Intracranial Hypotension

A proposal to create new codes for intracranial hypotension was previously presented at the March 2018 ICD-10 Coordination and Maintenance Committee meeting. This proposal has been modified based on comments from that prior presentation. Intracranial hypotension results from a loss of cerebrospinal fluid volume, and it is an under-recognized and under-diagnosed central nervous system disorder. Intracranial hypotension is most often associated with a cerebrospinal fluid leak at the level of the spine and is not causally associated with cerebrospinal fluid leaks arising from the skull base.

The causes of intracranial hypotension include:

- Spontaneous cerebrospinal fluid leaks at the level of the spine (most under-recognized category)
- Iatrogenic holes or defects in the spinal dura from:
  - Intentional diagnostic or therapeutic spinal dural punctures
  - Inadvertent spinal dural puncture during epidural injection procedures
  - Inadvertent or intentional spinal durotomy during spinal or other surgeries
- Over-drainage of cerebrospinal fluid shunting devices
- Traumatic spinal dural tears or defects resulting in spinal cerebrospinal fluid leaks

While headache is the most common symptom of intracranial hypotension, the presence of headache is not universal, nor is it the only symptom. A range of symptoms, signs and complications may occur as a result of effects on the brain and other intracranial structures, cranial nerve roots, spinal cord and spinal nerve roots. Using the term “intracranial hypotension” is more precise and inclusive. When serious complications occur, such as subdural hematomas, stroke, cerebral venous thrombosis, fronto-temporal dementia, or syringomyelia, these should also be coded. It is also of note that cranial MRI findings, or spinal imaging findings may or may not be evident.

While the headache and other symptoms associated with intracranial hypotension often have a positional component, not all patients with a positional (orthostatic) component to their symptoms have intracranial hypotension. An additional code for “headache with orthostatic component, not elsewhere classified” is being proposed for cases in which the diagnosis remains unclear.

The International Classification of Headache Disorders, 3rd edition, recognizes spontaneous intracranial hypotension as a secondary headache disorder, as well as classifying iatrogenic and traumatic cases.

Currently, intracranial hypotension is captured in ICD-10-CM using a number of codes that are not specific, such as those related to CSF leaks, and headaches. Iatrogenic causes of spinal CSF leaks are currently included at category G97. Intraoperative and postprocedural complications and disorders of nervous system, not elsewhere classified; some are also classified related to obstetrics, including subcategories O29.4, Spinal and epidural anesthesia induced headache during pregnancy; and O29.5, Other complications of spinal and epidural anesthesia during pregnancy; and codes O89.4, Spinal and epidural anesthesia-induced headache during the puerperium; O89.5, Other complications of spinal and epidural anesthesia during the puerperium; O74.5, Spinal and epidural anesthesia-induced headache during labor and delivery; and O74.6, Other complications of spinal and epidural anesthesia during labor and delivery.

These proposed changes have the potential to improve diagnostic accuracy, be supportive of improved understanding of requirements for diagnostic testing and treatments, and to better track prevalence of intracranial hypotension, and outcomes. They will also enable better differentiation of cranial and spinal
cerebrospinal fluid leaks, which have different underlying causes, symptoms, complications, diagnostic testing and treatments.

Specific codes and expansion in ICD-10-CM has been requested by a team of key opinion leaders, including Timothy Amhrein, MD, Peter G. Kranz, MD and Linda-Gray Leithe, MD, Neuroradiology / Spine Intervention at Duke University Medical Center; Ian Carroll, MD, MS, Anesthesiology / Pain Medicine at Stanford; Connie Deline, MD, General Medicine, Spinal CSF Leak Foundation; Charles Louy, PhD, MD, MBA, Anesthesiology / Pain Medicine at Cedars-Sinai; Marcel Maya, MD, Neuroradiology at Cedars-Sinai; Wouter Schievink, MD, Neurosurgery at Cedars-Sinai; Stephen Silberstein, MD, Headache Neurology at Jefferson Headache Center; and Deborah Friedman, MD, MPH, Neurology and Ophthalmology at UT Southwestern Medical Center.

References:


**TABULAR MODIFICATIONS**

<table>
<thead>
<tr>
<th>G96</th>
<th>Other disorders of central nervous system</th>
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<tbody>
<tr>
<td>G96.0</td>
<td>Cerebrospinal fluid leak</td>
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<tr>
<td>Add</td>
<td>Code also spontaneous intracranial hypotension, if applicable (G96.81)</td>
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<tr>
<td>Add</td>
<td>Excludes1: cerebrospinal fluid leak from spinal puncture (G97.0)</td>
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<tr>
<td>New code</td>
<td>G96.00</td>
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<tr>
<td>New code</td>
<td>G96.01</td>
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rhinorrhea due to CSF leak

New code G96.02 Spinal cerebrospinal fluid leak, spontaneous
  Cerebrospinal fluid leak from spine

New code G96.09 Other cerebrospinal fluid leak

G96.1 Disorders of meninges, not elsewhere classified

G96.11 Dural tear
Add Code also spontaneous intracranial hypotension, if applicable (G96.81)

G96.8 Other specified disorders of central nervous system

New sub-subcategory G96.81 Intracranial hypotension

Add Code also any associated diagnoses, such as:
  Brachial amyotrophy (G54.5)
  Cerebrospinal fluid leak from spine (G96.02)
  Cranial nerve disorders in diseases classified elsewhere (G53)
  Nerve root and compressions in diseases classified elsewhere (G55)
  Nonpyogenic thrombosis of intracranial venous system (I67.6)
  Nontraumatic subdural hemorrhage (I62.0-)
  Nontraumatic intracerebral hemorrhage (I61.-)
  Other and unspecified cord compression (G95.2-)
  Other secondary parkinsonism (G21.8)
  Reversible cerebrovascular vasoconstriction syndrome (I67.841)
  Spinal cord herniation (G95.89)
  Stroke (I63.-)
  Syringomyelia (G95.0)

New code G96.810 Intracranial hypotension, unspecified

New code G96.811 Intracranial hypotension, spontaneous

New code G96.819 Other intracranial hypotension

New code G96.89 Other specified disorders of central nervous system

G97 Intraoperative and post-procedural complications and disorders of nervous system, not elsewhere classified

G97.0 Cerebrospinal fluid leak from spinal puncture
Add Code also any associated diagnoses or complications, such as:
Add Intracranial hypotension following a procedure (G97.83)
G97.1 Other reaction to spinal and lumbar puncture
   Add Other reaction to spinal dural puncture
   Add Code also if applicable any associated headache with orthostatic component (R51.0)

G97.2 Intracranial hypotension following ventricular shunting
   Add Code also any associated diagnoses or complications

G97.4 Accidental puncture and laceration of a nervous system organ or structure during a procedure
   G97.41 Accidental puncture or laceration of dura during a procedure
   Incidental (inadvertent) durotomy
   Add Code also any associated diagnoses or complications

G97.8 Other intraoperative and postprocedural complications and disorders of nervous system
   Use additional code to further specify disorder
   New code G97.83 Intracranial hypotension following lumbar cerebrospinal fluid shunting
   Add Code also any associated diagnoses or complications

   New code G97.84 Intracranial hypotension following other procedure
   Add Code also, if applicable:
   Add Accidental puncture or laceration of dura during a procedure (G97.41)
   Add Cerebrospinal fluid leak from spinal puncture (G97.0)

G44.8 Other specified headache syndromes
   Add Excludes2: Headache with orthostatic or positional component, not elsewhere classified (R51.0)

R51 Headache
   Delete Facial pain NOS
   Delete Excludes1: atypical face pain (G50.1)
   Delete migraine and other headache syndromes (G43-G44)
   Delete trigeminal neuralgia (G50.0)
   Add Excludes2: atypical face pain (G50.1)
   Add migraine and other headache syndromes (G43-G44)
   Add trigeminal neuralgia (G50.0)

   New code R51.0 Headache with orthostatic component, not elsewhere classified
   Add Headache with positional component, not elsewhere classified

   New code R51.9 Headache, unspecified
   Add Facial pain NOS