging with more advanced imaging procedures to definitely identify or exclude a more subtle spinal CSF leak source [7,53,68-70].

CT Head Cisternography

There is no relevant literature to support the use of CT head cisternography in the subsequent imaging of suspected SIH. The spine has been shown to represent the anatomical source of most symptomatic CSF leaks and venous fistulas, such that the imaging investigation of leak source should be directed primarily toward the spine and not intracranially [22].

CT Myelography Dynamic Complete Spine

Dynamic CT myelography plays an important role in the subsequent imaging workup of SIH after initial brain and spine imaging [37,38]. Results from an initial spine MRI or conventional CT myelogram provide useful information on how a subsequent dynamic CT myelogram will be performed, such as prone positioning for suspected ventral dural defect or decubitus positioning for suspected leaking meningeal diverticulum or CSF-venous fistula [24,37-39]. Dynamic CT myelography involves an initial scan followed by delayed phase scans in immediate succession and, when performed in the decubitus position, may require 2 separate contrast injections due to the transient temporal characteristics associated with CSF-venous fistula visualization as well as limitations in contrast dosing [40-44].

DTPA Cisternography

DTPA cisternography of the spine can detect CSF leaks with similar accuracy to conventional CT myelography [45]. Due to its limited spatial resolution, a positive DTPA cisternogram may require further investigation with subsequent dynamic CT myelography or dynamic digital subtraction myelography to definitively localize leak for treatment planning [45,46].

MR Myelography Complete Spine

An MR myelogram with intrathecal gadolinium administration has been used in several studies to increase sensitivity for the detection of slow leaking dural and meningeal diverticular defects [47-49]. Some limited evidence suggests a potential role for MR myelography in the detection of CSF-venous fistulas [50]. It should be noted that intrathecal use of gadolinium is currently off label. Although studies have suggested that intrathecal gadolinium can

be administered safely in small doses, the safety profile of such imaging is not formally approved for such use, and special dosing caution is required to avoid potential of gadolinium induced neurotoxicity [51].

Radiographic Myelography Digital Subtraction Complete Spine

Dynamic digital subtraction myelography plays an important role in the subsequent imaging workup of SIH after initial brain and spine imaging [53]. Results from an initial spine MRI or conventional CT myelogram provide useful information on how a subsequent dynamic digital subtraction myelogram will be performed, such as prone positioning for suspected ventral dural defect or decubitus positioning for suspected leaking meningeal diverticulum or CSF-venous fistula [24,53,54]. Dynamic digital subtraction myelography involves continuous real-time fluoroscopic x-ray imaging of the entire spine or of a focused region of the spine where there is suspicion for CSF leak. When performed in the decubitus position, 2 separate contrast injections may be required due to the transient temporal characteristics associated with CSF-venous fistula visualization as well as limitations in contrast dosing [40-44].

Variant 6: Adult. Rebound headache following epidural blood patch or fibrin glue patch treatment for suspected intracranial hypotension. Initial imaging.

Rebound headaches are known to occur in about a quarter of patients following treatment of a CSF leak and are characterized by a postprocedural elevation in CSF pressure, usually occurring within the first 1 to 2 postprocedural days [71]. The clinical syndrome of rebound headaches can include a development of a phenotypically new headache with characteristic reversal of a patient's preprocedural orthostatic symptoms, whereas the patient feels relief of headache in the upright position and exacerbation of symptoms in the recumbent position [10]. Headache location may also change, from the characteristic occipital region predominant in SIH headaches to a more anteriorly located frontal, periorbital, or retroorbital location. Additional features of rebound headache include nausea, emesis, and blurry vision. It is important to recognize the clinical features of rebound headache because mistaking these symptoms as refractory SIH may subject the patient to unnecessary imaging or unnecessary repeat blood/fibrin patch procedures that could raise CSF pressure further and exacerbate the condition. Rebound headaches are usually self-limited and minor and can be managed conservatively. Acetazolamide, which lowers CSF production, may be prescribed in the postprocedural period to ameliorate symptoms of rebound headache. CSF drainage via lumbar puncture or lumbar drain can be used in more severe cases that are refractory to medical management [10].

CT Complete Spine With IV Contrast

There is no relevant literature to support the use of CT complete spine with IV contrast in the initial imaging of post–SIH treatment rebound headaches.

CT Complete Spine Without and With IV Contrast

There is no relevant literature to support the use of CT complete spine without and with IV contrast in the initial imaging of post–SIH treatment rebound headaches.

CT Complete Spine Without IV Contrast

There is no relevant literature to support the use of CT complete spine without IV contrast in the initial imaging of post–SIH treatment rebound headaches.

CT Head Cisternography

There is no relevant literature to support the use of CT head cisternography in the initial imaging of post–SIH treatment rebound headaches.

CT Head With IV Contrast

There is no relevant literature to support the use of CT head with IV contrast in the initial imaging of post–SIH treatment rebound headaches.

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 imaging of post–SIH treatment rebound headaches.

CT Head Without IV Contrast

There is no relevant literature to support the use of CT head without IV contrast in the initial imaging of post–SIH treatment rebound headaches.

CT Myelography Complete Spine

There is no relevant literature to support the use of CT myelography complete spine in the initial imaging of post—SIH treatment rebound headaches.

CT Myelography Dynamic Complete Spine

There is no relevant literature to support the use of dynamic CT myelography complete spine in the initial imaging of post–SIH treatment rebound headaches.

DTPA Cisternography

There is no relevant literature to support the use of DTPA cisternography in the initial imaging of post-SIH treatment rebound headaches.

MR Myelography Complete Spine

There is no relevant literature to support the use of MR myelography complete spine in the initial imaging of post—SIH treatment rebound headaches.

MRI Complete Spine With IV Contrast

There is no relevant literature to support the use of MRI complete spine with IV contrast in the initial imaging of post–SIH treatment rebound headaches.

MRI Complete Spine Without and With IV Contrast

There is no relevant literature to support the use of MRI complete spine without and with IV contrast in the initial imaging of post–SIH treatment rebound headaches.

MRI Complete Spine Without IV Contrast

There is no relevant literature to support the use of MRI complete spine without IV contrast in the initial imaging of post–SIH treatment rebound headaches.

MRI Head With IV Contrast

There is no relevant literature to support the use of MRI head with IV contrast in the initial imaging of post-SIH treatment rebound headaches.

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 initial imaging of post–SIH treatment rebound headaches.

MRI Head Without IV Contrast

There is no relevant literature to support the use of MRI head without IV contrast in the initial imaging of post–SIH treatment rebound headaches.

Radiographic Myelography Digital Subtraction Complete Spine

There is no relevant literature to support the use of dynamic digital subtraction myelography complete spine in the initial imaging of post–SIH treatment rebound headaches.

Summary of Highlights

- Variant 1: In an adult with an orthostatic headache from suspected intracranial hypotension, without recent spinal intervention that could cause CSF leakage, an MRI of the brain and the complete spine, either without and with IV contrast or without IV contrast, are both required to assist in localizing a potential CSF leak.
- Variant 2: In an adult with an orthostatic headache from suspected intracranial hypotension within 72 hours of dural puncture or other spinal intervention that could cause CSF leakage, imaging is not typically indicated because postdural puncture headaches are typically self-limited, with most symptoms fully resolving within 1 week without any treatment. The initial management of postdural puncture headaches is conservative medical management, with consideration of an epidural blood patch if symptoms are severe or not beginning to resolve by 2 to 3 days postdural puncture.
- Variant 3: In an adult with an orthostatic headache from suspected intracranial hypotension without improvement after 72 hours of dural puncture or other spinal intervention that could cause CSF leakage, imaging is not typically indicated because the subsequent management step for such patients involves an epidural blood patch procedure directed at the level of dural puncture.

- Variant 4: In an adult who is obtunded with initial brain imaging features of suspected intracranial hypotension, an MRI of the complete spine, either without and with IV contrast or without IV contrast, is required to assist in localizing a potential CSF leak. An intrathecal saline infusion procedure may also be considered as a temporizing measure to restore CSF pressures and reverse symptoms. This scenario represents a severe clinical manifestation of SIH, which may represent the sequela of more severe mechanical traction forces from brain sagging in CSF hypovolemia. The subsequent management of such patients is otherwise like that of patients with SIH without impaired consciousness. It focuses on spine imaging to localize a source of leak that may be targeted for definitive treatment.
- Variant 5: In an adult with chronic daily headache from suspected intracranial hypotension with negative initial brain and spine imaging but with history and clinical examination suggesting CSF leakage, either dynamic CT or dynamic digital subtraction myelography is indicated in the subsequent imaging workup of SIH because negative initial imaging should not preclude continued diagnostic workup. For instance, CSF-venous fistulas and slow meningeal diverticular leaks are often subtle imaging findings that may not be readily detectable using conventional imaging techniques with limited temporal resolution and may require follow-up imaging with more advanced imaging procedures to identify or exclude a more subtle spinal CSF leak source.
- Variant 6: Imaging is not typically indicated in an adult with rebound headache following epidural blood patch or fibrin glue patch treatment for suspected intracranial hypotension. Rebound headaches are usually self-limited and minor and can be managed conservatively.

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 go to www.acr.org/ac.

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.

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 [72].

Relative Radiation Level Designations		
Relative Radiation Level*	Adult Effective Dose Estimate Range	Pediatric Effective Dose Estimate Range
0	0 mSv	0 mSv
€	<0.1 mSv	<0.03 mSv
⊕⊕	0.1-1 mSv	0.03-0.3 mSv
♥♥	1-10 mSv	0.3-3 mSv
***	10-30 mSv	3-10 mSv
♥♥♥♥♥	30-100 mSv	10-30 mSv

^{*}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."

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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.