



ICD-10 Coordination and Maintenance Committee Meeting
March 6-7, 2018
Diagnosis Agenda

Welcome and announcements
Donna Pickett, MPH, RHIA
Co-Chair, ICD-10 Coordination and Maintenance Committee

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ICD-10-CM TIMELINE

A timeline of important dates in the ICD-10 process is described below:

March 6-7, 2018

ICD-10 Coordination and Maintenance Committee Meeting.

Those who wish to attend the ICD-10 Coordination and Maintenance Committee meeting **must have registered for the meeting online by February 23, 2018**. You must bring an official form of picture identification (such as a driver's license) in order to be admitted to the building.

In compliance to The Real ID Act, enacted in 2005, (<http://www.dhs.gov/real-id-enforcement-brief>) the following states/territories: Maine, Minnesota, Missouri, Montana and Washington State **will not** gain access into any Federal Agencies using the **above states** driver's license or ID. This means CMS visitors from these states/territories will need to provide alternative proof of identification (**such as a passport**) to gain entrance into Baltimore-based and Bethesda CMS buildings, as well as the Humphrey Building in Washington.

Because of increased security requirements, those wishing to attend the March 6-7, 2018 ICD-10 Coordination and Maintenance Committee meeting must register for the meeting online at:

<https://www.cms.gov/apps/events/default.asp>

Attendees must register online by February 23, 2018; failure to do so may result in lack of access to the meeting.

March 2018

Webcast of the March 6-7, 2018 ICD-10 Coordination and Maintenance Committee meeting will be posted on the CMS webpage as follows:

<https://www.cms.gov/Medicare/Coding/ICD9ProviderDiagnosticCodes/meetings.html>

April 1, 2018

There were no requests for ICD-10 codes to capture new diagnoses or new technology for implementation on April 1, 2018. Therefore, there will be no new ICD-10 diagnosis or procedure codes implemented on April 1, 2018.

April 2018

Notice of Proposed Rulemaking to be published in the Federal Register as mandated by Public Law 99-509. This notice will include references to the finalized FY 2019 ICD-10-CM diagnosis and ICD-10-PCS procedure

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codes to date. It will also include proposed revisions to the MS-DRG system based on ICD-10-CM/PCS codes on which the public may comment. The proposed rule can be accessed at:

<http://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/AcuteInpatientPPS/index.html?redirect=/AcuteInpatientPPS/IPPS/list.asp>

May 11, 2018

Deadline for receipt of public comments on proposed new codes and revisions discussed at the March 6-7, 2018 ICD-10 Coordination and Maintenance Committee meetings for implementation on October 1, 2019.

June 2018

Final addendum posted on web pages as follows:

Diagnosis addendum - <http://www.cdc.gov/nchs/icd/icd10cm.htm>

Procedure addendum -

<http://cms.hhs.gov/Medicare/Coding/ICD10/index.html>

July 13, 2018

Deadline for requestors: Those members of the public requesting that topics be discussed at the September 11-12, 2018 ICD-10 Coordination and Maintenance Committee meeting must have their requests submitted to CMS for procedures and NCHS for diagnoses.

August 1, 2018

Hospital Inpatient Prospective Payment System final rule to be published in the Federal Register as mandated by Public Law 99-509. This rule will also include links to all the final codes to be implemented on October 1, 2018.

This rule can be accessed at:

<http://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/AcuteInpatientPPS/index.html?redirect=/AcuteInpatientPPS/IPPS/list.asp>

August 2018

Tentative agenda for the Procedure part of the September 11-12, 2018 ICD-10 Coordination and Maintenance Committee meeting will be posted on the CMS webpage at –

<https://www.cms.gov/Medicare/Coding/ICD9ProviderDiagnosticCodes/ICD-9-CM-C-and-M-Meeting-Materials.html>

Tentative agenda for the Diagnosis part of the September 11-12, 2018 ICD-10 Coordination and Maintenance Committee meeting will be posted on the NCHS webpage at -

http://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm

Federal Register notice for the September 11-12, 2018 ICD-10 Coordination and Maintenance Committee meeting will be published. This will include the tentative agenda.

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- August 3, 2018** **On-line registration opens for the September 11-12, 2018 ICD-10 Coordination and Maintenance Committee meeting at:**
<https://www.cms.gov/apps/events/default.asp>
- September 3, 2018 Because of increased security requirements, those wishing to attend the September 11-12, 2018 ICD-10 Coordination and Maintenance Committee meeting must register for the meeting online at:
<https://www.cms.gov/apps/events/default.asp>
Attendees must register online by September 3, 2018; failure to do so may result in lack of access to the meeting.
- September 11-12, 2018 ICD-10 Coordination and Maintenance Committee Meeting.
- Those who wish to attend the ICD-10 Coordination and Maintenance Committee meeting **must have registered for the meeting online by September 3, 2018.** You must bring an official form of picture identification (such as a driver's license) in order to be admitted to the building.
- September 2018 Webcast of the September 11-12, 2018 ICD-10 Coordination and Maintenance Committee meeting will be posted on the CMS webpage as follows:
<https://www.cms.gov/Medicare/Coding/ICD9ProviderDiagnosticCodes/meetings.html>
- October 1, 2018 New and revised ICD-10-CM and ICD-10-PCS codes go into effect along with DRG changes. Final addendum available on web pages as follows:
Diagnosis addendum - <http://www.cdc.gov/nchs/icd/icd10cm.htm>
Procedure addendum –
<http://www.cms.gov/Medicare/Coding/ICD10/>
- October 12, 2018 **Deadline for receipt of public comments on proposed new codes discussed at the September 11-12, 2018 ICD-10 Coordination and Maintenance Committee meetings for implementation on April 1, 2019.**
- November 2018 Any new ICD-10 codes required to capture new technology that will be implemented on the following April 1 will be announced. Information on any new codes to be implemented April 1, 2019 will be posted on the following websites:
<http://www.cdc.gov/nchs/icd/icd10cm.htm>
<http://www.cms.gov/Medicare/Coding/ICD10/>

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November 12, 2018

Deadline for receipt of public comments on proposed new codes and revisions discussed at the September 11-12, 2018 ICD-10 Coordination and Maintenance Committee meetings for implementation on October 1, 2019.

NOTE: In compliance to The Real ID Act, enacted in 2005, the following states/territories: American Samoa, Louisiana, Minnesota, New Hampshire, and New York **will not** gain access into any Federal Agencies using the **above states** driver's license or ID. This means CMS visitors from these states/territories will need to provide alternative proof of identification (**such as a passport**) to gain entrance into Baltimore-based CMS building.

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Contact Information

Mailing address:

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ICD-9-CM Coordination and Maintenance Committee
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Comments on the diagnosis proposals presented at the ICD Coordination and Maintenance Committee meeting should be sent to the following email address: nchsicd10CM@cdc.gov

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Traci Ramirez	(301) 458-4454

NCHS Classifications of Diseases web page:

<http://www.cdc.gov/nchs/icd.htm>

Please consult this web page for updated information

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Continuing Education Credits

Continuing education credits may be awarded by the American Academy of Professional Coders (AAPC) or the American Health Information Management Association (AHIMA) for participation in CMS/NCHS ICD-10 Coordination and Maintenance (C&M) Committee Meeting.

Continuing Education Information for American Academy of Professional Coders (AAPC)

If you plan to attend or participate via telephone the ICD-10 Coordination and Maintenance (C&M) Committee Meeting, you should be aware that CMS /NCHS do not provide certificates of attendance for these calls. Instead, the AAPC will accept your printed topic packet as proof of participation. Please retain a your topic packet copy as the AAPC may request them for any conference call you entered into your CEU Tracker if you are chosen for CEU verification. Members are awarded one (1) CEU per hour of participation.

Continuing Education Information for American Health Information Management Association (AHIMA)

AHIMA credential-holders may claim 1 CEU per 60 minutes of attendance at an educational program. Maintain documentation about the program for verification purposes in the event of an audit. A program does not need to be pre-approved by AHIMA, nor does a CEU certificate need to be provided, in order to claim AHIMA CEU credit. For detailed information about AHIMA's CEU requirements, see the Recertification Guide on AHIMA's web site.

Please note: The statements above are standard language provided to NCHS by the AAPC and the AHIMA. If you have any questions concerning either statement, please contact the respective organization, not NCHS.

Acne Vulgaris

Acne is one of the most common disorders treated by dermatologists and other health care providers. Acne vulgaris typically affects the face, upper chest and back. It is characterized by noninflammatory and inflammatory lesions.

Currently, ICD-10-CM has a unique code (L70.0) for Acne vulgaris. More recent treatment paradigms by national and international consensus committees further define and subcategorize acne vulgaris as mild, moderate and severe acne.

Sun Pharmaceutical Industries, Inc. is requesting the following new codes to better differentiate the levels of severity for this condition.

TABULAR MODIFICATIONS

	L70	Acne	
New subcategory	L70.0	Acne vulgaris	
New code	L70.00	Acne vulgaris, unspecified	
Add		Acne vulgaris NOS	
New code	L70.01	Acne vulgaris, mild	
New code	L70.02	Acne vulgaris, moderate	
New code	L70.03	Acne vulgaris, severe	

BRCA

BRCA1 and BRCA2 are the best known links to breast cancer risk. BRCA1 and BRCA2 mutations can be inherited from either parent and can affect the risk of cancers in both women and men. Women who have a BRCA gene mutation tend to develop breast and ovarian cancers at younger ages than women who do not have these mutations.

Given the limitations of current ovarian cancer screening approaches, prophylactic oophorectomy is recommended for patients with the BRCA1 or BRCA2 genetic mutation by the age of 40 or after the conclusion of childbearing. Prophylactic salpingo-oophorectomy reduces the risk of breast cancer by 40-70%. This procedure has been shown to reduce the risk of ovarian, fallopian tube and peritoneal cancer by approximately 85-90% in women with known mutations in BRCA1 or BRCA2. The American College of Obstetricians and Gynecologists Practice Bulletin Number 103, reaffirmed in 2015 on BRCA1 and BRCA2 documents “For a woman with a BRCA1 mutation, the risk of ovarian cancer is 39-46%. For a woman with BRCA2 mutation, the risk ovarian cancer is 65-74%. For women with breast cancer, the 10-year actuarial risk of developing subsequent ovarian cancer is 12.7% for BRCA1 mutation carriers and 6.8% for BRCA2 mutation carriers”.

The following new codes are being requested to identify specific BRCA mutations for statistics, tracking and reporting.

TABULAR MODIFICATIONS

Z15 Genetic susceptibility to disease

Z15.0 Genetic susceptibility to malignant neoplasm

New

sub-subcategory

Z15.01 Genetic susceptibility to malignant neoplasm of breast

New code

Z15.011 BRCA1 genetic susceptibility to malignancy

New code

Z15.012 BRCA2 genetic susceptibility to malignancy

New code

Z15.018 Other genetic susceptibility to malignant
neoplasm of breast

Cyclical Vomiting Syndrome

This proposal was originally presented at the September 2017 Coordination and Maintenance (C&M) meeting at the request of the American Hospital Association's Coding Clinic Editorial Advisory Board. The American Academy of Pediatrics has reviewed and supports this proposal. The proposal has been revised based on public comments received following the September 2017 C&M meeting.

Cyclical vomiting syndrome is described by episodes of severe vomiting that have no noticeable cause. Episodes can last for days or hours and alternate with symptom-free periods of time. Each episode tends to start at the same time of day, last the same length of time and occur with the same symptoms and level of intensity. Cyclical vomiting syndrome may or may not be related to migraines. Treatment usually involves medications, including anti-nausea and migraine therapies, that may help lessen symptoms.

Currently, in ICD-10-CM, Cyclical vomiting is indexed to code G43.AO, Cyclical vomiting, not intractable. These codes fall within the code category of G43-, Migraine. In ICD-9-CM, Cyclical vomiting (not related to migraines) was captured under code 536.2, Persistent vomiting. Code 536.2 crosswalks to ICD-10-CM code R11.10, Vomiting, unspecified. This code does not adequately represent the clinical significance of the disorder in the treatment of cyclical vomiting syndrome not related to migraines.

The following tabular modifications are being proposed and changes from the previous presentation are shown in bold:

TABULAR MODIFICATIONS

	G43 Migraine
	G43.A Cyclical vomiting
Add	Excludes1: Cyclical vomiting syndrome unrelated to migraine (R11.15)
Revise	G43.A0 Cyclical vomiting, (migraine), not intractable Cyclical vomiting, without refractory migraine
Revise	G43.A1 Cyclical vomiting, (migraine), intractable Cyclical vomiting, with refractory migraine
	R11 Nausea and vomiting
	R11.1 Vomiting
	R11.10 Vomiting, unspecified Vomiting NOS
	R11.11 Vomiting without nausea
	R11.12 Projectile vomiting
	R11.13 Vomiting of fecal matter
	R11.14 Bilious vomiting Bilious emesis
New code	R11.15 Cyclical vomiting syndrome unrelated to migraine
Add	Cyclic vomiting syndrome

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Add	Excludes1: Cyclical vomiting (G43.A-)
Add	Excludes2: Diabetes mellitus due to underlying condition (E08.-)
Add	Bulimia nervosa (F50.2)

Deficiency of Adenosine Deaminase 2

Deficiency of Adenosine Deaminase 2 (DADA2), or adenosine deaminase 2 deficiency, is characterized by abnormal inflammation of various tissues, which may be associated with a mottled rash (livedo racemosa), early-onset strokes, other findings of vasculitis (consistent with polyarteritis nodosa), and sometimes immunodeficiency. It is autoinflammatory in nature, and besides the skin and nervous system, may affect the gastrointestinal system or kidneys, and may cause intermittent fevers. It may be associated with hepatosplenomegaly. Onset may be from early childhood to adulthood. Severity of the disorder varies.

While vasculopathy is common with adenosine deaminase 2 deficiency, it is not always present at diagnosis. Although some cases have been described as childhood-onset polyarteritis nodosa, since the diagnosis may be made without vasculopathy being present, this would not be inherent.

Although adenosine deaminase 2 deficiency was relatively recently discovered, researchers suspect that it may not be a rare disease. They are working to determine whether it may underlie other forms of vasculitis and stroke whose causes are now unknown.

Even though adenosine deaminase 2 deficiency may be associated with immunodeficiency, this is usually relatively mild, and it is usually not associated with a significantly increased risk of bacterial and viral infections. This is in contrast with adenosine deaminase deficiency type 1, which causes a severe combined immunodeficiency. Despite the similar terms used to identify these disorders, they are quite different clinically.

A request to create a specific ICD-10-CM code was received from Dr. Chip Chambers, Vanderbilt University Medical Center, Nashville, TN, and President of the DADA2 Foundation.

References

- . Adenosine deaminase 2 deficiency. Genetics Home Reference. 2015. NLM, NIH.
<https://ghr.nlm.nih.gov/condition/adenosine-deaminase-2-deficiency>
- Nanthapaisal S, Murphy C, Omoyinmi E, et. al. Deficiency of Adenosine Deaminase Type 2. A Description of Phenotype and Genotype in Fifteen Cases. *Arthritis & Rheumatology*. *Arthritis Rheumatol*. 2016 Sep;68(9):2314-22.
<https://doi.org/10.1002/art.39699>
- Nihira H, Nakagawa K, Izawa K, et. al. Fever of unknown origin with rashes in early infancy is indicative of adenosine deaminase type 2 deficiency. *Scand J Rheumatol*. 2018 Mar;47(2):170-172. Epub 2017 Jun 30.
<https://doi.org/10.1080/03009742.2017.1324912>
- Ombrello A, Stone D, Hoffmann P, et. al. The deficiency of adenosine deaminase type 2-results of therapeutic intervention. *Pediatr Rheumatol Online J*. 2015; 13(Suppl 1): O40. Epub 2015 Sep 28.
<https://doi.org/10.1186/1546-0096-13-S1-O40>
- Paul Brogan P, Eleftheriou D. Vasculitis update: pathogenesis and biomarkers. *Pediatr Nephrol*. 2018; 33(2): 187–198. Epub 2017 Aug 7.
<https://doi.org/10.1007/s00467-017-3597-4>
<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5769819/>

TABULAR MODIFICATIONS

D81 Combined immunodeficiencies

D81.3 Adenosine deaminase [ADA] deficiency

D81.30 Adenosine deaminase deficiency, unspecified
ADA deficiency NOS

D81.31 Adenosine deaminase deficiency with severe combined
immunodeficiency
ADA with SCID
Adenosine deaminase deficiency type 1

D81.32 Adenosine deaminase 2 deficiency
ADA2 deficiency
Adenosine deaminase deficiency type 2

Code also any associated manifestations, such as:
stroke (I63.-)
polyarteritis nodosa (M30.0)

D81.39 Other adenosine deaminase deficiency

Dravet Syndrome

The Dravet Syndrome Foundation is proposing a new code be created for Dravet syndrome. Dravet syndrome, previously known as severe myoclonic epilepsy in infancy (SMEI), is a genetic encephalopathy that presents in the first year of life. It is a rare disorder with an incidence estimated between 1:20,000 and 1:40,000 representing about 7% of all severe epilepsies starting before the age of 3 years.

Dravet syndrome is a part of a group of diseases known as SCN1A related seizure disorders. This syndrome lies at the severe end of the spectrum of SCN1A related disorders but can be associated with other mutations as well. Mutations of the SCN1A gene is the cause of 79% of diagnosed cases. This intractable (uncontrollable) epilepsy is characterized by unilateral clonic or tonic clonic (grand mal) seizures that may progress to status epilepticus.

Currently, there is no unique code for Dravet syndrome. It is currently being reported by using code G40.8-, Other epilepsy. Dravet syndrome has a unique phenotypic spectrum, and the requestor states it is important to differentiate between Dravet syndrome with the SCN1A mutation and without the SCN1A mutation.

The following new codes are being requested to identify this condition for research and reporting.

TABULAR MODIFICATIONS

G40 Epilepsy

G40.8 Other epilepsy and recurrent seizures

New
sub-subcategory

G40.83 Dravet syndrome
Polymorphic epilepsy in infancy (PMEI)
Severe myoclonic epilepsy in infancy (SMEI)

Add

Add

New code

G40.831 Dravet syndrome, intractable, with SCN1A
mutation, with status epilepticus

New code

G40.832 Dravet syndrome, intractable, without SCN1A
mutation, without status epilepticus

New code

G40.833 Dravet syndrome, intractable, without SCN1A
mutation, with status epilepticus

New code

G40.839 Dravet syndrome, intractable, with SCN1A
mutation, without status epilepticus

Add

Dravet syndrome NOS

Drowning/Submersion Occurring in Natural Bodies of Water

Drowning is the third leading cause of unintentional death worldwide.ⁱ It is also a leading cause of unintentional death in the U.S., resulting in more than 4,500 deaths in 2016.ⁱⁱ Nonfatal drowning is far more prevalent and can have lifelong, debilitating consequences. For every child who dies from drowning, another five receive emergency department care for nonfatal submersion injuries.ⁱⁱⁱ In 2015, there were approximately 6,300 emergency department visits and 2,500 hospitalizations for non-fatal drowning/submersions (all ages).^{iv}

Prevention is key to reducing the burden of drowning, but drowning prevention receives relatively little attention and few resources. Prevention takes various forms such as public education, learn-to-swim programs, and the provision of lifeguards. The ability to target resources to prevent drowning could be enhanced by a detailed understanding of where drownings occur. Drowning occurs both in human-made (e.g., bathtubs, pools, fountains, tanks) and natural bodies of water (e.g., lakes, rivers, oceans). More than half of fatal and nonfatal drownings in the U.S. among those 15 years and older (57% and 57% respectively) occurred in natural water settings.^v Studies have shown that the percentage of drownings in natural settings increases with age.^{vi}

While ICD-10-CM provides specific external cause of injury codes for drowning/submersion in different types of human-made locations (e.g. W16.011, W16.021, W16.031, W16.211, W16.221, W16.311, W16.321, W16.331, W16.511, W16.521, W16.531, W22.041, W65, W67, W73, X71.0-X71.2, X92.0-X92.2, Y21.0-Y21.3), the codes for drowning/submersion in natural settings are limited (i.e., refer only to “natural water” without greater detail). Y92 ‘Place of occurrence of external cause’ codes are also limited. The only Y92 code that refers to a natural water site is Y92.832: “Beach as the place of occurrence of the external cause.” However, a beach could exist at a small pond, a large lake, a river, or the ocean. As well, drowning does not actually occur on a beach, but in offshore water.

Ocean beaches are unique in that they feature surf and rip currents, among other hazards. Ocean bays do not feature surf and rip currents, but have unique hazards related to cyclical tidal currents and scouring of the bottom. The Great Lakes are large enough to generate surf and rip currents, but not as reliably as the oceanfront, where surf is generally larger and more prevalent. Rivers feature relentless currents (typically unaffected by tides) and bottom scouring, along with various obstructions (e.g. rocks, trees, etc.) Ponds, lakes, and reservoirs are generally more benign than the foregoing, lacking hazards related to currents or surf, but for this reason tend to attract less accomplished swimmers, and thus appear to be locations for substantial numbers of drownings. Flooding is the second leading cause of weather-related death, most typically due to drowning.^{vii} Separating deaths during flooded periods can allow specific preventive and response resources to be targeted.

The United States Lifesaving Association and the National Oceanic and Atmospheric Administration joined forces over 10 years ago to attempt to prevent rip current drowning, which occurs in the waters off beaches with surf, since surf is the primary generator of rip currents. The lack of data regarding the number of drowning injuries and deaths in the waters off surf beaches has made it very challenging to affix the magnitude of the problem, set goals, marshal prevention resources, and benchmark injury prevention success. This is equally true for other natural venues.

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The United States Lifesaving Association is requesting new codes for drowning/submersion that occur in the following types of natural settings:

- Oceanfront
- Ocean – protected bay or inland waterway
- Great Lakes
- Pond, lake, reservoir
- River
- Flooded area

TABULAR MODIFICATIONS

New subcategory	W69	Accidental drowning and submersion while in natural water
Delete		Accidental drowning and submersion while in lake
Delete		Accidental drowning and submersion while in open sea
Delete		Accidental drowning and submersion while in river
Delete		Accidental drowning and submersion while in stream
New code	W69.1	Accidental drowning and submersion while in open sea
New code	W69.2	Accidental drowning and submersion while in ocean bay
Add		Accidental drowning and submersion while in Inland waterway
New code	W69.3	Accidental drowning and submersion while in oceanfront water
New code	W69.4	Accidental drowning and submersion while in Great Lakes
New code	W69.5	Accidental drowning and submersion while in lake
Add		Accidental drowning and submersion while in pond
Add		Accidental drowning and submersion while in reservoir
New code	W69.6	Accidental drowning and submersion while in river
Add		Accidental drowning and submersion while in stream
New code	W69.7	Accidental drowning and submersion while in flooded area
New code	W69.8	Accidental drowning and submersion while in other natural water

ⁱ Fact Sheet. 2017. World Health Organization. <http://www.who.int/mediacentre/factsheets/fs347/en/>

ⁱⁱ WONDER. Accessed January 5, 2018.

ⁱⁱⁱ WISQARS. Accessed January 5, 2018.

^{iv} Unintentional drowning: Get the facts. Centers for Disease Control and Prevention.

<https://www.cdc.gov/homeandrecreationalafety/water-safety/waterinjuries-factsheet.html>. Accessed January 7, 2018.

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^v Nonfatal and Fatal Drownings in Recreational Water Settings -- United States, 2001-2002. MMWR. Morbidity and mortality weekly report, Vol. 53, no. 21, 447-452. June 4, 2004.

^{vi} Weather related fatalities. 2016. National Weather Service. <http://www.nws.noaa.gov/om/hazstats.shtml>

Ehlers-Danlos Syndromes

Ehlers-Danlos syndromes (EDS) are a clinically and genetically heterogeneous group of heritable connective tissue disorders characterized by articular hypermobility, skin hyperextensibility or laxity, and tissue fragility affecting virtually every organ system: skin, ligaments, joints, bone, muscle, blood vessels and various organs.

Over the past two decades, the Villefranche Nosology, with a simplified classification of EDS into 6 major types based on major and minor criteria, has been widely used as the standard for the clinical diagnosis of EDS and for clinical research on various aspects of these conditions. An International Consortium on EDS, created in 2012, convened experts, performed comprehensive literature reviews and developed an updated evidence-based nosology and diagnostic criteria for each type of EDS which was released in early 2017: the 2017 International Classification of EDS. A new classification of EDS was published in March 2017, defining 13 types of EDS.

Physicians providing care for and persons living with EDS request that a unique set of ICD-10-CM codes be established that assign a separate ICD-10-CM code for each of the 13 EDS types, consistent with the 2017 International Classification of EDS. The classification and manuscripts about EDS were published in the March 2017 Part C Seminars in Medical Genetics issue of the American Journal of Medical Genetics and are all available through the Ehlers-Danlos Society (<http://bit.ly/EDS2017papers>).

This new classification recognizes EDS to be a plural, Ehlers-Danlos Syndromes, multiple distinct syndromes linked by a common pathophysiologic cause (connective tissue disorder) with similar physical features. It has incorporated new information about genetics and clinical manifestations of the syndromes into 13 significant types defined by major and minor clinical criteria, as well as the genetic etiology(ies), which provides a “descriptive” diagnosis that the patient can identify.

Being able to differentiate the type of EDS present has additional value with respect to healthcare utilization and costs. People with EDS have multiple problems and potentially have a higher use of healthcare services. Treatment focuses on management of symptoms and prevention of complications, e.g. dislocations, joint damage, vascular or organ ruptures. Medical management includes physical rehabilitation, physical and occupational therapy to address prevention and adaptation for daily activities, pain management modalities, orthotics and splinting, and mental health support to improve coping. Specific type diagnosis and coding in the medical record allows for more accurate analysis of the healthcare utilization and costs associated with the different types. This will facilitate clinical utility analysis to assess the impact of new treatments designed specifically for EDS.

Additional benefits of having unique expansion of the codes for this condition include but not limited to clarity of diagnosis for patient and clinician with improved ability for education, guidance on future prognosis and life-planning, and anticipatory management of existing and associated manifestations; more accurate diagnosis coding in the EHR record for short- and long-term communication to the healthcare team as well as for use in clinical research, healthcare resource utilization; a better understanding of the natural history and epidemiology of EDS and the types with increased diagnostic accuracy, ease of data collections, and understanding of prevalence, incidence etc.

With a separately identifiable ICD-10-CM code, general and type-specific international registries will be able to better define the natural history, clinical experience and epidemiology of EDS. Specific ICD-10-CM diagnosis codes for each type will ease the process of identifying a homogenous population of

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potential candidates for participation in research studies as new diagnosis and treatments become available.

In addition, a specific ICD-10-CM code for each type will be of value to the patient and the clinician. Regardless of the type experienced, EDS is a life- long progressive condition that has a major impact on the lives and daily function of most living with EDS.

The clinical reality, as demonstrated by the 2017 Classification, is that EDS is not a single syndrome with a single set of manifestations; it has 13 defined types. Therefore, in order to support current improvement in the classification and diagnosis of EDS types and its positive impact on patient care and to remain consistent with the international consensus, it is being proposed that ICD-10-CM be expanded to delineate the 13 types of EDS in accordance with nosology provided in the 2017 International Classification of EDS.

This proposal is being submitted jointly by Brad Tinkle, MD PHD, Division Chief of Clinical Genetics at Advocate Children’s Hospital and member of the Steering Committee of the International Consortium on EDS and Kay Jewell, MD Consultant, Acer Therapeutics Pharmaceutical Company. This proposal has been reviewed by the American Academy of Pediatrics Council on Genetics.

TABULAR MODIFICATIONS

Q79 Congenital malformations of musculoskeletal system, not elsewhere classified

New / Revise subcategory Q79.6 Ehlers-Danlos Syndromes

New code Q79.60 Ehlers-Danlos Syndrome, unspecified

New code Q79.61 Classical Ehlers-Danlos Syndrome
Add Classical EDS (cEDS)

New code Q79.62 Classical-like Ehlers-Danlos Syndrome
Add Classical-like EDS (clEDS)

New code Q79.63 Cardiac-valvular Ehlers-Danlos Syndrome
Add Cardiac-valvular EDS (cvEDS)

New code Q79.64 Vascular Ehlers-Danlos Syndrome
Add Vascular EDS (vEDS)

New code Q79.65 Hypermobile Ehlers-Danlos Syndrome
Add Hypermobile EDS (hEDS)

New code Q79.66 Arthrochalasia Ehlers-Danlos Syndrome
Add Arthrochalasia EDS (aEDS)

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New code Add	Q79.67 Dermatosparaxis Ehlers-Danlos Syndrome Dermatosparaxis EDS (dEDS)
New code Add	Q79.68 Kyphoscoliotic Ehlers-Danlos Syndrome Kyphoscoliotic EDS (kEDS)
New code Add	Q79.6A Brittle Cornea Ehlers-Danlos Syndrome Brittle Cornea Syndrome (BCS)
New code Add	Q79.6B Spondylodyplastic Ehlers-Danlos Syndrome Spondylodysplastic EDS (spEDS)
New code Add	Q79.6C Musculocontractural Ehlers-Danlos Syndrome Musculocontractural EDS (mcEDS)
New code Add	Q79.6D Myopathic Ehlers-Danlos Syndrome Myopathic EDS (mEDS)
New code Add	Q79.6E Periodontal Ehlers-Danlos Syndrome Periodontal EDS (pEDS)

Encounter for Examination of Eyes and Vision with Abnormal Findings

Vision screening is a requirement of well-child primary care as described by Bright Futures, 3rd edition, a joint program of HRSA and the American Academy of Pediatrics. There is little information on exactly who fails vision screening. The addition of new codes to ICD-10-CM would allow this information to be collected, retrieved as needed for performance measurement and reported. The screening encounter could also be separately retrieved for the presence of abnormal findings.

The American Academy of Ophthalmology is requesting new codes for an encounter for examination of eyes and vision when patients fail vision screening in order to be able to identify and monitor this condition. This proposal was originally presented at the September 2015 Coordination and Maintenance meeting. Modifications have been made to the proposal based on public comments and is now presented for reconsideration.

References:

Marsh-Tootle WL, Russ SA, Repka MX, for the National Expert Panel to the National Center for Children's Vision and Eye Health. Vision and Eye Health in Children 36 to <72 Months: Proposed Tracking Definitions. . *Optometry and Visual Science*. 2015;92(1):17-23.

TABULAR MODIFICATIONS

Z01 Encounter for other special examination without complaint, suspected or reported diagnosis

Z01.0 Encounter for examination of eyes and vision

Z01.00 Encounter for examination of eyes and vision
without abnormal findings
Encounter for examination of eyes and vision NOS

Z01.01 Encounter for examination of eyes and vision
with abnormal findings
Use additional code to identify abnormal findings

New
sub-subcategory

Z01.03 Encounter for examination of eyes and
vision following failed vision screening

New code

Z01.030 Encounter for examination of eyes and
vision following failed vision screening without
abnormal findings

New code

Z01.031 Encounter for examination of eyes and
vision following failed vision screening
with abnormal findings

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Add

Use additional code to identify
abnormal findings

Exertional Heat Stroke

Exertional Heat Stroke (EHS) is the most severe form of Exertional Heat Illness.ⁱ Unlike classical or passive heat stroke, which typically develops over days and occurs in hot environments, Exertional Heat Stroke can develop within hours, and often in healthy individuals undergoing strenuous activity in hot, humid environments.ⁱⁱ EHS is defined by an elevated core body temperature, neurocognitive dysfunction, and recent exertion or physical activity.^{iii,iv} EHS represents a life-threatening medical emergency with major complications and risks, including: death, brain damage/injury, acute renal injury/insufficiency, liver damage, rhabdomyolysis, and disseminated intravascular coagulation.^{v,vi} A specific ICD-10-CM code to distinguish Exertional Heat Stroke (EHS) has been requested by Eagle Pharmaceuticals.

Standard treatments for EHS consist of body cooling by physical methods (e.g., water immersion, ice packs, and water mist) and other supportive measures (e.g., I.V. fluids and respiratory support). Despite aggressive cooling techniques, over 30% of EHS survivors have neurocognitive sequelae.

Currently there are ICD-10-CM codes for heat-related illnesses including heatstroke/sunstroke, heat syncope, heat cramp, heat exhaustion, heat fatigue and heat edema. Each of which are appended with the appropriate 7th digit character to identify encounter type (initial, subsequent or sequela).

There is currently no ICD-10-CM code to specifically identify Exertional Heat Stroke. The lack of specific coding has been cited by researchers as an impediment to analysis.^{viii} The clear differentiation between EHS and other heat related illness is a necessity in evaluating all current and future treatment modalities. There is defined diagnostic criteria for EHS and the populations at risk appear to be different for EHS in comparison to “classic” heat stroke.

This requested modification will advance the capability for tracking all incidence and outcomes of exertional heat stroke. In keeping with the current tabular format, the addition of this code would be identified in an initial, subsequent, or sequela phase of diagnosis.

This proposal has been reviewed and supported by CDC Division of Analysis & Epidemiology.

TABULAR MODIFICATIONS

T67	Effects of heat and light
	Excludes1:erythema [dermatitis] ab igne (L59.0) malignant hyperpyrexia due to anesthesia (T88.3) radiation-related disorders of the skin and subcutaneous tissue (L55-L59)
	Excludes2:burns (T20-T31)

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sunburn (L55.-)
sweat disorder due to heat (L74-L75)

The appropriate 7th character is to be added to each code from category T67

A initial encounter
D subsequent encounter
S sequela

New subcategory	T67.0 Heatstroke and Sunstroke
Delete	Heat apoplexy
Delete	Heat Pyrexia
Delete	Siriasis
Delete	Thermoplegia

Use additional code(s) to identify any associated complications of heatstroke, such as:

coma and stupor (R40.-)
systemic inflammatory response syndrome (R65.1-)

New code	T67.01 Heatstroke and Sunstroke
Add	Heat apoplexy
Add	Heat Pyrexia
Add	Siriasis
Add	Thermoplegia

New Code: T67.02 Exertional Heat Stroke

New code T67.09 Other Heatstroke and Sunstroke

ⁱ Army Public Health Center. U.S. Army Public Health Command, Aberdeen Proving Ground, Maryland 2010

ⁱⁱ Moreno, *Int. J. Environ. Res. Public Health* **2016**, 13, 551

ⁱⁱⁱ Walter EJ, et al. *JICS*. 2012;13(4):304-330

^{iv} Leon LR, et al. *J Appl Physiol*. 2010;109(6):1980-1988.

^v Bouchama A, Knochel JP. *NEJM*. 2002;346:1978-1988.

^{vi} Leon LR, et al. *J Appl Physiol*. 2010;109(6):1980-1988.

^{vii} Sithinamsuwan P, et al. *Military Med*. 2009;174(5):496-502.

^{viii} Moreno, *Int. J. Environ. Res. Public Health* **2016**, 13, 551

Glucose-6-phosphate dehydrogenase deficiency without anemia

Glucose-6-phosphate dehydrogenase (G6PD) deficiency is an X-linked recessive genetic metabolic abnormality caused by deficiency of the enzyme G6PD. This enzyme is critical for the proper function of red blood cells. When the level of this enzyme is too low, red blood cells can break down prematurely (hemolysis). When the body cannot compensate for accelerated loss, anemia develops.

However, deficiency of this enzyme is not sufficient to cause hemolysis on its own. Additional factors are required to “trigger” the onset of symptoms. Triggers of hemolysis in G6PD-deficient persons include certain infectious diseases, exposure to certain drugs, and eating fava beans which can cause a potentially serious acute hemolytic anemia known as favism. Favism is a classic example of hemolytic anemia in a patient with G6PD deficiency, triggered by ingestion of fava beans.

G6PD can also be associated with a prolonged hyperbilirubinemia in newborns that increase the risk of developing kernicterus. The World Health Organization Working Group developed a classification for G6PD activity. Patients with class II or III G6PD deficiency are at the greatest risk for developing acute hemolytic anemias.

A majority of individuals with G6PD deficiency are asymptomatic throughout their life; however once the condition is diagnosed they should avoid triggers such as medications known to trigger the hemolytic anemia, e.g. primaquine, sulfanilamide, nitrofurantoin and methylene blue.

Currently, ICD-10-CM coding for patients with G6PD deficiency defaults to D55.0 Anemia due to glucose-6-phosphate dehydrogenase [G6PD] deficiency. There is no unique code to identify the majority of individuals who do not have anemia, but are at risk.

The American Academy of Pediatrics respectfully requests that a new code be established to identify patients who have G6PD deficiency without anemia.

TABULAR MODIFICATIONS

D55 Anemia due to enzyme disorders

 D55.0 Anemia due to glucose-6-phosphate dehydrogenase (G6PD) deficiency
 Favism
 G6PD deficiency anemia

Add Excludes1: Glucose-6-phosphate dehydrogenase (G6PD) deficiency without anemia (D75.A)

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D75 Other and unspecified diseases of blood and blood-forming organs

New Code D75.A Glucose-6-phosphate dehydrogenase (G6PD) deficiency without anemia

Add Excludes1: Glucose-6-phosphate dehydrogenase (G6PD) deficiency with anemia
(D55.0)

Immunocompromised Status

An immunocompromised status is a state in which a person's immune system is immunosuppressed or weakened. Individuals who are immunocompromised are less capable of battling infections because the immune system response is not functioning properly.

Treating a patient who is immunocompromised poses more risks and challenges, therefore it is very important to be able to identify a patient with this status.

These individuals are more prone to serious infections, opportunistic infections and other types of complications. A patient may be immunocompromised due to a specific clinical condition such as HIV, AIDS, certain cancers and genetic disorders. There are also external factors such as treatment with certain medications or exposure to radiation therapy, or a combination of both clinical conditions and external factors.

There are circumstances where a patient may be immune competent because of improvement of an underlying condition that can affect the immune system, but become immunocompromised because of an acute illness, new treatment or medication, e.g. bone marrow transplant with a fever.

A patient whose immune system is suppressed because of illness or external factors generally requires greater resource utilization. These patients are at increased risk because of fevers, non-environmental hypothermia, or injury thus requiring more interventions such as laboratory testing and medications than those with normally functioning immune systems.

Clinicians routinely document in the medical record when a patient's immune system may be compromised by using terms such as "immunodeficiency," "immunosuppressed" or "immunocompromised." Conditions within category D80-D89, Certain disorders involving the immune mechanism, do not indicate that a patient is immunocompromised and are generally specific to the type of immune deficiency. The codes D84.8, Other specified immunodeficiencies and D89.89, Other specified disorders involving the immune mechanism, not elsewhere classified, are not specific enough to capture the details as to why a patient's immune system status is compromised and which places the patient at greater health risks.

Since this information cannot easily be inferred by other contributing diagnoses, the American Academy of Pediatrics (AAP) proposes that new codes be created to indicate when a patient is immunocompromised. This topic was originally presented at the March C&M meeting. However, in response to public comment and reevaluation of the code placement, a revised proposal is being presented.

The American Academy of Pediatrics requests the following tabular modifications:

TABULAR MODIFICATIONS

D84 Other immunodeficiencies

New subcategory	D84.8 Other specified immunodeficiencies
Add	Immunocompromised
Add	Immunodeficient
Add	Immunosuppressed
New code	D84.81 Immunodeficiency due to conditions classified elsewhere
Add	Code first underlying disease, if known, such as:
Add	acquired absence of spleen (Z90.81)
Add	chromosomal abnormalities (Q90-Q99)
Add	congenital absence and malformations of spleen (Q89.0)
Add	diabetes mellitus (E08-E13)
Add	human immunodeficiency virus (B20)
Add	malignant neoplasms (C00-C96)
Add	transplanted organ and tissue (Z94)
Add	Excludes1: combined immunodeficiencies (D81.-)
Add	common variable immunodeficiency (D83.-)
Add	defects in the complement system (D84.1)
Add	immunodeficiency associated with other major defects (D82.-)
Add	immunodeficiency with predominantly antibody defects (D80.-)
Add	lymphocyte function antigen-1 [LFA-1] defect (D84.0)
New sub-subcategory	D84.82 Immunodeficiency due to drugs and external causes
New code	D84.821 Immunodeficiency due to drugs
Add	Code also drug or medication such as:
Add	encounter for antineoplastic chemotherapy and immunotherapy (Z51.1)
Add	long term (current) drug therapy (Z79.-)

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New code D84.822 Immunodeficiency due to external causes
Add Code also external cause such as:
Add encounter for antineoplastic radiation
therapy (Z51.0)
Add exposure to ionizing radiation (W88)
Add other contact with and (suspected)
exposures hazardous to health (Z77)

New code D84.89 Other specified immunodeficiencies, not elsewhere
classified
Add Immunocompromised NOS
Add Immunodeficient NOS
Add Immunosuppressed NOS

Intracranial Hypotension

Intracranial hypotension results from a loss of cerebrospinal fluid volume. Intracranial hypotension is an under-recognized and under-diagnosed central nervous system disorder.

The causes 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 durotomies during spinal or other surgeries
- Over-drainage of cerebrospinal fluid shunting devices
- Traumatic spinal dural tears or defects resulting in spinal cerebrospinal fluid leaks

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.

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, they should be coded additionally. It is also of note that cranial MRI findings, or spinal imaging findings may or may not be evident. This proposal does not suggest code changes related to intracranial hypotension due to a traumatic etiology.

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

The International Classification of Headache Disorders, 3 beta, 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

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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.

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TABULAR MODIFICATIONS

	G96	Other disorders of central nervous system
	G96.0	Cerebrospinal fluid leak
Add		Code also spontaneous intracranial hypotension, if applicable (G96.81) Excludes1: cerebrospinal fluid leak from spinal puncture (G97.0)
New code	G96.00	Cerebrospinal fluid leak, unspecified

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New code G96.01 Cerebrospinal fluid leak from skull base, spontaneous

New code G96.02 Spinal cerebrospinal fluid leak, spontaneous

New code G96.09 Other cerebrospinal fluid leak

G96.1 Disorders of meninges, not elsewhere classified

Add G96.11 Dural tear
Code also spontaneous intracranial hypotension, if applicable (G96.81)
Excludes1:accidental puncture or laceration of dura during a procedure (G97.41)

G96.8 Other specified disorders of central nervous system

New code G96.81 Intracranial hypotension, spontaneous, with or without confirmed spinal cerebrospinal fluid leak

Add Code also any associated diagnoses, such as:
Brachial amyotrophy (G54.5)
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)
Other symptoms and signs involving cognitive functions and awareness (R41.-)
Reversible cerebrovascular vasoconstriction syndrome (I67.841)
Somnolence, stupor and coma (R40.-)
Spinal cord herniation (G95.89)
Stroke (I63.-)
Syringomyelia (G95.0)

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

Add G97.0 Cerebrospinal fluid leak from spinal puncture
Code also any associated diagnoses or complications, such as:
Add Intracranial hypotension following a procedure (G97.83)

Add G97.1 Other reaction to spinal and lumbar puncture
Headache due to lumbar puncture
Other reaction to spinal dural puncture

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Revise	G97.2	Intracranial hypotension following ventricular <u>or lumbar cerebrospinal fluid</u> 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
	G97.81	Other intraoperative complications of nervous system
	G97.82	Other postprocedural complications and disorders of nervous system
New code	G97.83	Intracranial hypotension following 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
New code	G44.86	Headache with orthostatic or positional component, not elsewhere classified

Left Against Medical Advice

Currently in ICD-10-CM, the code title for Z53.21 is Procedure and treatment not carried out due to patient leaving prior to being seen by health care provider. Following the alphabetical index, Procedure not done, because of patient's decision, left against medical advice (AMA) is also indexed to this code.

When a patient has left AMA, it is inferred that the patient was seen by a health care provider.

The indexing has caused confusion when trying to code AMA to the appropriate code. It has been requested that a new code be created to separately identify those patients that leave against medical advice from those that leave before being seen by a provider.

TABULAR MODIFICATIONS

Z53	Persons encountering health services for specific procedures and treatment, not carried out
	Z53.2 Procedure and treatment not carried out because of patient's decision for other and unspecified reasons
	Z53.20 Procedure and treatment not carried out because of patient's decision for unspecified reasons
	Z53.21 Procedure and treatment not carried out due to patient leaving prior to being seen by health care provider
New code	Z53.23 Procedure and treatment not carried out due to patient leaving after being seen by health care provider
	Z53.29 Procedure and treatment not carried out because of patient's decision for other reasons

INDEX MODIFICATIONS

	Procedure (surgical)
	- not done Z53.9
	- - because of
	- - - patient's decision Z53.20
	- - - - for reasons of belief or group pressure Z53.1
Revise	- - - - left against medical advice (AMA) Z53.21 Z53.23
	- - - - specified reason NEC Z53.29
	- - - specified reason NEC Z53.8

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New code	T50.914 Poisoning by multiple unspecified drugs, medicaments and biological substances, undetermined
New code	T50.915 Adverse effect of multiple unspecified drugs, medicaments and biological substances
New code	T50.916 Underdosing of multiple unspecified drugs, medicaments and biological substances

Orbital Roof and Wall Fracture

This topic was presented originally at the March 2017 Coordination and Maintenance meeting. This revised proposal is based on public comments received and further discussions with the American Academy of Ophthalmology. Orbital fractures may be defined in terms of anatomic location, including isolated fractures of the orbital floor, medial wall, temporal wall, and roof. These fractures are commonly seen with midfacial trauma.

Currently, there is only one code for orbital bone fractures, S02.3-, Fracture of orbital floor. There is no unique code in ICD-10-CM for capturing the diagnosis of an orbital roof fracture. These are reported using code S02.19, Other fracture of base of skull. There are three other walls of the orbit, including the roof, medial wall and temporal wall.

The American Academy of Ophthalmology is proposing the following tabular modifications for new codes to identify these specific types of fracture.

TABULAR MODIFICATIONS

	S02	Fracture of skull and facial bones
	S02.1	Fracture of base of skull
Delete		Excludes1: orbit NOS (S02.8)
Add		Excludes1: orbit NOS (S02.B)
Add		Excludes2: medial orbital wall (S02.A1)
Add		Excludes2: lateral orbital wall (S02.A2)
New sub-subcategory	S02.12	Fracture of orbital roof
New code	S02.121	Fracture of orbital roof, right side
New code	S02.122	Fracture of orbital roof, left side
New code	S02.129	Fracture of orbital roof, unspecified side
	S02.19	Other fracture of base of skull
Delete		Fracture of orbital roof
	S02.3	Fracture of orbital floor
Add		Fracture of inferior orbital wall
Delete		Excludes1: orbit NOS (S02.8)
Add		Excludes1: orbit NOS (S02.B)
Add		Excludes2: medial orbital wall (S02.A1)

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Add	Excludes2:	lateral orbital wall (S02.A2)
	S02.8	Fracture of other specified skull and facial bones
Delete		Fracture of orbit NOS
Delete		Excludes1: orbital roof (S02.1-)
Delete		Excludes1: orbital floor (S02.3-)
Add	Excludes2:	medial orbital wall (S02.A1)
Add	Excludes2:	lateral orbital wall (S02.A2)
Add	Excludes2:	orbital roof (S02.1-)
Add	Excludes2:	orbital floor (S02.3-)
New code	S02.A	Fracture of other orbital wall
Add	Excludes1:	orbit NOS (S02.B)
Add	Excludes2:	orbital roof (S02.1-)
Add	Excludes2:	orbital floor (S02.3-)
New sub-subcategory	S02.A1	Fracture of medial orbital wall
New code	S02.A11	Fracture of medial orbital wall, right side
New code	S02.A12	Fracture of medial orbital wall, left side
New code	S02.A19	Fracture of medial orbital wall, unspecified side
New sub-subcategory	S02.A2	Fracture of lateral orbital wall
New code	S02.A21	Fracture of lateral orbital wall, right side
New code	S02.A22	Fracture of lateral orbital wall, left side
New code	S02.A29	Fracture of lateral orbital wall, unspecified side
New code	S02.B	Fracture of orbit, unspecified
Add		Fracture of orbit NOS
New code	S02.C	Fracture of orbital wall, unspecified
Add		Fracture of orbit wall NOS

Personal History of In-situ Neoplasms

Carcinoma in-situ is a group of abnormal cells that are found only in the place where they first formed in the body, which may become cancer and spread to nearby normal tissue. The American Joint Committee on Cancer (AJCC) includes carcinoma in-situ (or tumor in-situ, Tis) in the staging system as it denotes the important relationship of the “T” status and overall risk of recurrence or progression. This was presented in March 2017; this update is based on comments.

There are currently specific codes in ICD-10-CM for personal history of carcinoma in-situ of the breast (Z86.000), cervix uteri (Z86.001), and other site (Z86.008). The Alliance of Dedicated Cancer Centers (ADCC) has requested expansion, to include codes for carcinoma in-situ of other additional specific sites. In some cases, patients may be treated for more than one type of cancer and history of in-situ neoplasms. This requested detail would enable better specificity and more accurate reporting of these diagnoses, and support assessment of risk for recurrence or potential need for future screening in particular cases.

TABULAR MODIFICATIONS

	Z86	Personal history of certain other diseases
	Z86.0	Personal history of in-situ and benign neoplasms and neoplasms of uncertain behavior
	Z86.00	Personal history of in-situ neoplasm
	Z86.000	Personal history of in-situ neoplasm of breast
Add		Conditions classifiable to D05
	Z86.001	Personal history of in-situ neoplasm of cervix uteri
Add		Conditions classifiable to D06
New code	Z86.002	Personal history of in-situ neoplasm of other and unspecified genital organs
		Personal history of vaginal intraepithelial neoplasia III [VAIN III]
		Personal history of vulvar intraepithelial neoplasia III [VIN III]
		Personal history of high-grade prostatic intraepithelial neoplasia III [HGPIN III]
		Conditions classifiable to D07

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New code	Z86.003	Personal history of in-situ neoplasm of oral cavity, esophagus and stomach Conditions classifiable to D00
New code	Z86.004	Personal history of in-situ neoplasm of other and unspecified digestive organs Personal history of anal intraepithelial neoplasia (AIN III) Conditions classifiable to D01
New code	Z86.005	Personal history of in-situ neoplasm of middle ear and respiratory system Conditions classifiable to D02
New code	Z86.006	Personal history of melanoma in-situ Conditions classifiable to D03 Excludes 2: sites other than skin- code to personal history of in-situ neoplasm of the site
New code	Z86.007	Personal history of in-situ neoplasm of skin Personal history of carcinoma in situ of skin Conditions classifiable to D04
Delete	Z86.008	Personal history of in-situ neoplasm of other site Personal history of vaginal intraepithelial neoplasia III [VAIN III]
Delete		Personal history of vulvar intraepithelial neoplasia III [VIN III] Conditions classifiable to D09

Post Endometrial Ablation Syndrome

The American Congress of Obstetricians and Gynecologists (ACOG) is requesting a new code to report post endometrial ablation syndrome.

This proposal was presented at the September 2016 C&M meeting. Based on public comments, changes have been made and the proposal is being resubmitted for consideration.

Global endometrial ablation is a procedure that is commonly performed for reproductive-aged women with menstrual disorders to include menorrhagia and menometrorrhagia. This procedure has been used in clinical practice for over two decades. Post endometrial ablation syndrome is a condition that may occur in up to 10% of women who undergo endometrial ablation that includes cyclic pain and hematometra. This condition occurs most commonly in women who have previously had fallopian tube occlusion performed for sterilization purposes.

Although ICD-10-CM allows one to code for the signs and symptoms related to this condition (e.g. pelvic pain, hematometra), post endometrial ablation syndrome occurs frequently enough that a separate code is warranted for better coding specificity and tracking purposes.

ACOG proposes the following tabular modification.

TABULAR MODIFICATION

N99	Intraoperative and postprocedural complications and disorders of genitourinary system, not elsewhere classified
N99.8	Other intraoperative and postprocedural complications and disorders of genitourinary system
New code	N99.85 Post endometrial ablation syndrome

Prader-Willi Syndrome

Prader-Willi syndrome (PWS) is a complex neurodevelopment disorder that affects many parts of the body. The Foundation for Prader-Willi Research (FPWR), the Prader-Willi Syndrome Association (USA) (PWSA(USA)), and the International Prader-Willi Syndrome Organisation (IPWSO) have requested that a specific ICD-10-CM code be created for PWS.

During infancy, PWS is characterized by hypotonia, feeding difficulties, poor growth, and delayed development. Beginning in childhood, affected individuals develop an insatiable appetite, which leads to chronic overeating (hyperphagia) and obesity. PWS is the most common genetic syndrome causing obesity. Some people with Prader-Willi syndrome, particularly those with obesity, also develop type 2 diabetes.

People with Prader-Willi syndrome typically have mild to moderate intellectual impairment and learning disabilities. Behavioral problems are common, including temper outbursts, stubbornness, and compulsive behavior such as picking at the skin. Sleep abnormalities can also occur. Additional features of this condition include distinctive facial features such as a narrow forehead, almond-shaped eyes, and a triangular mouth; short stature; and small hands and feet. Some people with Prader-Willi syndrome have unusually fair skin and light-colored hair. Both affected males and affected females have underdeveloped genitals. Puberty is delayed or incomplete, and most affected individuals are infertile.

PWS is caused by lack of expression of genes in the paternally inherited chromosome 15q11.2-q13 region, which has been referred to as the PWS/Angelman syndrome region (Angelman syndrome involves lack of expression of genes in the maternally inherited chromosome 15). The specific genetic mechanisms that cause PWS include paternal 15q microdeletion (estimated about two thirds of cases), chromosome 15 maternal uniparental disomy (UPD) (estimated about one third of cases), and imprinting defects (estimated 1-3% of cases). PWS due to deletions may be further divided into type I and type II, with type I deletions being larger (~7Mb, compared to ~5Mb for type II). Rarely, PWS is caused by chromosomal translocations. The estimated prevalence of PWS is 1:10,000 to 1:30,000.

PWS previously had a unique code in ICD-9-CM (759.81), but is now coded to Q87.1, Congenital malformation syndromes predominantly associated with short stature. It is grouped together with a number of different syndromes with which it shares little, other than short stature. With regard to PWS treatment and short stature, it is now generally treated with growth hormone, and as a result, most people with PWS do not have short stature, and also can have partially normalized facial growth (vertically), although certain distinctive facial characteristics remain.

Creation of a unique ICD-10-CM code for PWS would facilitate communication and research related to PWS, including research and clinical trials on effectiveness of new and novel interventions. Having PWS grouped with other syndromes causing short stature is confusing for some health care providers, particularly since most patients now take growth hormone and thus do not have short stature, as well as the fact that most cases are recognized to be due to chromosomal deletions, which are generally coded elsewhere. There are important genetic subtype differences that have been identified with respect to specific clinical features of PWS and inheritance/recurrence risk. Those with PWS due to UPD or an imprinting defect have increased susceptibility to autism and psychosis. In addition, differences in IQ scores and behavioral manifestations have been reported between type I and type II deletion patients,

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with additional studies needed for confirming and evaluating this finding. Thus the phenotypic features unique to the molecular mechanism of disease have important implications for treatment of behavioral concerns. It is also important to note the differences in genetic counseling that may be required depending on the molecular mechanism of PWS. An imprinting defect or translocation could carry a very high risk (up to 50%) of recurrence in a future pregnancy, whereas UPD and deletion cases carry a very low recurrence risk. A unique code for PWS will facilitate research to better understand the clinical implications of the different genetic subtypes.

It is proposed to differentiate PWS from “Prader-Willi-like syndrome” (PWLS), which has been described in the medical literature, sharing a number of features with PWS (hypotonia, obesity, hyperphagia, short extremities, and delayed development). However, despite similarities, PWLS is clinically and genetically heterogeneous, and differs from PWS. At this time, it is proposed to create a specific code for PWLS, although another option would be to group PWLS with the proposed new code for other congenital malformation syndromes predominantly associated with short stature. Some of those individuals with PWLS are diagnosed with genetic SIM1 deficiency (single gene at chromosome 6 q16.3), or with Schaaf-Yang syndrome (involving an abnormality in a single gene on chromosome 15, which is also involved in PWS), among other specific genetic disorders, although some do not have a disorder identified.

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TABULAR MODIFICATIONS

Q87 Other specified congenital malformation syndromes affecting multiple systems

Q87.1 Congenital malformation syndromes predominantly associated with short stature

Delete	Aarskog syndrome
Delete	Cockayne syndrome
Delete	De Lange syndrome
Delete	Dubowitz syndrome
Delete	Noonan syndrome
Delete	Prader-Willi syndrome
Delete	Robinow-Silverman-Smith syndrome
Delete	Russell-Silver syndrome
Delete	Seckel syndrome

New code	Q87.11	Prader-Willi syndrome
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New code	Q87.12	Prader-Willi-like syndrome
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New code	Q87.19	Other congenital malformation syndromes predominantly associated with short stature
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Add	Aarskog syndrome
Add	Cockayne syndrome
Add	De Lange syndrome
Add	Dubowitz syndrome
Add	Noonan syndrome
Add	Robinow-Silverman-Smith syndrome
Add	Russell-Silver syndrome
Add	Seckel syndrome

Presence of Other Specified Functional Implants

Neurostimulators are relatively common implanted devices and are used to treat symptoms associated with a variety of disorders, including multiple sclerosis, Parkinson's disease, chronic intractable pain, urinary and fecal incontinence, and gastroparesis among others. Similar to cardiac pacemakers and defibrillators, neurostimulator systems consist of an electrical generator in the subcutaneous tissue attached to a lead (or leads) situated at the nerves being stimulated. These nerves include, but are not limited to, nerves in the brain, peripheral nerves, vagus nerve, spinal cord, sacral nerves, or stomach. ICD-10-CM codes exist for the presence of cardiac pacemakers and defibrillators, as well as for other cardiac and vascular implants. It would be useful to create a corresponding code for the presence of neurostimulators. The existing ICD-10-CM code for attention to neurostimulators, assigned for routine device replacement for example, uses the outdated term "neuropacemaker." This is the only place in ICD-10-CM where this term appears. It is requested to update the wording to "neurostimulator" for consistency. It is also requested that inclusion terms be added to encompass the full range of neurostimulator sites.

The American Academy of Neurology (AAN) has reviewed and supports this request.

TABULAR MODIFICATIONS

Z45 Encounter for adjustment and management of implanted device

Includes: removal or replacement of implanted device

Excludes1: malfunction or other complications of device - see Alphabetical Index
presence of prosthetic and other devices (Z95-Z97)

Z45.4 Encounter for adjustment and management of implanted nervous system device

Revise Z45.42 Encounter for adjustment and management of ~~neuropacemaker~~
neurostimulator (brain) (peripheral nerve) (spinal cord) (vagus nerve)
(sacral nerve)(gastric)

Z96 Presence of other functional implants

Z96.8 Presence of other specified functional implants

Excludes2: complications of internal prosthetic devices, implants and grafts (T82-T85)
fitting and adjustment of prosthetic and other devices (Z44-Z46)

New code Add Z96.82 Presence of neurostimulator
Presence of brain, peripheral nerve, spinal cord, vagus nerve,
sacral nerve, gastric neurostimulator

Z96.89 Presence of other specified functional implants

Pyuria

Pyuria is the presence of white blood cells in the urine. It is a laboratory finding in many diseases, most commonly found in urinary tract infections. Sometimes urine can be white or cloudy and look as though there might be white blood cells in it, but may just be phosphates in a concentrated urine. However, there could be other causes of pyuria. White blood cells in the urine can be due to inflammation, kidney stone, tumor, etc.

Pyuria is currently indexed to code N39.0, Urinary tract infection. There is no unique code for reporting pyuria. Sterile pyuria is the finding when a patient has pyuria and no cause can be found (no tumor, no true urinary tract infection, and no stone disease).

A practitioner's office has requested new codes to identify this finding when clinical exploration is being done to determine the underlying diagnosis.

TABULAR MODIFICATIONS

R82 Other and unspecified abnormal findings in urine

New
subcategory

R82.8 Abnormal findings on cytological and histological examination of
urine

New code
Add

R82.81 Pyuria
Sterile pyuria

Rheumatoid Arthritis in Remission

Rheumatoid arthritis (RA) is the most common autoimmune inflammatory arthritis in adults. RA has a significant negative impact on the ability to perform daily activities, including work and household tasks.

The American College of Rheumatology (ACR) last published a guideline for RA management in 2015, due to rapid accrual of evidence and new therapies, advancement of guideline development methodologies, and the need to broaden the scope of its 2012 RA recommendations. The guideline includes 6 measures, used as instruments for determining RA disease activity. Each of the 6 has a threshold of disease activity used to define remission. These thresholds are used to determine if tapering or discontinuation of disease-modifying antirheumatic drug (DMARD) therapy is appropriate.

Currently, ICD-10-CM does not include a code to recognize RA patients in remission. Patients in remission may not require the same treatment as patients with active RA. For example, some patients in remission do not need to be on a disease-modifying anti-rheumatic drug (DMARD). Currently, DMARD use is one of the criteria for providing quality care in the Healthcare Effectiveness Data and Information Set (HEDIS), as well as a measure of quality care tracked in the ACR registry.

Kaiser Permanente is requesting the following new codes to differentiate between active RA and clinically inactive (i.e. – RA in remission), for correct application of ACR treatment recommendations and accurate reporting and tracking of clinical outcomes.

The American College of Rheumatology has reviewed and supports this proposal.

TABULAR MODIFICATIONS

	M05	Rheumatoid arthritis with rheumatoid factor
	M05.9	Rheumatoid arthritis with rheumatoid factor, unspecified
New code	M05.A	Rheumatoid arthritis with rheumatoid factor in remission
	M06	Other rheumatoid arthritis
	M06.9	Rheumatoid arthritis, unspecified
New code	M06.A	Rheumatoid arthritis in remission
	M08	Juvenile arthritis
	M08.1	Juvenile ankylosing spondylitis
Add		Excludes1: juvenile arthritis in remission (M08.A)
	M08.9	Juvenile arthritis, unspecified

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New code
Add

M08.A Juvenile arthritis in remission
Excludes1: juvenile ankylosing spondylitis (M08.1)

Sickle Cell Disease

Currently in ICD-10-CM, patients with sickle cell vasoocclusive crisis not associated with acute chest syndrome or splenic sequestration are coded as “with crisis, unspecified”. In the majority of these encounters, the vasoocclusive pain crisis is the problem that requires medical intervention as other major complications may not be present. Therefore, the vasoocclusive pain is considered inherent and not a manifestation.

Currently there is no adequate means to track patients with other types of complications in addition to acute chest syndrome and splenic sequestration. Other complications include but not limited to acute gall bladder involvement, priapism or fever.

Cerebral infarcts are also a major complication in patients with sickle cell disease. According to the CDC, about 10% of children with sickle cell disease will have a symptomatic stroke.

The American Academy of Pediatrics requests tabular modifications for sickle cell disorders with crisis to identify patients without major complications but who are in crisis. This proposal was originally presented at the September 2016 C&M meeting. However based on public comments, the proposal is being represented for consideration.

TABULAR MODIFICATIONS

D57 Sickle-cell disorders
Use additional code for any associated fever (R50.81)

D57.0 Hb-SS disease with crisis
Sickle-cell disease NOS with crisis
Hb-SS disease with vasoocclusive pain

Revise D57.00 Hb-SS disease with crisis with unspecified complication
Add Hb-SS with crisis NOS

Revise D57.01 Hb-SS disease with crisis with acute chest syndrome

Revise D57.02 Hb-SS disease with crisis with splenic sequestration

New code D57.03 Hb-SS disease with crisis with cerebral vascular complication
Add Code also: cerebral infarction (I63)

New code D57.04 HB-SS disease with crisis without complication

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- New code D57.09 HB-SS disease with crisis with other specified complication
Add Code also manifestations, such as:
Add cholelithiasis (K80)
Add priapism (N48.32)
- D57.2 Sickle-cell/Hb-C disease
Hb-SC disease
Hb-S/Hb-C disease
- D57.21 Sickle-cell/Hb-C disease with crisis
- Revise D57.211 Sickle-cell/Hb-C disease with crisis with acute chest syndrome
- Revise D57.212 Sickle-cell/Hb-C disease with crisis with splenic sequestration
- New code D57.213 Sickle-cell/Hb-C disease with crisis with cerebral vascular involvement
Add Code also: cerebral infarction (I63)
- New code D57.214 Sickle-cell/Hb-C disease with crisis without other complications
- New code D57.218 Sickle-cell/Hb-C disease with crisis with other specified complication
Add Code also manifestations, such as:
Add cholelithiasis (K80)
Add priapism (N48.32)
- D57.219 Sickle-cell/Hb-C disease with crisis, unspecified
Sickle-cell/Hb-C disease with crisis NOS
- D57.4 Sickle-cell thalassemia
Sickle-cell beta thalassemia
Thalassemia Hb-S disease
- D57.41 Sickle-cell thalassemia with crisis
Sickle-cell thalassemia with crisis with vasoocclusive pain
- Revise D57.411 Sickle-cell thalassemia with crisis with acute chest syndrome

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- Revise D57.412 Sickle-cell thalassemia with crisis with splenic sequestration
- New code D57.413 Sickle-cell/Hb-C disease with crisis with cerebral vascular involvement
- Add Code also: cerebral infarction (I63)
- New Code D57.414 Sickle-cell thalassemia with crisis without other manifestations
- New code D57.418 Sickle-cell thalassemia with crisis with other specified complication
- Add Code also manifestations, such as:
- Add cholelithiasis (K80)
- Add priapism (N48.32)
- D57.419 Sickle-cell thalassemia with crisis, unspecified
Sickle-cell thalassemia with crisis NOS
- D57.8 Other sickle-cell disorders
Hb-SD disease
Hb-SE disease
- D57.81 Other sickle-cell disorders with crisis
- Revise D57.811 Other sickle-cell disorders with crisis with acute chest syndrome
- Revise D57.812 Other sickle-cell disorders with crisis with splenic sequestration
- New code D57.813 Sickle-cell/Hb-C disease with crisis with cerebral vascular involvement
- Add Code also: cerebral infarction (I63)
- New code D57.814 Other sickle-cell disorders with crisis without other complications
- New code D57.818 Other sickle-cell disorders with crisis with other specified complication
- Add Code also manifestations, such as:
- Add cholelithiasis (K80)
- Add priapism (N48.32)
- D57.819 Other sickle-cell disorders with crisis, unspecified
Other sickle-cell disorders with crisis NOS

Traumatic Brain Herniation

Brain herniation occurs when brain tissue, cerebrospinal fluid, and blood vessels are moved or pushed away from their usual position inside the skull. Pressure resulting in such movement can be due to brain swelling from a head injury, stroke, brain tumor, abscess, hydrocephaly, or other underlying cause. Brain herniation can occur between areas inside the skull, such as those separated by a rigid membrane like the tentorium or falx, or to the outside of the skull, through the foramen magnum, or through a craniotomy opening, or other defect, whether traumatic or congenital. Traumatic brain injury is one of the most common causes of brain herniation.

Different parts of the brain may herniate, each causing a different clinical syndrome. Subfalcine herniation occurs when the cingulate gyrus herniates under the falx cerebri, resulting in a midline shift. Uncal herniation involves the medial temporal lobe herniating through the tentorium (also called a lateral transtentorial herniation). Central descending transtentorial herniation occurs with downward herniation of both temporal lobes through the tentorium. Ascending transtentorial herniation occurs in the opposite direction, with the cerebellum pushing upward through the tentorium. Tonsillar herniation occurs when the lower part of the cerebellum and brainstem herniates downward into the foramen magnum.

Brain herniation can cause a number of signs and symptoms (e.g., pupillary dilation), and sometimes can be fatal in a short time if not treated. The presence or absence of brain herniation is very important clinically.

Nontraumatic brain herniation is currently being captured with the use of code G93.5, Compression of brain. However, traumatic compression of brain is excluded from there, to codes at S06.2, Diffuse traumatic brain injury, and S06.3, Focal traumatic brain injury. It would also be possible to have brain herniation related to other codes within category S06. However, at this time, it is not possible to differentiate whether or not brain herniation is present using these codes.

Traumatic brain injury is also an active and important area of research. Having codes for traumatic herniation has potential to help with future research that could advance the care of these incredibly ill patients. A request to provide specific codes for traumatic brain herniation has been received from the University of Utah Health, Neurology Department.

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TABULAR MODIFICATIONS

S06 Intracranial injury

S06.8 Other specified intracranial injuries

New
 sub-subcategory

S06.83 Traumatic brain compression

Add

Traumatic cerebral compression

Add

Code first the underlying traumatic brain injury, such as:

Add

Diffuse traumatic brain injury (S06.2)

Add

Focal traumatic brain injury (S06.3-)

Add

Traumatic subdural hemorrhage (S06.5-)

Add

Traumatic subarachnoid hemorrhage (S06.6-)

New code

S06.830

Traumatic brain compression with no herniation

Add

Traumatic cerebral compression with no herniation

New code

S06.831

Traumatic brain compression with tonsillar
 herniation

Add

Traumatic cerebral compression with tonsillar
 herniation

Add

Traumatic cerebellar compression with tonsillar
 herniation

Add

Traumatic brainstem compression with tonsillar
 herniation

Add

Coning

New code

S06.832

Traumatic brain compression with ascending
 transtentorial herniation

Add

Traumatic cerebellar compression with ascending
 transtentorial herniation

Add

Reverse coning

New code

S06.833

Traumatic brain compression with subfalcine
 herniation

Add

Traumatic cerebral compression with subfalcine
 herniation

New code

S06.834

Traumatic brain compression with central
 transtentorial herniation

Add

Traumatic cerebral compression with central
 transtentorial herniation

New code

S06.835

Traumatic brain compression with uncal herniation

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Add		Traumatic cerebral compression with uncal herniation
Add		Traumatic brain compression with lateral transtentorial herniation
Add		Traumatic cerebral compression with lateral transtentorial herniation
New code	S06.836	Traumatic brain compression with transcalvarial herniation
Add		Traumatic cerebral compression with transcalvarial herniation
New code	S06.838	Other traumatic intracranial compression
Add		Traumatic brain compression with other herniation
Add		Traumatic cerebral compression with other cerebral herniation
New code	S06.839	Unspecified traumatic intracranial compression
Add		Traumatic brain compression with unspecified herniation
Add		Traumatic cerebral compression with unspecified cerebral herniation

Travel Counseling

While there are a number of codes for a variety of counseling services, currently there are no unique ICD-10-CM codes for counseling services related to travel. Patients and caregivers often seek travel counseling services without any signs or symptoms and /or unrelated to preventive medical care.

For example, when planning a trip to a particular country or region, the parent (or caregiver) may want to review with the physician potential risk factors such as safe drinking water or disease prevention.

The American Academy of Pediatrics (AAP) reports that there have been an increase in the number of patients seen for these services. AAP is requesting specific new codes to identify travel related encounters and to be able to track these encounters.

TABULAR MODIFICATIONS

Z71.8 Other specified counseling

New Code Z71.84 Travel counseling

Add Counseling for international travel

Vertigo of Central Origin

Central vertigo by definition is vertigo due to a disease originating with the central nervous system (CNS). It may be caused by CNS tumors, infection, trauma and cerebellar hemorrhage. Often it is unknown what specific CNS structure is causing the vertigo. Central vertigo of the left, right, bilateral or unspecified ear codes under subcategory H81.4, Vertigo of central origin is not clinically valid. It would not be appropriate to report laterality for vertigo of central origin.

Intelligent Medical Objects, Inc. (IMO) is requesting the following tabular modifications. The American Academy of Neurology supports this request.

TABULAR MODIFICATIONS

H81 Disorders of vestibular function

H81.4 Vertigo of central origin

Delete	H81.41	Vertigo of central origin, right ear
Delete	H81.42	Vertigo of central origin, left ear
Delete	H81.43	Vertigo of central origin, bilateral
Delete	H81.49	Vertigo of central origin, unspecified ear

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All proposed effective October 1, 2019

- A18 Tuberculosis of other organs
A18.4 Tuberculosis of skin and subcutaneous tissue
Excludes2:lupus erythematosus (L93.-)
Delete ~~lupus NOS (M32.9)~~
Revise systemic lupus erythematosus (M32.-)
- B97 Viral agents as the cause of diseases classified elsewhere
B97.4 Respiratory syncytial virus as the cause of diseases classified elsewhere
Add RSV as the cause of diseases classified elsewhere
- D21 Other benign neoplasms of connective and other soft tissue
D21.6 Benign neoplasm of connective and other soft tissue of trunk, unspecified
Revise Benign neoplasm of connective and other soft tissue back, NOS
- D36 Benign neoplasm of other and unspecified sites
D36.7 Benign neoplasm of other specified sites
Add Benign neoplasm of back NOS
- I21 Acute myocardial infarction
I21.A Other type of myocardial infarction
I21.A9 Other myocardial infarction type
Code also complication, if known and applicable, such as:
Revise (acute) stent stenosis (T82.8575-)
- I50 Heart failure
I50.9 Heart failure, unspecified
Revise Excludes2: fluid overload unrelated to congestive heart failure (E87.70)
- J12 Viral pneumonia, not elsewhere classified
J12.1 Respiratory syncytial virus pneumonia
Add RSV pneumonia
- J20 Acute bronchitis
J20.5 Acute bronchitis due to respiratory syncytial virus
Add Acute bronchitis due to RSV
- J21 Acute bronchiolitis
J21.0 Acute bronchiolitis due to respiratory syncytial virus

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- Add Acute bronchiolitis due to RSV
- J44 Other chronic obstructive pulmonary disease
- Revise J44.0 Chronic obstructive pulmonary disease with (acute) lower respiratory infection
- K59.0 Constipation
- Delete Use ~~additional code for adverse effect, if applicable, to identify drug (T36-T50 with fifth or sixth character 5)~~
- L50 Urticaria
- Revise Excludes1: urticaria pigmentosa (~~Q82.2~~) (D47.01)
- M24 Other specific joint derangements
- M24.2 Disorder of ligament
- Revise Excludes2: internal derangement of knee (~~M23.5-M23.89~~ M23.8X9)
- M34 Systemic sclerosis [scleroderma]
- Revise Excludes1: neonatal scleroderma (~~P83.8~~) (P83.88)
- M47 Spondylosis
- M47.1 Other spondylosis with myelopathy
- Revise Excludes1: vertebral subluxation (~~M43.3-M43.59~~ M43.5X9)
- O36 Maternal care for other fetal problems
- O36.8 Maternal care for other specified fetal problems
- O36.83 Maternal care for abnormalities of the fetal heart rate or rhythm
- Add Maternal care for depressed fetal heart rate tones
- Add Maternal care for fetal bradycardia
- Add Maternal care for fetal heart rate decelerations
- Add Maternal care for fetal heart rate irregularity
- Add Maternal care for fetal heart rate abnormal variability
- Add Maternal care for fetal tachycardia
- Add Maternal care for non-reassuring fetal heart rate or rhythm
- O99 Other maternal diseases classifiable elsewhere but complicating pregnancy, childbirth and the puerperium
- O99.3 Mental disorders and diseases of the nervous system complicating pregnancy, childbirth and the puerperium

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- 099.34 Other mental disorders complicating pregnancy, childbirth,
and the puerperium
Revise Conditions in F01-F09, ~~and~~ F20-F99 and F54-F99
- 099.8 Other specified diseases and conditions complicating pregnancy, childbirth and
the puerperium
Revise Excludes2: infection of genitourinary tract following delivery (O86.1-
~~O86.3~~ O86.4)
- P91 Other disturbances of cerebral status of newborn
P91.8 Other specified disturbances of cerebral status of newborn
P91.811 Neonatal encephalopathy in diseases classified elsewhere
Code first underlying condition, if known, such as:
Revise congenital cirrhosis (of liver) (~~P78.71~~) (P78.81)
- R63 Symptoms and signs concerning food and fluid intake
Delete Excludes1: ~~eating disorders of nonorganic origin (F50.)~~
Delete ~~malnutrition (E40-E46)~~
- T27 Burn and corrosion of respiratory tract
T27.5 Corrosion involving larynx and trachea with lung
Add Code first (T51-T65) to identify chemical and intent
- T28 Burn and corrosion of other internal organs
T28.4 Burns of other and unspecified internal organs
Delete Code first (T51-T65) to identify chemical and intent
- T79 Certain early complications of trauma, not elsewhere classified
T79.A Traumatic compartment syndrome
Revise Excludes2: traumatic ischemic infarction of muscle (T79.6)
- V86 Occupant of special all-terrain or other off-road motor vehicle, injured in transport
accident
V86.0 Driver of special all-terrain or other off-road motor vehicle injured in traffic
accident
V86.09 Driver of other special all-terrain or other off-road motor vehicle injured
in traffic accident
Delete ~~Driver of dirt bike injured in traffic accident~~
V86.1 Passenger of special all-terrain or other off-road motor vehicle injured in traffic
accident

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- V86.19 Passenger of other special all-terrain or other off-road motor vehicle injured in traffic accident
Delete ~~Passenger of dirt bike injured in traffic accident~~
- V86.2 Person on outside of special all-terrain or other off-road motor vehicle injured in traffic accident
V86.29 Person on outside of other special all-terrain or other off-road motor vehicle injured in traffic accident
Delete ~~Person on outside of dirt bike injured in traffic accident~~
- V86.3 Unspecified occupant of special all-terrain or other off-road motor vehicle injured in traffic accident
V86.39 Unspecified occupant of other special all-terrain or other off-road motor vehicle injured in traffic accident
Delete ~~Unspecified occupant of dirt bike injured in traffic accident~~
- V86.4 Person injured while boarding or alighting from special all-terrain or other off-road motor vehicle
V86.49 Person injured while boarding or alighting from other special all-terrain or other off-road motor vehicle
Delete ~~Person injured while boarding or alighting from dirt bike~~
- V86.5 Driver of special all-terrain or other off-road motor vehicle injured in nontraffic accident
V86.59 Driver of other special all-terrain or other off-road motor vehicle injured in nontraffic accident
Delete ~~Driver of dirt bike injured in nontraffic accident~~
- W25 Contact with sharp glass
Revise Excludes1: fall on same level due to slipping, tripping and stumbling with subsequent striking against sharp glass (~~W01.10~~ W01.110)
- Z45 Encounter for adjustment and management of implanted device
Z45.0 Encounter for adjustment and management of cardiac device
Z45.01 Encounter for adjustment and management of cardiac pacemaker
Z45.018 Encounter for adjustment and management of other part of cardiac pacemaker
Add Excludes1: presence of other part of cardiac pacemaker (Z95.0)
Revise Excludes2: presence of prosthetic and other devices (~~Z95-Z97~~) Z95.1-Z95.5, Z95.811-Z97
- Z69 Encounter for mental health services for victim and perpetrator of abuse

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	Z69.8	Encounter for mental health services for victim or perpetrator of other abuse
	Z69.81	Encounter for mental health services for victim of other abuse
Delete		Encounter for mental health services for perpetrator of non-spousal adult abuse
	Z69.82	Encounter for mental health services for perpetrator of other abuse
Add		Encounter for mental health services for perpetrator of non-spousal adult abuse
	Z90	Acquired absence of organs, not elsewhere classified
	Z90.4	Acquired absence of other specified parts of digestive tract
	Z90.41	Acquired absence of pancreas
		Use additional code to identify any associated:
Revise		insulin use (Z79.4) diabetes mellitus, postpancreatectomy (E13.-)
Revise		diabetes mellitus, postpancreatectomy (E13.-) <u>insulin use (Z79.4)</u>
	Z97	Presence of other devices
Delete		Excludes1: fitting and adjustment of prosthetic and other devices (Z44-Z46)
Add		Excludes2: fitting and adjustment of prosthetic and other devices (Z44-Z46)

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All proposed effective October 1, 2019

Abscess

- Revise - gingival - see ~~Peridontitis~~ Periodontitis, aggressive, localized
- Revise - gum - see ~~Peridontitis~~ Periodontitis, aggressive, localized
- Revise - parodontal - see ~~Peridontitis~~ Periodontitis, aggressive, localized
- Revise - pericemental - see ~~Peridontitis~~ Periodontitis, aggressive, localized
- Revise - pericoronal - see ~~Peridontitis~~ Periodontitis, aggressive, localized
- Revise - peridental - see ~~Peridontitis~~ Periodontitis, aggressive, localized
- Revise - periodontal (parietal) - see ~~Peridontitis~~ Periodontitis, aggressive, localized
- tooth, teeth (root) K04.7
- Revise - - supporting structures NEC - see ~~Peridontitis~~ Periodontitis, aggressive, localized

Abuse

- Revise - amphetamine (or related substance) - see also Abuse, drug, stimulant NEC
- Add - - stimulant NEC F15.10
- Add - - - with
- Add - - - - anxiety disorder F15.180
- Add - - - - intoxication F15.129
- Add - - - - - with
- Add - - - - - delirium F15.121
- Add - - - - - perceptual disturbance F15.122

Revise Acrocyanosis ~~I73.8~~ I73.89

Adhesions, adhesive

Revise - peritoneum, peritoneal (postinfective) K66.0

Anastomosis

- aneurysmal - see Aneurysm
- arteriovenous ruptured brain I60.8
- Add - - intracerebral I61.8
- Add - - intraparenchymal I61.8
- Add - - intraventricular I61.5
- Add - - subarachnoid I60.8

Aneurysm

- arteriovenous (congenital) - see also Malformation, arteriovenous
- - acquired I77.0
- - - brain I67.1
- Add - - - - ruptured - see Aneurysm, arteriovenous, brain, ruptured
- - - brain Q28.2
- - - ruptured I60.8
- Add - - - - intracerebral I61.8
- Add - - - - intraparenchymal I61.8

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Add - - - - intraventricular I61.5

Add - - - - subarachnoid I60.8

- brain I67.1

- - arteriovenous (congenital) (nonruptured) Q28.2

- - - acquired I67.1

Revise - - - - ruptured I60.8 - see Aneurysm, arteriovenous, brain, ruptured

Revise - - - - ruptured I60.8 - see Aneurysm, arteriovenous, brain, ruptured

- retina - see also Disorder, retina, microaneurysms

Revise - - diabetic - ~~see Diabetes, microaneurysms, retinal~~ see E08-E13 with .3-

Atrophy, atrophic (of)

- spinal (acute) (cord) G95.89

- - paralysis

Revise - - - - meaning progressive muscular atrophy ~~G12.21~~ G12.25

Brill(-Zinsser) disease (recrudescence typhus) A75.1

Delete ~~flea-borne~~ A75.2

Delete ~~louse-borne~~ A75.4

Burn

Revise - partial thickness - code as Burn, ~~unspecified~~ second degree, by site

Calculus, calculi, calculous

- pyelitis (impacted) (recurrent)

Revise - - with hydronephrosis ~~N13.2~~ N13.6

- pyelonephritis (impacted) (recurrent)

Revise - - with hydronephrosis ~~N13.2~~ N13.6

Cardiomyopathy

- amyloid E85.4, [I43]

Add - - transthyretin-related (ATTR) familial E85.4 [I43]

Add - specified NEC I42.8

Add - non-ischemic (see also by cause) I42.8

Add Cecoureterocele Q62.32

Chiari's

- disease or syndrome (hepatic vein thrombosis) I82.0

Revise - - type II - ~~see Spina bifida~~ Q07.0-Need to verify

Delirium

- due to (secondary to)

Revise - - unknown etiology ~~F05~~ R41.0

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Disease

- Revise - Rossbach's (hyperchlorhydria) ~~K30~~ K31.89
- Add - - psychogenic F45.8
- Revise - systemic tissue mast cell ~~E96.20~~ D47.02

Double

- uterus Q51.2
- Revise - - in pregnancy or childbirth ~~O34.59~~ O34.0-

Failure

- heart (acute) (senile) (sudden) I50.9
- - with
- Add - - - hypertension See Hypertension, heart

Findings

- radiologic (X-ray) R93.8
- Add - - musculoskeletal
- Add - - limbs R93.6
- Add - - - other than limb R93.7

Fistula (cutaneous) L98.8

- arteriovenous (acquired) (nonruptured) I77.0
- - brain I67.1
- - - congenital Q28.2
- Revise - - - - ruptured ~~I60.8~~ - see Fistula, arteriovenous, brain, ruptured
- - - - ruptured I60.8
- Add - - - - intracerebral I61.8
- Add - - - - intraparenchymal I61.8
- Add - - - - intraventricular I61.5
- Add - - - - subarachnoid I60.8
- - congenital (peripheral) - see also Malformation, arteriovenous
- - - brain Q28.2
- Revise - - - - ruptured ~~I60.8~~ - see Fistula, arteriovenous, brain, ruptured
- brain G93.89
- Revise - - arteriovenous (acquired) (see also Fistula, arteriovenous, brain) I67.1
- - - congenital Q28.2

Fracture, traumatic

- finger (except thumb) S62.60-
- - index S62.60-
- Revise - - - ~~medial~~ middle phalanx (displaced) S62.62-
- - little S62.60-
- Revise - - - ~~medial~~ middle phalanx (displaced) S62.62-
- Revise - - ~~medial~~ middle phalanx (displaced) S62.62-
- - middle S62.60-

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Revise - - - ~~medial~~ middle phalanx (displaced) S62.62-
- - ring S62.60-

Revise - - - ~~medial~~ middle phalanx (displaced) S62.62-
- toe S92.91-
- - lesser (displaced) S92.50-

Revise - - - ~~medial~~ middle phalanx (displaced) S92.52-

Revise Griesinger's disease ~~B76.9~~ B76.0

Revise Hookworm (~~anemia~~) (disease) (infection) (infestation) B76.9

Add - with anemia B76.9 [D63.8]

Hyperplasia

- breast - see also Hypertrophy, breast

Add - - atypical, atypia N60.9-

Revise - - ductal (~~atypical~~) N60.9-

Add - - lobular N60.9-

Revise - endometrium, endometrial (adenomatous) (~~benign~~) (cystic) (glandular) (glandular-cystic)
(polypoid) N85.00

Add --benign N85.01

Impingement

Add - joint- see-Disorder, joint, specified type NEC

Infection

- periodental, periodontal K05.20

Revise - - generalized - see ~~Peridontitis~~ Periodontitis, aggressive, generalized

Revise - - localized - see ~~Peridontitis~~ Periodontitis, aggressive, localized

Revise - ureter ~~N28.86~~ (see Ureteritis)

Insufficiency

- pulmonary J98.4

Revise - - newborn ~~P28.5~~ P28.8

Intoxication

Revise - amphetamine (without dependence) - see also Abuse, drug, stimulant, with intoxication

Revise - - with dependence - see Dependence, drug, stimulant, with intoxication

Add - - stimulant NEC F15.10

Add - - - with

Add - - - - anxiety disorder F15.180

Add - - - - intoxication F15.129

Add - - - - - with

Add - - - - - delirium F15.121

Add - - - - - perceptual disturbance F15.122

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Malformation

- arteriovenous, aneurysmatic (congenital) Q27.30
- - brain Q28.2
- Add - - - ruptured I60.8
- Add - - - - intracerebral I61.8
- Add - - - - intraparenchymal I61.8
- Add - - - - intraventricular I61.5
- Add - - - - subarachnoid I60.8
- Revise - - cerebral (see also Malformation, arteriovenous, brain) Q28.2

- Chiari

- Revise - - Type II ~~Q07.01~~ Q07.0

Narrowing

- artery
- Revise - - choroidal - see Occlusion, artery, ~~cerebral precerebral~~, specified NEC
- Revise - - communicating posterior - see Occlusion, artery, ~~cerebral precerebral~~, specified NEC
- Revise - - hypophyseal - see Occlusion, artery, ~~cerebral precerebral~~, specified NEC
- Revise - - pontine - see Occlusion, artery, ~~cerebral precerebral~~, specified NEC

Osteomyelitis

- acute M86.10
- Revise - - ilium ~~M86.159~~ M86.18
- Revise - - ischium ~~M86.159~~ M86.18

Paralysis, paralytic

- muscle, muscular NEC G72.89
- progressive G12.21
- Add - - progressive, spinal G12.25
- Add - - spinal progressive G12.25
- spinal (cord) G83.9
- - progressive G12.21
- Add - - - muscle G12.25

Pericementitis (chronic) (suppurative) - see also Periodontitis

- Revise - - generalized - see ~~Peridontitis~~ Periodontitis, aggressive, generalized
- Revise - - localized - see ~~Peridontitis~~ Periodontitis, aggressive, localized

- Revise Petit mal seizure - see Epilepsy, ~~generalized, specified NEC~~ childhood, absence

Poisoning

- Revise - food (~~acute~~) (~~diseased~~) (~~infected~~) (~~noxious~~) NEC ~~T62.9~~ A05.9

Polyneuropathy

- amyloid (Portuguese) E85.1 [G63]
- Add - - transthyretin-related (ATTR) familial E85.1 [G63]

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Pregnancy

- complicated by (care of) (management affected by)
- - fetal (maternal care for)

Revise - - - heart rate irregularity (bradycardia) (decelerations) (tachycardia) ~~Ø76~~ O36.83-

Prolonged

Revise -QT interval ~~I45.81~~ R94.31

Pyelitis

- with
- - calculus

Revise - - - with hydronephrosis ~~N13.2~~ N13.6

- chronic
- - with calculus

Revise - - - with hydronephrosis ~~N13.2~~ N13.6

Pyelonephritis

- with calculus

Revise - - with hydronephrosis ~~N13.2~~ N13.6

- calculus

Revise - - with hydronephrosis ~~N13.2~~ N13.6

- chronic
- - with calculus

Revise - - - with hydronephrosis ~~N13.2~~ N13.6

Rheumatic (acute) (subacute) (chronic)

Revise - coronary arteritis ~~I01.9~~ I01.8

Add - chronic I09.89

Rupture, ruptured

Revise - arteriovenous fistula, brain ~~I60.8~~ - see Fistula, arteriovenous, brain, ruptured

Separation

Add -muscle (nontraumatic) – see Diastasis, muscle

Sclerosis

- spinal (cord) (progressive) G95.89

Revise - - lateral (amyotrophic) ~~G12.23~~ G12.21

Add - - - progressive G12.23

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Screening (for)

- Revise - developmental handicap Z13.4~~2~~
- Revise - - in early childhood Z13.4~~2~~
- Revise - - infant Z13.4~~4~~
- Revise - disability, intellectual Z13.4~~2~~
- Revise - - infant Z13.4~~4~~
 - disease or disorder Z13.9
- Revise - - developmental Z13.4~~2~~
- Revise - - - in child Z13.4~~2~~
- Revise - - - infant Z13.4~~4~~
- Revise -intellectual disability Z13.4~~2~~
- Revise - - infant Z13.4~~4~~

Suppuration

- gum K05.20
- Revise - - generalized - see Peridontitis Periodontitis, aggressive, generalized
- Revise - - localized - see Peridontitis Periodontitis, aggressive, localized

Seizure(s)

- Add - absence G40.A-
- Revise - petit mal ~~G40.409~~ G40.A-

Syndrome

- Add - Churg-Strauss
 - right
- Revise - - ventricular obstruction – see Failure, heart, ~~congestive~~, right
- Add - Glass Q87.89
- Revise - premenstrual dysphoric ~~F32.89~~ F32.81
- Add - SATB2-associated Q87.89

Thrombosis

- mesenteric (artery) (with gangrene) (see also Infarct, intestine) K55.069
- Revise - - vein (inferior) (superior) ~~I84~~ K55.0

Unstable

- Revise - lumbosacral joint (congenital) see subcategory M53.2
- Delete —acquired— see subcategory M53.2

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Varix

-with

Add --bleeding I83.899

Add --ruptured I83.899

Weak, weakening, weakness (generalized) R53.1

Revise - foot (double) – see also Weak, arches

Revise Zellweger's syndrome ~~Q87.89~~ E71.510

ICD-10-CM Table of Drugs and Chemicals

Revise Clozapine ~~T42.4X1~~ T43.501 ~~T42.4X2~~ T43.502 ~~T42.4X3~~ T42.503 ~~T42.4X4~~ T42.504 ~~T42.4X5~~ T42.505 ~~T42.4X6~~ T42.506