ICD-10 Coordination and Maintenance Committee Meeting
September 9-10, 2025
Diagnosis Agenda

Register in advance for this webinar:
https://cms.zoomgov.com/webinar/register/WN_cfwiQyYuR4uyU1x0HZKCKA
After registering, you will receive a confirmation email containing information about joining the webinar.

Welcome and announcements
Captain Monica Leonard
Co-Chair, ICD-10 Coordination and Maintenance Committee

Diagnosis Topics:

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September 9, 2025, ICD-10 C & M Diagnosis Timed Agenda
Order of Presentation (Day 1)

***Please dial in 45 mins before the topic of interest in the event ahead of schedule, presentation times subject to change**

Topic*	Presenter
Opening Remarks, CMS	Mady Hue
Welcome, CDC, Diagnosis Day 1 **Time: 9:05-910amET**	Monica Leonard
Topic(s): Nipple Ischemia and Nipple Necrosis **Time: 9:10 – 9:25amET**	Traci Ramirez
Topic: Personal History of Clostridioides Difficile Infection **Time: 9:25-9:40amET**	Traci Ramirez
Topic: Vanishing Twin Syndrome **Time: 9:40-9:55amET**	Traci Ramirez
Topic: Toxic Stress **Time: 9:55-10:10amET**	Traci Ramirez Sarah C DeSilvey
Topic: Low Body Mass Index **Time: 10:10-10:25amET**	David Berglund Dr. Patrick Romano
Topic: Alzheimer's Biomarker **Time: 10:25-10:40amET**	Cheryl Bullock Dr. Katherine Coerver
Topic: Pneumothorax that occurs after CPR **Time: 10:40-10:55amET**	Cheryl Bullock
Topic: Apragmatism **Time: 10:55-11:10amET**	Traci Ramirez Dr. Jamila Minga
Topic: Ectopic Pregnancy **Time: 11:10-11:25amET**	Traci Ramirez

Topic: Pediatric Pyloric Stenosis **Time: 11:25-11:40amET**	Desiree Abrams
Topic: Catatonia **Time: 11:40-11:55amET**	Cheryl Bullock
Topic: Sleep Inertia **Time: 11:55-12:10pmET**	Cheryl Bullock
Topic: Potts Puffy Tumor **Time: 12:10-12:25pmET**	Shannon McConnell-Lamptey Dr. Matthew J Molloy
LUNCH BREAK	12:30-1:30
Topic: Risk of Malignancy Status **Time: 1:30-1:45pmET**	Shannon McConnell-Lamptey Dr. Jonathan Rubenstein
Topic: Blast Overpressure Exposure **Time: 1:45-2:00pmET**	Traci Ramirez Dr. Jessica Bock
Topic: Gadolinium Induced Gout **Time: 2:00-2:15pmET**	Cheryl Bullock Dr. Regina Sutton
Topic(s): Glanzmann thrombasthenia **Postprocedural Open Deep Wound** **Time: 2:15-2:30pmET**	Shannon McConnell-Lamptey Dr. Michael Recht Shannon McConnell-Lamptey
Topic: Screening of Diabetes Mellitus **Time: 2:30-2:45pmET**	Shannon Mconnell-Lamptey Dr. Brigitte Frohnert
Topic: Pancreatic Contusion **Time: 2:45-3:00pmET**	Traci Ramirez Dr. Bradley Dennis

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Topic: Familial-genetic Dilated Cardiomyopathy **Time: 3:00-3:15pmET**	Dr. David Berglund Dr. Jack Lawrence
Topic: Medetomidine Withdrawal Syndrome **Time: 3:15-3:30pmET**	Shannon McConnell-Lamptey Dr. Daniel Teixeira da Silva
Topic: Hepatic Fibrosis **Time: 3:30-3:45pmET**	Shannon McConnell-Lamptey Dr. Shaun Chandna
Topic: Okur-Chung Neurodevelopmental Syndrome (OCNDS) **Time: 3:45-4:00pmET**	Dr. David Berglund Dr. Wendy Chung
Topic: Lynch Syndrome **Time: 4:00-4:15pmET**	Dr. David Berglund Dr. Peter Stanich
Topic: Obesity Due to Disruption of MC4R Pathway **Time: 4:15-4:30pmET**	Dr. David Berglund Dr. Ali Mohamadi
Topic: Koolen-de Vries Syndrome **Time: 4:30-4:45pmET**	Dr. David Berglund Dr. Heather Mefford
Topic: GRI Neurodevelopmental Disorders **Time: 4:45-5:00pmET**	Dr. David Berglund Dr. Tim Benke

ICD-10 Coordination and Maintenance Committee Meeting
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September 9, 2025, ICD-10 C & M Diagnosis Timed Agenda
Order of Presentation (Day 2)

***Please dial in 45 mins before the topic of interest in the event ahead of schedule, presentation times subject to change**

Topic*	Presenter
Welcome, Diagnosis Day 2 **Time: 9:00-9:10amET**	Monica Leonard
Topic: Hypertriglyceridemia **Time: 9:10-9:25amET**	Shannon McConnell-Lamptey Dr. Leticia Ferri
Topic: Skin Changes Due to Skin Failure **Time: 9:25-9:40amET**	Shannon McConnell-Lamptey Dr. Diane Krasner
Topic: Doxy PEP **Time: 9:40-9:55amET**	Shannon McConnell-Lamptey Dr. Laura Quilter
Topic: Lipedema and Lipolymphedema **Time: 9:55-10:10amET**	Shannon McConnell-Lamptey Dr. Karen Herbst
Topic: Secondary Neoplasm of the Oral Cavity, Larynx and Pharynx **Time: 10:10-10:25amET**	Traci Ramirez Jennifer LaBay
Topic: Vitiligo **Time: 10:25-10:40amET**	Shannon McConnell-Lamptey Vineeth R. Vaidyula
Topic: Estrogen Receptor Low Status (ER+/1-10% expression) **Time: 10:40-10:55amET**	Traci Ramirez Dr. Eleanor Faherty
Topic: Pediatric Healthcare: Impact of Parental Mental Health **Time: 10:55-11:10amET**	Cheryl Bullock Dr. Katherine Minaya
Topic: Pediatric Healthcare: Impact of Social Circumstances **Time: 11:10-11:25amET**	Cheryl Bullock Dr. Katherine Minaya

Topic: Pediatric Healthcare: Screening for and Preventing Child Maltreatment **Time: 11:25-11:40amET**	Cheryl Bullock Dr. Katherine Minaya
Topic: Loeys-Dietz Syndrome **Time: 11:40 -11:55amET**	Cheryl Bullock Dr. Hal Dietz
Topic: Gender Identity Disorder, in Remission (desistance) **Time: 11:55-12:10pmET**	Dr. Kurt Miceli
Topic: Vexas Syndrome **Time: 12:10-12:25pmET**	Dr. David Berglund Dr. Peter Grayson
LUNCH BREAK	12:30-1:30
Topic: Chronic Hand Eczema **Time: 1:30-1:45pmET**	Shannon McConnell-Lamptey Dr. Raj Chovatiya
Topic: Amyloid-related imaging abnormalities (ARIA) **Time: 1:45-2:00pmET**	Dr. David Berglund Dr. Danielle Abraham
Topic: Topical Steroid Withdrawal **Time: 2:00-2:15pmET**	Shannon McConnell-Lamptey
Topic: Post Bariatric Hypoglycemia **Time: 2:15-2:30pmET**	Shannon McConnell-Lamptey Dr. Colleen Craig
Topic: METALD **Time: 2:30-2:45pmET**	Shannon McConnell-Lamptey Dr. Jeff Swanson
Topic: Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) **Time: 2:45-3:00pmET**	Dr. David Berglund Dr. Andrew Landstrom
Topic: Arrhythmogenic Cardiomyopathy (ACM) **Time: 3:00-3:15pmET**	Dr. David Berglund Brittney Murray

Topic: Brugada Syndrome **Time: 3:15-3:30pmET**	Dr. David Berglund Dr. Dan Roden
Topic: Alkene Exposure **Time: 3:30-3:45pmET**	Dr. David Berglund Laree LaPierre
Topic: Cycloparaffin Exposure **Time: 3:45-4:00pmET**	Dr. David Berglund Laree LaPierre
Topic: Hexamethylene Diisocyanate Exposure **Time: 4:00-4:15pmET**	Dr. David Berglund Laree LaPierre
Topic(s): Burn Pit Exposure **Agent Orange** **Time: 4:15-4:30pmET**	Desiree Abrams Laree LaPierre Desiree Abrams Laree LaPierre
Topic(s): Odontogenic Sinusitis **Ledderhose Disease/Plantar Fibromatosis and Plantar Fasciitis** **Time: 4:30-4:45pmET**	Shannon McConnell-Lamptey Shannon McConnell-Lamptey
Topic(s): Ventricular Bigeminy **Pulmonary Mycetoma** **Prevesical Abscess** **Time: 4:45-5:00pmET**	Dr. David Berglund Dr. David Berglund Dr. David Berglund

ICD-10 TIMELINE

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

September 9-10, 2025	The diagnosis code portion of the September 2025 ICD-10 Coordination and Maintenance Committee Meeting is anticipated to be fully virtual by zoom and dial-in. Those who wish to attend must participate via Zoom Webinar or by dialing in. The procedure code topics will be open for public comment.
September 2025	Recordings and slide presentations of the September 9-10, 2025 ICD-10 Coordination and Maintenance Committee Meeting will be posted on the following web pages: **Diagnosis code portion of the recording and related materials–** https://www.cdc.gov/nchs/icd/icd-10-maintenance/meetings.html **Procedure code portion of the recording and related materials–** https://www.cms.gov/medicare/coding-billing/icd-10-codes/icd-10-coordination-maintenance-committee-materials
October 1, 2025	New and revised ICD-10-CM and ICD-10-PCS codes go into effect along with MS-DRG changes. Final addenda available on web pages as follows: **Diagnosis addendum –** https://www.cdc.gov/nchs/icd/icd-10-cm/files.html **Procedure addendum –** https://www.cms.gov/medicare/coding-billing/icd-10-codes
October 10, 2025	**Deadline for receipt of public comments on proposed new codes and revisions discussed at the September 9-10, 2025 ICD-10 Coordination and Maintenance Meeting being considered for implementation on April 1, 2026.**
November 2025	Any new ICD-10 codes that will be implemented on the following April 1 will be announced. Information on any new codes to be implemented April 1, 2026 will be posted on the following websites: https://www.cdc.gov/nchs/icd/icd-10-cm/files.html https://www.cms.gov/medicare/coding-billing/icd-10-codes

November 14, 2025	**Deadline for receipt of public comments on proposed new codes and revisions discussed at the September 9-10, 2025 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2026.**
December 5, 2025	**Deadline for requestors: Those members of the public requesting that topics be discussed at the March 17-18, 2026 ICD-10 Coordination and Maintenance Committee Meeting must have their requests submitted to CMS for procedures and to NCHS for diagnoses by this date.** **Procedure code requests should be directed to CMS at:** https://mearis.cms.gov. **Diagnosis code requests should be directed to NCHS at:** nchsicd10cm@cdc.gov. **Please be advised that new guidance on proposal submissions for diagnosis-related topics will be available by mid-October 2025 at: https://www.cdc.gov/nchs/icd/icd-10-maintenance/proposals.html.** Requestors should indicate if they are submitting their code request for consideration for an April 1, 2027 implementation date, or an October 1, 2027 implementation date. The ICD-10 Coordination and Maintenance Committee will make efforts to accommodate the requested implementation date for each request submitted, however, the Committee will determine which requests will be presented for consideration for an April 1, 2027 implementation date or an October 1, 2027 implementation date.
January 2026	Federal Register notice for the March 17-18, 2026 ICD-10 Coordination and Maintenance Committee Meeting will be published. This will include the tentative agenda.
February 2026	Tentative agenda for the Procedure portion of the March 17, 2026 ICD-10 Coordination and Maintenance Committee Meeting posted on CMS webpage at: https://www.cms.gov/medicare/coding-billing/icd-10-codes/icd-10-coordination-maintenance-committee-materials Tentative agenda for the Diagnosis portion of the March 18, 2026, ICD-10 Coordination and Maintenance Committee Meeting posted on NCHS homepage at: https://www.cdc.gov/nchs/icd/icd-10-maintenance/meetings.html

February 1, 2026	ICD-10 MS-DRG Grouper software and related materials posted on CMS webpage at: https://www.cms.gov/medicare/payment/prospective-payment-systems/acute-inpatient-pps/ms-drg-classifications-and-software Any updates to the ICD-10-CM and ICD-10-PCS Coding Guidelines will be posted on the following websites: https://www.cdc.gov/nchs/icd/icd-10-cm/files.html https://www.cms.gov/medicare/coding-billing/icd-10-codes All ICD-10-CM and ICD-10-PCS code update files (includes April 1 update and full files from prior October 1) will be posted on the following websites: https://www.cdc.gov/nchs/icd/icd-10-cm/files.html https://www.cms.gov/medicare/coding-billing/icd-10-codes
March 17-18, 2026	The ICD-10 Coordination and Maintenance Committee Meeting is anticipated to be fully virtual by zoom and dial-in. Those who wish to attend must participate via Zoom Webinar or by dialing in.
March 2026	Recordings and slide presentations of the March 17-18, 2026 ICD-10 Coordination and Maintenance Committee Meeting will be posted on the following web pages: **Diagnosis code portion of the recording and related materials–** https://www.cdc.gov/nchs/icd/icd-10-maintenance/meetings.html **Procedure code portion of the recording and related materials–** https://www.cms.gov/medicare/coding-billing/icd-10-codes/icd-10-coordination-maintenance-committee-materials
April 1, 2026	Any new or revised ICD-10 codes will be implemented on April 1, 2026.
April 17, 2026	**Deadline for receipt of public comments on proposed new codes and revisions discussed at the March 17-18, 2026, ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on April 1, 2027**.
April 2026	Notice of Proposed Rulemaking to be published in the Federal Register as mandated by the Omnibus Budget Reconciliation Act of 1986, Public Law 99-509 (Pub. L. 99-509). This notice will include references to the FY 2027 ICD-10-CM diagnosis and ICD-10-PCS procedure codes finalized 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: https://www.cms.gov/medicare/payment/prospective-payment-systems/acute-inpatient-pps
May 18, 2026	**Deadline for receipt of public comments on proposed new diagnosis codes and revisions discussed at the March 17-18, 2026, ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2027**.
May/June 2026	Final addenda posted on web pages as follows: **Diagnosis addendum -** https://www.cdc.gov/nchs/icd/icd-10-cm/files.html **Procedure addendum -** https://www.cms.gov/medicare/coding-billing/icd-10-codes
June 5, 2026	**Deadline for requestors: Those members of the public requesting that topics be discussed at the September 15-16, 2026, ICD-10 Coordination and Maintenance Committee Meeting must have their requests submitted to CMS for procedures and NCHS for diagnoses.** **Procedure code requests should be directed to CMS at:** https://mearis.cms.gov. **Diagnosis code requests should be directed to NCHS at:** nchsicd10cm@cdc.gov Requestors should indicate if they are submitting their code request for consideration for an April 1, 2027, implementation date or an October 1, 2027, implementation date. The ICD-10 Coordination and Maintenance Committee will make efforts to accommodate the requested implementation date for each request submitted, however, the Committee will determine which requests will be presented for consideration for an April 1, 2027, implementation date or an October 1, 2027, implementation date.
July 2026	Federal Register notice for the September 15-16, 2026, ICD-10 Coordination and Maintenance Committee Meeting will be published. This will include the tentative agenda.
August 1, 2026	Hospital Inpatient Prospective Payment System final rule expected to be published in the Federal Register as mandated by Pub. L. 99-509. This rule will also include links to all the final codes to be implemented on October 1, 2026.

	This rule can be accessed at: https://www.cms.gov/medicare/payment/prospective-payment-systems/acute-inpatient-pps
August 2026	Tentative agenda for the Procedure portion of the September 15, 2026, ICD-10 Coordination and Maintenance Committee Meeting will be posted on the CMS webpage at – https://www.cms.gov/medicare/coding-billing/icd-10-codes/icd-10-coordination-maintenance-committee-materials Tentative agenda for the Diagnosis portion of the September 16, 2026, ICD-10 Coordination and Maintenance Committee Meeting will be posted on the NCHS webpage at: https://www.cdc.gov/nchs/icd/icd-10-maintenance/meetings.html
September 15-16, 2026	The September 2026 ICD-10 Coordination and Maintenance Committee Meeting is anticipated to be fully virtual by zoom and dial-in. Those who wish to attend must participate via Zoom Webinar or by dialing in.
September 2026	Recordings and slide presentations of the September 15-16, 2026, ICD-10 Coordination and Maintenance Committee Meeting will be posted on the following web pages: **Diagnosis code portion of the recording and related materials–** https://www.cdc.gov/nchs/icd/icd-10-maintenance/meetings.html **Procedure code portion of the recording and related materials–** https://www.cms.gov/medicare/coding-billing/icd-10-codes/icd-10-coordination-maintenance-committee-materials
October 1, 2026	New and revised ICD-10-CM and ICD-10-PCS codes go into effect along with MS-DRG changes. Final addenda available on web pages as follows: **Diagnosis addendum -** https://www.cdc.gov/nchs/icd/icd-10-cm/files.html **Procedure addendum -** https://www.cms.gov/medicare/coding-billing/icd-10-codes

Contact Information

ICD-10 Coordination and Maintenance Committee Meeting
September 9-10, 2025

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

Captain Monica Leonard	(404) 718-6443
Desiree Abrams	(301) 458-4384
David Berglund, MD	(301) 458-4095
Cheryl Bullock	(301) 458-4297
Shannon McConnell-Lamptey	(301) 458-4612
Traci Ramirez	(301) 458-4454

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 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.

Alkene Exposure

Alkenes (or olefins) are important industrial chemicals used in high volumes as petrochemicals as well as in the manufacturing of plastics and synthetic rubbers.[1,2] They are a significant constituent of jet propellent fuel 5 (JP-5).

Exposure to alkenes can occur via ingestion, inhalation, or skin contact. Symptoms of acute exposure can include central nervous system symptoms (headache, fatigue, dizziness, concentration problems), gastrointestinal symptoms (nausea, vomiting, abdominal pain), and dermatologic issues (skin rashes, irritation). Exposure to certain alkenes, such as butadiene, can cause lymphohematopoietic cancers in occupationally exposed workers.[1,3]

Documentation of toxic substances and exposures during or associated with military service is a high priority for both the Department of Defense (DOD) and the Department of Veteran Affairs (VA) due to their potential long term health consequences and directives for care of these exposures in recent legislation in the Promise to Address Comprehensive Toxics (PACT) Act of 2022.[4]

A proposal was received from the Federal Electronic Health Record Modernization (FEHRM) Office to create a specific ICD-10-CM code for reporting exposures to alkenes. This will facilitate tracking and documentation of alkene exposure in the electronic health records of active duty service members within the DOD and VA, benefiting them and their family members, as well as others in the general public who may come in contact with these substances.

References
1. Zhang XY, Elfarra AA. Myeloperoxidase-mediated bioactivation of olefins. Chapter 4 in Advances in Molecular Toxicology, Volume 12. Edited by James C. Fishbein, Jacqueline M. Heilman. Pp. 123-150. 2018. https://doi.org/10.1016/B978-0-444-64199-1.00005-1
2. Henderson RF. Species differences in the metabolism of olefins: implications for risk assessment. Chemico-Biological Interactions 135–136:53-64, 1 June 2001. https://doi.org/10.1016/s0009-2797(01)00170-3
3. Zhang XY, Elfarra AA. Potential roles of myeloperoxidase and hypochlorous acid in metabolism and toxicity of alkene hydrocarbons and drug molecules containing olefinic moieties. Expert Opinion on Drug Metabolism & Toxicology 13(5):513-524, 2017, published online: 16 Dec 2016. https://doi.org/10.1080/17425255.2017.1271413
4. United States Congress. (2022). Sergeant First Class Heath Robinson Honoring our Promise to Address Comprehensive Toxics Act of 2022. https://www.congress.gov/117/plaws/publ168/PLAW-117publ168.pdf

TABULAR MODIFICATIONS

	T52	Toxic effect of organic solvents	
	T52.8	Toxic effects of other organic solvents	
New sub-Subcategory	T52.81	Toxic effects of alkenes	
New code Add	T52.811	Toxic effect of alkenes, accidental (unintentional) Toxic effects of alkenes NOS	
New code	T52.812	Toxic effect of alkenes, intentional self-harm	
New code	T52.813	Toxic effect of alkenes, assault	

New code		T52.814	Toxic effect of alkenes, undetermined
Delete	T52.8X	~~Toxic effects of other organic solvents~~	
Delete		~~T52.8X1~~	~~Toxic effect of other organic solvents, accidental (unintentional)~~
Delete			~~Toxic effects of other organic solvents NOS~~
Delete		~~T52.8X2~~	~~Toxic effect of other organic solvents, intentional self-harm~~
Delete		~~T52.8X3~~	~~Toxic effect of other organic solvents, assault~~
Delete		~~T52.8X4~~	~~Toxic effect of other organic solvents, undetermined~~
New sub-subcategory	T52.89	Toxic effects of other organic solvents	
New code		T52.891	Toxic effect of other organic solvents, accidental (unintentional)
Add			Toxic effects of other organic solvents NOS
New code		T52.892	Toxic effect of other organic solvents, intentional self-harm
New code		T52.893	Toxic effect of other organic solvents, assault
New code		T52.894	Toxic effect of other organic solvents, undetermined

Amyloid-Related Imaging Abnormalities

Amyloid-related imaging abnormalities (ARIA) are findings that may be secondary to amyloid beta-directed monoclonal antibody treatment in Alzheimer's Disease (AD). This is a repeat presentation, with prior presentation having been in September 2024.

ARIA is diagnosed on neuroimaging. It is thought to be the result of reduced vascular integrity due to an inflammatory response and impaired perivascular amyloid beta clearance. ARIA presents in two forms, ARIA-edema/effusion (ARIA-E) and ARIA-hemosiderin deposition (ARIA-H). ARIA-E is the consequence of vascular leakage of proteinaceous fluid and presents as parenchymal vasogenic edema and sulcal effusion in the leptomeningeal/subpial space. ARIA-H is a consequence of blood leakage and deposition of blood degradation products into brain parenchyma and leptomeningeal/subpial space presenting as microhemorrhages and superficial siderosis, respectively.

Presence of symptoms associated with ARIA impacts clinical decision-making and treatment.

The term ARIA is usually reserved for neuroimaging findings in the context of amyloid beta-directed monoclonal antibody treatment of AD. ARIA may be asymptomatic or associated with a range of neurological findings and symptoms. However, similar imaging changes may occur spontaneously with other diseases leading to amyloid accumulation in the brain, such as cerebral amyloid angiopathy (CAA) and AD, which may co-exist. However, CAA has distinct diagnostic criteria that rely on neuroimaging and/or neuropathologic findings. In addition, ARIA and CAA may have different clinical presentations when symptomatic and may be managed differently. Concurrent CAA (with or without neuroimaging evidence) is thought to be an underlying risk factor for ARIA in AD patients treated with amyloid beta-directed monoclonal antibodies.

The Center for Drug Evaluation and Research, U.S. Food and Drug Administration, has proposed new codes for ARIA. The updated proposal would include codes for ARIA under the ICD-10-CM category for "Other abnormal findings on diagnostic imaging of central nervous system". When applicable, these codes should be used in conjunction with the ICD-10-CM code for CAA and/or adverse effect of immunoglobulin. The proposed codes distinguish ARIA-E from ARIA-H, as well as subtypes of ARIA-H. This updated revision would capture whether ARIA is symptomatic based on use of separate codes for the symptoms.

The addition of the proposed codes for ARIA would allow population-based monitoring of this important safety risk in the context of treatment with amyloid beta-directed monoclonal antibodies.

TABULAR MODIFICATIONS

G30 Alzheimer's disease

Use additional code, if applicable, to identify:
Add Amyloid-related imaging abnormalities (ARIA) (R90.83-)

R90 Abnormal findings on diagnostic imaging of central nervous system

R90.8 Other abnormal findings on diagnostic imaging of central nervous system

New sub-subcategory R90.83 Amyloid-related imaging abnormalities [ARIA]
Add Code also, if applicable, for:
Add adverse effect of immunoglobin (T50.Z15)
Add cerebral amyloid angiopathy (I68.0)
Add symptoms and signs resulting from ARIA (may be coded if applicable) including, but not limited to, the following:
Add seizure (G40)
Add status epilepticus (G41.9)
Add unspecified encephalopathy (G93.40)
Add visual disturbances (H53)
Add dizziness (R42)
Add headache (R51.9)
Add gait abnormality (R26.9)
Add nausea (R11.0)
Add vomiting (R11.10)

New code R90.830 Amyloid-related imaging abnormalities with edema/effusion [ARIA-E]

New code R90.831 Amyloid-related imaging abnormalities with hemosiderin deposition [ARIA-H]

New code R90.8310 Amyloid-related imaging abnormalities with hemosiderin deposition [ARIA-H] unspecified

New code R90.8311 Amyloid-related imaging abnormalities with hemosiderin deposition [ARIA-H] with microhemorrhage

New code R90.8312 Amyloid-related imaging abnormalities with hemosiderin deposition [ARIA-H] with superficial siderosis

New code R90.839 Amyloid-related imaging abnormalities, unspecified

Apragmatism

Stroke is the leading cause of disability in the United States with a nearly 50% chance of damage to either hemisphere. While left hemisphere damage after stroke results in the language disorders of aphasia, right hemisphere damage (RHD) results in apragmatism with up to 78% of survivors experiencing deficits that affect pragmatic, contextualized use of language.

Speech intelligibility, grammar, and sentence structure of language after RHD are not customarily impaired like in aphasia. Rather, the ability to organize, plan, use, and understand verbal or nonverbal language for the communicative context and communication partners is impaired. Language use after RHD may not follow conventions of turn-taking with reduced interpretation and use of non-verbal, gestural, and vocal cues important to the context. It can be perceived as impolite, off-topic, and self-centered, and may contain few questions, poor initiation and management of topics, contextually inappropriate and tangential comments and humor. Aberrant pragmatic language skills after right hemisphere stroke are distinctly atypical well within the chronic phase of recovery and contribute to personal and professional communication challenges that cause irreparable damage to relationships.

Within the ICD-10-CM classification, pragmatic language disorders are restricted to a developmental code (F80.82). Due to the lack of availability of more specific coding options to capture apragmatism, clinicians are constrained to use ICD-10-CM codes which are established for cognitive communicative disorders (CCD) such as R48.8 – (other symbolic dysfunction), R41.84 (cognitive communication disorders), and I69.31 and I69.315 (cognitive deficits following cerebral infarction) to support inpatient and outpatient clinical encounters. These ICD-10-CM codes, however, are neither specific to nor representative of apragmatism as an acquired language disorder after RHD. Social isolation, unemployment, damaged personal and professional relationships are at the core of the socio-economic and emotional toll experienced by survivors of RHD. A unique ICD-10-CM code would assist to develop population specific treatments, focused curricula, and professional development to mitigate the disabling and chronic effects of apragmatism. An ICD-10-CM diagnostic code for apragmatism can promote greater recognition, treatment, and empirical pursuits for right hemisphere language disorders. The ability to accurately report these conditions is crucial for early identification and multidisciplinary rehabilitation to optimize patient outcomes of disability associated with stroke.

This proposal is submitted by the Ad Hoc Apragmatism ICD-10 Code Committee at Duke University School of Medicine, a multidisciplinary group comprised of clinician scientists in speech-language pathology, neurology, and psychology with the support of the American Speech-Language and Hearing Association's Healthcare Economics Committee (ASHA HCEC), Duke Stroke Center, the Academy of Neurologic Communication Disorders and Sciences (ANCDS), and the North Carolina Stroke Advisory Council.

References
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TABULAR MODIFICATIONS

I69 Sequelae of cerebrovascular disease
I69.0 Sequelae of nontraumatic subarachnoid hemorrhage
I69.02 Speech and language deficits following nontraumatic subarachnoid hemorrhage
New code I69.024 Apragmatism following nontraumatic subarachnoid hemorrhage

I69.1 Sequelae of nontraumatic intracerebral hemorrhage
I69.12 Speech and language deficits following nontraumatic intracerebral hemorrhage
New code I69.124 Apragmatism following nontraumatic intracerebral hemorrhage
I69.2 Sequelae of other nontraumatic intracranial hemorrhage
I69.22 Speech and language deficits following other nontraumatic intracranial hemorrhage

New code I69.224 Apragmatism following other nontraumatic intracranial hemorrhage

I69.3 Sequelae of cerebral infarction

I69.32 Speech and language deficits following cerebral infarction

New code I69.324 Apragmatism following cerebral infarction

I69.8 Sequelae of other cerebrovascular diseases

I69.82 Speech and language deficits following other cerebrovascular disease

New code I69.824 Apragmatism following other cerebrovascular disease

I69.9 Sequelae of unspecified cerebrovascular diseases

I69.92 Speech and language deficits following unspecified cerebrovascular disease

New code I69.924 Apragmatism following unspecified cerebrovascular disease

R47 Speech disturbances, not elsewhere classified

R47.8 Other speech disturbances

New code R47.83 Apragmatism

Arrhythmogenic Cardiomyopathy

Arrhythmogenic Cardiomyopathy (ACM) also referred to as Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) is a genetic cardiomyopathy disease that is characterized by the replacement of normal cardiac muscle with fibrofatty tissue that affects the right, left, or both ventricles and which contributes to the development of dangerous ventricular arrhythmias and sudden cardiac death (SCD). ACM affects roughly 1 in 2,000 to 1 in 5,000 people.

The clinical hallmark of ACM is ventricular arrhythmias that often arise from the right ventricle due to the fibrofatty replacement of ventricular myocardium (per McKenna et al., 1994). Diagnosis usually occurs at a young age. Affected individuals have reported experiencing premature ventricular contractions, which may be experienced as palpitations, fainting or syncope. In some patients, the first sign of ACM is sudden cardiac arrest. Approximately 20% of patients are diagnosed following their first cardiac arrest, and the first arrest may be sudden cardiac death.

The diagnosis and management of ACM requires a multidisciplinary approach involving cardiologists, electrophysiologists, and genetic counselors. Lifestyle modifications, such as restriction from certain sports and strenuous physical activity, may be recommended to reduce the risk of arrhythmias and sudden cardiac events. Genetic counseling is advised for patients and family members, as ACM is inherited in an autosomal dominant pattern, and at-risk relatives may benefit from genetic testing and regular cardiac evaluations.

ACM is a progressive disease with no known cure. Treatment of ACM focuses on mitigating progression, preventing arrhythmias, cardiac arrest, and sudden death. Beta blockers are used to lower heart rate and blood pressure; ACE inhibitors may also be helpful. Treatment may also include ablation procedures and/or implantable cardioverter-defibrillators (ICDs).

Historically, the paradigm for diagnosing and treating cardiomyopathies relied on multiple criteria being present, because no diagnostic test was considered specific enough to establish a definitive diagnosis (per Migliore et al., 2021). Over time, clinical studies demonstrated that the initial ACM diagnostic criteria were highly specific but lacked sensitivity in mild cases. Consequently, the revised 2010 Task Force (TF) criteria added new ECG parameters, quantitative measurements in echocardiogram parameters, and magnetic resonance (MR) imaging to enhance diagnostic sensitivity (per Migliore et al., 2021). In 2019, an International Expert Report pointed out that the 2010 TF did not include criteria for the diagnosis of Arrhythmogenic Left Ventricular Cardiomyopathy (ALVC) to identify fibrofatty scar at the LV level. (Towbin et al., 2019) It was in this context that the 2020 International Expert consensus document upgraded the diagnostic criteria for ACM by adding the Padua criteria for the diagnosis of ACM phenotypes, including the left-sided forms. Today, the term “Arrhythmogenic Cardiomyopathy” is the preferred terminology used for the diagnosis of ACM.

There has been an evolution of knowledge regarding ACM and diagnostic advancements, with development of 2020 international criteria, labeled the Padua criteria, which use cardiac MR findings for characterization (per Corrado et al., 2021).

There are several ongoing trials using adeno-associated virus (AAV)-based gene therapy to treat ACM caused by mutations in the PKP2 gene. One company has received orphan drug designation from the FDA, indicating that ACM is distinct from other forms of cardiomyopathy.

Creation of a specific ICD-10-CM code for arrhythmogenic cardiomyopathy has been proposed by the Sudden Arrhythmia Death Syndromes (SADS) Foundation.

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TABULAR MODIFICATIONS

	I42	Cardiomyopathy
	I42.8	Other cardiomyopathies
New code	I42.81	Arrhythmogenic cardiomyopathy
New code	I42.89	Other cardiomyopathies not elsewhere classified

INDEX MODIFICATIONS

	Cardiomyopathy
Add	- arrhythmogenic I42.81
Add	- - biventricular I42.81
Add	- - ventricular (left) (right) I42.81

Dysplasia – see also Anomaly
Revise - arrhythmogenic right ventricular I42.81
Revise - right ventricular, arrhythmogenic I42.81

Biomarkers for Alzheimer's Disease

Biomarkers are biological indicators that can be identified and measured in blood, other body fluids, and tissues. They can be linked to increased risk for certain diagnoses or used to make or confirm a specific diagnosis. Once a diagnosis is established, biomarkers can also be used to assess disease progression and to inform clinical decision-making.

Biomarkers play a major role in detecting underlying pathology of Alzheimer's disease. Amyloid plaques and tau tangles in the brain are key pathological features of Alzheimer's disease that disrupt normal function of neurons. Amyloid plaques are abnormally folded beta-amyloid Aβ peptides that clump together in the brain. Aβ peptides are derived from amyloid precursor protein, which performs various neuronal development, signaling and stability functions [1].
Phosphorylated tau (p-tau) occurs when phosphate molecules bind to tau, a structural protein, and can lead to misfolded hyperphosphorylated tau tangles in the brain. Alzheimer's disease pathology may be detected up to two decades before the emergence of clinical symptoms, such as cognitive decline [2].

In the past, amyloid plaques and tau tangles could be identified only through post-mortem brain autopsy. Later, it was found that they could be reliably detected and measured using PET scans with specific radiotracers. Abnormal levels of Aβ peptides and tau in cerebrospinal fluid and blood correlate with amyloid plaques and tau tangles in the brain [3]. These can be detected through cerebrospinal fluid obtained via lumbar puncture or, more recently, with blood- based biomarker tests. In May 2025, the FDA cleared the first blood-based biomarker test for the detection of Alzheimer's pathology in individuals with cognitive decline [4].

While there is robust clinical discussion over whether biomarkers represent an early stage of Alzheimer's disease or a risk factor for the disease, there is consensus that they are present - and potentially clinically significant - even before onset of symptoms [3, 5-6]. As individuals move either toward ruling out or establishing a diagnosis, the presence of biomarkers provides a way to clearly describe the rationale for continued evaluation.

In the absence of clinical symptoms, the presence of biomarkers also impacts management of individuals, including addressing modifiable risk factors and determining appropriate follow-up. Lifestyle modifications may reduce or delay cognitive decline from Alzheimer's disease [7].
Clinical studies are also underway to identify drugs and other interventions in cognitively intact but biomarker-positive individuals to slow or prevent progression to symptomatic Alzheimer's disease [8-9].

After reviewing laboratory or imaging findings of Alzheimer's pathology, physicians may document the presence of biomarkers using phases such as "biomarker evidence of amyloid pathology in the brain" or "elevated p-tau in CSF concerning for Alzheimer's disease." At present, there are no specific codes for Alzheimer's biomarkers.

Given the clinical significance of biomarkers, they should be distinctly identified in the data to reflect accurate rationale for continuing clinical evaluations and work-up, identify possible participants in future clinical trials for asymptomatic patients with abnormal biomarkers, avoid misuse of diagnosis codes for Alzheimer's disease, and track evolving management.

This proposal has been submitted by The Global CEO Initiative on Alzheimer's Disease (CEOi), which is a convened consortium of the non-profit patient advocacy organization UsAgainstAlzheimer's. Founded in 2013, CEO brings together private-sector leaders and other collaborators to provide leadership in the fight against Alzheimer's.

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TABULAR MODIFICATIONS

R77 Other abnormalities of plasma proteins
Excludes1:disorders of plasma-protein metabolism (E88.0-)

R77.0 Abnormality of albumin
R77.1 Abnormality of globulin
Hyperglobulinemia NOS
R77.2 Abnormality of alphafetoprotein
R77.8 Other specified abnormalities of plasma proteins

New code R77.81 Abnormality in beta-amyloid and tau biomarkers
Add Elevated level of beta-amyloid in plasma
Add Elevated level of tau in plasma
Add Excludes1: Alzheimer's disease (G30.-)

New code R77.89 Other specified abnormalities of plasma proteins

R77.9 Abnormality of plasma protein, unspecified

R83 Abnormal findings in cerebrospinal fluid

R83.8 Other abnormal findings in cerebrospinal fluid

Delete ~~Abnormal chromosomal findings in cerebrospinal fluid~~

New code R83.81 Abnormality in beta-amyloid and tau biomarkers in cerebrospinal fluid

Add Elevated beta-amyloid in cerebrospinal fluid

Add Elevated tau in cerebrospinal fluid

Add Excludes1: Alzheimer's disease (G30.-)

New code R83.89 Other abnormal findings in cerebrospinal fluid

Add Abnormal chromosomal findings in cerebrospinal fluid

R90 Abnormal findings on diagnostic imaging of central nervous system

R90.8 Other abnormal findings on diagnostic imaging of central nervous system

New code R90.84 Beta-amyloid and tau biomarkers of brain

Add Presence of beta-amyloid plaques of brain

Add Presence of tau tangles of brain

Add Excludes1: Alzheimer's disease (G30.-)

Brugada Syndrome

Brugada syndrome is a rare, autosomal dominant genetic disorder that causes ventricular tachycardia. Brugada syndrome is 8-10 times more prominent in males than in females. The mean age of sudden death is 40 years, but the youngest reported case was at 2 days of life. Individuals with Brugada syndrome are more likely to develop ventricular fibrillation as compared to the general population. Brugada syndrome accounts for up to 20% of all sudden deaths in individuals with a presumed normal heart. Brugada syndrome affects approximately 3 to 5 individuals per 10,000 in the general population.

Clinically, Brugada syndrome is established by the following clinical pathway.
A syncopal episode results in ventricular arrhythmia identification/confirmation, with a family history of sudden cardiac arrest or sudden cardiac death. Brugada pattern is identified on electrocardiogram, with characteristic ST and T wave changes in the precordial leads.

There are no known cures for Brugada syndrome. Management of the condition is non-specific and emphasizes the prevention of sudden cardiac death. The only effective therapy requires the implantation of a cardioverter defibrillator (ICD). Asymptomatic patients may be monitored until symptoms develop, but the first symptom can be sudden cardiac death.

Currently, there are over 30 clinical trials in various stages for Brugada syndrome. Some trials seek additional information on the condition, while others seek to decrease the risk of sudden cardiac death. One clinical trial is investigating a new drug for the treatment of Brugada syndrome.

Brugada syndrome is currently reported with the nonspecific ICD-10-CM code: I49.8 Other specified cardiac arrhythmias.

A specific ICD-10-CM code would facilitate clinical research and enable collection of standardized data for Brugada syndrome, enabling the tracking of patient outcomes, which is crucial for determining the effectiveness or ineffectiveness of treatment options. Importantly, with clinical trials studying treatments for Brugada syndrome, a more specific ICD-10-CM code would enable the medical records and study data to be more precise and comprehensive, with the potential to help improve patient care and lead to more effective interventions. Additionally, specific ICD-10-CM codes facilitate clear and standardized communication among all healthcare providers and insurers, enabling better support for specific medication or therapy, as well as reducing the risk of errors in patient care.

Creation of a specific ICD-10-CM code for Brugada syndrome has been proposed by the Sudden Arrhythmia Death Syndromes (SADS) Foundation.

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TABULAR MODIFICATIONS

	I49	Other cardiac arrhythmias	
	I49.8	Other specified cardiac arrhythmias	
Delete		~~Brugada syndrome~~	
Delete		~~Coronary sinus rhythm disorder~~	
Delete		~~Ectopic rhythm disorder~~	
Delete		~~Nodal rhythm disorder~~	
New code	I49.81	Brugada syndrome	
New code	I49.89	Other specified cardiac arrhythmias not elsewhere classified	
Add		Coronary sinus rhythm disorder	
Add		Ectopic rhythm disorder	
Add		Nodal rhythm disorder	

Catatonia

Catatonia is a syndrome of clinical importance involving primarily psychomotor disturbances, characterized by the co-occurrence of several symptoms of decreased, increased, or abnormal psychomotor activity. It can occur in the context of certain medical conditions (e.g., stroke, anti-NMDA receptor encephalitis). The condition can be caused by substance or medication use (cocaine, phencyclidine). Catatonia can occur in the context of several mental disorders including schizophrenia, bipolar disorder and major depressive disorder.

Currently, ICD-10-CM includes catatonia due to a medical condition (F06.1), catatonia associated with schizophrenia (F20.2), and stupor (R40.1).

The Diagnostic and Statistical Manual of Mental Disorders, Text Revision (DSM-5-TR), published by the American Psychiatric Association (APA), is a clinical classification with relevant terminology designed to facilitate reliable and consistent mental and behavioral health conditions diagnosis. The classification provides diagnostic criteria approved for routine clinical use and corresponding ICD-10-CM diagnostic codes.

In the DSM-5-TR, it is noted that catatonia can manifest in the context of various mental health disorders, including neurodevelopmental, psychotic, bipolar, and depressive disorders, as well as medical conditions.

This proposal aims to rectify the absence of an ICD-10-CM code for catatonia attributable to a mental disorder, distinct from F06.1, which is appropriate only when catatonia results from a medical condition.

According to the American Psychiatric Association (APA), the inclusion of terms under F06.1, Catatonic disorder due to known physiologic condition, was a provisional measure to accommodate these conditions within the code set. However, these terms are not a genuine clinical inclusion terms for this code subcategory.

The Permanente Federation along with the support of the American Psychiatric Association request the following tabular modification to the code set to attain alignment between the code set and the diagnostic manual and promote accurate and complete coding.

ICD-10 Coordination and Maintenance Committee Meeting
September 9-10, 2025

TABULAR MODIFICATIONS

F06 Other mental disorders due to known physiological condition

F06.1 Catatonic disorder due to known physiological condition

Delete ~~Catatonia associated with another mental disorder~~
Delete ~~Catatonia NOS~~
Add Catatonia disorder due to another medical condition
Excludes1: catatonic stupor (R40.1)
stupor NOS (R40.1)
Add catatonia (R46.82)

R46 Symptoms and signs involving appearance and behavior

Excludes1: appearance and behavior in schizophrenia, schizotypal and delusional disorders (F20-F29)
mental and behavioral disorders (F01-F99)

R46.8 Other symptoms and signs involving appearance and behavior

R46.81 Obsessive-compulsive behavior
Excludes1: obsessive-compulsive disorder (F42.-)

New code R46.82 Catatonia
Add Catatonia associated with another mental disorder
Add Catatonia induced by substances or medications
Add Catatonia NOS
Add Excludes1: catatonic disorder due to known physiological condition (F06.1)

Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)

Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) is a genetic disorder that leads to ventricular tachycardia. It is estimated to affect about one in 10,000 individuals in the general population, although the true prevalence remains unknown.

Clinically, CPVT is established through the following clinical pathway:
Syncope episode during exercise or emotion-induced bidirectional or polymorphic ventricular tachycardia in a patient with a structurally normal heart and normal resting EKG.
Genetic testing can confirm the diagnosis. The International Guidelines on Sudden Cardiac Death indicate that the diagnosis is established if a patient has heterozygous or biallelic pathogenic variants, with 75% of CPVT cases caused by variants in eight genes (*CALM1, CALM2, CALM3, CASQ2, KCNJ2, RYR2, TECRL, TRDN*). CPVT can be inherited in an autosomal dominant or recessive pattern.

Onset of CPVT syncopal episodes typically occurs between the ages of 7 and 12 years. CPVT can lead to spontaneous recovery or ventricular fibrillation, which may cause sudden death. Approximately 30% of affected individuals experience at least one cardiac arrest. The first manifestation of CPVT can be sudden death.

Currently, there are no known cures for CPVT. The Heart Rhythm Society, the European Heart Rhythm Society, and the European Society of Cardiology have established guidelines for managing ventricular arrhythmias. These guidelines are not specific to CPVT and are intended to prevent symptoms. Management options may include beta-blocker therapy, antiarrhythmic medications, implantable cardioverter defibrillator (ICD), and left cardiac sympathetic denervation. Although ICDs can prevent sudden cardiac death, their implantation may result in inappropriate shocks, multiple surgeries, and post-traumatic stress disorder due to previous shocks.

Currently, CPVT may be reported with a non-specific ventricular tachycardia ICD-10-CM code. Specific diagnosis codes enhance clinical research by providing standardized data that can be used to study treatment effectiveness, disease epidemiology, and patient outcomes. Accurate and detailed diagnosis codes are crucial for tracking disease prevalence.

There are now at least eight clinical trials focused on therapies for catecholaminergic polymorphic ventricular tachycardia (CPVT). These trials aim to repair leaky RyR2 channels or inhibit the protein kinase CaMKII. The FDA has designated CPVT as a rare pediatric disease and granted orphan drug status to one therapy.

A specific ICD-10-CM code would facilitate clinical research and enable collection of accurate and more comprehensive data, and would have potential to enhance patient care by accurately conveying the subtype of the disease and potentially leading to more effective interventions. The creation of unique ICD-10-CM codes for CPVT is likely to improve the identification of patients with CPVT, facilitate genetic testing, and enhance the clinical care of patients, ultimately bolstering research efforts. Establishing a specific ICD-10-CM code is expected to enhance clinical patient care and facilitate advancements in

research by enabling the tracking of patient outcomes, which is crucial for informing the effectiveness or ineffectiveness of current treatment options.

Creation of a specific ICD-10-CM code has been proposed by the Sudden Arrhythmia Death Syndromes (SADS) Foundation.

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TABULAR MODIFICATIONS

I47 Paroxysmal tachycardia

I47.2 Ventricular tachycardia

New Code Add	I47.22 Catecholaminergic polymorphic ventricular tachycardia [CPVT] Familial polymorphic ventricular tachycardia [FPVT]

Chronic Hand Eczema

Chronic hand eczema, also called chronic hand dermatitis, is a common inflammatory skin disease that lasts for more than three months or relapses twice or more within a year. It is the most common occupational disorder of the skin and creates a significant burden unlike those of other forms and sites of eczema, greatly impacting quality of life for affected individuals.

The disorder is characterized by erythema, edema, and vesicles, leading to scaling, thickening or hyperkeratosis, lichenification, and fissuring of the hands. Nail changes may also occur, including loss of the cuticle and thickening of the nail plate.

Chronic hand eczema is a very common skin disorder, with a general prevalence of about 4-5% at any given time and a lifetime prevalence of about 14%. It is more prevalent in people with certain occupations that involve frequent exposure of the hands to water, irritants, chemicals, and allergens, such as nurses, hairdressers, butchers, construction workers, metalworkers, and florists.

There are three subtypes of chronic hand eczema, although it is not uncommon for chronic hand eczema to involve more than one subtype at a time.

- *Atopic dermatitis* tends to arise at younger ages and in addition to the hands, may be seen in association with atopic dermatitis elsewhere on the body.

- *Allergic contact dermatitis* is due to direct exposure of the hands to specific allergens including chemicals, dyes, cosmetics, food, plants and animal hair. Diagnosis involves identifying the culprit allergen, for which the gold standard is patch testing.

- *Irritant contact dermatitis* is due to direct exposure of the hands to specific irritants such as detergents, solvents, chemicals, and metals. This is typically a diagnosis of exclusion when the other subtypes have been ruled out and is the most common subtype of chronic hand eczema.

Some classifications separately identify protein contact dermatitis in which contact with animal or plant proteins can produce a hypersensitivity allergic reaction; hyperkeratotic hand eczema with thickening of the palms; dyshidrosis, also called recurrent vesicular eczema, which is characterized by eruption of blister-like vesicles; and nummular eczema in which coin-shaped lesions appear on the back of the hands.

Management of chronic hand eczema involves basic skincare steps including avoiding the allergens and irritants identified, using specific handwashing and drying techniques, and taking skin protection measures such as gloves, barrier creams, and emollients. Treatment frequently involves topical anti-inflammatories, such as corticosteroids, and, in some cases, calcineurin inhibitors, such as tacrolimus. In moderate to severe disease, systemic therapies may be needed, including oral corticosteroids. Immunosuppressants, such as cyclosporine and methotrexate, and retinoids are also used. However, these have significant side effects, including teratogenicity in retinoids, which may preclude use as a long-term treatment.

Phototherapy may also be used, largely on the basis of its effectiveness in other skin disorders. Notably, none of these therapies are FDA-approved specifically for chronic hand eczema in the US.

LEO Pharma, Incorporated, a pharmaceutical company, is requesting new codes to identify individuals with chronic hand eczema for future research and treatment. Currently, there is no unique code in ICD-10-CM. This proposal has been reviewed and supported by the American Academy of Dermatology.

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TABULAR MODIFICATION

L20 Atopic dermatitis
Add Code also, if applicable, chronic hand eczema (L30.81)

L23 Allergic contact dermatitis
Add Code also, if applicable, chronic hand eczema (L30.81)

L24 Irritant contact dermatitis
Add Code also, if applicable, chronic hand eczema (L30.81)

L30 Other and unspecified dermatitis
Excludes2: contact dermatitis (L23-L25)
dry skin dermatitis (L85.3)
small plaque parapsoriasis (L41.3)
stasis dermatitis (I87.2)

L30.0 Nummular dermatitis
Add Code also, if applicable, chronic hand eczema (L30.81)

L30.1 Dyshidrosis [pompholyx]
Add Vesicular eczema

Add Code also, if applicable, chronic hand eczema (L30.81)

New subcategory L30.8 Other specified dermatitis

New code L30.81 Chronic hand eczema

Add Code also, if applicable:
Add allergic contact dermatitis (L23.-)
Add atopic dermatitis (L20.-)
Add dyshidrosis (L30.1)
Add hyperkeratosis (L85.9)
Add irritant contact dermatitis (L24.-)
Add nummular dermatitis (L30.0)

New code L30.89 Other specified dermatitis

L85 Other epidermal thickening

L85.9 Epidermal thickening, unspecified
Add Hyperkeratosis NOS

Add Code also, if applicable, chronic hand eczema (L30.81)

Contusion of Pancreas

Currently, ICD-10-CM classifies a traumatic splenic contusion based on whether it is minor, major, or unspecified. However, a traumatic pancreatic contusion is classified in ICD-10-CM solely based on the anatomical location: head, body, tail, or unspecified, and does not delineate between minor, major, or unspecified.

Both the American Association for the Surgery of Trauma (AAST) and Abbreviated Injury Scale (AIS) grading systems make a distinction between major and minor contusion for spleen and pancreatic contusions. It is proposed to add new ICD-10-CM codes to further specify the severity of pancreatic contusions that are minor, major, or unspecified.

The traumatic pancreatic injury is usually a sequela of penetrating trauma. Blunt trauma (i.e., bicycle handlebar injuries in children, steering wheel injury in an adult motor vehicle collision, or direct kick in an assault) causes a sudden localized impact to the abdomen which results in compression of the intraabdominal organs against the vertebral column and can lead to pancreatic injury. Main pancreatic duct disruption is crucial to identify as it is the primary cause of delayed complications. Pancreatic injuries are rarely solitary as 90% of cases usually involve damage to another abdominal organ with duodenal-pancreatic injury being the most common.

While anatomic location is an important consideration for management and prognosis of pancreatic injury, severity of injury is a significant consideration in management of pancreatic trauma. In fact, the involvement of the pancreatic duct is the most important consideration in the evaluation and management of these injuries, even more important than anatomical location. As severity of contusion increases, the likelihood of pancreatic duct involvement increases.

This proposal was submitted by Vanderbilt University Medical Center.

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ICD-10 Coordination and Maintenance Committee Meeting
September 9-10, 2025

TABULAR MODIFICATIONS

S36 Injury of intra-abdominal organs
S36.2 Injury of pancreas
S36.22 Contusion of pancreas

Revise	S36.220	Contusion of head of pancreas, unspecified severity
Add		Contusion of head of pancreas, unspecified involvement of pancreatic duct
Revise	S36.221	Contusion of body of pancreas, unspecified severity
Add		Contusion of body of pancreas, unspecified involvement of pancreatic duct
Revise	S36.222	Contusion of tail of pancreas, unspecified severity
Add		Contusion of tail of pancreas, unspecified involvement of pancreatic duct
Revise	S36.229	Contusion of unspecified part of pancreas, unspecified severity
Add		Contusion of unspecified part of pancreas, unspecified involvement of pancreatic duct
New code	S36.22A	Minor contusion of head of pancreas
Add		Contusion of head of pancreas without pancreatic duct injury
Add		Contusion of head of pancreas without pancreatic duct involvement
New code	S36.22B	Minor contusion of body of pancreas
Add		Contusion of body of pancreas without pancreatic duct injury
Add		Contusion of body of pancreas without pancreatic duct involvement
New code	S36.22C	Minor contusion of tail of pancreas
Add		Contusion of tail of pancreas without pancreatic duct injury
Add		Contusion of tail of pancreas without pancreatic duct involvement

New code	S36.22D	Minor contusion of unspecified part of pancreas
Add		Contusion of unspecified part of pancreas without pancreatic duct injury
Add		Contusion of unspecified part of pancreas without pancreatic duct involvement
New code	S36.22E	Major contusion of head of pancreas
Add		Contusion of head of pancreas with pancreatic duct injury
Add		Contusion of head of pancreas with pancreatic duct involvement
New code	S36.22F	Major contusion of body of pancreas
Add		Contusion of body of pancreas with pancreatic duct injury
Add		Contusion of body of pancreas with pancreatic duct involvement
New code	S36.22G	Major contusion of tail of pancreas
Add		Contusion of tail of pancreas with pancreatic duct injury
Add		Contusion of tail of pancreas with pancreatic duct involvement
New code	S36.22H	Major contusion of unspecified part of pancreas
Add		Contusion of unspecified part of pancreas with pancreatic duct injury
Add		Contusion of unspecified part of pancreas with pancreatic duct involvement

Cycloparaffin Exposure

Cycloparaffins (also known as naphthenes and cycloalkanes) are hydrocarbon solvents found in solvents, adhesives, and sealants.[1] It is also the second most common chemical by volume contained within jet propellant fuel 5 (JP-5) and is a significant component of jet propellant fuel 4 (JP-4).[2]

Exposure to cycloparaffin can occur via ingestion, inhalation, or skin contact. Symptoms of acute exposure include central nervous system symptoms (headache, fatigue, dizziness, concentration problems), gastrointestinal symptoms (nausea, vomiting, abdominal pain), and dermatologic issues (skin rashes, irritation).[3,4]

Documentation of cycloparaffin exposure is of particular significance for military personnel who may have been exposed to the substance during their military service due to their potential long term health consequences. Recent legislation in the Promise to Address Comprehensive Toxics (PACT) Act of 2022 expanded benefits for veterans exposed to toxic substances.[5] Currently no ICD-10-CM code exists that specifically records exposures to cycloparaffins.

A proposal was received from the Federal Electronic Health Record Modernization (FEHRM) Office to create a specific ICD-10-CM code for reporting exposures to cycloparaffins. This will facilitate tracking, documentation, and accurate reporting within the Department of Defense (DOD) and the Department of Veteran Affairs (VA), as well as others in the general public who may come in contact with these substances.

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TABULAR MODIFICATIONS

T52 Toxic effect of organic solvents

T52.8 Toxic effects of other organic solvents

New sub-subcategory	T52.82	Toxic effects of cycloparaffins	
Add		Toxic effects of naphthenes	
Add		Toxic effects of cycloalkanes	
New code		T52.821	Toxic effect of cycloparaffins, accidental (unintentional)
Add			Toxic effects of cycloparaffins NOS
New code		T52.822	Toxic effect of cycloparaffins, intentional self-harm
New code		T52.823	Toxic effect of cycloparaffins, assault
New code		T52.824	Toxic effect of cycloparaffins, undetermined

Doxycycline Post-Exposure Prophylaxis

Bacterial sexually transmitted infections (STIs) are common in the United States, with over 1.6 million cases of chlamydia, over 600,000 cases of gonorrhea, and over 200,000 cases of syphilis reported in 2023.[1] Gonorrhea and chlamydia can cause long-term health consequences such as pelvic inflammatory disease, chronic pelvic pain, or infertility and ectopic pregnancy, particularly when left untreated.[2] Syphilis can also cause serious clinical complications, including central nervous system disease and adverse pregnancy outcomes.[3] In June 2024, the Centers for Disease Control and Prevention (CDC) released its guidelines on the use of doxycycline as post-exposure prophylaxis (doxy PEP) for the prevention of bacterial STIs.[4] These guidelines detail how providers can prescribe doxycycline 200mg, taken once after sex, to significantly reduce the risk of acquiring a bacterial STI among certain populations disproportionately affected by STIs.

Doxy PEP represents the first new biomedical prevention tool for the prevention of bacterial STIs in decades. In three randomized controlled trials, doxy PEP demonstrated efficacy in reducing acquisition of bacterial STIs (syphilis, chlamydia, and gonorrhea).[5-7]

Doxy PEP has been well received, with interest from patients and uptake in a number of jurisdictions across the United States.[8-11] To date, at least 17 state and county public health departments have released guidance supporting the use of doxy PEP.[12] Early evidence also points to the efficacy of doxy PEP as a public health intervention to reduce STI rates. At the same time, evidence from clinicals trials shows that use of doxy PEP may be associated with increased antimicrobial resistance (AMR) among sexually transmitted pathogens such as gonorrhea and other pathogens, such as *Staphylococcus aureus*.[13] In both clinical trials and population studies, doxy PEP has been associated with increased high-level tetracycline resistance in gonorrhea.[6, 14]

Currently, there is no unique ICD-10-CM code addressing an indication for doxy PEP prescription. The use of ICD-10-CM code Z20.2, Contact with and (suspected) exposure to infections with a predominantly sexual mode of transmission, is not specific to the use of doxycycline as STI prophylaxis. The existing code Z20.2 is for a wide array of purposes related to screening, prevention, and treatment of STIs limits any interpretation of its use.

CDC Division of Sexually Transmitted Disease Prevention is requesting a new code to better capture prescription of doxycycline specifically for post-exposure prophylaxis to prevent the acquisition of bacterial STIs and to track the use of doxy PEP to understand its public health benefits and potential consequences for antimicrobial resistance.

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TABULAR MODIFICATIONS

Z29 Encounter for other prophylactic measures

Z29.8 Encounter for other specified prophylactic measures

New subcategory Z29.82 Encounter for doxycycline post-exposure prophylaxis

New code Z29.820 Encounter for doxycycline post-exposure prophylaxis bacterial sexually transmitted infection

New code Z29.828 Encounter for doxycycline post-exposure prophylaxis other infection

Add Encounter for doxycycline prescription following tick bite

Ectopic Pregnancies

Ectopic pregnancies (pregnancies that implant outside of the endometrial cavity) pose significant risk to a patient's health and are treated as medical emergencies, accounting for 4% of pregnancy-related deaths in the United States and 80% of pregnancy-related deaths in the first trimester.[1–4] Approximately 2% of all reported pregnancies are ectopic.[1,5] While over 90% of ectopic pregnancies occur within the fallopian tube (ICD-10 CM O00.1), ectopic pregnancies may implant in other sites within the abdomen and pelvis – referred to as non-tubal ectopic pregnancies and account for 5-8.3% of ectopic pregnancies.[4]

This proposal was presented at the September 2024 ICD-10 Coordination and Maintenance Meeting. In response to public comments, changes have been made and are noted in **bold**.

Cesarean scar ectopic pregnancy (CSEP) occurs when an embryo implants into the niche created by the incision of a previous cesarean delivery (CD).[5] It is estimated that cesarean scar ectopic pregnancy occurs in up to 1/1800 pregnancies and is likely underreported without a unique ICD-10-CM code.[2] Furthermore, the incidence is increasing due to rising rates of CD in the United States and improved awareness and recognition.[6]

CSEP has a 44% estimated rate of significant morbidity when untreated or if treatment is delayed, including uterine rupture, life-threatening hemorrhage, unplanned laparotomy and hysterectomy, and even mortality.[6] In 2020, the Society for Maternal-Fetal Medicine with the endorsement of The American College of Obstetricians and Gynecologists (ACOG) and the Society of Family Planning and support from the American Society for Reproductive Medicine, published recommendations to classify these pregnancies as ectopic pregnancies to highlight the distinction from normally implanted intrauterine pregnancies.[2] Other non-tubal ectopic pregnancy locations include the cervix and the interstitial portion of the tube (the section of the fallopian tube that traverses the myometrium), both of which are rare, but pose equally high risk of morbidity for patients. Cornual ectopic pregnancy is an interstitial ectopic pregnancy in a bicornuate uterus or within the rudimentary horn of an unicornuate uterus.

Non-tubal ectopic pregnancies are distinct in their anatomic implantation, and each have unique clinical considerations. While "other ectopic pregnancy" (O00.8) and for "ectopic pregnancy, unspecified" (O00.9) currently exist, they do not adequately capture each unique clinical entity. Without individual ICD-10-CM codes for each distinct type of ectopic pregnancy, clinicians are unable to capture the correct diagnoses and severely limits the ability to identify and study these cases to improve patient outcomes.

This proposal was submitted by The University of Colorado, Division of Complex Family Planning, Department of Obstetrics & Gynecology (Nancy Fang, MD MS) and Oregon Health & Science University, Division of Complex Family Planning, Department of Obstetrics & Gynecology (Jessica Reid, MD MCR) and is supported by the Society of Family Planning, the Society for Maternal-Fetal Medicine, and the American College of Obstetrics and Gynecology.

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TABULAR MODIFICATIONS

O00 Ectopic pregnancy

O00.1 Tubal pregnancy

New sub subcategory **O00.12 Interstitial ectopic pregnancy without intrauterine pregnancy**

New code **O00.121 Right interstitial ectopic pregnancy without intrauterine pregnancy**

New code **O00.122 Left interstitial ectopic pregnancy without intrauterine pregnancy**

New code **O00.129 Unspecified interstitial ectopic pregnancy without intrauterine pregnancy**

New sub subcategory **O00.13 Interstitial ectopic pregnancy with intrauterine pregnancy**

New code **O00.131 Right interstitial ectopic pregnancy with intrauterine pregnancy**

New code **O00.132 Left interstitial ectopic pregnancy with intrauterine pregnancy**

New code **O00.139 Unspecified interstitial ectopic pregnancy with intrauterine pregnancy**

New subcategory O00.3 Cesarean scar ectopic pregnancy

New code O00.31 Cesarean scar ectopic pregnancy without intrauterine pregnancy

New code O00.32 Cesarean scar ectopic pregnancy with intrauterine Pregnancy

New subcategory	O00.4 Cervical ectopic pregnancy
New code	O00.41 Cervical ectopic pregnancy without intrauterine pregnancy
New code	O00.42 Cervical ectopic pregnancy with intrauterine pregnancy

New subcategory	**O00.5 Cornual ectopic pregnancy**
New sub subcategory	**O00.51Cornual ectopic pregnancy without intrauterine pregnancy**
New code	**O00.511 Right cornual ectopic pregnancy without intrauterine pregnancy**
New code	**O00.512 Left cornual ectopic pregnancy without intrauterine pregnancy**
New code	**O00.519 Unspecified cornual ectopic pregnancy without intrauterine pregnancy**
New sub subcategory	**O00. 52 Cornual ectopic pregnancy with intrauterine pregnancy**
New code	**O00.521 Right cornual ectopic pregnancy with intrauterine pregnancy**
New code	**O00.522 Left cornual ectopic pregnancy with intrauterine pregnancy**
New code	**O00.529 Unspecified cornual ectopic pregnancy with intrauterine pregnancy**

	O00.8 Other ectopic pregnancy
Delete	~~Cervical pregnancy~~
Delete	~~Cornual pregnancy~~

ER-low (Estrogen Receptor-low [ER+/1-10% expression]), HER2-low immunohistochemistry [IHC]

ER-low (Estrogen Receptor-low [ER+/1-10% expression]), HER2-low immunohistochemistry ([IHC] score of 1+ or 2+ without amplification on in situ hybridization) and HER2-ultralow cancers (IHC 0+) represent clinically significant subtypes of breast cancer, but no unique codes in the ICD-10-CM specifically describe ER-low or HER2-low and ultralow. Unique codes for ER-low, HER2-low and ultralow, as well as Human Epidermal Growth Factor Receptor null status (IHC 0, without membrane staining), are needed to track ER-low, HER2-low and ultralow patients.

Breast cancer is the second most common cancer and one of the leading causes of cancer-related deaths worldwide. More than two million breast cancer cases were diagnosed in 2022 with more than 665,000 deaths globally. In the US, more than 300,000 cases of breast cancer are diagnosed annually. While survival rates are high for those diagnosed with early breast cancer, only about 30% of patients diagnosed with, or who progress to, metastatic disease are expected to live five years following diagnosis. HR-positive, HER2-negative is the most common breast cancer subtype, accounting for approximately 70% of all breast cancers. Endocrine therapies are widely given consecutively in the early lines of treatment for HR-positive metastatic breast cancer. However, after initial treatment, further efficacy from endocrine therapy is often limited.

The current standard of care following endocrine therapy is chemotherapy, which is associated with poor response rates and outcomes. HER2 is a tyrosine kinase receptor growth-promoting protein expressed on the surface of many types of tumors, including breast cancer. Patients with high levels of HER2 expression (IHC 3+ or 2+/ISH+) are classified as HER2-positive and treated with HER2-directed therapies, representing approximately 15-20% of all breast cancers. Historically, tumors that were not classified as HER2-positive were classified as HER2-negative.

ER-low tumors, those with 1-10% ER expression, represent a relatively small subgroup of breast cancer patients, with an estimated prevalence of 2-7%. These tumors are like ER-negative disease “in their molecular landscape, clinicopathological characteristics, prognosis, and response to therapy. Nevertheless, a proportion may retain some degree of ER signaling dependency, and the possibility of responding to some degree to endocrine therapy cannot be completely ruled out.”

While ER and HER2 expression are often discussed in the context of breast cancer, these subtypes are recognized in other cancers; non-small cell lung cancer (NSCLC) “harboring HER2 alterations is now considered a distinct molecular subtype” and HER2 has been identified as contributing to worse outcomes in prostate cancer.

By showing that a significant percentage of ER-low, HER2-low and ultralow breast cancers would previously have been deemed ER or HER2 negative, these data underscore the need for more detailed coding of ER-low, HER2-low and ultralow. By adding these codes to the Z17 code series, as opposed to within the C50 code set describing breast cancer, providers would be allowed to capture crucial information without extensive modification of the broader cancer neoplasms code sets. As HER2-

alterations become an important factor in the treatment of ovarian, lung, prostate or other cancers, these new Z17 codes will serve to better describe patients with these subtypes.

This proposal was submitted by Daiichi Sankyo, Inc., a pharmaceutical company.

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TABULAR MODIFICATIONS

Z17 Estrogen, and other hormones and factors receptor status

New code Z17.A Estrogen receptor low status
Add ER+/1-10% expression

Z17.3 Human epidermal growth factor 2 receptor

New code Z17.33 Human epidermal growth factor receptor 2 low status
Add IHC 1+, IHC 2+/ISH-

New code Z17.34 Human epidermal growth factor receptor 2 ultralow status
Add IHC 0+, with membrane staining

New code Z17.35 Human epidermal growth factor receptor 2 null status
Add IHC 0, without membrane staining

Z17.4 Combined receptor status
Z17.41 Hormone receptor positive

New code — Z17.412 Hormone receptor positive with human epidermal growth factor receptor 2 low status
Add — HR+ with HER2-low [IHC1+, IHC2+/ISH-]

New code — Z17.413 Hormone receptor positive with human epidermal growth factor receptor 2 ultralow status
Add — HR+ with HER2-ultralow [IHC0+, with membrane staining]

New code — Z17.414 Hormone receptor positive with human epidermal growth factor receptor 2 null status
Add — HR+ with HER2-null [IHC0, without membrane staining]

Z17.42 Hormone receptor negative

New code — Z17.422 Hormone receptor negative with human epidermal growth factor receptor 2 low status
Add — HR- with HER2-low [IHC1+, IHC2+/ISH-]

Exposure to Blast Overpressure

The Traumatic Brain Injury Center of Excellence (TBICoE) of the DOD Defense Health Agency is requesting new ICD-10-CM codes for exposure to high-level blast (HLB) overpressure and exposure to low-level blast (LLB) overpressure.

The Deputy Secretary of Defense issued a Department of Defense (DOD) memorandum, Managing Brain Health Risks from Blast Overpressure[2], on August 8, 2024.

The requirements include:

- Establish procedures to ensure personnel recognize blast overpressure (BOP) symptoms, report exposures to their command, and seek an evaluation from their medical provider if experiencing symptoms.
- Establish procedures to track and maintain oversight of BOP exposure risk management actions, including processes to request, and provide justification in writing for, any exceptions to the policies in this memorandum.

ICD-10-CM codes are also needed to track blast overpressure exposure. As described in the September Blast overpressure (BOP) injury codes presentation[1], the Deputy Secretary of Defense memorandum[2], and other research[3,4], brain health effects from BOP exposures are not yet fully understood. Medical providers should document accurate clinical information including BOP exposure, and the distinction between LLB and HLB. Identifying these exposures is necessary for surveillance and research to determine possible symptoms related to BOP exposure.

These BOP codes are intended to capture LLB and HLB exposures that do not meet the DOD criteria for traumatic brain injury (TBI), which is defined as an injury event that results in a blow or jolt to the head with concurrent loss of consciousness, alteration of consciousness, or post-traumatic amnesia. After LLB exposure, a service member might report vague symptoms (e.g., “don’t feel right”), especially after deployment and near retirement. Medical providers need to be able to document an encounter for BOP exposure with a SM whose symptoms may or may not be attributed to the exposure, or who may be asymptomatic.

ICD-10-CM Z codes represent reasons for encounters when circumstances other than a disease, injury, or external cause classifiable to categories. When some circumstance or problem is present which influences the person's health status but is not in itself a current illness or injury. More specifically, category Z77 indicates contact with and suspected exposures hazardous to health. These contact/exposure codes may be used to explain an encounter for testing, or, more commonly, to identify a potential risk. This aligns with verbiage from the Deputy Secretary of Defense’s memorandum on DOD Requirements for Managing Brain Health Risks from Blast Overpressure[2] as well as the DOD Warfighter Brain Health Research Strategy[5].

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TABULAR MODIFICATIONS

Z77	Other contact with and (suspected) exposures hazardous to health
New subcategory	Z77.4 Contact with and (suspected) exposure to blast (overpressure)
New code	Z77.40 Contact with and (suspected) exposure to unspecified blast (overpressure)
Add	Contact with and (suspected) exposure to blast (overpressure) NOS
New code	Z77.41 Contact with and (suspected) exposure to low-level blast (overpressure)
Add	Contact with and (suspected) exposure to LLB
New code	Z77.42 Contact with and (suspected) exposure to high-level blast (overpressure)
Add	Contact with and (suspected) exposure to HLB
New code	Z77.49 Contact with and (suspected) exposure to other blast (overpressure)

Familial-Genetic Dilated Cardiomyopathy (DCM)

Dilated cardiomyopathy (DCM) is the most common form of cardiomyopathy and is characterized by progressive thinning of the walls of the heart resulting in enlarged heart chambers that are unable to pump blood effectively. DCM affects approximately 1 in 250 individuals with primary etiologies of heart failure and is the leading cause of cardiac transplantation (Zheng et al, 2024; Myers et al, 2024).

A U.S. survey of 156,013 hospitalized patients (average age 75) found ischemic cardiomyopathy made up 59% of cases while nonischemic cardiomyopathy contributed 41%. Randomized heart failure trials report 30-40% of participants with nonischemic DCM compared to ischemic DCM (McNally et al, 2017).

Nonischemic DCM can stem from genetic factors, acquired conditions, or a combination of both. Familial-genetic DCM accounts for up to 50% of DCM patients, with up to 40% of familial patients having an identifiable genetic cause (Eldemire et al, 2024). Mutations in more than 250 genes (e.g., TTN, LMNA, BAG3, etc.) have been linked to DCM. Mutations causing DCM may impact force transmission, mechanical stress, signaling, desmosomal proteins, nuclear structure and function, ion channel activity, protein turnover, and calcium homeostasis. Based on the current U.S. population, the prevalence of familial-genetic DCM is estimated to be 276,341 patients (Newman et al, 2024).

Interestingly, a genetic cause is identified more frequently in pediatric patients than in adult patients (54% versus 27%). The 5-year survival rate for pediatric DCM with a familial cause is 94%, but this same group of patients has a relatively high 5-year transplantation rate of 38%, a significantly higher healthcare resource utilization. These trends differ in idiopathic DCM and myocarditis, highlighting the importance of genetic testing for all pediatric patients with DCM and proper identification (Eldemire et al, 2024).

The TTN gene produces titin, a crucial structural protein within muscle cells that plays an essential role in sarcomere function and muscle contraction. Truncating mutations in the sarcomeric protein, which represent approximately 50% of genetic variants, are the most common cause of familial-genetic DCM in adults (Grondin et al, 2025).

Patients with LMNA (Lamin A) variants represent approximately 11% of familial-genetic DCM cases and exhibit high disease penetrance (over 90%). These patients often experience a significant burden of electrophysiologic abnormalities, including atrioventricular block, atrial fibrillation, and malignant ventricular arrhythmias (Grondin et al, 2025; Newman et al, 2024).

Mutations in the BAG3 gene (BCL-2 associated athanogene 3) is associated with 2.3 to 3.6% of familial-genetic DCM across US, Europe and Japan, while a cohort of individuals with inherited cardiovascular disease in Canada identified mutations in BAG3 in 8% of DCM (Grondin et al, 2025). These mutations are highly penetrant, with approximately 80% of individuals carrying disease-causing genetic variants in the BAG3 gene developing early onset, rapidly progressing heart failure with 22% of patients progressing to heart transplant (Dominguez et al, 2018).

DCM patients with a genetic mutation are treated with drugs for heart failure, which includes angiotensin converting enzyme inhibitors, angiotensin receptor blockers, neprilysin inhibitors, beta-adrenergic

receptor antagonists, sodium-glucose cotransporter 2 (SGLT2) inhibitors, aldosterone antagonists and diuretics, along with certain lifestyle changes (Bozkurt et al, 2016; Patel et al, 2024). Patients who meet specific parameters may also undergo placement of an implantable cardioverter defibrillator, a cardiac resynchronization device or a combination of the two. These patients progress to end-stage heart failure and death more rapidly than patients with DCM not associated with genetic variants, highlighting a higher level of healthcare resource utilization. For example, patients require mechanical cardiac support, heart transplant, or have heart failure-related death, at nearly twice the rate of similarly staged non-familial non-genetic DCM patients (Dominguez et al, 2018; McNamara et al, 2001; Kubanek et al, 2013).

Recent advances in cardiogenetics and deep phenotyping have shifted the paradigm toward more personalized treatment strategies. With improved access to next-generation sequencing and the integration of genomic insights into clinical practice, there is growing potential for targeted therapies tailored to specific genetic subtypes such as TTN, LMNA or BAG3 associated DCM (Malinow et al 2024; Chao et al, 2024). Affinia is developing AFTX-201, an investigational AAV gene therapy product that carries a full-length human BAG3 complementary deoxyribonucleic acid (cDNA) coding for the expression of BAG3 protein.

Currently, the ICD-10-CM classification does not include a code specifically for familial-genetic DCM. Clinical genetic testing was previously uncommon, with less than 0.43% of DCM cases undergoing genetic testing (Ababio et al, 2023). However, professional society guidelines have incorporated genetic testing into standard of care (AHA/ACC/HFSA Guideline, 2022) and other organizations including the Genetic Cardiomyopathy Awareness Consortium have further advanced the cause for genetic testing. As a consequence, genetic testing is becoming more common.

An ICD-10-CM code specifically for 'Familial-Genetic DCM' will further support the advances in genetic testing leading to improved management and risk assessment for individuals and their families including prophylactic use of Implantable Cardioverter Defibrillator (ICD) to prevent sudden cardiac death. Additionally, the code will keep pace with the evolution in clinical practice and research focused on gene-targeted therapies. Clinicians will be able to identify eligible participants more efficiently for clinical trials, ensuring enrollment with individuals who meet the specific genetic criteria required for the study. This streamlined enrollment process would accelerate the pace of research and development in the field of gene-targeted therapies, ultimately leading to the discovery of new treatments and cures for genetic disorders. This framework also enhances healthcare by improving patient care, optimizing resource utilization, advancing health outcomes research, and supporting public health initiatives.

A proposal to add a specific code for familial-genetic dilated cardiomyopathy and a separate specific code for nonfamilial dilated cardiomyopathy was received from Affinia Therapeutics.

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TABULAR MODIFICATIONS

	I42	Cardiomyopathy
	I42.0	Dilated cardiomyopathy
Delete		~~Congestive cardiomyopathy~~
New Code	I42.00	Dilated cardiomyopathy, unspecified
Add		Congestive cardiomyopathy, NOS
New Code	I42.01	Familial-genetic dilated cardiomyopathy
New Code	I42.09	Other (nonfamilial) dilated cardiomyopathy

Gadolinium Induced Gout

Gadolinium toxicity causes many health problems specifically bone pain and muscle weakness in both children [9] and adults. Mechanism of gadolinium bone pain and disorder movement leads to falls and significant morbidity and mortality. Fibrotic scarring is the pathophysiologic mechanism of the bone pain. Similar to lead, individuals are more susceptible to gadolinium toxicity harm due to lifelong neurotoxicity. The bone pain results occur by multisystem pathophysiologic mechanisms involving gadolinium binding throughout the body. Gadolinium binds calcium sites in cortical bone([10]), and transmetallation occurs with other metals body wide.

Gadolinium also binds proteins of many types including cell membrane transporters and receptors, disrupts synaptic neurotransmission in the brain [1], and binds actin in [2] in muscle, as well as binding carbonates, phosphates, citrates in the body.

Gadolinium induces cytokine proteins [3] termed hypercytokinemia of the human immune response resulting in often severe immune system dysregulation illness and systemic organ fibrosis and skeletal fibrosis. All of these biologic chemical reactions cause bone and joint symptomatic disease [7] which is the justification for gadolinium gout code set. The proposed code set directly follows the lead gout pathology code set.

Gadolinium toxicity causes brain damage & nerve disorders inclusive of movement disorders [1], and the resultant muscle and joint & bone pain[6]. Blood gadolinium levels are measured[5]. Low testosterone caused by gadolinium additionally results in bone pain via Leydig cell damage[4] and this also affects musculoskeletal function.

Finding gadolinium in blood samples indicates that exposure has resulted in absorption with distribution to all cells in all organs in the body. Research conducted has summarized and demonstrated that the brain is a critical target organ for detrimental gadolinium effects as gadolinium especially causes frontotemporal brain dysfunction and also disrupts the deep cerebral nuclei for movement.

Similar to lead, gadolinium causes brain damage and compared to other etiologies of brain damage, gadolinium toxicity results in a newly discovered signature injury of brain cell synaptic transmission damage producing different patterns of impairments in different individuals. Skull and rib pain are signature symptoms of gadolinium toxicity caused by gadolinium induced macrophage damage and monocyte distortion in the bone marrow.[8] Surgical bone specimens and autopsied bone specimens contain gadolinium. At surgical joint reconstructive surgeries, the incidence of gadolinium presence in the operative bone specimens is microscopically identified in all surgically resected joints and bone resection specimens documenting the prevalence of gadolinium involvement in all bone and joint disease requiring operative intervention. Gadolinium is found in all bone surgical specimens resected at the time of operative resection of diseased joints undergoing joint reconstructive implant surgery in all people exposed to gadolinium.

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TABULAR MODFICATIONS

M1A Chronic gout

New subcategory	M1A.5 Gadolinium-induced chronic gout Code first toxic effects of gadolinium (T56.82-)
New code	M1A.50 Gadolinium-induced chronic gout, unspecified site
New sub-subcategory	M1A.51 Gadolinium- induced chronic gout, shoulder
New code	M1A.511 Gadolinium-induced chronic gout, right shoulder
New code	M1A.512 Gadolinium-induced chronic gout, left shoulder
New code	M1A.519 Gadolinium-induced chronic gout, unspecified shoulder

New sub-subcategory	M1A.52 Gadolinium-induced chronic gout, elbow
New code	M1A.521 Gadolinium-induced chronic gout, right elbow
New code	M1A.522 Gadolinium-induced chronic gout, left elbow
New code	M1A.529 Gadolinium-induced chronic gout, unspecified elbow
New sub-subcategory	M1A.53 Gadolinium-induced chronic gout, wrist
New code	M1A.531 Gadolinium induced chronic gout, right wrist
New code	M1A.532 Gadolinium-induced chronic gout, left wrist
New code	M1A.539 Gadolinium-induced chronic gout, unspecified wrist
New sub-subcategory	M1A.54 Gadolinium-induced chronic gout, hand
New code	M1A.541 Gadolinium-induced chronic gout, right hand
New code	M1A.542 Gadolinium-induced chronic gout, left hand
New code	M1A.549 Gadolinium-induced chronic gout, unspecified hand
New sub-subcategory	M1A.55 Gadolinium-induced chronic gout, hip
New code	M1A.551 Gadolinium-induced chronic gout, right hip
New code	M1A.552 Gadolinium-induced chronic gout, left hip
New code	M1A.559 Gadolinium-induced chronic gout, unspecified hip
New sub-subcategory	M1A.56 Gadolinium-induced chronic gout, knee
New code	M1A.561 Gadolinium induced chronic gout, right knee
New code	M1A.562 Gadolinium- induced chronic gout, left knee
New code	M1A.569 Gadolinium-induced chronic gout, unspecified knee

New sub-subcategory		M1A.57 Gadolinium-induced chronic gout, ankle and foot
New code		M1A.571 Gadolinium-induced chronic gout, right ankle and foot
New code		M1A.572 Gadolinium-induced chronic gout, left ankle and foot
New code		M1A.579 Gadolinium-induced chronic gout, unspecified ankle and foot
New code		M1A.58 Gadolinium-induced chronic gout, vertebrae
New code		M1A.59 Gadolinium-induced chronic gout, multiple sites
	M10	Gout Acute gout Gout attack Gout flare Podagra
New subcategory		M10.A Gadolinium-induced gout
Add		Code first toxic effects of gadolinium (T56.82-)
New code		M10.A0 Gadolinium-induced gout, unspecified site
New sub-subcategory		M10.A1 Gadolinium-induced gout, shoulder
New code		M10.A11 Gadolinium-induced gout, right shoulder
New code		M10.A12 Gadolinium-induced gout, left shoulder
New code		M10.A19 Gadolinium-induced gout, unspecified shoulder
New sub-subcategory		M10.A2 Gadolinium-induced gout, elbow
New code		M10.A21 Gadolinium-induced gout, right elbow
New code		M10.A22 Gadolinium-induced gout, left elbow
New code		M10.A29 Gadolinium-induced gout, unspecified elbow

New sub-subcategory	M10.A3 Gadolinium-induced gout, wrist
New code	M10.A31 Gadolinium-induced gout, right wrist
New code	M10.A32 Gadolinium-induced gout, left wrist
New code	M10.A39 Gadolinium-induced gout, unspecified wrist
New sub-subcategory	M10.A4 Gadolinium-induced gout, hand
New code	M10.A41 Gadolinium-induced gout, right hand
New code	M10.A42 Gadolinium-induced gout, left hand
New code	M10.A49 Gadolinium-induced gout, unspecified hand
New sub-subcategory	M10.A5 Gadolinium-induced gout, hip
New code	M10.A51 Gadolinium-induced gout, right hip
New code	M10.A52 Gadolinium-induced gout, left hip
New code	M10.A59 Gadolinium-induced gout, unspecified hip
New sub-subcategory	M10.A6 Gadolinium-induced gout, knee
New code	M10.A61Gadolinium-induced gout, right knee
New code	M10.A62 Gadolinium-induced gout, left knee
New code	M10.A69 Gadolinium-induced gout, unspecified knee
New sub-subcategory	M10.A7 Gadolinium -induced gout, ankle and foot
New code	M10.A71Gadolinium-induced gout, right ankle and foot
New code	M10.A72 Gadolinium-induced gout, left ankle and foot
New code	M10.A79 Gadolinium-induced gout, unspecified ankle and foot
New code	M10.A8 Gadolinium-induced gout, vertebrae
New code	M10.A9 Gadolinium-induced gout, multiple sites

R78 Findings of drugs and other substances, not normally found in blood
Revise Use additional code to identify ~~the~~ any retained foreign body, if applicable (Z18.-)
Excludes2: mental or behavioral disorders due to psychoactive substance use (F10-F19)

R78.7 Finding of abnormal level of heavy metals in blood
R78.71 Abnormal lead level in blood
Excludes1:lead poisoning (T56.0-)
New code R78.72 Abnormal gadolinium level in blood
Add Excludes1: toxic effect of gadolinium(T56.82-)
R78.79 Finding of abnormal level of heavy metals in blood

Z77 Other contact with and (suspected) exposures hazardous to health
Includes: contact with and (suspected) exposures to potential hazards to health

Z77.0 Contact with and (suspected) exposure to hazardous, chiefly nonmedicinal, chemicals
Z77.01 Contact with and (suspected) exposure to hazardous metals
Z77.010 Contact with and (suspected) exposure to arsenic
Z77.011 Contact with and (suspected) exposure to lead
Z77.012 Contact with and (suspected) exposure to uranium
Excludes1: retained depleted uranium fragments (Z18.01)
New code Z77.013 Contact with and (suspected) exposure to gadolinium

Gender Identity Disorder, in Remission (Desistance)

The literature on gender dysphoria encompasses cases of desistance, referring to a resolution of clinical symptoms associated with this mental health condition. Additionally, some individuals with gender dysphoria who underwent medical and surgical interventions to transition from their natal sex have later pursued detransition.[1,2] Many of these individuals, who once experienced incongruence between their experienced/expressed gender and natal sex, have come to later accept their biological sex. Whereas they once met the diagnostic criteria for gender dysphoria, they have now entered a period of remission (desistance) from this stated condition. Their cognitive experience of incongruence has remitted whereby experienced/expressed gender and natal sex are now aligned.

At the present, this reality is not accounted for in the ICD-10-CM. Similarly, those with a history of detransition or the reversal of gender transition, are not found in the classification system. The reasons for detransition may vary, though many now identify with their natal sex and have achieved remission from gender dysphoria. Others may fear medical complications from transition, while for some external pressures, such as ability to pay for transition, may be a factor.[3] We thus propose an additional code to account for the personal history of detransition.

Furthermore, the distress one may experience as a result of any adverse permanent and physiological effects of medical or surgical transition is not captured by the ICD-10-CM. The literature speaks to various feelings one may experience from regret to mourning. Data is limited. We seek to capture this clinical entity as posttransition distress which may include, but is not limited to, feelings of grief, regret, self-hatred, and/or isolation subsequent to and related to transition, that are clinically significant and impair social, school, occupational, or other important areas of functioning. This clinical distress is independent of desistance or detransition.

Without a code for desistance, detransition, or posttransition distress appropriate tracking of these clinical entities within the classification system is not possible. Their rates are uncertain. However, with increasing rates of gender dysphoria, evolving clinical and policy decisions, and a need to improve the healthcare for these individuals, it is more critical than ever to capture precisely these clinical entities.

The ICD-10-CM diagnosis codes we propose would provide a means to collect valuable health information to do research, improve patient care and safety, and inform the public health needs of this growing area of medicine. Such data could also be used to better inform public policy by allowing the medical community, and the public at large, to better understand the incidence and prevalence of remission and detransition. More so, it will allow providers to more accurately document the individual's clinical state, and better track such throughout the medical record for the purpose of delivering the very best care, including preventive health services. Lastly, establishing these diagnostic codes will move past the erroneous narrative that those with desistance and/or detransition do not exist. Currently, their clinical

status remains invisible to medicine's classification system, as well as the posttransition distress that can ensue regardless of whether one completes detransition or not.

Additionally, it is important to consider that cognitive acceptance of one's natal sex and remission from gender dysphoria does not necessarily resolve all psychological and physiological consequences of transition that one may have previously undertaken from one's natal sex to a different gender identity. Posttransition distress, as noted above, is one such consequence. To better capture the physiological effects, it is essential to more clearly elucidate within the coding hierarchy the historical actions those with gender dysphoria have taken in terms of transition or sex trait modification. To start, we propose that code Z87.890 Personal history of sex reassignment become a subcategory for two new codes – one for surgical procedures and another for medical interventions related to sex trait modifications. These would serve to better track the interventions one has undertaken.

Similarly, we do not seek to ignore the social transition which some have pursued. As this is outside the bounds of any medical or surgical "sex reassignment," we offer a new Z code for one's personal history of gender transition, social, i.e., those non-surgical and non-pharmacological steps one may take to live as a gender different from one's natal sex. This may include aspects of changing one's name or binding one's breasts, for example, the latter of which can result in medical complications. Ultimately, we believe these additional codes will better account for one's personal history of transition, recognizing there are surgical, medical, and social aspects, as well as complications which can arise from any of these.

To that end, much as it is important to understand the remission of gender dysphoria and to note any personal history of detransition, it is also important to capture the impact gender transition has on an individual. Depending on the type of sex trait modification or social transition, complications may include, but are not limited to, infertility/sterility, sexual dysfunction, impaired bone density accrual, cardiovascular disease and metabolic disorders, psychiatric disorders, surgical complications, complications to genital tucking and chest binding, and the aforementioned posttransition distress.[4,5,6] We seek to make the optional listing of any such complications part of the coding system. Such will allow for improved data collection and understanding, as well as provide an enhanced clinical picture, ultimately supporting the appropriate delivery of care.

We believe adding our proposed set of codes would set the United States apart in recognizing and supporting the care needs of those who have undergone desistance and/or detransition. We would be the first country with a classification system to formally monitor and track remission from gender dysphoria, a history of detransition, and any complications that arise from a "sex reassignment" or transition. With that, we anticipate better care, as well as a more comprehensive understanding of these diagnostic entities.

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TABULAR MODIFICATIONS

F64 Gender identity disorders

New code — F64.A Gender identity disorder, in remission
Add — Gender identity disorder, in remission (desistance)
New code — F64.B Posttransition Distress

Z87 Personal history of other diseases and conditions
Z87.8 Personal history of other specified conditions
Excludes2: personal history of self-harm (Z91.5-)

Z87.89 Personal history of other specified conditions

New sub-subcategory — Z87.890 Personal history of sex reassignment
Add — Code also: any manifestations such as
Add — acne (L70.-)
Add — drug-induced androgenic alopecia (L64.0)
Add — hyperlipidemia, unspecified (E78.5)
Add — metabolic syndrome (E88.810)
Add — other specified disorders of bone density and structure, unspecified site (M85.80)

New code — Z87.8901 Personal history of sex reassignment, surgical

New code	Z87.8902 Personal history of sex reassignment, medical
New code	Z87.893 Personal history of gender transition, social
Add	Code also: any manifestations such as
Add	other chest pain (R07.89)
Add	pain in thoracic spine (M54.6)
Add	rash and other nonspecific skin eruption (R21)
Add	shortness of breath (R06.02)
Add	testicular pain (N50.81-)
New code	Z87.894 Personal history of detransition
Add	Code also: any manifestations such as
Add	depressive episode (F32.-)
Add	female infertility, unspecified (N97.9)
Add	male infertility, unspecified (N46.9)
Add	menopausal and other perimenopausal disorders (N95.)
Add	voice and resonance disorders (R49.-)

Glanzmann Thrombasthenia

Glanzmann Thrombasthenia (GT) is a rare autosomal recessive disorder characterized by a deficiency or dysfunction of the αIIbβ3 integrin, leading to impaired platelet aggregation and mucocutaneous bleeding from early childhood. The prevalence of GT varies globally, ranging from 1 in 200,000 to 1 in 1,000,000 individuals, with higher incidences in regions with increased consanguinity. Diagnosis often requires specialized genetic and platelet function tests, distinguishing GT from other qualitative platelet disorders. The uniqueness of GT paired with life-threatening clinical outcomes warrants differentiation in ICD-10-CM from other platelet disorders.

The clinical phenotype of GT is characterized by a wide range of bleeding manifestations, from mild mucocutaneous bleeding to life-threatening hemorrhages. The clinical presentation is typically characterized by epistaxis, gingival bleeding, and menorrhagia most reported. Over 75% of people with GT experience life-threatening bleeding. GT is associated with significant morbidity with a substantial impact on day-to-day functioning, with approximately half of people with GT reporting daily bleeding , resulting in missed school and work as well as fatigue, cognitive impairment, and diminished quality of life secondary to iron deficiency. Bleeding is present throughout life.

Treatment recommendations for GT align with general measures for minor bleeding events akin to approaches for other platelet disorders. Nevertheless, substantial disparities emerge in management strategies. The management of GT, especially during pregnancy, demands specialized approaches due to the high risks of alloimmunization and severe bleeding. Platelet transfusions are used for the treatment of persistent, severe or life-threatening bleeding and to manage surgical bleeding, however with several risks, including allergic reactions, pathogen transmission, and the formation of allo-antibodies, resulting in refractoriness to platelets in the future. Pregnancy in people with GT carries significant risks of alloimmunization and life-threatening bleeding in the fetus, necessitating specialized care. The development of targeted therapies, such as bispecific antibodies and gene therapies, underscores the need for precise coding to monitor treatment efficacy and safety in GT patients.

The National Bleeding Disorders Foundation is requesting the following tabular modifications to facilitate accurate data collection, support epidemiological research, and treatment outcomes.

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TABULAR MODIFICATIONS

D69 Purpura and other hemorrhagic conditions

D69.1 Qualitative platelets defects

Delete		~~Bernard-Soulier [giant platelet] syndrome~~
Delete		~~Glanzmann's disease~~
Delete		~~Grey platelet syndrome~~
Delete		~~Thromboasthenia (hemmorrhagic) (hereditary)~~
Delete		~~Thrombocytopathy~~
New code	D69.11	Glanzmann thrombasthenia
Add		Glanzmann's disease
Add		Thromboasthenia (hemmorrhagic) (hereditary)
New code	D69.19	Other qualitative platelet defects
Add		Bernard-Soulier [giant platelet] syndrome
Add		Grey platelet syndrome
Add		Thrombocytopathy

Glutamate Receptor Ionotropic Neurodevelopmental Disorders

This is a repeat presentation for a subset of specific genetic Glutamate Receptor Ionotropic (GRI) neurodevelopmental disorders.

The CureGRIN Foundation has proposed new ICD-10-CM codes for certain distinct neurological disorders related to the ionotropic glutamate receptor, each with a specific genetic basis. These include GRIN1-Related Neurodevelopmental Disorder, GRIN2A-Related Neurodevelopmental Disorder, GRIN2B-Related Neurodevelopmental Disorder, GRIN2D-Related Neurodevelopmental Disorder, and GRIA2-Related Neurodevelopmental Disorder.

These disorders each have a unique phenotype that is related to their membership in a larger family of ionotropic glutamate receptors. These receptors mediate most excitatory synaptic transmission in the central nervous system. Each one is distinct in its genetics and function in the central nervous system, and all have been richly studied.[1] Ionotropic glutamate receptors play an important role in learning and memory, as well as, other critical biological functions including regulation of movements and biological rhythms such as breathing. The expression and function of each gene is differentially regulated during development, with distinct expression patterns pre- and postnatally. Each gene plays a specific role in central nervous system functions, with many involved in directing the wiring or connections of the central nervous system.[1] While all GRI genes are highly expressed in the brain, they are functionally and spatially distinct and each contributes uniquely to synaptic activity at different synapses. GRI genes are also variably expressed in other tissues.

The specific symptoms, temporal onset of symptoms, and severity of symptoms of each of these different disorders are related to the specific alteration of a specific gene. Each gene codes for a different receptor that has different functions at different synapses at different developmental time points in the brain. The GRIN1, GRIN2A, GRIN2B, and GRIN2D genes are among those that encode the subunits of the N-methyl-D-aspartate (NMDA) receptor.[1] NMDA receptors are assembled by these subunits in different combinations that are developmentally, functionally, and anatomically regulated. Variations of these receptors across the brain allow for the complexity that makes us human. Each of these is associated with a different spectrum of phenotypes ranging in severity. Symptoms are unique to each gene but often include developmental delay, intellectual disability, autism, speech deficiency, inability to walk, low muscle tone, gastrointestinal issues, feeding difficulties, cortical visual impairments, dystonia, seizures, movement disorders and paroxysmal sympathetic hyperactivity (PSH).[1-5]

GRIN1-related neurodevelopmental disorder often is associated with developmental delay, intellectual disability, hypotonia, spasticity of the limbs, movement disorders, behavior disorders, cortical visual impairment, feeding difficulties, gastrointestinal abnormalities, microcephaly, and malformations of cortical development.[2] GRIN1 variants are associated with epileptic encephalopathies, with seizure semiology including infantile spasms, tonic and atonic seizures, focal dyscognitive seizures, febrile seizures, hypermotor seizures, generalized tonic-clonic seizures, and status epilepticus.[2] The predicted incidence per 100,000 births is 5.45 for GRIN1.[6,7]

GRIN2A-related neurodevelopmental disorder may be associated with severe developmental and epileptic encephalopathy (DEE). GRIN2A variants also may be associated with epilepsy syndromes such as Landau-Kleffner syndrome.[8,9] Focal seizures are the predominant presentation of GRIN2A-related epilepsy, seen in more than half of affected individuals.[8] Generalized seizures can occur in up to 10% of cases and comprise atonic, atypical absence, myoclonic, and generalized tonic-clonic seizures. However, some individuals do not have seizures. Speech disorders are often seen in those with GRIN2A variants (over 90%), ranging from mild speech delay to severe speech and language difficulties.[8] About one-third of individuals have normal development and normal intellect.[9] The predicted incidence per 100,000 births is 3.23 for GRIN2A.[6,7]

GRIN2B-related neurodevelopmental disorder is often associated with developmental delay, intellectual disability, hypotonia, spasticity, movement disorders, behavior disorders, cortical visual impairment, feeding difficulties, and microcephaly.[4] The seizure semiology of GRIN2B-related epilepsy includes generalized seizures (mostly tonic or tonic-clonic), focal seizures, and epileptic spasms.[4] Two specific epilepsy syndromes that are evident in some individuals with GRIN2B variants include West syndrome and Lennox-Gastaut syndrome.[4] The prevalence of GRIN2B-related neurodevelopmental disorder among individuals with neurodevelopmental disorders/childhood-onset epilepsy is estimated at 0.2%.[10] The predicted incidence per 100,000 births is 5.91 for GRIN2B.[6,7]

GRIN2D-related neurodevelopmental disorder often has been associated with developmental delay, intellectual disability, hypotonia, spasticity, movement disorders, behavior disorders, cortical visual impairment, feeding difficulties, and microcephaly.[11] It presents with refractory epilepsy in early infancy, and DEE.[11] The predicted incidence per 100,000 births is 4.61 for GRIN2D.[6,7]

The GRIA2 gene encodes one of the subunits of the AMPA receptor.[1] GRIA2-related neurodevelopmental disorder patients display intellectual disability (ID) and neurodevelopmental abnormalities including autism spectrum disorder (ASD), Rett syndrome-like features, and seizures or developmental epileptic encephalopathy (DEE).[12] The predicted incidence per 100,000 births is 2.79 for GRIA2.[6,7]

Accurate and timely diagnosis is required to pursue adequate treatment and precision therapy for affected individuals. The proposed unique codes for each disorder will inform clinical management and drug discovery. Early evidence shows that therapies may be successful in some GRI diseases but unsafe or contraindicated in others. In addition, many affected individuals are dependent on the support and care of guardians and caregivers throughout life. Unique ICD-10-CM codes will assist with identifying affected individuals and enabling specific therapies as well as tracking patients and evaluating the impact of each of these disorders on healthcare systems and decision-making.

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TABULAR MODIFICATIONS

QA0 Neurodevelopmental disorders related to specific genetic pathogenic variants

QA0.0 Neurodevelopmental disorders related to pathogenic variants in specific genes

QA0.01 Neurodevelopmental disorders related to pathogenic variants in certain specific genes

QA0.011 Neurodevelopmental disorders, related to pathogenic variants in glutamate receptor genes

New code	QA0.0111	GRIN1-related neurodevelopmental disorder
New code	QA0.0112	GRIN2A-related neurodevelopmental disorder
New code	QA0.0113	GRIN2B-related neurodevelopmental disorder
New code	QA0.0114	GRIN2D-related neurodevelopmental disorder

New code	QA0.0115	GRIA2-related neurodevelopmental disorder
New code	QA0.0119	Other glutamate receptor, ionotropic, related neurodevelopmental disorder

Hepatic Fibrosis

In June 2023, as the result of a multi-national Delphi process with medical societies, patient groups, and clinicians, a new nomenclature for non-alcoholic fatty liver disease (NAFLD) and non-alcoholic steatohepatitis (NASH) was announced. Nonalcoholic fatty liver disease is now metabolic dysfunction-associated steatotic liver disease (MASLD) and encompasses patients who have hepatic steatosis and have at least one of five cardiometabolic risk factors. Metabolic dysfunction-associated steatohepatitis (MASH) is the replacement term for nonalcoholic steatohepatitis.[1]

With this change, updated consensus guidelines now characterize stages F2 and F3 hepatic fibrosis as the "at-risk" MASH population that may benefit from a pharmacotherapy intervention.[2] As of November 2024, five Phase 3 studies are ongoing in NASH/MASH with moderate to advanced fibrosis with the first-ever treatment for patients with MASH approved by the FDA in March 2024 for patients with noncirrhotic moderate to advanced liver fibrosis (consistent with stages F2 and F3).[3-7] Noninvasive tests (versus liver biopsies) are now recommended for the staging of hepatic fibrosis, leading to language identifying fibrosis as consistent with fibrosis stage 2.

The ICD-10-CM codes implemented in October 2020 identify hepatic fibrosis but are no longer consistent with current regulatory and consensus practice guidelines or clinical practice.

Fibrosis stage is used to identify "at-risk" MASH patients and those that may benefit from a pharmacotherapy intervention. Terminology characterizing the fibrosis stage has evolved and differs across regulatory and consensus practice guidelines. However, the commonality across all is the distinct differentiation of stages F2 and F3 (moderate to advanced fibrosis) from stage F1 (early fibrosis) and stage F4 (cirrhosis).

Another significant advancement in the identification of MASH is how staging of hepatic fibrosis is determined. In clinical trials, the traditional approach to defining disease and fibrosis severity in patients with MASH has been to perform an invasive liver biopsy, which has variability in sampling; is subject to intra- and interobserver variability; and, rarely, may be associated with severe and/or fatal procedural complications. [8]

Biopsies are rarely used in clinical practice for the treatment of patients with MASH. Noninvasive tests have emerged as validated tools to address the problem of early risk stratification in MASH.[1] As such, characterization of clinically significant or moderate to advanced non-cirrhotic fibrosis using noninvasive testing is often referred to as consistent with fibrosis stages F2 and/or F3. The use of "consistent with" to describe fibrosis stages is important. This approach acknowledges that clinical practice has shifted from biopsy to using biomarkers to noninvasively categorize NASH/MASH severity[1] and aligns with labeling language for the first recently approved medication for NASH/MASH, resmetirom, a thyroid hormone receptor-beta (THR-beta) agonist indicated in conjunction with diet and exercise for the treatment of adults with NASH with moderate to advanced liver fibrosis (consistent with stages F2 to F3 fibrosis)." [10]

With the ICD-10-CM codes that identify hepatic fibrosis no longer consistent with current regulatory and consensus practice guidelines or clinical practice, Global Liver Institute is requesting the following modifications.

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TABULAR MODIFICATION

K74 Fibrosis and cirrhosis of liver

K74.0 Hepatic fibrosis

K74.00 Hepatic fibrosis, unspecified

K74.01 Hepatic fibrosis, early fibrosis

Revise Hepatic fibrosis, stage F1 ~~or stage F2~~

Add Mild fibrosis

New code K74.0A Hepatic fibrosis, moderate fibrosis

Add Hepatic fibrosis, stage F2

Add Mild to moderate fibrosis

K74.02 Hepatic fibrosis, advanced fibrosis

Hepatic fibrosis, stage F3

Hexamethylene Diisocyanate Exposure

Hexamethylene diisocyanate, also known as HDI, is found in hardening agents for automobile paints.[1] The most common way a person can be exposed to HDI is by breathing air that contains it as a vapor or mist, like that made when spray-painting a car.[2,3] Chemical Agent Resistant Coating (CARC) paints make up the largest category of paints used on military vehicles and is one of several potentially harmful substances to which some service members were exposed during the Gulf War. Acute (short-term) exposure to high concentrations of hexamethylene diisocyanate in humans can cause pulmonary edema, coughing, and shortness of breath.[4] Hexamethylene diisocyanate is also extremely irritating to the eyes, nose, and throat. Human studies have suggested that chronic (long-term) exposure to hexamethylene diisocyanate may cause chronic lung problems.

Creation of a specific code for this will allow documentation of exposure to hexamethylene diisocyanate in the electronic health records of active-duty service members (Department of Defense, DOD) and Veterans (Department of Veteran Affairs, VA), as well as any others in the general public who may have such exposures. Documentation of toxic exposures during military service is a high priority for both Departments due to their potential long term health consequences as well as directives in recent legislation, Promise to Address Comprehensive Toxics (PACT) Act.[5]

Hexamethylene diisocyanate was the 26th most common hazard (out of 767), based on data pulled January 2023, tracked within the Individual Longitudinal Exposure Record (ILER) which tracks toxic exposures for the DOD and VA. This request is a submission on behalf of the DOD, VA, and the Office of Federal Electronic Health Record Modernization (FEHRM).

A consortium of clinicians, researchers, and advocates propose the following tabular modifications to aid clinical care of affected veterans, advance epidemiological tracking of this condition and improve health outcomes in all individuals experiencing adverse effects from toxic exposure to hexamethylene diisocyanate.

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TABULAR MODIFICATIONS

T59 Toxic effect of other gases, fumes and vapors

T59.8 Toxic effect of other specified gases, fumes and vapors

New sub-subcategory	T59.82	Toxic effects of hexamethylene diisocyanate	
New code		T59.821	Toxic effect of hexamethylene diisocyanate, accidental (unintentional)
Add			Toxic effects of hexamethylene diisocyanate NOS
New code		T59.822	Toxic effect of hexamethylene diisocyanate, intentional self-harm
New code		T59.823	Toxic effect of hexamethylene diisocyanate, assault
New code		T59.824	Toxic effect of hexamethylene diisocyanate, undetermined

Hypertriglyceridemia

The National Lipid Association and the American Society for Preventive Cardiology issued a joint expert clinical consensus statement in March of 2025 titled, "Recognition and management of persistent chylomicronemia: A joint expert clinical consensus by the National Lipid Association and the American Society for Preventive Cardiology." The consensus statement uses the terminology normal, mild to moderate, severe, and extreme for defining the spectrum of hypertriglyceridemia (HTG) and identification of chylomicronemia.

HTG is diagnosed based on a fasting serum (or plasma) triglyceride panel. Individuals are diagnosed with HTG when fasting plasma concentrations of triglycerides exceed a threshold value of 150 mg/dL. Performance of and results from a lipid panel are critical to assessing diagnosis, disease risks, and treatment of HTG in clinical practice guidelines. Optimal triglyceride levels are below 150 mg/dL. HTG occurs when triglyceride levels exceed 150 mg/dL, and severe HTG (sHTG) occurs when triglyceride levels exceed 500 mg/dL.

Both sHTG [TGs ≥ 500 mg/dL] and extreme HTG (eHTG) [TGs ≥ 880 mg/dL] are characterized by very high levels of triglycerides. People with sHTG and eHTG experience an increased risk of developing other medical conditions, including acute pancreatitis and cardiovascular diseases. In the United States, 1.5% of adults, equal to approximately 3.9 million people, suffer from sHTG, and 0.29% of adults, equal to approximately 750,000 people, are diagnosed with eHTG.

eHTG, also referred to as chylomicronemia, involves excessive accumulation of chylomicrons in the blood, resulting in extremely high triglyceride levels. eHTG includes multifactorial chylomicronemia syndrome (MCS), characterized by genetic variances that on top of environmental factors lead to eHTG, and familial chylomicronemia syndrome (FCS), a distinct genetic disorder characterized by a limited ability to breakdown triglycerides in chylomicrons and, therefore, accumulation of those chylomicrons in the blood. MCS is more common and presents in adulthood, while FCS is rare and patients may present with eHTG even in childhood. Individuals with either condition are at high lifetime risk of acute pancreatitis.

Ionis pharmaceuticals is requesting the following new codes to facilitate research and characterization of the patient population, disease management, and treatment of patients with HTG.

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TABULAR MODIFICATIONS

E78 Disorders of lipoprotein metabolism and other lipidemias

E78.1 Pure hyperglyceridemia
Elevated fasting triglycerides
Endogenous hyperglyceridemia
Fredrickson's hyperlipoproteinemia, type IV
Hyperlipidemia, group B
Hyperprebetalipoproteinemia

Add
Pure hyperglyceridemia NOS
Very-low-density-lipoprotein-type [VLDL] hyperlipoproteinemia

Add
Use additional code to identify hypertriglyceridemia level, if applicable (E78.A-)

New sub-subcategory
E78.A Hypertriglyceridemia level

New code
E78.A0 Hypertriglyceridemia, mild to moderate

New code
E78.A1 Hypertriglyceridemia, severe
Add
Hypertriglyceridemia, severe [sHTG]

E78.3 Hyperchylomicronemia

Delete
~~Chylomicron retention disease~~
Delete
~~Fredrickson's hyperlipoproteinemia, type I or V~~
Delete
~~Hyperlipidemia, group D~~
Delete
~~Mixed hyperglyceridemia~~

New code
E78.30 Hyperchylomicronemia
Add
Chylomicron retention disease
Add
Extreme hypertriglyceridemia [eHTG]
Add
Fredrickson's hyperlipoproteinemia, type V
Add
Hyperlipidemia, group D
Add
Mixed hyperglyceridemia
Add
Multifactorial chylomicronemia syndrome [MCS]

New code
E78.31 Familial chylomicronemia syndrome [FCS]
Add
Fredrickson's hyperlipoproteinemia, type I

Koolen-de Vries Syndrome

Koolen-de Vries Syndrome (KdVS) is caused by pathogenic variants in the KAT8 Regulatory NSL Complex Subunit One gene (*KANSL1*). KdVS has a clinically recognizable phenotypic presentation with a range of both neurological and non-neurological symptoms impacting several organ systems.[1] The primary hallmarks of KdVS were first described in 2006,[2–4] and consist of psychomotor developmental delays, mild-to-moderate intellectual disability, hypotonia, and a distinct facial gestalt.[1–3] Hypotonia and significant motor impairment are often the earliest apparent symptoms (~50-75% of individuals), and cause developmental issues with feeding ability, as well as protracted and disordered speech and language development.[1,5,6] Generalized hypotonia is present in most (~77%) cases.[7] One of the more severe medical effects is tracheo/laryngomalacia, which causes respiratory problems, which are often recurrent and result in further clinical complications.[7–9] Also common are scoliosis, joint hypermobility, and congenital malformations of the heart and urogenital system.[1,7]

Individuals with KdVS often present with a characteristic set of facial features including low-set ears, a bulbous, "pear-shaped" nose, and an open mouth which is attributed to facial hypotonia.[1,2,7] Their behavior is described as "friendly, amiable, and cooperative."[1] Communication disorder is a core feature of KdVS (100% of patients): delayed speech articulation development with speech disorders of apraxia and dysarthria are the most common, and some patients have transient stuttering.[5,6] Receptive and expressive language impairment is also present.[5,6] Approximately 30-50% of KdVS patients have epilepsy, typically focal seizures with childhood onset.[1,10] A recent study on KdVS found that nearly 25% of KdVS patients report some congenital heart defect including atrial and ventricular septal defects, prolapsed and leaky mitral valves, bicuspid aortic valves and cardiomyopathy.[7,11] Over 50% of male patients have cryptorchidism.[7,12] Finally, there is emerging evidence that individuals with KdVS have compromised immune systems, with high rates of chronic respiratory infections, autoimmune diseases and chronic neutropenia.[1,7,12]

Koolen-de Vries Syndrome is caused by haploinsufficiency of the KANSL1 protein due to a deleterious mutation involving one copy of the *KANSL1* gene.[1] These mutations can either affect the *KANSL1* gene alone, or can be larger deletions (up to 600 kilobases) encompassing the 17q21.31 region including *KANSL1*.[1,13,14] The *KANSL1* gene encodes for the KAT8 Regulatory NSL Complex Subunit One protein, which is a key component of the MOF histone acetyltransferase complex, which regulates transcriptional activation of a broad set of genes.[15,16] Pathogenic disruptions of KANSL1 are linked to gene expression changes that cause dysregulated autophagy, cell division, clearance of damaged mitochondria, and synaptic transmission and plasticity.[17–20]

In 2016 the prevalence was estimated at 1:55,000,[7] but more recent estimates suggest a higher prevalence up to 1:30,000, implying that there are expected to be over 11,000 individuals with KdVS in the United States alone.

Creation of a specific ICD-10-CM code for KdVS will enable improved tracking and data collection, and lead to better understanding of the natural progression of this syndrome. This will support analysis of health data that is vital for epidemiological research. In turn, this will be key for determining treatment windows and appropriate outcome measures. Having a specific code would aid in developing standard

protocols of care across clinical centers that treat individuals with Koolen-de Vries Syndrome. This is particularly important for the hypotonia-caused tracheo/laryngomalacia, weakness of the tracheal and laryngeal tissues which puts KdVS patients undergoing anesthesia at high risk for airway blockage leading to hypoxia; intubation is difficult due to the risk of laryngeal spasm. Case reports have been published by clinicians regarding this risk, as warnings that KdVS individuals will need to use extra precaution during intubation and extubation during anesthesia to avoid further damage.[1,8,9] In addition, having a specific ICD-10-CM code will help in selecting cohorts of patients eligible for future clinical trials across multiple healthcare centers. This will be key for developing clinical trials to test therapies that are currently under development by the researchers of the KdVS scientific community.[23]

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TABULAR MODIFICATIONS

QA0 Neurodevelopmental disorders related to specific genetic pathogenic variants

New subcategory

QA0.7 Neurodevelopmental disorders related to pathogenic variants in specific genes associated with proteins with other and multiple functions

New sub-subcategory

QA0.70 Neurodevelopmental disorders related to pathogenic variants in certain specific genes associated with proteins with other and multiple functions

New code

QA0.702 Koolen-de Vries Syndrome

Ledderhose Disease/Plantar Fibromatosis & Plantar Fasciitis

This topic was presented at the March 2024 and September 2024 ICD-10 Coordination and Maintenance meeting. Based on public comments, revisions to the proposal have been made for reconsideration. Changes are indicated in **bold**.

Ledderhose Disease is a genetic disease, in the same family of diseases as Dupuytren's contacture and Peyronie's Disease. It leads to growth of painful fibrous nodules known as fibromas on the plantar surface of the feet that can be disabling to many. The nodules are most commonly found on the medial and central band of the plantar fascia in the midfoot and forefoot. Nodules are consistently painful and worsening after long periods of standing or ambulation.

Plantar fasciitis is an inflammatory and, at times, degenerative condition of the plantar fascia related to chronic or repetitive strain. The plantar fascia, anatomically, originates at the plantar aspect of the calcaneal tuberosity and extends distally, attaching to soft tissue in proximity to the MTP joints/bases of the toes. Typically, symptoms present as pain first step in the morning with improvement in ambulation.

Unilateral codes for plantar fasciitis would allow the healthcare practitioner to specify the foot being treated. As such they would be used with a higher frequency than bilateral codes. If plantar fasciitis is the main patient concern, and if present bilaterally, a common practice would be to code plantar fasciitis, left and plantar fasciitis right. If plantar fasciitis was not the presenting concern but present upon examination, then it may be listed as an unspecified code.
Plantar fasciitis most commonly occurs at its origin at the infracalcaneal tuberosity but can occur at its midportion in the medial longitudinal arch, and occasionally at its insertion.
Further anatomical subdivision with respect to specific non-heel locations would have limited utility

Currently, both Ledderhose Disease/Plantar Fibromatosis & Plantar fasciitis share the same ICD-10-CM code of M72.2. In addition, there is no laterality assigned to this code. The two entities are very different in presentation, different in treatment and are of different causes. There has been difficulty in tracking Ledderhose Disease with respect to incidence and prevalence due to the lack of a unique diagnosis code.

Podiatrists, Dr. Paul Carroll and Dr. Eddie Davis, are requesting new ICD-10-CM codes to distinguish between the two conditions and better track these patients.

ICD-10 Coordination and Maintenance Committee Meeting
September 9-10, 2025

TABULAR MODIFICATIONS

M67 Other disorders of synovium and tendon

New subcategory M67.A Plantar fasciitis

New sub-subcategory	**M67.A0**	**Plantar fasciitis, foot**	
New code	**M67.A01**	**Plantar fasciitis, right foot**	
New code	**M67.A02**	**Plantar fasciitis, left foot**	
New code	**M67.A09**	**Plantar fasciitis, unspecified foot**	
Add		**Plantar fasciitis NOS**	

M72 Fibroblastic disorders

M72.2 Plantar fascial fibromatosis

Delete	~~Plantar fasciitis~~
Add	Ledderhose disease
New code	M72.20 Plantar fascial fibromatosis, unspecified foot
New code	M72.21 Plantar fascial fibromatosis, right foot
New code	M72.22 Plantar fascial fibromatosis, left foot

Lipedema and Lipolymphedema

This topic was presented at the September 2020 and September 2024 ICD-10 Coordination and Maintenance meeting. Based on comments received during the public comments period and additional clinical consultations, a revised proposal is being presented for consideration. Changes are indicated in **bold**.

Lipedema, initially described at the Mayo clinic in 1940,[1] [2] is a loose, connective-tissue (fat) disease (lipomatosis) with a pathological deposition of fibrotic fatty tissue on the limbs of women sparing the trunk, hands and feet,[3] [4] resulting in a disproportionate body habitus. Lipedema is thought to affect 11% of the female population.[5] There is no specific ICD-10-CM code for lipedema. Deposition of lipedema fat increases with stage and body mass index (BMI) and likely involves sex hormones during times when weight is gained (puberty, pregnancy and menopause). Lipedema is inherited in 60% of women likely through genes affecting microvessels resulting in excess fluid bound to glycosaminoglycans in the interstitial space.[6]

Unique to lipedema is fat that is highly resistant to loss by diet, exercise, or bariatric surgery.[7-9] Lipedema is often confused with secondary obesity or lymphedema. Because of the many signs and symptoms associated with lipedema, lipedema is also known as a syndrome.[2] [10-12]

There are four stages of lipedema:[13]

Stage 1: Smooth skin over an enlarged hypodermis often with palpable pearl-sized nodules

Stage 2: Indentations of the skin often with a mattress pattern appearance due to the presence of fibrosis in the fibers and interstitial space[5] overlying a hypertrophic hypodermis with pearl-size and larger masses the size of walnuts or larger

Stage 3: Indentations of the skin accompanied by lobules of skin and hypodermal tissue that often form over the elbow, at the waist, at the hips, on the medial inner thighs and around the knees accompanied by small to very large masses

Stage 4: Extensive lobular and pendulous adipose tissue folds affecting multiple regions of the body, including the arms, abdomen, hips, thighs, buttocks, and legs

Stage 4 lipedema should be diagnosed by the presence of extensive lobular and pendulous adipose tissue folds affecting multiple regions of the body, including the arms, abdomen, hips, thighs, buttocks, and legs. This presentation has been described in Földi and Földi's *Textbook of Lymphology* as "elephantiastic lobar lipedema," due to its clinical similarity to advanced elephantiasis observed in severe lymphedema.[14] The term "lipolymphedema" is sometimes used to describe Stage 4 lipedema; however, lymphedema can arise at any stage of lipedema.

Lipedema is also divided into five subtypes, or phenotypes, depending on the primary affected region:[15-17]

Subtype I: increased deposit of fat in gluteus, hips and thighs
Subtype II: lipedema extends from hips to knees with a fat pad in the internal zone of the knees
Subtype III: lipedema extends from hips to ankles
Subtype IV: upper limbs (arms) are affected
Subtype V: only the lower part of the legs is affected

Lipedema subtypes enable targeted treatment planning, particularly when the disease extends beyond the legs to include the arms and torso. It also informs surgical decision-making by distinguishing medically necessary procedures aimed at removing pathologic adipose tissue from elective cosmetic surgery. While lipedema staging offers a general overview of disease severity, it does not capture the anatomic distribution or dynamic progression of this chronic, life-long condition.

Lymphedema is a chronic and progressive swelling caused by a low output failure of the lymphatic system, resulting in the development of a high-protein edema in the tissues. Lymphedema is a lifelong condition for which no cure exists. Lymphedema can be either primary (hereditary) or secondary. Secondary lymphedema is the most common cause of the disease and affects approximately 1 in 1000 Americans. Complications of lymphedema include recurrent bouts of cellulitis and/or lymphangitis, bacterial and fungal infections, lymphangio-adenitis, deep venous thrombosis, poor wound healing, leg ulcers, severe functional impairment, disability, and necessary amputation. Patients with chronic lymphedema for 10 years have a 10% risk of developing lymphangiosarcoma.

Dr. Karen Herbst, with support from the American Vein & Lymphatic Society (AVLS), is submitting the following modifications to identify and track lipedema with and without lymphedema patients.

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TABULAR MODIFICATIONS

E88 Other and unspecified metabolic disorders

New subcategory	E88.2	Lipomatosis, not elsewhere classified
Delete		~~Lipomatosis NOS~~
Delete		~~Lipomatosis (Check) dolorosa [Dercum]~~
New code		E88.21 Dercum disease
Add		Lipomatosis dolorosa [Dercum]
New code		E88.29 Lipomatosis, not elsewhere classified
Add		Lipomatosis NOS
	E88.8	Other specified metabolic disorders
New sub-subcategory		E88.83 Lipedema
Add		**Lipedema syndrome**
Add		**Use additional code to identify the lipedema subtype (E88.8A-)**
Add		**Code also if applicable lymphedema (I89.0)**
New code		E88.831 Lipedema, Stage 1
New code		E88.832 Lipedema, Stage 2
New code		E88.833 Lipedema, Stage 3
New code		**E88.834 Lipedema, Stage 4**
Add		**Lipolymphedema**
New code		**E88.838 Other lipedema stage**
New code		E88.839 Lipedema, unspecified
Add		Lipedema NOS
New sub-subcategory		E88.8A Lipedema **subtype**
Add		Code first, if known, the lipedema stage (E88.83-)

New code E88.8A1 Lipedema **subtype I**
Add Lipedema of buttock to hips

New code E88.8A2 Lipedema **subtype II**
Add Lipedema buttock to knees

New code E88.8A3 Lipedema **subtype III**
Add Lipedema of buttock to ankles

New code E88.8A4 Lipedema **subtype IV**
Add Lipedema of arms

New code E88.8A5 Lipedema **subtype V**
Add Lipedema of lower legs

New code E88.8A8 Other lipedema **subtype**

New code **E88.8A9 Lipedema, unspecified subtype**

E88.89 Other specified metabolic disorders
Delete ~~Launois-Bensaude adenolipomatosis~~

New code E88.891 Multiple symmetric lipomatosis
Add Launois Bensaude adenolipomatosis
Add Madelung's disease

New Code E88.898 Other specified metabolic disorders

I89 Other noninfective disorders of lymphatic vessels and lymph nodes

I89.0 Lymphedema, not elsewhere classified
Elphantiasis (nonfilarial) NOS
Lymphangiectasis
Obliteration, lymphatic vessel
Delete ~~Praecox lymphedema~~
Secondary lymphedema

New code **I89.A Idiopathic lymphedema**

Q82 Other congenital malformations of skin

Q82.0 Hereditary lymphedema
Add Praecox lymphedema

Loeys-Dietz Syndrome

Loeys-Dietz syndrome (LDS) is a multisystem disorder with autosomal dominant inheritance, caused by pathogenic variants in genes that encode effectors of the transforming growth factor-beta (*TGFβ*) signaling pathway that include *TGFBR1, TGFBR2, SMAD2, SMAD3, TGFB2*, and *TGFB3*.[1-6] LDS can lead to aggressive and early-onset aortic and arterial aneurysms and dissections, along with craniofacial anomalies (hypertelorism, bifid uvula, cleft palate), skeletal abnormalities (scoliosis, pectus deformity, clubfoot, cervical spine instability), and cutaneous features (translucent skin, easy bruising and abnormal scarring).[7,8] LDS can also include a predisposition for severe allergic manifestations (e.g., asthma, eczema, environmental and food allergies, anaphylaxis) and inflammatory gastrointestinal disorders, including eosinophilic esophagitis and inflammatory bowel disease.[9,10]

LDS is clinically and genetically distinct from other vascular disorders such as Marfan syndrome and vascular Ehlers-Danlos syndrome. Despite sharing some overlapping phenotypic features with these conditions, pathogenic variants in some of the LDS-related genes tend to cause vascular complications at smaller aortic diameters and younger ages. In contrast to Marfan syndrome, where the base of the aorta (i.e., aortic root) is the usual site of vascular involvement and complications, LDS is characterized by a strong predisposition for aneurysms and vascular tear or rupture throughout the entire arterial tree, so that frequent imaging of the entire arterial system is an essential part of clinical surveillance.

Clinical management guidelines, including those from the American Heart Association, recommend lower thresholds for surgical intervention for LDS due to its aggressive vascular course, which reflects the genetic heterogeneity.[11] As a result, clinical surveillance, surgical thresholds, and treatment strategies differ significantly from those for Marfan syndrome and vascular EDS and among those with LDS due to different causative genes. In this context, accurate diagnostic coding is essential.[7,11]

Current best estimates for the prevalence of LDS are in between those for Marfan syndrome (1:10,000) and Vascular Ehlers-Danlos syndrome (1:50-200,000)[12,13] These syndromes have ICD-10-CM codes and similar multisystemic manifestations but unique management guidelines.

Currently, LDS lacks a dedicated ICD-10-CM code. This leads to inconsistent classification under broader categories such as Marfan syndrome (Q87.4) or connective tissue disorder NOS (M35), which impairs clinical management, research, reimbursement, public health tracking, and rare disease registry development. It limits the ability to track and analyze LDS's unique clinical characteristics, associated medical complications, and healthcare utilization. As a result, important data related to the prevalence, treatment outcomes, and healthcare requirements for LDS remain obscured.

A dedicated code improves patient care, facilitates efficient healthcare standards and utilization, and accelerates research. A unique code would also support the goals of the Loeys-Dietz Syndrome Foundation, other patient advocacy groups, clinicians, and researchers, all of whom frequently describe barriers to care due to the absence of an LDS-specific ICD-10-CM code.

This proposal is submitted jointly by the Loeys-Dietz Syndrome Foundation, the Marfan Foundation, and the GenTAC Alliance. The proposal has been reviewed and supported by the American Academy of Pediatrics.

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Q87 Other specified congenital malformation syndromes affecting multiple systems
Use additional code(s) to identify all associated manifestations

New code
Add
Q87.A Loeys-Dietz syndrome
Excludes2: Arterial tortuosity syndrome (Q87.82)
Ehlers-Danlos syndrome (Q79.6)
Marfan syndrome (Q87.40–Q87.43)
other systemic involvement of connective tissue (M35)

Add
Use additional code, if applicable, to identify**:**
aortic aneurysm (I71.4–I71.9)
aortic dissection (I71.00–I71.03)
craniofacial anomalies (Q67.–)
other disorders involving the immune mechanism, not elsewhere classified (D89)
other specific joint derangements (M24)
scoliosis (M41)

Low Body Mass Index (BMI)

The University of California Davis Center for Healthcare Policy and Research has requested creation of a new code for Body Mass Index (BMI) less than 18.5. ICD-10-CM Category Z68 includes codes for BMI. Currently, the lowest BMI code is Z68.1 "Body mass index [BMI] 19.9 or less, adult". However, there is a need for further specificity for BMIs less than 19.9. ESPEN (the European Society of Clinical Nutrition and Metabolism) uses BMI<18.5 to define malnutrition.[1] CDC defines "underweight" as BMI <18.5.[2] WHO also defines "underweight" as BMI <18.5.[3]

Underweight (BMI < 18.5 kg/m2) is detrimental to population health. When compared to normal weight individuals (BMI 18.5-25 kg/m2), underweight individuals have significantly higher death rates with a Hazard Ratio of 2.27 and 95 percent confidence intervals (CI) = 1.78, 2.90.[4] Individuals with a BMI < 18.5kg/m2 have been shown to be at a higher risk for adverse events, postoperative infection, and/or mortality following a surgical procedure.[5-8] BMI below normal parameters is a risk factor for developing severe illness from respiratory infections such as influenza and COVID-19.[9,10] BMI below normal parameters can negatively impact both male and female fertility.[11,12]

Poor nutrition or underlying health conditions can result in underweight.[13] The National Health and Nutrition Examination Survey (NHANES) results from 2007-2010 indicate that women are more likely to be underweight than men.[13] However, all patients should be equally screened for underweight and followed up with nutritional counseling or another clinically appropriate intervention to reduce mortality and morbidity associated with underweight.

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TABULAR MODIFICATIONS

Z68 Body mass index [BMI]

Z68.1 Body mass index [BMI] 19.9 or less, adult

New code	Z68.18	Body mass index [BMI] 18.4 or less, adult
New code	Z68.19	Body mass index [BMI] 18.5-19.9, adult

Lynch Syndrome and Other Genetic Cancer Syndromes

This proposal is a repeat presentation related to Lynch syndrome, together with newly proposed codes for other genetic cancer risk conditions, including BRCA1 and BRCA2, and Li Fraumeni syndrome. This is based on a request from Fight Colorectal Cancer (Fight CRC) and its Genetics and Family History Advisory Council, which includes national leaders in the field of hereditary gastrointestinal cancers from multiple disciplines and institutions, with support from American Cancer Society, American College of Gastroenterology, American Society of Gastrointestinal Endoscopy, Collaborative Group of the Americas on Inherited Gastrointestinal Cancer, Lynch Syndrome Screening Network, and other groups, as well as individual experts in the field, with revisions based on comments and input from other sources. Since the cancer risks vary dramatically by gene, it has been proposed to create specific ICD-10-CM codes for these, and with particular interest in certain of these conditions which are common rather than rare.

Prior proposals to create codes for Lynch syndrome were presented in March 2024 and in September 2024. Further clinical details are available from the prior proposals. Certain information is repeated here for convenience, and some new information is included.

Lynch syndrome is the most common cause of inherited colorectal and endometrial cancer and accounts for about 4% of colorectal cancers and 3% of endometrial cancers. It is estimated to affect one in 279 people in the general population, which projects it to affect over 1.18 million people in the U.S. It is caused by pathogenic variants in genes in the DNA mismatch repair pathway that maintain fidelity during replication (including the four DNA mismatch repair genes *MLH1, MSH2, MSH6,* and *PMS2,* along with *EPCAM*). Deletions in the *EPCAM* gene lead to silencing *MSH2* and thus causing Lynch syndrome.[1-3] These are inherited in an autosomal dominant fashion.

Lynch syndrome is characterized by increased risk of cancer in multiple organ systems. The lifetime cancer risks vary dramatically by gene and includes elevated risks of colorectal cancer up to 46-61%, endometrial cancer up to 34-54%, ovarian cancer up to 8-38%, urinary tract cancer up to 2.2 - 28%, gastric cancer up to 9% as well as increased but less defined risks of small bowel, pancreas, biliary tract, and prostate cancer.

For those with MLH1 Lynch syndrome and MSH2 Lynch syndrome, as well as EPCAM Lynch syndrome, colorectal cancer screening is recommended every one to two years starting at age 20 to 25 years, or for those with family history earlier than 25 years, starting at an age two to five years earlier than the youngest such history. [1,2] For those with MSH6 Lynch syndrome and PMS2 Lynch syndrome, colorectal cancer screening can be delayed until 30 to 35 years of age, and done every one to three years, provided that there is no family history of earlier onset cancer.[1,2] Other specific cancer screening is recommended to be performed starting at specific ages, as determined based on the specific genetic type of Lynch syndrome, together with the known family history for the individual.[1,2]

Hereditary breast and ovarian cancer syndrome is a genetic syndrome with increased risk for developing certain specific cancers.[4] "Breast and ovarian cancer are present in several autosomal dominant cancer syndromes, although they are most strongly associated with highly penetrant germline pathogenic variants in *BRCA1* and *BRCA2*."[5] The prevalence of a germline pathogenic variant in *BRCA1* or *BRCA2* is

approximately 1 in 400 to 1 in 800 individuals in the general population.[5] This projects to a range of between 412,000 to 825,000 people affected in the U.S. As well as being associated with breast and ovarian cancer in females (including early-onset breast cancer), pathogenic *BRCA1* and *BRCA2* mutations are also associated with fallopian tube cancer, primary peritoneal cancer, male breast cancer, prostate cancer, and pancreatic cancer.[6,7]

Li-Fraumeni syndrome (LFS) is a cancer disorder characterized by premenopausal breast cancer, sarcoma, brain tumors, leukemia, and adrenocortical carcinoma, with pathogenic germline *TP53* mutations and autosomal dominant inheritance.[4,5] It has generally been thought to be rare, and previous general population prevalence estimates ranged from 1 in 5,000 to 1 in 20,000.[7,8] This would correspond to a projected range of 16,500 to 66,000 people affected in the U.S. However, a 2017 study found 34 different potentially pathogenic TP53 variants in 131/63,983 individuals,[8] thus about 1 in 488 people. This would project to LFS potentially affecting just over 675,000 people in the U.S.

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TABULAR MODIFICATIONS

Chapter 17
Congenital malformations, deformations, chromosomal abnormalities, and genetic disorders (Q00-QA0)

This chapter contains the following blocks:
…

Revise — QA0-QA1 Genetic disorders, not elsewhere classified

Revise — Genetic disorders, not elsewhere classified (QA0-QA1)

New category — QA1 Genetic disorders associated with neoplasms, not elsewhere classified

Add — Code also, if applicable, any associated conditions, such as:
Add — genetic susceptibility to malignant neoplasm by site (Z15.0-)
Add — malignant neoplasms (C00.0-C96.9)
Add — personal history of malignant neoplasm (Z85.-)

Add — Excludes2: Multiple endocrine neoplasia [MEN] syndromes (E31.2-)

New subcategory — QA1.7 Inherited neoplasm predisposition syndromes involving multiple systems, not elsewhere classified

New sub-subcategory — QA1.71 Lynch syndrome
Add — Hereditary nonpolyposis colorectal cancer susceptibility

New code — QA1.710 Lynch syndrome due to MLH1

New code — QA1.711 Lynch syndrome due to MSH2
Add — Lynch syndrome due to EPCAM (with MSH2 silencing)

New code — QA1.718 Other Lynch syndrome
Add — Lynch syndrome due to MSH6
Add — Lynch syndrome due to PMS2

New code — QA1.719 Lynch syndrome, unspecified

New sub-subcategory	QA1.79	Other inherited neoplasm predisposition syndrome of multiple systems	
New code		QA1.790	Familial cancer syndrome with pathogenic BRCA1 mutation
Add			BRCA1-cancer predisposition syndrome
Add			Hereditary breast and ovarian cancer syndrome with pathogenic BRCA1 mutation
New code		QA1.791	Familial cancer syndrome with pathogenic BRCA2 mutation
Add			BRCA2-cancer predisposition syndrome
Add			Hereditary breast and ovarian cancer syndrome with pathogenic BRCA2 mutation
New code		QA1.792	Li Fraumeni syndrome
New code		QA1.798	Other inherited neoplasm predisposition syndrome of multiple systems

Medetomidine Withdrawal Syndrome

Medetomidine is an alpha-2-agonist veterinary sedative medication that has been detected alongside illicitly manufactured fentanyl (IMF) with serious clinical consequences for people who use drugs (PWUD).[1–3] In May 2025, the Centers for Disease Control and Prevention (CDC) published two Morbidity and Mortality Weekly Reports (MMWR) characterizing the emerging syndrome of medetomidine withdrawal.[1,2] This syndrome is characterized by severe autonomic dysfunction, such as severe hypertension and tachycardia, as well as nausea, vomiting, tremor, and altered mental status.[1,2] Almost all patients suspected of having Medetomidine Withdrawal Syndrome required admission to an intensive care unit and nearly a quarter were intubated.[1,2] Accurate diagnosis and documentation of Medetomidine Withdrawal Syndrome is fundamental to improving treatment and health outcomes for PWUD, as well as monitoring the clinical implications of the introduction of medetomidine to IMF, but is not yet available. This proposal addresses the need for a diagnostic code for Medetomidine Withdrawal Syndrome.

Medetomidine is a sedative medication, but characterization of Medetomidine Withdrawal Syndrome does not align with diagnostic criteria for sedative withdrawal. Diagnostic criteria for sedative withdrawal includes developing two or more of the following cessation or reduction in use: autonomic hyperactivity, hand tremor, insomnia, nausea or vomiting, transient hallucinations or illusions, psychomotor agitation, anxiety, or grand mal seizures.[4] The coding note in the Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition, uses the ICD 10-CM code F13.239 for sedative withdrawal without perceptual disturbances, which is the closest diagnostic code for Medetomidine Withdrawal Syndrome but is not appropriate due to the ways this differs from other sedative withdrawal.[4] First, Medetomidine Withdrawal Syndrome has not been associated with seizure activity.[1–3,5] Second, Medetomidine Withdrawal Syndrome has been associated with altered mental status that may be secondary to Posterior Reversible Encephalopathy Syndrome (PRES), but has not been associated with hallucinations or illusions.[1,3] Third, Medetomidine Withdrawal Syndrome has been associated with end organ damage, such as PRES and Non-ST Elevation Myocardial Infarction, in the setting of severe hypertension which is not a characteristic of other sedative withdrawal syndromes. Lastly, the tremor associated with Medetomidine Withdrawal Syndrome has not been described as “hand tremor”.[1–3] Withdrawal is a criterion for diagnosing substance use disorders.[4] Inaccurate coding of Medetomidine Withdrawal Syndrome may lead to inaccurate diagnosis of substance use disorders and limit the ability of clinicians to provide appropriate treatment.

Medetomidine was first detected in Philadelphia’s illicit fentanyl supply in May, 2025, and recognized by the Philadelphia Department of Public Health (PDPH) to be associated with a severe withdrawal syndrome in December, 2025.[3,6] Since the introduction of medetomidine to Philadelphia’s IMF supply the number of people presenting to the emergency department for withdrawal has more than doubled.[7] The efficacy of withdrawal management in Philadelphia’s health systems has been shown to decrease with the

introduction of medetomidine, which has required updates to withdrawal protocols to effectively treat Medetomidine Withdrawal Syndrome.[8] Accurate diagnostic coding for Medetomidine Withdrawal Syndrome is needed to continue to monitor changes in withdrawal presentation and improve treatment protocols.

Prior to medetomidine being detected, xylazine was the most prevalent alpha-2-agonist veterinary sedative medication in Philadelphia's illicit fentanyl supply. In 2023, nearly 100% of illicit fentanyl in Philadelphia contained xylazine.[9] At the end of 2024, xylazine was present in 30-40% of illicit fentanyl and medetomidine was present in over 70% of illicit fentanyl.[3,10] Medetomidine is rapidly proliferating across the United States, and is present in the illicit fentanyl supply in several states.[11] Accurate diagnostic coding for Medetomidine Withdrawal Syndrome is fundamental to provide appropriate clinical care for PWUD, and provides a critical public health tool for surveillance of clinical implications of a changing IMF supply in the United States.

In addition to a unique withdrawal syndrome, medetomidine use has also presented changes in overdose presentations. Medetomidine involved opioid overdoses have been characterized by prolonged sedation and bradycardia.[12,13] A case series of 11 patients who presented with medetomidine involved opioid overdoses in Philadelphia described a median (interquartile range [IQR]) lower heart rate of 41 beats/minute (38.5-40) for a median (IQR) duration of 3.4 hours (2.7-9.3). This is consistent with MMWR of 38 patients who presented with medetomidine involved overdoses in Chicago where 87% had bradycardia. Prolonged sedation in opioid overdoses involving medetomidine has been described as sedation that does not respond to the opioid overdose reversal agent, naloxone.[13] Bradycardia and sedation not responsive to naloxone are symptoms of medetomidine toxicity that require changes to overdose reversal strategies. A modification to ICD-10 to include medetomidine toxicity will ensure accurate capture of medetomidine involved opioid overdoses, as well as appropriate overdose reversal response.

Daniel Teixeira da Silva, MD, MSHP of the Philadelphia Department of Public Health's Division of Substance Use Prevention and Harm Reduction Division is requesting the following tabular modifications for monitoring and tracking Medetomidine Withdrawal Syndrome.

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TABULAR MODIFICATIONS

F13 Sedative, hypnotic, or anxiolytic related disorders
F13.2 Sedative, hypnotic, or anxiolytic dependence with withdrawal
F13.23 Sedative, hypnotic, or anxiolytic dependence with withdrawal

New code — F13.238 Sedative, hypnotic, or anxiolytic dependence with withdrawal with other disturbances

New subcategory — F13.A Specific sedative, hypnotic, or anxiolytic drugs causing related disorders

New code — F13.A1 Medetomidine causing related disorders

Add — Code first if applicable, for medetomidine withdrawal related to dependence (F13.238)

Add — Use additional code to identify associated manifestations, such as:
- drug-induced tremor (G25.1)
- hypertensive crisis (I16)
- non-ST elevation (NSTEMI) myocardial infarction (I21.4)
- posterior reversible encephalopathy syndrome (I67.83)
- tachycardia NOS (R00.0)
- vomiting (R11.10)

T65 Toxic effect of other and unspecified substances
The appropriate 7th character is to be added to each code from category T65
A-initial encounter
D-subsequent encounter
S-sequela

T65.8 Toxic effect of other specified substances

New sub-subcategory — T65.85 Toxic effect of Medetomidine

Add — Use additional code to identify associated manifestations, such as:
- bradycardia NOS (R00.1)
- somnolence, stupor and coma (R40)

New code — T65.851 Toxic effect of medetomidine, accidental (unintentional)

Add — Toxic effect of medetomidine NOS

New code — T65.852 Toxic effect of medetomidine, intentional self-harm

New code — T65.853 Toxic effect of medetomidine, assault

New code — T65.854 Toxic effect of medetomidine, undetermined

Metabolic Dysfunction and Alcohol Associated Liver Disease

An addendum was proposed at the March 2024 Coordination and Maintenance meeting to add two new inclusion terms for existing nonalcoholic liver disease classifications, creating inclusion terms and indexing to support both MASH (metabolic dysfunction-associated steatohepatitis) and MASLD (metabolic dysfunction-associated steatotic liver disease).

One of the newly adopted terms from the global NAFLD nomenclature steering committee was not included: Metabolic dysfunction- and alcohol-associated liver disease (MetALD). MetALD applies to patients with liver steatosis, at least one metabolic risk factor, and a history of moderate alcohol intake. This category recognizes that liver steatosis can result from both metabolic factors and alcohol consumption. Defined as MASLD with moderate alcohol consumption (30-60 g/day for men and 20-50 g/day for women), MetALD currently lacks an ICD-10-CM code due to its recent identification. The consensus suggests coding MetALD using the ICD-10-CM for either MASLD or alcoholic liver disease (ALD), depending on the predominant factor in each case.

The Permanente Federation is requesting the following tabular modification to differentiate specific types of liver diseases based on the new nomenclature.

TABULAR MODIFICATIONS

K70 Alcoholic liver disease

New subcategory

K70.8 Other alcoholic liver disease

New code

K70.81 Alcoholic liver disease, not elsewhere classified
Add
Metabolic dysfunction- and alcohol associated liver disease (MetALD)

New code

K70.89 Other specified alcoholic liver disease

K76 Other diseases of liver

K76.0 Fatty (change of) liver, not elsewhere classified
Nonalcoholic fatty liver disease (NAFLD)
Metabolic dysfunction-associated steatotic liver disease (MASLD)

Nipple Ischemia and Nipple Necrosis

Nipple ischemia and nipple necrosis can happen after surgical procedures for the management of patients with breast cancer. [1,2] Since the late 1990's, attention to preserving the nipple areolar complex for optimal cosmetic outcomes has been explored with increasing popularity and is now a routine procedure in many centers. [3,4] Loss of blood flow to the nipple areolar complex from surgery (nipple necrosis) is one of the most common complications of these operations. [5,6,7] Nipple ischemia, which is insufficient blood flow to a lesser degree, or nipple necrosis (more substantial loss of blood flow resulting in tissue loss) can result in significant healthcare resource utilization for the purpose of nipple areolar complex salvage and/or management of related complications including medications, interventions with hospital stay (hyperbaric oxygen therapy), angiography and subsequent surgical procedures for debridement and/or nipple resection with or without nipple reconstruction. In the surgical field, extensive time, discussion, and research has been dedicated to defining the rates of nipple necrosis in these operations. [5,7,8,9,10] As future cosmetic procedures emerge and while we evaluate our current surgical approaches for safety and feasibility, monitoring rates of nipple ischemia and/or nipple necrosis for surgical quality improvement is essential.

This proposal was presented at the September 2024 ICD-10 Coordination and Maintenance Meeting. In response to public comments, changes have been made.

Nipple ischemia and necrosis rates for nipple sparing mastectomy range from 0-48%. [5,6] Nipple necrosis for other procedures (lumpectomy, female to male chest masculinization for gender re-assignment and others) can occur in as high as 20% of procedures. [11,12]

Currently, impaired blood flow to the nipple areolar complex is only captured under unspecified ICD-10-CM codes. N64 defined as "other disorders of the breast" with N64.5 indicating "other signs and symptoms in breast." N64.9 encompasses "nipple disorder" but this implies a disorder of the nipple inherent to the patient, not a result of iatrogenic interventions. Whether the nipple areolar complex is categorized as breast or skin, there is no defined ICD-10-CM code currently to represent impaired blood flow to the nipple areolar complex. A modification to the ICD-10-CM code set is needed to describe nipple ischemia and nipple necrosis to align clinical documentation, support effective and granular disease tracking, and to facilitate accurate monitoring of diagnosis and longitudinal patient management.

The proposal was submitted by Intuitive Surgical; a biotechnology company. The American Society of Plastic Surgeons has reviewed and supports the proposal.

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TABULAR MODIFICATIONS

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 sub-subcategory
N99.86 Intraoperative and postprocedural complications and disorders of breast

New code
N99.860 Intraoperative and postprocedural nipple ischemia

New code
N99.861 Intraoperative and postprocedural nipple necrosis

Nonsegmental and Segmental Vitiligo

Vitiligo is an autoimmune skin disorder characterized by a selective loss of melanocytes, resulting in depigmented, white patches on the skin.[1] Vitiligo is traditionally classified as nonsegmental vitiligo (NSV) or segmental vitiligo (SV). NSV and SV are clinically distinct subtypes of vitiligo, differing in presentation, underlying mechanisms, and associated comorbidities.

The defining feature of NSV (also called generalized vitiligo) is its bilateral and often symmetrical distribution of depigmented patches. NSV usually appears later in life,[2] involves a higher number of lesions,[3] affects a larger body surface area,[3] and is more associated with autoimmune comorbidities, particularly thyroid disorders.2-4 NSV is driven by systemic autoimmunity, including reduced regulatory T cells.[5]

SV is unilateral and often asymmetrical in distribution, typically with an early onset,[2,6] fewer lesions,[3] and lower body surface area involvement.[3] It stabilizes rapidly and shows lower rates of Koebnerization.[2,3] SV is associated with localized cytotoxic immune responses and minimal systemic autoimmunity.[5] Family history is less common in SV compared to NSV.[2]

These differences are clinically important for diagnosis, prognosis, and guiding treatment, but in the current ICD-10-CM code set, NSV and SV are not specifically identified. When coded, they are placed under the general category of L80, Vitiligo. This limitation hinders the ability to tailor treatment based on disease subtype and restricts the collection of reliable epidemiological data.

Roopal Kundu, MD, Professor of Dermatology and Vineeth Vaidyula are requesting the following new codes to distinguish the two types of vitiligo. This proposal has been reviewed and supported by the American Academy of Dermatology.

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TABULAR MODIFICATIONS

L80 Vitiligo
Excludes2: vitiligo of eyelids (H02.73-)
vitiligo of vulva (N90.89)

New code	L80.A	Nonsegmental vitiligo
Add		Generalized vitiligo
New code	L80.B	Segmental vitiligo
New code	L80.C	Vitiligo, unspecified
Add		Vitiligo NOS

Obesity Due to Disruption of MC4R Pathway

Rhythm Pharmaceuticals, Inc. has proposed the creation of new ICD-10-CM diagnosis codes for further describing the different presentations of obesity due to MC4R pathway disruption: acquired hypothalamic obesity, congenital hypothalamic obesity, syndromic obesity and rare monogenic forms of obesity due to disruption of the MC4R pathway. The latter can result from mutations in the following genes: leptin (LEP), leptin receptor (LEPR), proopiomelanocortin (POMC), proprotein convertase subtilisin/kexin type 1 (PCSK1), Melanocortin 4 receptor (MC4R), Src homology 2B adaptor signaling protein 1 (SH2B1) or Nuclear Receptor Coactivator 1 (NCOA1).

This proposal is related to a prior proposal from March 2023, monogenic forms of obesity, which resulted in creation of the single code E88.82, Obesity due to disruption of MC4R pathway. It is also related to prior proposals regarding hypothalamic obesity, presented in March 2024 and September 2024.

By way of additional background, general obesity is the most common form of obesity and results from a combination of lifestyle, diet, and environmental factors. However, general obesity is distinct from MC4R related obesity of which there are distinct forms.

Acquired hypothalamic obesity can result from physical damage to the hypothalamus where the MC4R pathway is mostly located. This includes instances where the hypothalamus is damaged due to the presence of tumors, treatment of these tumors, traumatic brain injury or stroke.

Syndromic obesity includes Prader-Willi Syndrome, which is caused by gene imprinting errors and Bardet Beidl Syndrome which is multisystemic and involves clinical manifestations in multiple organ systems.

Congenital hypothalamic obesity occurs when the physical damage to hypothalamus formation is congenital in nature due to inborn errors during in utero development, such as for example in septo-optic dysplasia that leads to obesity.

Monogenic obesity was described in detail in the March 2023 ICD-10 Coordination and Maintenance Committee Meeting proposal titled, “Monogenic Forms of Obesity.” Please reference that for detailed information related to this and the genes noted above. Prevalence information related to each of these was included there also supportive of creating more detailed codes.

The connection between hypothalamic obesity and MC4R pathway disruption is supported by converging evidence from neurobiology, clinical imaging, and pharmacologic trials. Both genetic MC4R deficiency and acquired hypothalamic injury result in similar phenotypes: early-onset obesity, hyperphagia, and decreased basal metabolic rate. This phenotypic overlap reinforces the role of MC4R dysfunction in hypothalamic obesity. Damage to hypothalamic nuclei that regulate MC4R signaling — whether by injury or disease — disrupts appetite regulation and metabolic rate, leading to the persistent obesity seen in hypothalamic obesity (HO). There is also direct functional evidence that restoring MC4R activity can ameliorate HO symptoms, in that the drug setmelanotide, an MC4R agonist (FDA-approved in 2020 for certain types of monogenic obesity), has demonstrated significant reductions in hunger and weight in

patients with HO. In addition, functional imaging in HO patients shows blunted hypothalamic activation in response to feeding cues. This suggests impaired MC4R-mediated satiety signaling.

TABULAR MODIFICATIONS

E88 Other and unspecified metabolic disorders

E88.8 Other specified metabolic disorders

E88.82 Obesity due to disruption of MC4R pathway

Add — Code also obesity class, if known (E66.81-)

New code — E88.820 Acquired hypothalamic obesity due to disruption of MC4R pathway

Add — Code also, if applicable, specific associated disorder or injury associated, such as:

Add — hypothalamic dysfunction, not elsewhere classified (E23.3)

Add — malignant neoplasm of brain (C71.-)

Add — other sequelae of cerebral infarction (I69.398)

Add — personal history of irradiation (Z92.3)

Add — postsurgical status (Z98.890)

Add — sequela of focal traumatic brain injury (S06.3- with seventh character S)

New code — E88.821 Congenital hypothalamic obesity due to disruption of MC4R pathway

Add — Code first causative congenital disorder, such as:

Add — septo-optic dysplasia of brain (Q04.4)

New code — E88.822 Syndromic obesity due to disruption of MC4R pathway

Add — Code first causative syndrome, such as:

Add — Bardet-Biedl syndrome (Q87.83)

Add — Prader-Willi syndrome (Q87.11)

New code — E88.823 Monogenic obesity due to disruption of MC4R pathway

New code — E88.829 Obesity due to disruption of MC4R pathway, unspecified

Add — Hypothalamic obesity, NOS

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INDEX MODIFICATIONS

	Obesity E66.9
Revise	- due to disruption of MC4R pathway E88.823
Revise	- due to gene mutation leptin (LEP) E88.823
Revise	- due to gene mutation leptin receptor (LEPR) E88.823
Revise	- due to gene mutation melanocortin 4 Receptor (MC4R) E88.823
Revise	- due to gene mutation nuclear receptor coactivator 1 (NCOA1) E88.823
Revise	- due to gene mutation proopiomelanocortin (POMC) E88.823
Revise	- due to gene mutation proprotein convertase subtilisin/kexin type 1 (PCSK1) E88.823
Revise	- due to gene mutation src homology 2B adaptor signaling protein (SH2B1) E88.823

Odontogenic Sinusitis

This topic was presented at the September 2024 ICD 10 Coordination and Maintenance meeting. Based on public comments, revisions to the proposal have been made for reconsideration. Changes are indicated in **bold**.

Odontogenic sinusitis (ODS) refers to bacterial maxillary sinusitis, with or without extension to other paranasal sinuses, secondary to either adjacent infectious maxillary dental pathology, or iatrogenic injury from dental procedures.[1] ODS is distinct from all other forms of rhinosinusitis, and despite being more common than previously thought, ODS is still underrecognized and has no ICD-10-CM diagnostic code. While previously reported to represent 10% of sinusitis, more recent studies suggest it represents 25-40% of patients with maxillary sinusitis.[2,3] More importantly, ODS usually presents unilaterally as opposed to most rhinosinusitis being bilateral, and 45-75% of unilateral sinusitis has been shown to be due to ODS.[4-6] Unfortunately, ODS only represents about 1% of the sinusitis literature over the last 20 years,[7] and therefore it has not been represented adequately in national and international sinusitis guidelines.[8,9]

ODS management depends on whether or not there is treatable infectious dental pathology. If there is no treatable dental pathology, antibiotics can be attempted, but patients will still usually require endoscopic sinus surgery to drain the infection. If there is treatable dental pathology, it can be controversial whether to treat the teeth or sinuses first. While the infectious dental source will need to be treated, this only resolves the purulent sinusitis in 40-60% of cases, and therefore many of these patients require both dental treatment and sinus surgery.[10,11]

Dr. John R. Craig and Dr. Alberto M. Saibene are rhinologists (subspecialty-trained otolaryngologists) who have been studying multiple aspects of ODS clinically and translationally, and these works have led to recent multidisciplinary national and international consensus statements on diagnosis and management.[1,10] Awareness of ODS prevalence amongst otolaryngologists and dental providers has been increasing, but many clinicians, both medical and dental, are still either unaware of the condition or of the optimal diagnostic and therapeutic interventions.

One major issue is that while higher quality evidence has been mounting on ODS in the last 5-10 years, the lack of a unique ICD-10-CM code has hindered both research pursuits and patient care. While ODS can present acutely (J01.0-J01.9), chronically (J32.0-J32.9), possibly as recurrent acute sinusitis (J01.01-J01.91), and possibly with nasal polyps (J33), none of the aforementioned codes capture the odontogenic cause of the sinusitis. Evidence to date suggests that the treatment of ODS does not change based on duration of symptoms (acute or chronic). It is most important that providers recognize the ODS condition.

Rhinologists, Dr. John R. Craig and Dr. Alberto M. Saibene are requesting new ICD-10-CM codes to distinguish ODS from other forms of sinusitis and to help facilitate future research for patient care.

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TABULAR MODIFICATIONS

J34 Other and unspecified disorders of nose and nasal sinuses

J34.8 Other and unspecified disorders of nose and nasal sinuses

New sub-subcategory	**J34.83** Odontogenic sinusitis
New code	**J34.830** Odontogenic sinusitis, maxillary sinus
New code	**J34.831** Odontogenic sinusitis, ethmoid sinus
New code	**J34.832** Odontogenic sinusitis, frontal sinus
New code	**J34.833** Odontogenic sinusitis, sphenoid sinus
New code	**J34.839** Odontogenic sinusitis, unspecified
Add	Odontogenic sinusitis NOS

Okur-Chung Neurodevelopmental Syndrome (OCNDS)

Okur-Chung Neurodevelopmental Syndrome (OCNDS) is a genetic, autosomal-dominant disorder with specific potential findings that may affect multiple areas. This proposal is based on a request from the CSNK2A1 Foundation, which is a not-for-profit organization composed of volunteers, advocates, and researchers focused on finding a cure for OCNDS.

"OCNDS typically presents with developmental delays, mild- to- moderate intellectual impairment, hypotonia, . . . distinctive facial features [(facial dysmorphism characterized by a round face and short, broad nasal tip)], speech delay" or the inability to speak, cortical malformations, and low muscle tone (hypotonia).[1-4] Individuals also frequently have gastrointestinal issues, like constipation, and feeding and swallowing difficulties.[2] OCNDS is also associated with learning disability and autism spectrum disorder.[5] "Less common findings may include kyphoscoliosis, postnatal short stature, disrupted circadian rhythm leading to sleep disturbance, seizures, and poor coordination."[2] "Developmental delay[s] affect[] all areas of development, but language is more impaired than gross motor skills in most individuals."[2] Data also suggests that OCNDS manifests with unusual primary teeth, contributing to poor oral health.[6]

OCNDS is caused by heterozygous mutation in the *CSNK2A1* gene localized on chromosome 20p13.1.[5, 7] The *CSNK2A1* gene encodes the alpha subunit of protein kinase (enzyme) CK2, a well-characterized and conserved serine/threonine protein kinase[8] that is important for development of neurons and synaptic transmission in the brain.[9] The *CSNK2A1* mutations associated with OCNDS lead to missense or deletion/truncating variants in the encoded protein; with 56 different missense mutations identified to date in association with OCNDS.[10] OCNDS has been most frequently associated with the mutation of lysine-198 to arginine (K198R), which has been speculated to result in a loss of kinase function and shown to result in substantial changes to kinase specificity for CK2.[8] 64 patients with the K198R variant have registered with the Foundation.[10] Individuals with a null variant of *CSNK2A1* have been observed to exhibit a milder phenotype compared to patients with missense variants, displaying reduced symptoms like language impairment, dysmorphic facial features, and intellectual disability.[1]

OCNDS is diagnosed through molecular genetic testing, most commonly through genetic blood tests such as Whole Exome Sequencing (WES) or Whole Genome Sequencing (WGS).[11] Although the exact prevalence of OCNDS is not currently known (in part due to the lack of adequate tracking due to the absence of unique diagnosis codes), it has been estimated to have a prevalence of 1 in every 100,000 people, and the number of diagnoses has risen rapidly in recent years, since the disorder was first identified in 2016 and especially as WES is increasingly utilized worldwide.[12] Additionally, the CSNK2A1 Foundation has partnered with Simons Searchlight on a natural history study in an effort to develop clinical consensus guidelines that is actively searching for additional patients and would benefit from a unique diagnosis code.

A specific diagnosis code would help researchers reach OCNDS patients and better understand what types of treatment and management work best to help patients lead rich and fulfilling lives. While there are dedicated researchers that have made breakthroughs in OCNDS research, there is also much left to learn on the condition's incidence, characteristics, and treatment. Many of the patients that researchers have

identified are young children, and a diagnosis code would allow clinicians to track patients' development and clinical outcomes throughout their lifetime.

Currently, there is no cure for OCNDS, and clinicians are limited to treating the manifestations of the condition. Treatment typically includes a combination of physical, occupational, and behavioral health therapy, along with pharmacologic treatments for common OCNDS-related conditions (e.g. standard anti-seizure treatment for epilepsy), and management of common OCNDS-related physical abnormalities (e.g. scoliosis, congenital heart defects, renal anomalies).

There is also an especially heightened need for a unique diagnosis code in light of ongoing studies regarding anticipated OCNDS-specific pharmacological interventions. Currently, the CSNK2A1 Foundation has two ongoing drug repurposing studies to identify treatment options for patients, and creation of a unique diagnosis code will help enable researchers to reach patients that could benefit from potential clinical breakthroughs.[13, 14] The CSNK2A1 Foundation is currently anticipating availability of repurposed small molecule drugs for OCNDS-specific therapeutic intervention as early as 2025, and a unique diagnosis code will facilitate tracking and intervention for purposes of these anticipated treatments.

Surveillance of OCNDS is also important for effective management, including measuring growth and nutritional status, neurologic changes, developmental progress and educational needs, and assessing behavioral tendencies.[2] A diagnosis code will assist with tracking for such treatment purposes and will also help to facilitate research that could ultimately result in the development of a therapy targeting the root genetic cause of OCNDS. A specific code is vital to supporting existing research initiatives into OCNDS, helping OCNDS patients get the treatment and support they need, and discovering new OCNDS breakthroughs.

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TABULAR MODIFICATIONS

	QA0	Neurodevelopmental disorders related to specific genetic pathogenic variants	
New subcategory	QA0.7	Neurodevelopmental disorders related to pathogenic variants in specific genes associated with proteins with other and multiple functions	
New sub-subcategory	QA0.70	Neurodevelopmental disorders related to pathogenic variants in certain specific genes associated with proteins with other and multiple functions	
New code	QA0.701	Okur-Chung neurodevelopmental syndrome	

Pediatric Healthcare: Impact of Parental Mental Health

Children whose parents struggle with mental health conditions are at heightened risk for developmental, emotional, and behavioral challenges. These children are often affected not only by the symptoms of a parent's condition but also by the caregiving environment shaped by that condition. The impact can be particularly pronounced during the perinatal period, when secure attachment and early relational health are critical to a child's lifelong development.

Despite the well-established risks, the ICD-10-CM classification system does not adequately capture the specific challenges faced by children of parents with mental health conditions—particularly those affected by perinatal mood disorders such as postpartum depression and psychosis. Existing codes focus almost exclusively on the adult, leaving a substantial gap in documenting the pediatric implications of these parental diagnoses.

This gap hinders clinicians' ability to:

- Identify children at risk before symptoms appear,
- Justify early intervention strategies (distinct from formal Early Intervention services),
- Coordinate care across providers,
- And collect data needed to inform public health strategies.

Children of parents with postpartum depression, for example, are at increased risk for delayed development, emotional dysregulation, behavioral problems, and impaired attachment. Some of these children may ultimately qualify for Early Intervention services if delays manifest.

However, early intervention—preventive action taken *before* delays or dysfunction emerge—is critical for reducing the likelihood of those outcomes in the first place. Without a specific pediatric code, children living in high-risk contexts often go unrecognized by systems designed to support them.

Perinatal mood disorders are far too common to be ignored in pediatric risk assessments. Approximately 15–20% of new mothers experience perinatal depression (Rafferty et al.), and many more experience related conditions such as anxiety, postpartum psychosis, or postpartum blues. These disorders have direct implications for children's health and development, including increased risk of:

- Abuse or neglect
- Feeding and breastfeeding challenges,
- Poor early parent-infant bonding,
- Disruptions in physical, emotional, cognitive, and social development (Rafferty et al.).

To address this gap, the creation of pediatric-specific ICD-10-CM codes that would capture a child's risk due to parental mental health conditions, particularly those on the Perinatal Depression Spectrum. These codes would not indicate a diagnosis in the child but would instead identify that they are living in a context of elevated developmental and psychosocial risk—a crucial distinction for prevention-based pediatric care.

Such codes would:

- Enable pediatricians to document observed or reported parental mental health conditions that may affect the child;
- Alert other providers to monitor for developmental or behavioral concerns;
- Justify referrals not only to Early Intervention when delays are identified, but also to early intervention supports—such as parenting education, dyadic therapy, and home visiting—*before* symptoms arise;
- Facilitate interprofessional collaboration between pediatricians, mental health providers, and social services;
- Allow healthcare systems and to track prevalence and allocate necessary support services accordingly.

This addition would support the broader pediatric mission of prevention, early detection, and equity. Clinicians need the ability to code for the real-world contexts that shape child health—especially when those contexts are clinically significant but not yet diagnosable within the child.

This proposal has been submitted by Katherine Minaya MD, MST, FAAP of the Bronx Health Center and Foster Care Services. The proposal has been reviewed and supported by the American Academy of Pediatrics.

TABULAR MODIFICATIONS

Z62 Problems related to upbringing
Includes: current and past negative life events in childhood
current and past problems of a child related to upbringing
Excludes2: maltreatment syndrome (T74.-)
problems related to housing and economic circumstances (Z59.-)

New subcategory Z62.A Child at risk due to parent mental (behavioral) health condition
Exclude2: substance addiction in family (Z63.72)

New sub-subcategory Z62.A1 Child at risk due to biological parent mental (behavioral) health condition

New code Z62.A10 Child at risk due to biological parent mental (behavioral) health condition

New code	Z62.A11 Child at risk due to biological parental substance use
Add	Child at risk due to biological parental prescription medication use
New code	Z62.A12 Child at risk due to biological parental alcohol use
New code	Z62.A13 Child at risk due to maternal post-partum depression or another post-partum mood disorder
New code	Z62.A18 Child at risk due to biological parental with other mental (behavioral) health condition
New code	Z62.A19 Child at risk due to biological parental unspecified mental (behavioral) health condition
New sub-subcategory	Z62.A2 Child at risk due to adopted parent mental (behavioral) health condition
New code	Z62.A20 Child at risk due to adopted parent mental (behavioral) health condition
New code	Z62.A21 Child at risk due to adopted parental substance use
Add	Child at risk due to adopted parental prescription medication use
New code	Z62.A22 Child at risk due to adopted parental alcohol use
New code	Z62.A28 Child at risk due to adopted parental or other mental (behavioral) health condition
New code	Z62.A29 Child at risk due to adopted parental unspecified mental (behavioral) health condition
New sub-subcategory	Z62.A3 Child at risk due to foster parent mental (behavioral) health condition
New code	Z62.A30 Child at risk due to foster parent mental (behavioral) health condition

New code	Z62.A31 Child at risk due to foster parental substance use
Add	Child at risk due to foster parental prescription medication use
New code	Z62.A32 Child at risk due to foster parental alcohol use
New code	Z62.A38 Child at risk due to foster parental other mental (behavioral) health condition
New code	Z62.A39 Child at risk due to foster parental unspecified mental (behavioral) health condition
New sub-subcategory	Z62.A4 Child at risk due to step-parent mental (behavioral) health condition
New code	Z62.A40 Child at risk due to step-parent mental (behavioral) health condition
New code	Z62.A41 Child at risk due to step-parent substance use
Add	Child at risk due to step-parent prescription medication use
New code	Z62.A42 Child at risk due to step-parent alcohol use
New code	Z62.A48 Child at risk due to step-parent other mental (behavioral) health condition
New code	Z62.A49 Child at risk due to step-parent unspecified mental (behavioral) health condition
New sub-subcategory	Z62.A5 Child at risk due to non-parental relative mental (behavioral) health condition
New code	Z62.A50 Child at risk due to non-parental relative mental (behavioral) health condition
New code	Z62.A51 Child at risk due to non-parental relative substance use
Add	Child at risk due to non-parental relative prescription medication use
New code	Z62.A52 Child at risk due to non-parental relative alcohol use

New code	Z62.A58 Child at risk due to non-parental relative with other mental(behavioral) health condition
New code	Z62.A59 Child at risk due to non-parental relative unspecified mental (behavioral) health condition

Pediatric Healthcare: Impact of Parental Social Circumstance

Children with adolescent parents face challenges that are often different than older parents may face. The families are “at increased risk for medical, psychological, developmental, and social problems” (Powers et al.). Arguably, growing up with a teen parent is an Adverse Childhood Experience (ACE) as it can have long-lasting consequences for a child's education as well as physical and mental health. It is of great clinical value to capture this information, and a unique ICD-10-CM code is being requested.

Children born to teen mothers may experience higher rates of illness, mortality, and low birth weight due to factors like prenatal care, nutrition, and substance use. Children of teen parents often face challenges in education, including lower academic achievement and higher dropout rates.

Teen parents may struggle with emotional maturity, leading to difficulties in forming stable relationships and providing consistent emotional support. Teen parents may lack the social support and resources necessary for successful parenting, leading to increased stress and challenges. Teen parents are more likely to experience mental health problems, such as depression, anxiety, and substance abuse, as compared to their nonparent counterparts, which can significantly impact their child’s emotional and social environment. Lack of parenting skills can put the children of teen parents at a higher risk of neglect or abuse.

More likely to live in poverty, teen parents are more likely to experience limited access to resources, healthcare, and educational opportunities. Moreover, growing up with a teen parent can mean an increased risk for chronic health conditions like obesity, heart disease, diabetes, and mental health issues later in life.

There is currently no ICD-10-CM code that adequately captures that the patient (child) is at risk as a direct result of having an adolescent parent. This can impact pediatric clinicians’ capacity to adequately document risk factors, assess for qualification for and connect to resources. These then lead to decreased capacity to provide appropriate health and legal rights education and health care for both the child adolescent parent.

A new ICD-10-CM code would acknowledge (capture) the impact of having an adolescent parent is needed. With a more specific code, healthcare providers can better track the prevalence of these issues, identify trends, and advocate for increased support.

Equally important, a specific code would allow for the identification of disparities in health outcomes for children of adolescent parents, leading to targeted interventions and programs. By tracking the prevalence

of these issues and the outcomes of interventions, policymakers can evaluate the effectiveness of programs designed to support adolescent parents and their children.

Moreover, with a code that is more specific, "pediatric clinicians can shape the health of adolescent parents and their children because they are optimally trained to provide comprehensive care for infants, children, and adolescents and they understand the importance of creating a medical home for all patients, including the adolescent parent" (Powers et al.).

This proposal was submitted by Katherine Minaya MD, MST, FAAP of the Bronx Health Center and Foster Care Services. The proposal has been reviewed and supported by the American Academy of Pediatrics.

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TABULAR MODIFICATIONS

Z62	Problems related to upbringing		
	Z62.8	Other specified problems related to upbringing	
		Z62.89	Other specified problems related to upbringing
			Z62.890 Parent-child estrangement NEC
			Z62.891 Sibling rivalry
			Z62.892 Runaway [from current living environment] Child leaving living situation without permission
New code			Z62.893 Child with an adolescent parent(s)
			Z62.898 Other specified problems related to upbringing

Pediatric Healthcare: Screening for and Preventing Child Maltreatment

Child maltreatment is defined as physical, sexual, emotional, psychological abuse, or neglect—is often difficult to prevent. In many cases, abuse or neglect is only recognized after a child has endured significant trauma. Pediatric providers are often the first and sometimes only point of contact for children at risk, and they play a critical role in identifying and intervening before maltreatment escalates. It is not uncommon for clinicians to carry the burden of wondering what might have changed if warning signs had been recognized earlier— before a child is found to have nonaccidental injury or before another dies from malnutrition.

Currently, ICD-10-CM codes in categories T74 (Adult and child abuse, neglect and other maltreatment, confirmed) and T76 (suspected) are available—but these are limited to cases where abuse is already suspected or confirmed. They do not capture the routine, proactive screening efforts performed by pediatric providers to identify risk factors or early warning signs of maltreatment before a formal suspicion is documented.

Pediatricians are uniquely trained to recognize these early signs. They understand that maltreatment is often rooted in environments where caregivers lack the resources, education, mental health support, or social networks needed to provide a safe, nurturing environment. As Masarik et al. note, maltreatment frequently stems from systemic inequities and unmet family needs. Prevention, as emphasized by Garner et al. and Schofield et al., hinges on fostering relational health—stable, supportive relationships that buffer against adversity and build resilience in children.

Routine screening is especially critical for children with disabilities or chronic medical conditions, those too young to verbalize distress, those living in poverty, and those whose caregivers face mental health challenges—populations statistically at greater risk.

To clarify: screening tests differ fundamentally from diagnostic tests in purpose, timing, and population: screening is preventive; diagnosis is reactive.

- Screening is the process of evaluating asymptomatic individuals to identify risk or early signs of a condition—before symptoms appear, and often before there is any specific concern.
- Diagnostic testing, by contrast, is used to confirm or rule out a condition in individuals who are already symptomatic or after an abnormal screening result.

Clinicians screen universally for conditions like developmental delays, heart disease, and depression, screening for maltreatment must become a normalized, standardized component of pediatric care. Universal screening is not about expressing suspicion. It’s about ensuring that every child, has access to the same protections and early interventions.

Child maltreatment is a public health crisis with lasting effects. The American Academy of Pediatrics (AAP) emphasizes that 1 in 4 children experience some form of abuse or neglect in their lifetime, with 1 in 7 affected annually. The lifelong physical and mental health consequences for survivors and the economic impact on healthcare systems are profound.

Currently, no ICD-10-CM code exists to document that a pediatrician has screened a child for abuse or neglect in the absence of specific suspicion. Without this code:

- Data cannot be collected on screening practices.
- Populations at risk remain invisible in public health surveillance.

The addition of a new ICD-10-CM code to denote that screening for child maltreatment was performed would support:

- Epidemiologic tracking of high-risk populations,
- Research on the effectiveness of early interventions,
- Equitable access to protective care across demographics,
- And greater investment in child abuse prevention infrastructure.

This proposal has been submitted by Katherine Minaya MD, MST, FAAP of the Bronx Health Center and Foster Care Services. The proposal has been reviewed and supported by the American Academy of Pediatrics.

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TABULAR MODIFICATIONS

Z13 Encounter for screening for other diseases and disorders

Screening is the testing for disease or disease precursors in asymptomatic individuals so that early detection and treatment can be provided for those who test positive for the disease.

Excludes1: encounter for diagnostic examination-code to sign or symptom

Z13.8 Encounter for screening for other specified diseases and disorders
Excludes2: screening for malignant neoplasms (Z12.-)

New Code Z13.86 Encounter for screening for child abuse, neglect and other maltreatment

Pediatric Hypertrophic Pyloric Stenosis

Pediatric pyloric stenosis, also known as infantile hypertrophic pyloric stenosis (IHPS), and hypertrophic pyloric stenosis (HPS) is condition in infants characterized by abnormal thickening of the pylorus muscles in the stomach, leading to gastric outlet obstruction. The blockage leads to forceful vomiting and dehydration and causes a functional gastric outlet obstruction because of hypertrophy and hyperplasia of the muscular layers of the pylorus.

Pediatric hypertrophic pyloric stenosis is a condition that affects young infants. In infants, hypertrophic pyloric stenosis is the most common cause of gastric outlet obstruction and the most common surgical cause of vomiting. Clinically, infants are well at birth. Then, at 3 to 6 weeks of age and up to 6 months of age, they present with "projectile" vomiting, potentially leading to dehydration and weight loss.

This condition is rare in children after infancy. In an older child, causes of gastric outlet obstruction (GOO) such as primary acquired GOO, peptic ulcer disease, pyloric stricture (PS) due to granulomatous or eosinophilic gastroenteritis, ingestion of caustic substances, or neoplasia, such as gastrinoma or primary gastric tumors, must be ruled out in order to diagnose idiopathic HPS.

The exact etiology of IHPS is unknown. Some studies have shown that young infants treated with macrolide antibiotics had an increased incidence of IHPS. Postnatal exposure to erythromycin has also been associated with an increased risk for the development of pyloric stenosis. Other risk factors include bottle feeding, preterm birth, cesarean section delivery, and first-born infants (30% to 40% of cases). Diagnosis is confirmed by ultrasound and treated surgically.

Children’s Hospital Association is requesting a specific ICD-10-CM code for this condition as there is currently no unique code for pediatric hypertrophic pyloric stenosis. There are unique codes for adult hypertrophic pyloric stenosis (K31.1) and congenital hypertrophic pyloric stenosis (Q40.0). There are many pediatric patients seen each year in children’s hospitals and pediatric centers for treatment of hypertrophic pyloric stenosis.

This new code would provide clinical information for health care providers and researchers regarding frequency, treatment protocols and complications. In addition, the new code would provide clarification for HIM Coding professionals.

This proposal has been reviewed and supported by the American Academy of Pediatrics.

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TABULAR MODIFICATIONS

K31 Other diseases of stomach and duodenum
Includes: functional disorders of stomach
Excludes2: diabetic gastroparesis (E08.43, E09.43, E10.43, E11.43, E13.43)
diverticulum of duodenum (K57.00-K57.13)

K31.1 Adult hypertrophic pyloric stenosis
Add — Acquired hypertrophic pyloric stenosis
Add — Adult pyloric stenosis
Pyloric stenosis NOS
Revise — Excludes1: congenital ~~or infantile~~ pyloric stenosis (Q40.0)

New code — K31.B Pediatric hypertrophic pyloric stenosis
Add — Infantile hypertrophic pyloric stenosis
Add — Pediatric pyloric stenosis
Add — Excludes1: congenital pyloric stenosis (Q40.0)

Q40 Other congenital malformations of upper alimentary tract

Q40.0 Congenital hypertrophic pyloric stenosis
Revise — Congenital ~~or infantile~~ constriction
Revise — Congenital ~~or infantile~~ hypertrophy
Revise — Congenital ~~or infantile~~ spasm
Revise — Congenital ~~or infantile~~ stenosis
Revise — Congenital ~~or infantile~~ stricture
Add — Excludes 1: pediatric hypertrophic pyloric stenosis (K31.B)

Personal history of Clostridioides Difficile Infection

Clostridioides difficile is a bacterium that causes an infection of the colon, the longest part of the large intestine. Symptoms can range from diarrhea to life-threatening damage to the colon. The bacterium is often called C. difficile or C. diff.

Illness from C. difficile often occurs after using antibiotic medicines. It mostly affects older adults in hospitals or in long-term care settings. People not in care settings or hospitals also can get C. difficile infection. Some strains of the bacterium that can cause serious infections are more likely to affect younger people.

A patient's personal history of *Clostridioides* (formerly known as *Clostridium*) *difficile* (C diff) infection (CDI) is clinically significant as it is a risk factor for its recurrence and may influence the choice of antibiotic usage for the patient. ICD-10-CM identifies patients who have 'recurrent CDI' with code A04.71 Enterocolitis due to Clostridium difficile, recurrent, but there is no unique code to identify the personal history of C. difficile infection. Currently it is coded in the general code Z86.19, Personal history of other infectious and parasitic diseases.

A history of C diff is a risk factor for its recurrence. Patients who experience one episode of C. difficile colitis often develop another one. "A meta-analysis found that 13% to 50% of patients with *C. difficile* infection had at least one recurrence." [1] Since the presence of C diff is significant and can be complicated, it is relevant to identify a patient who is at high risk for the disease.

A new code is being requested by Optum 360, to specifically identify a personal history of C. difficile infection.

References

Clostridioides difficile Infection: Update on Management | AAFP
https://www.mayoclinic.org/diseases-conditions/c-difficile/symptoms-causes/syc-20351691

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TABULAR MODIFICATIONS

Z86 Personal history of certain other diseases
Z86.1 Personal history of infectious and parasitic diseases
New Code Z86.17 Personal history of Clostridioides difficile infection
Add Personal history of Clostridium difficile colitis (fulminant) (nonfulminant)
Add Personal history of foodborne intoxication by Clostridium difficile
Add Personal history of Pseudomembraneous colitis
Add Excludes1: enterocolitis due to Clostridium difficile, recurrent (A04.71)

INDEX MODIFICATIONS

History
- personal (of) - see also History, family (of)
- - infection NEC Z86.19
Add - - - Clostridioides (Clostridium) difficile (colitis) (infection) Z86.17

Pneumothorax that occurs after CPR

Cardiopulmonary resuscitation (CPR) involves artificially circulating the blood through repeated sternal compression to rescue the patient from cardiac arrest. This can exert excessive physical force on the chest of the patient which can result in chest injuries. Studies have reported that CPR-related chest injuries range from non-complicated skeletal injuries to life-threatening injuries, such as mediastinal hemorrhage or large pneumothorax. [1-4]

Wijck, et al. (2024), performed a systematic review and meta-analysis on the prevalence of rib fractures and other injuries resulting from CPR. They performed database searches to identify studies reporting on CPR-related injuries in patients who underwent chest compression for a non-traumatic cardiopulmonary arrest and included 74 studies encompassing a total of 16,629 patients. They found CPR-related injury documented in 60% of patients, with fractures identified as the most common injury.[2]

Pneumothorax resulting from CPR-related rib fractures is a known risk that may result from this life-saving procedure. In one hospital study of 237 cases, pneumothorax occurred in about 11% of patients after CPR. In the same study, the condition was more common - about 23% - in patients with history of obstructive lung disease.[3] In a similar study by another hospital, the most common thoracic injuries found in patients after CPR were those associated with rib and sternal fractures, including pneumothorax which was found in 10% of the patients.[4]

A unique code for fractures due to CPR (M96.A, Fracture of ribs, sternum and thorax associated with compression of the chest and cardiopulmonary resuscitation) was implemented in ICD-10-CM in October 2022. However, there is currently no unique code to represent pneumothorax due to CPR. The ICD-10-CM code currently being used for this, J95.811 (postprocedural pneumothorax), is not specific to CPR-related incidence of pneumothorax.

The requesting facility recently implemented a dedicated sudden cardiac arrest (SCA) - whole-body computed tomography (WBCT) protocol to evaluate SCA patients with return of spontaneous circulation (ROSC) following cardiopulmonary resuscitation (CPR). They seek a new code to allow for accurate reporting of CPR-related chest injuries, including not only fractures but also pneumothorax.

A unique code to identify CPR-related pneumothorax, as distinct from other causes, is important for clinical care and quality reporting. The proposed new code for Pneumothorax due to rib fracture associated with chest compression and cardiopulmonary resuscitation will help accurately report the clinical picture and promote better data mining.

This proposal was submitted by Mercy Health Systems.

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TABULAR MODIFICATIONS

J93 Pneumothorax and air leak

J93.8 Other pneumothorax and air leak

New code J93.84 Pneumothorax due to rib fracture associated with chest compression and cardiopulmonary resuscitation.

Add Code also associated rib fracture with chest compression and cardiopulmonary resuscitation (M96.A-)

M96.A Fracture of ribs, sternum and thorax associated with compression of the chest and cardiopulmonary resuscitation.

Add Code also any associated pneumothorax due to rib fracture associated with chest compression and cardiopulmonary resuscitation (J93.84)

Post-Bariatric Hypoglycemia

Post-bariatric hypoglycemia (PBH) is a chronic condition that develops in some individuals who have undergone bariatric surgery.[1-6] PBH is believed to be caused by changes in hormonal and glycemic patterns, including an excessive insulin and glucagon-like peptide-1 (GLP-1) response, as a result of altered nutrient transit post-surgery.[2-5] Individuals with PBH experience recurrent, debilitating hypoglycemia, the clinical relevance of which can be significant as patient safety, nutrition, cognition, and quality of life can be compromised.[7-9] Recurrent hypoglycemia can lead to defective counter-regulatory responses, reduced hypoglycemia awareness, impaired cognitive function, cardiac arrhythmia, and even death.[6,10]

Current coding options do not distinguish PBH from other types of hypoglycemia, resulting in a loss of essential clinical information that could inform and improve patient care in this unique group. There are many potential causes of non-diabetic hypoglycemia with different diagnostic and treatment strategies including congenital hyperinsulinism, severe sepsis, insulin autoimmune syndrome, insulinoma, and anorexia nervosa.[11,12] Accurate identification of individuals with PBH is also not possible using existing codes (i.e., E16.1 Other Hypoglycemia and Z98.84 Bariatric Surgery Status). Up to 75% of individuals will experience hypoglycemia after bariatric surgery.[13] However, only a minority of those individuals will have recurrent hypoglycemic events classified as PBH.[14] The lack of a PBH-specific code limits the ability to track and analyze PBH's unique clinical characteristics and related healthcare utilization. As a result, important data related to prevalence, risk factors, burden, and treatment outcomes remain unavailable, and patient care is impacted.

Despite the advent of GLP-1 receptor agonists for obesity management, bariatric surgery is expected to remain a mainstay of treatment given the magnitude and durability of the metabolic and weight-loss effect in addition to secondary health economics.[15-22] PBH prevalence rates are, therefore, not expected to decrease over time and may in fact rise with increasing PBH awareness and more standardized diagnostic testing.[23]

Physician scientist, Colleen Craig, MD, is proposing a new post bariatric hypoglycemia code to facilitate the collection of data in this population, accelerating research into risk factors, burden of the condition, and therapeutic interventions.

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TABULAR MODIFICATIONS

E89 Postprocedural endocrine and metabolic complications and disorders, not elsewhere classified

E89.8 Other postprocedural endocrine and metabolic complications and disorders

New sub-subcategory	E89.83	Postprocedural hypoglycemia following a procedure
Add		Use additional code, if applicable, for hypoglycemia level (E16.A-)
New code	E89.830	Post bariatric hypoglycemia
New code	E89.838	Other postprocedural hypoglycemia
Add		Post Nissen fundoplication hypoglycemia

Postprocedural Open Deep Wound without Disruption

This topic was presented at the September 2024 ICD-10 Coordination and Maintenance meeting. Based on public comments, revisions to the proposal have been made for reconsideration.

In certain circumstances, the surgical wound is temporarily left open at the end of a procedure. For some scenarios, an abdominal incision may intentionally be left open at the completion of surgery to allow an abdominal infection to resolve, and to facilitate a "second look" laparotomy at a later time. Other scenarios involve open chest, open fractures, and others where there is a high risk of postoperative compartment syndrome or deep surgical site infection. The initial operation may have occurred in the same hospital (i.e., not present on admission) or at a different hospital (i.e., present on admission). Having an open surgical wound after a prior operation is a high-risk situation that strongly affects resource use and care in the hospital, at least until the wound is closed. This is an important, clinically significant concept, but there currently is no way to account for it in ICD-10-CM.

The Agency for Healthcare Research and Quality (AHRQ) is requesting the following tabular modifications to report intended postprocedural state wherein the surgical wound is deliberately left open, to be closed at a later time.

TABULAR MODIFICATIONS

T81 Complications of procedures, not elsewhere classified

T81.3 Disruption of wound, not elsewhere classified
Disruption of any suture materials or other closure methods
Add Excludes1: postprocedural open surgical wound (Z98.88-)

Z98 Other postprocedural states

Z98.8 Other specified postprocedural states

New sub-subcategory Z98.88 Postprocedural open surgical wound

New code Z98.880 Postprocedural open abdomen
Add Delayed abdominal closure
Add Intentional delayed closure of abdominal surgical wound
Add Postprocedural temporary open abdomen

New code	Z98.881	Postprocedural open chest
Add		Delayed chest closure
Add		Intentional delayed closure of chest surgical wound
Add		Postprocedural temporary open chest
New code	Z98.888	Other postprocedural open surgical wound
Add		Other delayed surgical wound closure
Add		Other intentional delayed closure of surgical wound
Add		Other postprocedural temporary open surgical wound
New code	Z98.889	Postprocedural open surgical wound, unspecified
Add		Intentional delayed surgical wound closure NOS

Potts' Puffy Tumor

Pott's puffy tumor (PPT) is a serious, potentially life-threatening complication of chronic rhinosinusitis. While rare, it can pose serious risks to patients affected. PPT is characterized by localized swelling of the forehead with edema caused by osteomyelitis of the anterior wall of the frontal sinus and frontal bone subperiosteal abscess. It typically starts from untreated, poorly managed or undiagnosed frontal sinusitis. This condition can also develop after trauma or insect bites.

The incidence rate of annual cases in pediatric patients (aged 21 years and under) identified in Cosmos is 0.69 cases per 10,000 pediatric hospitalizations in 2024, and the incidence rate appears to have doubled in the past 5 years. Cosmos is a large database from participating healthcare organizations that use Epic.

Diagnostic studies such as CT scan and MRI of the head and sinuses are crucial in the diagnostic process to visualize the extent of the infection and any intracranial involvement. Treatment involves the early administration of antibiotics, surgical interventions including endoscopic sinus surgery, subperiosteal drainage, and if necessary, drainage of intracranial or intraorbital abscesses.

Currently, the ICD-10-CM index for Pott's tumor, puffy states -*see* "Osteomyelitis, specified type NEC. There is no option for frontal bone of skull. The closest option (code selection) is code M86.8X8 Other osteomyelitis, other site.

Since this ICD-10-CM code is non-specific and assigned for patients with osteomyelitis of many other bones or body sites, it is difficult for researchers to determine accurately how many patients get the life-threatening complication of Pott's puffy tumor or osteomyelitis of the frontal bone of the skull.

Children's Hospital Association is requesting the addition of a unique ICD-10-CM code for Pott's puffy tumor to better delineate this condition.

References:

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TABULAR MODIFICATIONS

M86 Osteomyelitis
M86.8 Other osteomyelitis
Brodie's abscess
M86.8X Other osteomyelitis
M86.8X0 Other osteomyelitis, multiple sites

New sub-subcategory	M86.8X1 Other osteomyelitis, shoulder
New code	M86.8X11 Other osteomyelitis, right shoulder
New code	M86.8X12 Other osteomyelitis, left shoulder
New code	M86.8X19 Other osteomyelitis, unspecified shoulder
New sub-subcategory	M86.8X2 Other osteomyelitis, upper arm
New code	M86.8X21 Other osteomyelitis, right upper arm
New code	M86.8X22 Other osteomyelitis, left upper arm
New code	M86.8X29 Other osteomyelitis, unspecified upper arm
New sub-subcategory	M86.8X3 Other osteomyelitis, forearm
New code	M86.8X31 Other osteomyelitis, right forearm
New code	M86.8X32 Other osteomyelitis, left forearm
New code	M86.8X39 Other osteomyelitis, unspecified forearm
New sub-subcategory	M86.8X4 Other osteomyelitis, hand
New code	M86.8X41 Other osteomyelitis, right hand
New code	M86.8X42 Other osteomyelitis, left hand
New code	M86.8X49 Other osteomyelitis, unspecified hand
New sub-subcategory	M86.8X5 Other osteomyelitis, thigh
New code	M86.8X51 Other osteomyelitis, right thigh
New code	M86.8X52 Other osteomyelitis, left thigh
New code	M86.8X59 Other osteomyelitis, unspecified thigh

New sub-subcategory	M86.8X6 Other osteomyelitis, lower leg
New code	M86.8X61 Other osteomyelitis, right lower leg
New code	M86.8X62 Other osteomyelitis, left lower leg
New code	M86.8X69 Other osteomyelitis, unspecified lower leg
New sub-subcategory	M86.8X7 Other osteomyelitis, ankle and foot
New code	M86.8X71 Other osteomyelitis, right ankle and foot
New code	M86.8X72 Other osteomyelitis, left ankle and foot
New code	M86.8X79 Other osteomyelitis, unspecified ankle and foot
New sub-subcategory	M86.8X8 Other osteomyelitis, other site
New code	M86.8X80 Other osteomyelitis, skull
New code Add	M86.8X81 Other osteomyelitis, face and sinuses Pott's puffy tumor
New code	M86.8X89 Other osteomyelitis, other site
	M86.8X9 Other osteomyelitis, unspecified sites

Prevesical Abscess and Other Pelvic Abscess

An abscess can develop in the space in front of the bladder, and behind the pubic bone. This is called the prevesical space, or the retropubic space (although there may be some distinctions used in identifying this region). The prevesical space is the largest potential space within the pelvic extraperitoneal space.[1,2]

An infection may start and spread from a number of different places to the prevesical space, and lead to an abscess forming. These can include the genitourinary organs (male or female), the musculoskeletal system (muscle, bone or joint in the region), the gastrointestinal system (such as from a diverticulitis), or even more distant spread via connecting spaces, such as from the retroperitoneal space. [1-7]

There are other pelvic spaces, and there is potential utility in identifying a pelvic abscess in such spaces as well. In addition, other types of pathology may occur, such as blood collection (hematoma) or other fluid collection.[1,2,7] Other codes are proposed for completeness.

This proposal is based on internal review and discussion.

References
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TABULAR MODIFICATIONS

Chapter 11
Diseases of the digestive system (K00-K95)

This chapter contains the following blocks:
…

Revise K65-K6AK68 Diseases of peritoneum, and retroperitoneum and pelvis

K57 Diverticular disease of intestine

Revise Code also, if applicable: peritonitis K65.-
Add pelvic abscess (K6A.0-)
Add peritonitis K65.-

Revise Diseases of peritoneum, ~~and~~ retroperitoneum and pelvis (K65- K6A~~K68~~)

K65 Peritonitis

K65.1 Peritoneal abscess
Add Excludes2: pelvic abscess (extraperitoneal) (K6A.0-)

New Category K6A Diseases of the pelvis, not elsewhere classified

New subcategory K6A.0 Pelvic abscess
Add Code also, if applicable:
Add diverticular disease of intestine (K57.-)
Add female pelvic inflammatory disease (N73.-)
Add Use additional code (B95-B97), to identify infectious agent, if known
Add Excludes2: peritoneal abscess (K65.1)
Add retroperitoneal abscess (K68.1-)

New code K6A.01 Prevesical abscess
Add Retropubic abscess

New code K6A.09 Other pelvic abscess
Add Pelvic abscess, unspecified

New code K6A.8 Other diseases of the pelvis, not elsewhere classified

Inflammatory diseases of female pelvic organs (N70-N77)
Add Code also, if applicable, pelvic abscess (K6A.0-)

INDEX MODIFICATIONS

Abscess (connective tissue) (embolic) (fistulous) (infective) (metastatic) (multiple) (pernicious) (pyogenic)(septic) L02.91
Revise - cul-de-sac (Douglas') (posterior) – see Abscess, pelvic, female, peritoneal ~~Peritonitis, pelvic, female~~
Revise - Douglas' cul-de-sac or pouch – see Abscess, pelvic, female, peritoneal ~~Peritonitis, pelvic, female~~
- extraperitoneal K68.19
Add - - meaning pelvic – see Abscess, pelvic
Revise - pelvis, pelvic K6A.09

Revise - - female –(see also Disease, pelvis, inflammatory) K6A.09
Add - - - peritoneal (see also Peritonitis, pelvic, female) K65.1
Revise - - male~~, peritoneal K65.1~~ K6A.09
Add - - - peritoneal K65.1
Revise - pouch of Douglas – see see Abscess, pelvic, female, peritoneal ~~Peritonitis, pelvic, female~~

Hematoma (traumatic) (skin surface intact) – see also Contusion
Revise - pelvis K6A.8 ~~(female) (nontraumatic) (nonobstetric) N94.89~~
Add - - female (nontraumatic) (nonobstetric) N94.89
- - obstetric O71.7
- - traumatic – see Injury, by site

Peritonitis (adhesive) (bacterial) (fibrinous) (hemorrhagic) (idiopathic) (localized) (perforative) (primary) (with adhesions) (with effusion) K65.9
Revise - pelvic (see also, if applicable, Abscess, abdominopelvic)

Pulmonary Mycetoma

A pulmonary mycetoma is a fungal ball in the lungs. Most commonly it is related to aspergillosis, but other fungi may also be related, including candida and coccidiomycosis. In most cases a pulmonary mycetoma grows at the site of a previous condition, such as following pulmonary tuberculosis.

Pulmonary mycetoma should not be confused with a mycetoma of the skin, such as Madura foot, which can be caused by a fungus (eumycetoma) or bacteria (actinomycetoma).

This proposal is the result of internal review following external questions raised related to pulmonary mycetoma.

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TABULAR MODIFICATIONS

B37 Candidiasis

B37.1 Pulmonary candidiasis
Add Code also, if applicable:
Add pulmonary mycetoma due to coccidioidomycosis (J4B)

B38 Coccidioidomycosis

B38.0 Acute pulmonary coccidioidomycosis
Add Code also, if applicable:
Add pulmonary mycetoma due to coccidioidomycosis (J4B)

B38.1 Chronic pulmonary coccidioidomycosis
Add Code also, if applicable:
Add pulmonary mycetoma due to coccidioidomycosis (J4B)

B38.2 Pulmonary coccidioidomycosis, unspecified
Add Code also, if applicable:
Add pulmonary mycetoma due to coccidioidomycosis (J4B)

B44 Aspergillosis
Delete ~~Includes: aspergilloma~~
Add Code also, if applicable:
Add aspergilloma of other site – code to chronic infection of site
Add pulmonary aspergilloma (J4B)
Add pulmonary aspergillosis fungal ball (J4B)
Add pulmonary mycetoma due to aspergillosis (J4B)
Add sinus aspergilloma (J32.-)

B47 Mycetoma
Add Excludes2: pulmonary mycetoma (J4B)

Chapter 10
Diseases of the respiratory system (J00-J99)

This chapter contains the following blocks:
…
Revise J40-J4B ~~J4A~~ Chronic lower respiratory diseases

Revise Chronic lower respiratory diseases (J40- J4B ~~J4A~~)

New code J4B Pulmonary mycetoma
Add Pulmonary fungal ball
Add Code also, if known, associated infection, such as:
Add aspergillosis (B44.-)
Add chronic pulmonary coccidioidomycosis (B38.1)
Add pulmonary candidiasis (B37.1)

Risk Stratification of Cancer

Risk stratification of patients with cancer conditions allows providers to maximize appropriate treatment plans for the patient's cancer upon the patient's cancer risk (e.g., low, intermediate, high risk). For many cancer types (including prostate and non-muscle invasive bladder cancer), guidelines recommend risk stratification upon diagnosis and treatment protocols are then advised based upon that risk category. For example, the American Urological Association (AUA)/Society of Urologic Oncology, Inc. Guidelines [1] which are supported by the National Comprehensive Cancer Network (NCCN) Guidelines [2] recommend that "At the time of each occurrence/recurrence, a clinician should assign a clinical stage and classify a patient accordingly as "low-," "intermediate-," or "high-risk.". Patients with low-risk cancer are typically put on surveillance protocols, those with intermediate risk status may be eligible for intravesical chemotherapy or immunotherapy, and those with high-risk status will often undergo repeat surgeries and intensive intravesical therapies. Similarly, in prostate cancer, both the AUA [3] and NCCN [4] recommend risk-stratifying patients into low, intermediate, and high-risk groups. Treatment protocols are based off that risk stratification.

Patients with low-risk conditions are typically placed into surveillance protocols and typically imaging and further lab tests are not needed, whereas those with intermediate risk disease will often undergo further lab or radiology testing and typically undergo active treatment, and those at high risk may undergo genetic testing and intensive treatments.

If a patient has a diagnosed bladder cancer and based upon certain criteria (size of tumor, invasiveness, pathologic findings) they are felt to be at "low risk" of recurrence, it is best practice to assign that patient that status; they then follow the "low risk" pathway (cystoscopies, urine tests etc.); if based upon the criteria they are "intermediate risk" then that should be noted and they then follow the intermediate risk pathway (more aggressive cystoscopies and bladder therapies), and if they are deemed to be "high risk" then that is the most aggressive follow-up and unique therapies that are only indicated in patients with that risk. Therefore, having ICD-10-CM codes that can further describe a patient's risk status will help clinicians provide the most optimal care of individual patients with cancer.

The American Urological Association is requesting new codes that can provide further risk stratification of a patient's cancer that can be used in addition to the code for their cancer.

References:

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ICD-10 Coordination and Maintenance Committee Meeting
September 9-10, 2025

TABULAR MODIFICATIONS

Chapter 2
Neoplasms (C00-D49)

Add Use additional code, where applicable, to identity:
Add Risk of malignancy status (Z78.2-)

Z78 Other specified health status

New subcategory Z78.2 Risk of malignancy status
Add Code first neoplasm, if known (C00-D49)

New code	Z78.20	Low risk malignancy status
New code	Z78.21	Intermediate risk malignancy status
New code	Z78.22	High risk malignancy status

Screening for Diabetes

Type 1 diabetes (T1D) results from chronic autoimmune destruction of insulin-producing pancreatic beta cells. Early-stage T1D can be first identified by markers of immune activation (multiple islet autoantibodies: (Stage 1 T1D) which are present long before development of glucose abnormalities. Ongoing loss of beta cell mass is associated with progression from milder hyperglycemia (Stage 2 T1D) to symptomatic hyperglycemia meeting ADA and WHO definitions for clinical diabetes (Stage 3 T1D).[1,2]

Prompt identification of children and adults with early-stage (presymptomatic) T1D is beneficial to both short- and long-term health outcomes:

1) Identification and monitoring of children at early-stage T1D significantly lowers rates of diabetic ketoacidosis (DKA) at onset of stage 3 T1D.[3,4] Beyond acute morbidity and mortality risk, diagnosis prior to DKA has been associated with durable improvement in achieving glucose targets.[5]

2) Individuals at stage 2 T1D may be eligible for treatment with recently FDA-approved teplizumab (Tzield®) to delay the need for insulin therapy;[6] other interventions are under active investigation.

3) DKA rates at onset of stage 3 T1D are known to be higher in historically marginalized populations. Equitable screening for early-stage T1D has the potential to ameliorate this inequity.[7,8]

4) Early diagnosis allows for education and psychosocial support before onset of need for insulin and avoids the traumatic experience of a DKA event.[9,10]

Presymptomatic T1D has been identified using screening for islet autoantibodies in 0.3% of general population children ages 2 to 5 years with increased prevalence likely at older ages.[11] In addition to autoantibody screening, population analysis of genetic risk scores specific to type 1 vs type 2 diabetes indicate that genetic tools may be useful to identify populations at high risk for T1D.[12,13]

Multiple institutions in the US and other countries are engaged in screening of general-population children. [11,14,15–19] In 2023, Italy became the first country to mandate autoantibody screening for T1D and celiac disease in the general population of children through unanimous vote by their parliament.[20]

Universal screening for T1D risk in children is spurred by the need to identify those who may benefit from advances in early-stage treatment options. New codes for screening will help identify patterns in screening for T1D, thus facilitating understanding of the best approaches for screening and evaluation of equity in screening across all sociodemographic groups. Further, codifying the distinction between screening for early-stage T1D versus pre-T2D, will facilitate broader understanding in the medical community of the differences in populations at risk as well as the differences in tempo and management strategy for these two disease states.

Monogenic diabetes includes both Maturity Onset Diabetes of the Young (MODY) and Neonatal Diabetes Mellitus (NDM). The overall prevalence of MODY is estimated to be 1% to 3% of all diabetes. NDM is further divided into Transient Neonatal Diabetes Mellitus (TNDM) and Permanent Neonatal Diabetes Mellitus (PNDM), each representing about 50% of the cases. Estimated incidence of NDM ranges between 1.11-0.38 per100,000 live births. The correct identification of monogenic forms of diabetes is critical for proper treatment and can often result in successful treatment with oral therapy in lieu of insulin.

Brigitte Frohnert, MD, PhD, Associate Professor of Pediatrics, Barbara Davis Center of Diabetes, is proposing the following tabular modifications to differentiate screening for T1D, T2D, and other types of diabetes.

References:

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TABULAR MODIFICATIONS

Z13 Encounter for screening for other diseases and disorders
Z13.1 Encounter for screening for diabetes mellitus

New code	Z13.11	Encounter for screening for Type 1 diabetes mellitus
New code	Z13.12	Encounter for screening for Type 2 diabetes mellitus
New code	Z13.13	Encounter for screening for diabetes mellitus due to underlying condition
New code	Z13.14	Encounter for screening for drug or chemical induced diabetes mellitus
New code	Z13.15	Encounter for screening for monogenic diabetes
New code	Z13.19	Encounter for screening for other specified diabetes mellitus

Secondary Metastatic Cancers of the Oral Cavity, Larynx, and Pharynx

The American Association of Post-Acute Care Nursing (AAPACN) is requesting to create unique ICD-10-CM diagnosis codes for secondary metastatic cancers of the oral cavity, larynx, and pharynx that are currently classified under the broad and nonspecific code C79.89, Secondary malignant neoplasm of other specified sites, and C78.39, Secondary malignant neoplasm of other respiratory organs.

At present, only malignant primary ICD-10-CM codes are mapped to the Speech Language Pathology (SLP) comorbidity classification related to oral and laryngeal cancers. Unfortunately, this excludes patients with metastatic disease to these regions, despite their equal or greater clinical need for speech therapy interventions.

Their generality undermines both clinical specificity and accurate reflection of disease burden. As a result, secondary cancers of the oral cavity and larynx are not recognized, potentially limiting appropriate rehabilitative care. This situation presents a significant clinical gap.

Malignant secondary neoplasms of the oral cavity, larynx, and pharynx are often more severe and debilitating than their primary counterparts, due to both disease progression and the aggressive treatments needed. The presence of metastatic lesions in these critical anatomical regions frequently compromises speech, swallowing, and overall airway protection, requiring intensive speech language pathology services.

TABULAR MODIFICATIONS

C78 Secondary malignant neoplasm of respiratory and digestive organs
C78.3 Secondary malignant neoplasm of other and unspecified respiratory organs
New code C78.31 Secondary malignant neoplasm of larynx

New code C78.32 Secondary malignant neoplasm of pharynx

C79 Secondary malignant neoplasm of other and unspecified sites
C79.8 Secondary malignant neoplasm of other specified sites

New code C79.83 Secondary malignant neoplasm of oral cavity

Skin Changes Due to Skin Failure

This topic was originally presented at the September 2024 ICD 10 Coordination and Maintenance meeting. Based on public comments, revisions to the proposal have been made for reconsideration.

On October 1, 2023, the Centers for Medicare and Medicaid Services (CMS) implemented new guidance in the Long-Term Care Facility Resident Assessment Instrument 3.0 User's Manual Version 1.18.11 October 2023 for Section M of the MDS (page M-6):

> "Skin changes at the end of life (SCALE), also referred to as Kennedy Terminal Ulcers (KTUs) and skin failure, are not primarily caused by pressure and are not coded in Section M."

Numerous ICD-10-CM categories exist to properly classify various types of organ failures for the body's major organs (heart, lungs, kidneys, liver), but no category exists for failure of the largest organ of the body, the skin. With the recent CMS guidance recognizing that skin failure and end-of-life wounds are not pressure ulcers, there are currently no unique ICD-10-CM diagnosis codes to classify the condition.

The skin failing may be due to other organs shutting down, reduced skin and soft tissue blood perfusion (hypoperfusion), a decreased resistance to external pressure, the skin's reduced ability to remove metabolic waste, or processes related to death or dying.

Acute skin failure is an event in which skin and underlying tissue die due to hypoperfusion concurrent with a critical illness.[1] Chronic skin failure is an event in which skin and underlying tissue die due to hypoperfusion concurrent with chronic illness.[1] End-stage skin failure is an event in which skin and underlying tissue die due to hypoperfusion concurrent with the end-of-life.[1] Based on a whole patient/resident assessment and a skin/wound assessment, skin failure due to hypoperfusion can be distinguished from pressure injury due to pressure or from wounds of other etiologies. Identification of the primary cause/etiology of the skin changes/wounds and of concurrent conditions guides healthcare professionals in accurately identifying skin failure.

The Post Acute Wound and Skin Integrity Council (PAWSIC) along with the American Academy of Dermatology Association (AADA) are requesting the following tabular modifications to address the gap in nomenclature and coding of these specific skin changes. PAWSIC has convened a working group of thirty-five international experts and thought leaders in the area of skin failure to develop recommendations for clinical practice and research related to skin failure and will collaborate with other organizations to educate healthcare professionals, patients, families and caregivers.

PAWSIC, in collaboration with the AADA, will assist with developing coding guidelines to aid in understanding the appropriate selection and utilization of the new skin failure codes. Additionally, educational resources will be disseminated through stakeholder organizations (e.g., AHA, AADA, national and international wound organizations) to ensure the correct application and use of these codes.

Reference:
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TABULAR MODIFICATIONS

L98 Other disorders of skin and subcutaneous tissue, not elsewhere classified

L98.8 Other specified disorders of the skin and subcutaneous tissue

New code	L98.80	Skin changes due to skin failure
Add		Code also underlying condition
Add		Excludes2: candidiasis (B37)
Add		erythema intertrigo (L30.4)
Add		incontinence-Associated Dermatitis (L24.A2)
Add		laceration S31.00-, S31.02-, S31.11-, S31.12-, S31.80-, S31.81-, S31.82-)
Add		moisture-associated skin damage (MASD) (L24.A-, L24.B-)
Add		non-pressure chronic ulcer (pressure area) (L97.-)
Add		pressure ulcer (pressure area) (L89.-)
New sub-subcategory	L98.801	Acute skin changes due to skin failure
New code	L98.8010	Acute skin changes due to skin failure, unspecified thickness
New code	L98.8011	Acute skin changes due to skin failure, skin intact
New code	L98.8012	Acute skin changes due to skin failure, partial thickness
New code	L98.8013	Acute skin changes due to skin failure, full thickness
New sub-subcategory	L98.802	Chronic skin changes due to skin failure
New code	L98.8020	Chronic skin changes due to skin failure, unspecified thickness

New code		L98.8021	Chronic skin changes due to skin failure, skin intact
New code		L98.8022	Chronic skin changes due to skin failure, partial thickness
New code		L98.8023	Chronic skin changes due to skin failure, full thickness
New sub-subcategory	L98.803	End-stage skin changes due to skin failure	
New code		L98.8030	End-stage skin changes due to skin failure, unspecified thickness
New code		L98.8031	End-stage skin changes due to skin failure, skin intact
New code		L98.8032	End-stage skin changes due to skin failure, partial thickness
New code		L98.8033	End-stage skin changes due to skin failure, full thickness
New sub-subcategory	L98.809	Unspecified skin changes due to skin failure	
New code		L98.8090	Unspecified skin changes due to skin failure, unspecified thickness
New code		L98.8091	Unspecified skin changes due to skin failure, skin intact
New code		L98.8092	Unspecified skin changes due to skin failure, partial thickness
New code		L98.8093	Unspecified skin changes due to skin failure, full thickness
New code	L98.89	Other specified disorders of the skin and subcutaneous tissue	

Sleep Inertia

Sleep inertia is a physiological state of impaired cognitive and sensory-motor performance that is present immediately after awakening. It persists during the transition of sleep to wakefulness, where an individual will experience feelings of drowsiness, disorientation and a decline in motor dexterity. Impairment from sleep inertia may take several hours to dissipate. In most cases, morning sleep inertia is experienced for 15 to 30 minutes after waking.

Sleep inertia is the transition state during which alertness and cognitive performance are temporarily impaired after awakening. This period of impairment is of concern to individuals (i.e. pilots,) who sleep or nap during on-call work hours or in-flight rest, then need to perform safety-critical tasks soon after waking. This condition is a paradoxical phenomenon of "waking up tired" - a period of impaired cognitive performance and grogginess experienced after waking, which dissipates as time awake increases.

A more gradual awakening, however, may also be protective given the complexity of neural circuitry in transitioning from one state to another. Sleep inertia may, therefore, be an adaptive mechanism to promote sleep upon awakening so that sleep is maintained when the awakening is undesired. For example, as with the timing of the circadian nadir, sleep inertia may help to maintain sleep in the last part of a nocturnal sleep episode when homeostatic sleep pressure has largely dissipated. Sleep inertia has been added to the Bordely's two-process model of sleep regulation to improve the understanding of experimental study observations.

It is important for all health care providers to be able to collect data on patients experiencing sleep inertia. This is important to both health care providers and those affected by this diagnosis. There are many individual shift workers, including active-duty personnel, who may be encouraged to take naps during their shifts, but need to be able to awaken quickly to full cognitive performance to perform critical task.

This proposal is submitted jointly by Jeanne Yoder, Defense Health Agency, Health Informatics and Mike Dubik, MD, Sleep Medicine, Naval Medical Center, Portsmouth Virginia.

References:
Aerospace medicine and human performance vol. 95, no. 4 April 2024
https://health.clevelandclinic.org/sleep-inertia
https://pubmed.ncbi.nlm.nih.gov/38486319/

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TABULAR MODIFICATIONS

G47 Sleep disorders
Excludes2: nightmares (F51.5)
nonorganic sleep disorders (F51.-)
sleep terrors (F51.4)
sleepwalking (F51.3)

G47.8 Other sleep disorders

Delete ~~Other specified sleep-wake disorder~~

New code G47.81 Sleep inertia

New code G47.89 Other sleep disorders
Add Other specified sleep-wake disorder

Topical Steroid Withdrawal

This topic was presented at the March 2024 and September 2024 ICD-10 Coordination and Maintenance meetings. Based on public comments, revisions to the proposal have been made for reconsideration. Changes are indicated in **bold**.

Topical corticosteroids (TCS) are first-line therapies for Atopic Dermatitis (AD) and other inflammatory dermatological conditions [1]. While short-term low- to mid-potency TCS monotherapy is likely safe and efficacious, there are well-established adverse effects to higher-potency or longer-term us of TCS such as skin atrophy, telangiectasia, and striae, as well as systemic side effects, including Hypothalamus-Pituitary-Adrenal (HPA) Axis suppression [2][3]. In recent decades, there has been a concerning rise in severe systemic adverse reactions due to long-term use and abrupt cessation of moderate- to high-potency TCS, commonly referred to as Topical Steroid Withdrawal (TSW) [4][5][6]. TSW is the most common term used to describe this syndrome, there are alternate names such as “Red Skin Syndrome” and “Topical Steroid Withdrawal Syndrome,” among others. The National Eczema Association, additionally, acknowledges TSW as a separate clinical entity from AD and highlights the absence of a formal diagnostic criteria for TSW [4]. This lack of standardized criteria and inconsistency in naming create constraints in conducting population studies.

The most up-to-date literature on TSW demonstrates an increased prevalence of TSW in adult (83.1%) females (78.9%) [4]. The primary indication for TCS use was cosmetic (61.4%), atopic dermatitis (14.2%) and acne (11.2%) and location of TCS use was also predominantly on the face (97.4%) [4]. Potency of TCS used was mostly moderate (68.9%) or high (20.85%) and duration of usage was typically 6 months or more [4]. The etiology of TSW in literature is strongly correlated with the potency and duration of topical corticosteroid (TCS) use.

Clinical findings associated with TSW that are distinct from atopic dermatitis include thermodysregulation, neurogenic pain, burning sensation, telangiectasia, skin atrophy, oozing containing a metallic smell, and specific cutaneous signs such as the "red sleeve sign" and "elephant wrinkles" [4][8]. In a recent study, discoveries show that TSW is clinically and biochemically distinct from atopic dermatitis in that the condition uniquely involves an overproduction of mitochondrial NAD+ and excessive breakdown of tryptophan causing neurologic pains while shared features include the microbiome and IL-4/IL-13 excess). Major diagnostic criteria derived in this study include burning, flushing, and temperature dysregulation [9].

These symptoms and morphological features are often notably distinct from the patient's primary dermatoses and may manifest in regions of the body where TCS were never applied. Two subtypes of TSW have been proposed: an erythematous-edematous subtype in patients with underlying eczematous dermatosis and a papulopustular subtype in patients who used TCS for cosmetic or acneiform conditions [4][7].

The management and treatment of TSW focus on symptom relief and promoting skin healing. Strategies include gradual tapering of TCS, regular use of moisturizers and emollients to repair skin barrier and alleviate dryness, wet wrap therapy, dupilumab, psychological support and lifestyle adjustments. However, no current therapy appears to offer more than symptomatic control.

Currently, there is not a unique ICD-10-CM code for TSW. Diagnosis often relies on clinical evaluation, patient history of TCS use, and exclusion of other potential causes of the symptoms. The distinct clinical features and course of TSW is inadequately characterized by existing diagnosis codes as symptoms are not a flare of existing AD or other skin condition, but of another clinical condition. Patients with eczematous dermatitis after >4-6 months of TCS use who have additional symptoms of thermodysregulation and cutaneous neurogenic pain should be diagnosed under this new code instead of the existing codes for "atopic dermatitis" or "rash and other nonspecific skin eruption."

The International Topical Steroid Awareness Network, with the support of National Eczema Association, Allergy & Asthma Network, and National Institute of Allergy and Infectious Disease allergy training program leadership is requesting the following tabular modifications.

References:

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9**.** Shobnam, Nadia, et al**.** "Topical Steroid Withdrawal Is a Targetable Excess of Mitochondrial NADD." *Journal of Investigative Dermatology*, vol. 145, no. 8, 2025, pp. 1953–1968, doi:10.1016/j.jid.2024.11.026.

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TABULAR MODIFICATIONS

L30 Other and unspecified dermatitis

L30.8 Other specified dermatitis

New code L30.81 **Chronic dermatitis following topical steroid use**

Add Use additional code for adverse effect, if applicable, to identify drug (T49.0X5-)

Add **Red skin syndrome**

Add **Topical steroid withdrawal**

Add Topical steroid withdrawal syndrome

New code L30.89 Other specified dermatitis

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Toxic Stress

The Gravity Project is requesting ICD-10-CM needed terms and codes from the project's consensus, evidence-based analysis of toxic stress.

The risk of toxic stress physiology and the role of adverse childhood experiences (ACEs) are well-documented in the literature. Based on the bedrock of many decades of study, approaches to addressing ACEs are woven into standardized national, state, and clinical efforts. However, to date, there has been insufficient terminology to document the core ten categories of adverse childhood experiences and risk of toxic stress physiology for the purposes of clinical documentation and surveillance.

The foundational Kaiser Permanente and Center for Disease Control Adverse Childhood Experiences study identified ten primary experiences that have the potential to contribute to the development of toxic stress physiology. It is these ten categories that are identified, addressed, and studied over time: physical, emotional, and sexual abuse, physical and emotional neglect, and the household challenges of parental separation or divorce, household member mental health issues, household member substance use issues, household member incarceration, and violence and abuse of household members.

This proposal aims to ensure that there are clear paths to reporting and capturing each of these core concerns within both current reporting for children toward identification and possible risk mitigation, and historical reporting for adults toward understanding and treatment.

TABULAR MODIFICATIONS

	Z62 Problems related to upbringing
	Z62.8 Other specified problems related to upbringing
New sub-subcategory	Z62.84 Personal history of household challenge in childhood
Add	Excludes 2: Current household member challenges (Z63.73-)
New code	Z62.840 Personal history of household challenge in childhood, parental separation or divorce
Add	Excludes 2: disruption of family by separation and divorce (Z63.5)
New code	Z62.841 Personal history of household challenge in childhood, violence or abuse of household member
Add	Excludes1: current child physical abuse (T74.12, T76.12)
Add	current child sexual abuse (T74.22, T76.22)

New code	Z62.842 Personal history of household challenge in childhood, household member mental health issue
New code	Z62.843 Personal history of household challenge in childhood, household member substance use problem
Add	Excludes 2: alcoholism and drug addiction in family (Z63.72)
New code	Z62.844 Personal history of household challenge in childhood, household member incarceration
	Z63 Other problems related to primary support group, including family circumstances
	Z63.3 Absence of family member
New code	Z63.33 Absence of parent or caregiver due to incarceration
	Z63.5 Disruption of family by separation and divorce
New code	Z63.50 Disruption of family by separation and divorce, unspecified
New code	Z63.51 Disruption of family by separation and divorce, absence of parent or caregiver
New code	Z63.59 Other disruption of family by separation and divorce
	Z63.7 Other stressful life events affecting family and household
New sub-subcategory	Z63.73 Household member challenges
Add	Excludes 2: Personal history of household challenge in childhood (Z62.84-)
New code	Z63.730 Living in household with violence or abuse of household member
New code	Z63.731 Living with a household member with a mental health issue
New code	Z63.732 Living with household member with a substance use problem
New code	Z63.733 Household member incarceration
New code	Z63.738 Other household member challenges
New code	Z63.739 Household member challenges, unspecified

	Z91 Personal risk factors, not elsewhere classified
	Z91.8 Other specified personal risk factors, not elsewhere classified
New sub-subcategory	Z91.86 Toxic stress physiology risk factors
New code	Z91.861 Risk for toxic stress physiology, low
New code	Z91.862 Risk for toxic stress physiology, moderate
New code	Z91.863 Risk for toxic stress physiology, high
New code	Z91.869 Risk for toxic stress physiology, unspecified

Vanishing Twin Syndrome

Vanishing twin syndrome (VTS) is a miscarriage that causes a pregnancy involving twins or multiples to become a pregnancy involving one embryo. According to the Cleveland Clinic, VTS occurs when an embryo is detected during an ultrasound but cannot be found on a future ultrasound. VTS happens when the vanishing twin's tissue gets absorbed by the surviving embryo(s) and the parent. VTS can happen in the first-third trimester.

The management and treatment may vary depending on when the embryo disappears. VTS can't be treated or prevented. Getting both the clinical and emotional support is essential to progress through the pregnancy. Vanishing twin syndrome may be more common (7-36%) with in vitro fertilization pregnancies. Research shows that women over age 30 are at risk.

Currently, there is no ICD-10-CM code to report and data mine the continuation of a pregnancy following a VTS.

Mercy Health Systems is requesting the following new codes to capture VTS. American College of Obstetricians and Gynecologists (ACOG) has reviewed and supports this proposal.

References
https://my.clevelandclinic.org/health/diseases/23023-vanishing-twin-syndrome

TABULAR MODIFICATIONS

	O31	Complications specific to multiple gestation
New subcategory		O31.4 Continuing pregnancy after vanishing twin syndrome of one fetus or more
New code		O31.40 Continuing pregnancy after vanishing twin syndrome of one fetus or more, unspecified trimester
New code		O31.41 Continuing pregnancy after vanishing twin syndrome of one fetus or more, first trimester
New code		O31.42 Continuing pregnancy after vanishing twin syndrome of one fetus or more, second trimester
New code		O31.43 Continuing pregnancy after vanishing twin syndrome of one fetus or more, third trimester

Ventricular Bigeminy

In ventricular bigeminy, there are alternating normal sinus heart beats, and premature ventricular complexes (PVCs), or premature ventricular contractions. PVCs are ectopic beats that arise from within the ventricles, rather than from the sinus node.

Ventricular bigeminy may be accompanied by symptoms such as dizziness or syncope. While ventricular bigeminy is not uncommon, there are a number of different possible causes, some of which can be relatively common, and others more uncommon. One clinically significant issue for ventricular bigeminy is the potential to go from this to ventricular tachycardia, with the potential for that to become unstable. While in some cases ventricular bigeminy can be relatively benign, it can require careful evaluation and sometimes treatment for underlying causes.

This proposal originated from internal discussions following review of related external questions.

References:
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TABULAR MODIFICATIONS

Note: A separate proposal for Brugada syndrome is also related to expansion at category I49.

	I49		Other cardiac arrhythmias
		I49.8	Other specified cardiac arrhythmias
Delete			~~Brugada syndrome~~
Delete			~~Coronary sinus rhythm disorder~~
Delete			~~Ectopic rhythm disorder~~
Delete			~~Nodal rhythm disorder~~
New code		I49.82	Ventricular bigeminy
Add			Code also, if applicable, associated symptoms such as:
Add			dizziness (R42)
Add			syncope (R55)
New code		I49.89	Other specified cardiac arrhythmias not elsewhere classified
Add			Coronary sinus rhythm disorder
Add			Ectopic rhythm disorder
Add			Nodal rhythm disorder

VEXAS Syndrome

Vacuoles, E1 enzyme, X-linked, Autoinflammatory, and Somatic mutations (VEXAS) syndrome, is a rare, recently discovered inflammatory disorder primarily affecting biological males. It is caused by mutations in the UBA1 gene on the X chromosome, leading to abnormal immune responses. The syndrome manifests in middle-aged or older adults and is characterized by somatic mutations, which are acquired rather than inherited. Patients with VEXAS syndrome experience a wide array of inflammatory symptoms affecting multiple organs. Common manifestations include painful skin rashes, pain and swelling in cartilaginous structures like the ear and nose, cough and shortness of breath in the lungs, joint swelling and pain, and potentially severe vessel inflammation. Additional symptoms include fever, extreme fatigue, anemia, low platelets, and an increased risk of blood clots. Patients may also have associated diagnoses such as relapsing polychondritis, polyarteritis nodosa, sweet syndrome, and myelodysplastic syndrome. VEXAS syndrome has unique symptoms and underlying genetic causes that distinguish it from other inflammatory and rheumatologic disorders. [1,2] A 2023 study found VEXAS syndrome more common than previously known, with prevalence about 1 in 13,500 [3].

Creation of a specific code for VEXAS syndrome will enable healthcare providers and public health organizations to collect reliable data on disease incidence, prevalence, demographics, and geographical spread. It will help to prevent misclassification with similar diseases. A unique ICD-10 CM code ensures that healthcare providers can precisely document the condition.

The proposal was submitted by the National Institutes of Health.

References:
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2. Beck DB, Ferrada MA, Sikora KA, et al. Somatic Mutations in UBA1 and Severe Adult-Onset Autoinflammatory Disease. N Engl J Med 2020; 383(27):2628-2638. http://doi.org/10.1056/NEJMoa2026834
3. Beck DB, Bodian DL, Shah V, et al. Estimated Prevalence and Clinical Manifestations of UBA1 Variants Associated With VEXAS Syndrome in a Clinical Population. JAMA 2023; 329(4):318-324. http://doi.org/10.1001/jama.2022.24836

TABULAR MODIFICATIONS

	M04	Autoinflammatory syndromes
New code	M04.3	VEXAS syndrome
Add		Vacuoles, E1 ubiquitin-activating enzyme, X-linked, autoinflammatory, somatic syndrome
Add		Code also any associated findings and disorders

War Theater Exposure: Burn Pit Emissions and Agent Orange

Burn Pit Emissions

Burn pits were commonly used by the United States military during operations in Iraq, Afghanistan, and other regions to dispose of waste, including plastics, metals, chemicals, and medical waste (Kim et al., 2021; Woskie et al., 2023; Trembley et al., 2024). These pits were often operated in open-air conditions, where large volumes of waste were burned without controls, leading to the release of a complex mix of hazardous chemicals and pollutants into the air ("Reassessment of the Department of Veterans Affairs Airborne Hazards and Open Burn Pit Registry," 2022). The pollutants released from burn pits include polycyclic aromatic hydrocarbons (PAHs), volatile organic compounds (VOCs), dioxins, and heavy metals, along with known carcinogens (Woskie et al., 2023; Kim et al., 2021). Symptoms and health effects of burn pit exposure may include respiratory issues such as chronic bronchitis and asthma, headaches, and fatigue. Long-term exposure has been associated with increased risks of respiratory diseases, cardiovascular issues, and cancers, particularly lung cancer (Savitz et al., 2024; Sico et al. ,2024; Kim et al., 2021; Trembley et al., 2024).

Currently, there is no ICD-10-CM code which adequately encompasses the impacts of open burn pit exposures. Without a specific ICD-10-CM code for burn pits, researchers and providers experience several challenges, including but not limited to inconsistent reporting, difficulty identifying eligible exposed Veterans, and a potentially inaccurate understanding of the prevalence of burn pit exposure among military personnel. Establishing a dedicated ICD-10-CM code could increase reporting and correct classification, significantly improving research and epidemiologic analyses resulting in improved health care outcomes for affected military service members, Veterans, DOD civilian employees and contractors, and all other exposed personnel. It will improve the accuracy of medical records and will significantly enhance research efforts resulting in improved health care outcomes for those affected.

Agent Orange

Agent Orange was developed by the United States military as a method to remove foliage from trees and expose enemy forces in the Vietnam War. The U.S. government defines Agent Orange exposure as service in Vietnam during the years when Agent Orange was in widespread use (ie between January 9, 1962 to May 7,1975) (Pagadala et al., 2024). It is created by combining various herbicides, most notably 2,4-dichlorophenoxyacetic acid (2,4-D) and 2,4,5-trichlorophenoxyacetic acid (2,4,5-T). During the manufacturing process, a highly toxic dioxin called 2,3,7,8-tetrachlorobenzo-p-dioxin (TCDD) is created as a byproduct. TCDD is known to manipulate DNA and bind to hormone receptors within the body causing several health issues. In fact, it is classified as a group 1 known human carcinogen by the International Agency for Research on Cancer (IARC) and shows a positive association to soft-tissue sarcomas, non-Hodgkin lymphomas, and lung cancer (National Academies Press (US), 2016).

One study suggests a possible association between Agent Orange exposure and prostate cancer (Pagadala et al., 2024) and another shows a slight increased risk of bladder cancer among the exposed (Williams et al., 2023). There is also a higher likelihood of developing dementia among exposed Veterans, and at an

earlier age (Martinez et al., 2021). The U.S. Department of Veterans Affairs (VA) has acknowledged the dangers of Agent Orange and other herbicide exposures and has created presumptions connecting Agent Orange exposure to the following health concerns: AL Amyloidosis, Chronic B-cell Leukemias, Chloracne, Diabetes Mellitus Type 2, Hodgkin's Disease, Ischemic Heart Disease, Multiple Myeloma, Non-Hodgkin's Lymphoma, Parkinson's Disease, Early- Onset Peripheral Neuropathy, Porphyria Cutanea Tarda, Prostate Cancer, Respiratory Cancers, and Soft Tissue Sarcomas. The VA also associates certain birth defects in children of Vietnam and Korean War Veterans with the service members' qualifying military service (US Department of Veterans Affairs, Veterans Health Administration, 2020).

Establishing a dedicated ICD-10-CM code for Agent Orange exposure is critical for improving the accuracy of clinical documentation and surveillance. It will improve the accuracy of medical records and will significantly enhance research efforts resulting in improved health care outcomes for those affected. Without a specific ICD-10-CM code for Agent Orange, researchers and providers experience several challenges, including but not limited to inconsistent reporting, difficulty identifying eligible exposed Veterans, and a potentially inaccurate understanding of the prevalence of Agent Orange exposure among military personnel.

Accurate documentation of burn pit and Agent Orange exposure is essential for prioritizing this issue, as emphasized in recent legislation such as the Promise to Address Comprehensive Toxics Act of 2022 [PACT] (United States Congress, 2022) and the Consolidated Appropriations Act, 2023. Establishing a dedicated ICD-10-CM code for these exposures is critical to ensuring that U.S. service members, veterans, and DOD civilians and contractors receive appropriate care and recognition.

This proposal, submitted by Laree LaPierre, MPH on behalf of Mr. John Short, Chief Data Interoperability Officer from the Federal Electronic Health Record Modernization (FEHRM), advocates for creating a new ICD-10-CM code to improve tracking, facilitate treatment, and monitor outcomes for affected individuals.

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TABULAR MODIFICATIONS

* Certain codes shown will become effective Oct. 1, 2025*

Z77 Other contact with and (suspected) exposures hazardous to health

Z77.3 Contact with and (suspected) exposure to war theater

*Z77.31 Contact with and (suspected) exposure to Gulf War theater
Contact with and (suspected) exposure to Persian Gulf War theater

New code — Z77.32 Contact with and (suspected) exposure to burn pits in war theater

New code — Z77.33 Contact with and (suspected) exposure to Agent Orange

*Z77.39 Contact with and (suspected) exposure to other war theater

Delete — ~~Agent Orange exposure~~

ICD-10 Coordination and Maintenance Committee Meeting
September 9-10, 2025

TABULAR MODIFICATIONS PROPOSED ADDENDA

All approved modifications will be effective April 1, 2026

A09 Infectious gastroenteritis and colitis, unspecified
Delete Excludes1: ~~diarrhea NOS (R19.7)~~
Add Excludes2: diarrhea NOS (R19.7)

D18 Hemangioma and lymphangioma, any site
Delete Excludes1: ~~benign neoplasm of glomus jugulare (D35.6)~~
Delete ~~blue or pigmented nevus (D22.-)~~
Delete ~~nevus NOS (D22.-)~~
Delete ~~vascular nevus (Q82.5)~~

Add Excludes2: benign neoplasm of glomus jugulare (D35.6)
Add blue or pigmented nevus (D22.-)
Add nevus NOS (D22.-)
Add vascular nevus (Q82.5)

D49 Neoplasms of unspecified behavior
Delete ~~Excludes1: neoplasms of uncertain behavior (D37-D44, D48)~~
Add Excludes2: neoplasms of uncertain behavior (D37-D44, D48)

D51 Vitamin B12 deficiency anemia
Delete ~~Excludes1: vitamin B12 deficiency (E53.8)~~
Add Excludes2: vitamin B12 deficiency (E53.8)

E53 Deficiency of other B group vitamins
E53.8 Deficiency of other specified B group vitamins
Delete ~~Excludes1: vitamin B12 deficiency anemia (D51.-)~~
Add Excludes2: vitamin B12 deficiency anemia (D51.-)

D65 Disseminated intravascular coagulation [defibrination syndrome]
Excludes1: disseminated intravascular coagulation (complicating):
Delete ~~abortion or ectopic or molar pregnancy (O00-O07, O08.1)~~
Delete ~~pregnancy, childbirth and the puerperium (O45.0, O46.0, O67.0, O72.3)~~

Excludes2: disseminated intravascular coagulation (complicating):
Add abortion or ectopic or molar pregnancy (O00-O07, O08.1)
Add pregnancy, childbirth and the puerperium (O45.0, O46.0, O67.0, O72.3)

D72 Other disorders of white blood cells
Delete Excludes1: ~~neutropenia (D70)~~
Add Excludes2: neutropenia (D70.-)
D72.8 Other specified disorders of white blood cells

D72.81 Decreased white blood cell count
Delete Excludes1: ~~neutropenia (D70.-)~~
Add Excludes2: neutropenia (D70.-)

D72.819 Decreased white blood cell count, unspecified
Delete ~~Excludes1: malignant leukopenia (D70.9)~~

E21 Hyperparathyroidism and other disorders of parathyroid gland
E21.2 Other hyperparathyroidism
Tertiary hyperparathyroidism
Delete ~~Excludes1: familial hypocalciuric hypercalcemia (E83.52)~~
Add Excludes2: familial hypocalciuric hypercalcemia (E83.52)

E29 Testicular dysfunction
E29.1 Testicular hypofunction
Delete ~~Excludes1: postprocedural testicular hypofunction (E89.5)~~
Add Excludes2: postprocedural testicular hypofunction (E89.5)

E55 Vitamin D deficiency
E55.0 Rickets, active
Delete Excludes~~1~~: ~~hereditary vitamin D-dependent rickets (E83.32)~~
Delete ~~vitamin D-resistant rickets (E83.31)~~
Add Excludes2: hereditary vitamin D-dependent rickets (E83.32)
Add vitamin D-resistant rickets (E83.31)

Metabolic disorders (E70-E88)

Delete Excludes1: ~~5-alpha-reductase deficiency (E29.1)~~
Add Excludes2: 5-alpha-reductase deficiency (E29.1)

E83 Disorders of mineral metabolism
E83.3 Disorders of phosphorus metabolism and phosphatases
E83.31 Familial hypophosphatemia
Delete ~~Excludes1: vitamin D-deficiency rickets (E55.0)~~
Add Excludes2: vitamin D-deficiency rickets (E55.0)

F07 Personality and behavioral disorders due to known physiological condition
F07.8 Other personality and behavioral disorders due to known physiological condition
F07.81 Postconcussional syndrome
Add Code also, if applicable, sequela of concussion (S06.0X- with seventh character S)
Revise Excludes1: current concussion (brain) (S06.0-<u>with seventh character A</u>)

G35 Multiple sclerosis
G35.A Relapsing-remitting multiple sclerosis
Revise Excludes1: demyelinating disease of central nervous system, unspecified (G37.9)
G36 Other acute disseminated demyelination
G36.0 Neuromyelitis optica [Devic]
Demyelination in optic neuritis
Delete ~~Excludes1: optic neuritis NOS (H46)~~
Add Excludes2: optic neuritis NOS (H46)

G43 Migraine
Delete ~~Excludes1:headache NOS (R51.9)~~
Add Excludes2: headache NOS (R51.9)

H40 Glaucoma
H40.8 Other glaucoma
H40.84 Neovascular secondary angle closure glaucoma
Revise Code ~~first~~ also the underlying condition such as:

I06 Rheumatic aortic valve diseases
Delete Excludes1: ~~aortic valve disease not specified as rheumatic (I35.-)~~
Delete ~~aortic valve disease with mitral and/or tricuspid valve involvement (I08.-)~~

Add Excludes2: aortic valve disease not specified as rheumatic (I35.-)
Add aortic valve disease with mitral and/or tricuspid valve involvement (I08.-)

I11 Hypertensive heart disease
I11.0 Hypertensive heart disease with heart failure
Hypertensive heart failure
Revise ~~Use additional~~ Code also to identify type of heart failure (I50.-)

I16 Hypertensive crisis
I16.1 Hypertensive emergency
Revise ~~Use additional~~ Code also, if applicable, to identify specific organ dysfunction, such as:

I27 Other pulmonary heart diseases
I27.8 Other specified pulmonary heart diseases
I27.84 Fontan related circulation
I27.841 Fontan-associated lymphatic dysfunction
Code also associated conditions such as:
Revise Fontan associated protein-losing enteropathy (~~K90.89~~ K90.49)

Cerebrovascular diseases (I60-I69)

Add Code also, if applicable, presence of:
Add hypertension (I10-I1A)

Use additional code to identify presence of:
Delete ~~hypertension (I10-I1A)~~

J95 Intraoperative and postprocedural complications and disorders of respiratory system, not elsewhere classified
J95.8 Other intraoperative and postprocedural complications and disorders of respiratory system, not elsewhere classified
J95.82 Postprocedural respiratory failure
Delete ~~Excludes1: Respiratory failure in other conditions (J96.-)~~
Add Excludes2: Respiratory failure in other conditions (J96.-)

K52 Other and unspecified noninfective gastroenteritis and colitis
K52.9 Noninfective gastroenteritis and colitis, unspecified
Delete ~~Excludes1: diarrhea NOS (R19.7)~~
Delete ~~functional diarrhea (K59.1)~~
Delete ~~infectious gastroenteritis and colitis NOS (A09)~~
Delete ~~psychogenic diarrhea (F45.8)~~

Add Excludes2: diarrhea NOS (R19.7)
Add functional diarrhea (K59.1)
Add infectious gastroenteritis and colitis NOS (A09)
Add psychogenic diarrhea (F45.8)

K60 Fissure and fistula of anal and rectal regions
Delete ~~Excludes1: fissure and fistula of anal and rectal regions with abscess or cellulitis (K61.-)~~

N87 Dysplasia of cervix uteri
Delete Excludes1: ~~abnormal results from cervical cytologic examination without histologic confirmation (R87.61-)~~
Delete ~~HGSIL of cervix (R87.613)~~

Add Excludes2: abnormal results from cervical cytologic examination without histologic confirmation (R87.61-)
Add HGSIL of cervix (R87.613)

N89 Other noninflammatory disorders of vagina
Delete Excludes1: ~~senile (atrophic) vaginitis (N95.2)~~
Add Excludes2: senile (atrophic) vaginitis (N95.2)

Revise QA0 Neurodevelopmental disorders related to specific genetic pathogenic -variants

CHAPTER 15

Pregnancy, childbirth and the puerperium (O00-O9A)

Delete Excludes1: ~~supervision of normal pregnancy (Z34.-)~~
Add Excludes2: supervision of normal pregnancy (Z34.-)

R12 Heartburn
Delete ~~Excludes1: dyspepsia NOS (R10.13)~~
Add Excludes2: dyspepsia NOS (R10.13)

R52 Pain, unspecified
Delete ~~Excludes1: joint pain (M25.5-)~~
Add Excludes2: joint pain (M25.5-)

R53 Malaise and fatigue
R53.8 Other malaise and fatigue
Delete Excludes1: ~~exhaustion and fatigue due to recurrent depressive episode (F33)~~
R53.83 Other fatigue
Add Excludes2: exhaustion and fatigue due to recurrent depressive episode (F33)

R63 Symptoms and signs concerning food and fluid intake
R63.5 Abnormal weight gain
Delete Excludes1: ~~obesity (E66.-)~~
Add Excludes2: obesity (E66.-)

R92 Abnormal and inconclusive findings on diagnostic imaging of breast
R92.3 Mammographic density found on imaging of breast
R92.30 Dense breasts, unspecified

R92.31 Mammographic fatty tissue density of breast
Revise Breast density category within Imaging Reporting and Data System (BI-RADS): A
Delete ~~Breast Imaging Reporting and Data System (BI-RADS): 1~~

R92.32 Mammographic fibroglandular density of breast
Revise Breast density category within Imaging Reporting and Data System (BI-RADS): B
Delete ~~Breast Imaging Reporting and Data System (BI-RADS): 2~~

R92.33 Mammographic heterogeneous density of breast
Revise Breast density category within Imaging Reporting and Data System (BI-RADS): C
Delete ~~Breast Imaging Reporting and Data System (BI-RADS): 3~~

R92.34 Mammographic extreme density of breast
Revise Breast density category within Imaging Reporting and Data System (BI-RADS): D
Delete ~~Breast Imaging Reporting and Data System (BI-RADS): 4~~

T43 Poisoning by, adverse effect of and underdosing of psychotropic drugs, not elsewhere classified
Delete Excludes1: ~~appetite depressants (T50.5-)~~
Delete ~~barbiturates (T42.3-)~~
Delete ~~benzodiazepines (T42.4-)~~
Delete ~~methaqualone (T42.6-)~~
Delete ~~psychodysleptics [hallucinogens] (T40.7-T40.9-)~~

Add Excludes2: appetite depressants (T50.5-)
Add barbiturates (T42.3-)
Add benzodiazepines (T42.4-)
Add methaqualone (T42.6-)
Add psychodysleptics [hallucinogens] (T40.7-T40.9-)

Other and unspecified effects of external causes (T66-T78)

Add Code also, if applicable, such as:
Add highway rest stop (Y92.523)
Add school (private) (public) (state) as the place of occurrence of the external cause (Y92.21)
Add service areas as the place of occurrence of the external cause (Y92.52-)
Add single-family non-institutional (private) house as the place of occurrence of the external cause (Y92.01-)
Add transport vehicle as the place of occurrence of the external cause (Y92.81-)

Z00 Encounter for general examination without complaint, suspected or reported diagnosis
Delete ~~Excludes1: encounter for examination for administrative purposes (Z02.-)~~
Add Excludes2: encounter for examination for administrative purposes (Z02.-)

Z01 Encounter for other special examination without complaint, suspected or reported diagnosis
Delete ~~Excludes1: encounter for examination for administrative purposes (Z02.-)~~
Add Excludes2: encounter for examination for administrative purposes (Z02.-)

Z01.4 Encounter for gynecological examination
Z01.41 Encounter for routine gynecological examination
Delete Excludes1: ~~screening cervical pap smear not a part of a routine gynecological examination (Z12.4)~~
Add Excluses2: screening cervical pap smear not a part of a routine gynecological examination (Z12.4)

Z12 Encounter for screening for malignant neoplasms
Z12.4 Encounter for screening for malignant neoplasm of cervix
Delete ~~Excludes1: when screening is part of general gynecological examination (Z01.4-)~~
Add Excludes2: when screening is part of general gynecological examination (Z01.4-)

Z34 Encounter for supervision of normal pregnancy
Delete Excludes1: ~~any complication of pregnancy (O00-O9A)~~
Add Excludes2: any complication of pregnancy (O00-O9A)

Z79 Long term (current) drug therapy
Z79.8 Other long term (current) drug therapy
Z79.89 Other long term (current) drug therapy
Z79.891 Long term (current) use of opiate analgesic
Delete Excludes1 ~~methodone use NOS (F11.9-):~~
Delete ~~use of methodone for treatment of heroin addiction (F11.2-)~~

Add Excludes2: methadone use NOS (F11.9-)
Add use of methadone for treatment of heroin addiction (F11.2-)

ICD-10 Coordination and Maintenance Committee Meeting
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INDEX MODIFICATION PROPOSED ADDENDA

All approved modifications will be effective April 1, 2026

	Disorder
	- eye H57.9
Revise	- - postprocedural - see Complication, ~~postprocedural,~~ eye, postprocedural
	Drip, postnasal (chronic) R09.82
	- due to
Revise	- - other known condition - code to condition
	Parkinsonism (idiopathic) (primary) G20.C
Add	- due to
Add	- - Parkinson's Disease – see Disease Parkinson's
Revise	Parkinson's disease~~, syndrome or tremor -see Parkinsonism~~ – see Disease, Parkinson's
Add	Parkinson's syndrome or tremor -see Parkinsonism
	Pneumonitis (acute) (primary) -see also Pneumonia J98.4
Add	- chemotherapy (J70.4)
Add	- - acute J70.2
Add	- - chronic J70.3
	Postnasal drip R09.82
	- due to
Revise	- - other known condition - code to condition
	Syndrome - see also Disease
	- myelodysplastic D46.9
	- - with
Add	- - - pancytopenia (acquired) D61.818
	Tumor
Revise	- neuroendocrine ~~D3A.8~~ C7A.-

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TABULAR MODIFICATIONS PROPOSED ADDENDA
All approved modifications will be effective October 1, 2026

CHAPTER 1
Certain infectious and parasitic diseases (A00-B99)

Delete ~~Excludes1: certain localized infections - see body system-related chapters~~
Add Excludes2: certain localized infections - see body system-related chapters

A49 Bacterial infection of unspecified site
Delete Excludes1: ~~chlamydial infection NOS (A74.9)~~
Add Excludes2: chlamydial infection NOS (A74.9)

D05 Carcinoma in situ of breast
Delete ~~Excludes2: malignant neoplasm of breast (C50.-)~~
Add Excludes1: malignant neoplasm of breast (C50.-)

D68 Other coagulation defects
D68.3 Hemorrhagic disorder due to circulating anticoagulants
D68.31 Hemorrhagic disorder due to intrinsic circulating anticoagulants, antibodies, or inhibitors

D68.312 Antiphospholipid antibody with hemorrhagic disorder
Lupus anticoagulant (LAC) with hemorrhagic disorder
Systemic lupus erythematosus [SLE] inhibitor with hemorrhagic disorder
Delete Excludes1: antiphospholipid antibody, finding without diagnosis (R76.0)
Delete ~~antiphospholipid antibody syndrome (D68.61)~~
Delete ~~antiphospholipid antibody with hypercoagulable state (D68.61)~~
Delete lupus anticoagulant (LAC) finding without diagnosis (R76.0)
Delete ~~lupus anticoagulant (LAC) with hypercoagulable state (D68.62)~~
Delete systemic lupus erythematosus [SLE] inhibitor finding without diagnosis (R76.0)
Delete ~~systemic lupus erythematosus [SLE] inhibitor with hypercoagulable state (D68.62)~~

Add Excludes2: antiphospholipid antibody syndrome (D68.61)
Add antiphospholipid antibody with hypercoagulable state (D68.61)

Add lupus anticoagulant (LAC) with hypercoagulable state (D68.62)
Add systemic lupus erythematosus [SLE] inhibitor with hypercoagulable state (D68.62)

Certain disorders involving the immune mechanism (D80-D89)

Delete ~~Excludes1: human immunodeficiency virus [HIV] disease (B20)~~
Add Excludes2: human immunodeficiency virus [HIV] disease (B20)

E29 Testicular dysfunction
E29.1 Testicular hypofunction
Revise 5-~~delta~~Alpha-~~R~~reductase deficiency (with male pseudohermaphroditism)

E72 Other disorders of amino-acid metabolism
E72.1 Disorders of sulfur-bearing amino-acid metabolism
E72.11 Homocystinuria
Add Homocystinemia

E35 Disorders of endocrine glands in diseases classified elsewhere
Delete ~~Use additional code, if applicable, to identify:~~
~~sequelae of tuberculosis of other organs (B90.8)~~

E83 Disorders of mineral metabolism
E83.5 Disorders of calcium metabolism
Delete Excludes1: ~~hyperparathyroidism (E21.0-E21.3)~~
Add Excludes2: hyperparathyroidism (E21.0-E21.3)

E87 Other disorders of fluid, electrolyte and acid-base balance
E87.2 Acidosis
E87.22 Chronic metabolic acidosis
Chronic lactic acidosis
Revise Code first, if applicable, underlying etiology~~, if applicable~~

F02 Dementia in other diseases classified elsewhere
Code first the underlying physiological condition, such as:
Delete ~~frontotemporal dementia (G31.09)~~
Revise other frontotemporal neurocognitive disorder ~~(G31.90)~~ (G31.09)

F05 Delirium due to known physiological condition
Add Delirium NOS
Delete ~~Excludes1: delirium NOS (R41.0)~~

G93 Other disorders of brain
G93.5 Compression of brain
Add Midline shift of brain

J42 Unspecified chronic bronchitis
Add Code also, if applicable, chronic respiratory conditions due to chemicals, gases, fumes and vapors (J68.4)

J69 Pneumonitis due to solids and liquids
Revise Code~~s~~ also, if applicable, other types of pneumonias, such as:
Add bacterial pneumonia (J13-J15.-)
Add viral pneumonia (J12.-)

J68 Respiratory conditions due to inhalation of chemicals, gases, fumes and vapors
J68.4 Chronic respiratory conditions due to chemicals, gases, fumes and vapors
Code also, if applicable, chronic conditions, such as:
Add chronic bronchitis (J42)

J99 Respiratory disorders in diseases classified elsewhere
Code first underlying disease, such as:
Add bronchomycosis (B49)

Dentofacial anomalies [including malocclusion] and other disorders of jaw (M26-M27)

Delete Excludes1: ~~hemifacial atrophy or hypertrophy (Q67.4)~~
Delete ~~unilateral condylar hyperplasia or hypoplasia (M27.8)~~

Add Excludes2: hemifacial atrophy or hypertrophy (Q67.4)

M26 Dentofacial anomalies [including malocclusion]
M26.0 Major anomalies of jaw size
Add Excludes2: unilateral condylar hyperplasia or hypoplasia (M27.8)

O35 Maternal care for known or suspected fetal abnormality and damage
O35.1 Maternal care for (suspected) chromosomal abnormality in fetus
O35.11 Maternal care for (suspected) chromosomal abnormality in fetus, Trisomy 13
Add Maternal care for (suspected) chromosomal abnormality in fetus, Patau Syndrome
O35.12 Maternal care for (suspected) chromosomal abnormality in fetus, Trisomy 18
Add Maternal care for (suspected) chromosomal abnormality in fetus, Edwards Syndrome

O35.13 Maternal care for (suspected) chromosomal abnormality in fetus, Trisomy 21

Add Maternal care for (suspected) chromosomal abnormality in fetus, Down Syndrome

O35.15 Maternal care for (suspected) chromosomal abnormality in fetus, sex chromosome abnormality

Add Maternal care for (suspected) chromosomal abnormality in fetus, Trisomy X

Add Maternal care for (suspected) chromosomal abnormality in fetus, Klinefelter Syndrome

Add Maternal care for (suspected) chromosomal abnormality in fetus, XYY Syndrome

CHAPTER 17
Congenital malformations, deformations and chromosomal abnormalities (Q00-Q99)

Revise Note: Codes from this chapter are not for use on maternal records, if applicable

Add Codes from this chapter are not for use on maternal records for conditions in the fetus.

Add Codes from this chapter could appropriately be used on maternal charts for conditions that directly involve the mother, if applicable.

R10 Abdominal and pelvic pain

R10.8 Other abdominal pain

R10.85 Abdominal pain of multiple sites

Revise Excludes1: localized abdominal pain (R10.1-~~R10.4~~ R10.3-)

R26 Abnormalities of gait and mobility

R26.2 Difficulty in walking, not elsewhere classified

Excludes1: falling (R29.6)

Delete ~~unsteadiness on feet (R26.81)~~

Add Excludes2: unsteadiness on feet (R26.81)

R29 Other symptoms and signs involving the nervous and musculoskeletal systems

R29.7 National Institutes of Health Stroke Scale (NIHSS) score

Revise Code first the type of cerebral infarction (I60.-, I61.-, I62.-,I63.-)

R41 Other symptoms and signs involving cognitive functions and awareness

R41.0 Disorientation, unspecified

Confusion NOS

Delete ~~Delirium NOS~~

R79 Other abnormal findings of blood chemistry

Delete Excludes1: ~~hyperglycemia NOS (R73.9)~~

Add Excludes2: hyperglycemia NOS (R73.9)

S06 Intracranial injury
S06.A Traumatic brain compression and herniation
Add Traumatic midline shift of brain

S22 Fracture of rib(s), sternum and thoracic spine
Add Code first, if applicable, spinal cord injury (S24.0-, S24.1-)
Code also, if applicable, any associated condition such as:
Delete ~~spinal cord injury (S24.0 , S24.1)~~

S23 Dislocation and sprain of joints and ligaments of thorax
S23.4 Sprain of ribs and sternum
S23.42 Sprain of sternum
Delete ~~S23.420 Sprain of sternoclavicular (joint) (ligament)~~

T45 Poisoning by, adverse effect of and underdosing of primarily systemic and hematological agents, not elsewhere classified
T45.A Poisoning by, adverse effect of and underdosing of immune checkpoint inhibitors and immunostimulant drugs
T45.AX Poisoning by, adverse effect of and underdosing of immune checkpoint inhibitors and immunostimulant drugs
T45.AX1 Poisoning by immune checkpoint inhibitors and immunostimulant drugs, accidental (unintentional)
Revise Poisoning by immune checkpoint inhibitors and ~~immunosuppressive~~ immunostimulant drugs NOS

T79 Certain early complications of trauma, not elsewhere classified
T79.1 Fat embolism (traumatic)
Add Code also, if applicable:
Add fat embolism of pulmonary artery with acute cor pulmonale (I26.04)
Add fat embolism of pulmonary artery without acute cor pulmonale (I26.96)
Excludes1: fat embolism complicating:
abortion or ectopic or molar pregnancy (O00-O07, O08.2)
Delete ~~pregnancy, childbirth and the puerperium (O88.8)~~

Add Excludes2: fat embolism complicating:
Add pregnancy, childbirth and the puerperium (O88.8)

Z29 Encounter for other prophylactic measures
Z29.1 Encounter for prophylactic immunotherapy
Revise Z29.14 Encounter for prophylactic rabies immune ~~globin~~ globulin

Z36 Encounter for antenatal screening of mother
Z36.0 Encounter for antenatal screening for chromosomal anomalies
Add Encounter for antenatal screening for Down syndrome
Add Encounter for antenatal screening for fetal aneuploidy
Add Encounter for antenatal screening for Patau syndrome
Add Encounter for antenatal screening for Trisomy 13
Add Encounter for antenatal screening for Trisomy 21

Z51 Encounter for other aftercare and medical care
Z51.5 Encounter for palliative care
Add Encounter for comfort care
Add Encounter for hospice care

Z91 Personal risk factors, not elsewhere classified
Z91.0 Allergy status, other than to drugs and biological substances
Z91.01 Food allergy status
Z91.014 Allergy to mammalian meats
Add Alpha-gal syndrome (AGS)

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INDEX MODIFICATION PROPOSED ADDENDA

All approved modifications will be effective October 1, 2026

Angioma - see also Hemangioma, by site
Add - cherry, see also Hemangioma

Bite(s) (animal) (human)
Revise - amphibian (venomous) – see ~~Venom, bite, amphibian~~ Table of Drugs and Chemicals, by animal or substance, poisoning
- animal - see also Bite, by site
Revise - - venomous - see ~~Venom~~ Table of Drugs and Chemicals, by animal or substance, poisoning
Revise - arthropod NEC – see ~~Venom, bite, arthropod~~ Table of Drugs and Chemicals, by animal or substance, poisoning
Revise - centipede - see ~~Toxicity, venom, arthropod, centipede~~ Table of Drugs and Chemicals, by animal or substance, poisoning
Revise - lizard (venomous)- see ~~Venom, bite, reptile~~ Table of Drugs and Chemicals, by animal or substance, poisoning
Revise - marine animals (venomous) - see ~~Toxicity~~, ~~venom, marine animal~~ Table of Drugs and Chemicals, by animal or substance, poisoning
Revise - poisonous – see ~~Venom~~ Table of Drugs and Chemicals, by animal or substance, poisoning
Revise - reptile NEC - see ~~also~~Venom, bite, reptile Table of Drugs and Chemicals, by animal or substance, poisoning
Revise - reptile NEC snake -see ~~Venom, bite, snake~~ Table of Drugs and Chemicals, by animal or substance, poisoning
Revise - sea-snake (venomous) - see ~~Toxicity, venom, snake, sea snake~~ Table of Drugs and Chemicals, by animal or substance, poisoning
Revise - snake -see ~~also Venom, bite, snake~~ Table of Drugs and Chemicals, by animal or substance, poisoning
Revise - spider (venomous)- see ~~Toxicity, venom, spider~~ Table of Drugs and Chemicals, by animal or substance, poisoning
Revise - venomous - see ~~Venom~~ Table of Drugs and Chemicals, by animal or substance, poisoning

Bronchitis (diffuse) (fibrinous) (hypostatic) (infective) (membranous)
- chemical (acute) (subacute) J68.0
Revise - - chronic ~~J42~~ J68.4 ~~– see also Disease, respiratory, chronic, due to chemials, gases, fumes or vapors J42~~
Revise - - due to fumes or vapors ~~J42~~ J68.0
- chronic J42
Revise - - chemical (due to fumes or vapors) J68.4 ~~see also Disease, respiratory, chronic, due to chemicals, gases, fumes or vapors J42~~
- - due to
Revise - - - chemicals, gases, fumes or vapors (inhalation) J68.4 ~~see also Disease, respiratory, chronic, due to chemicals, gases, fumes or vapors J42~~

Care (of) (for) (following)
Add - comfort Z51.5
Add - hospice Z51.5

Change(s) (in) (of) - see also Removal
- bone - see also Disorder, bone
Revise - - diabetic - see Diabetes, ~~bone change~~ by type, with complication, specified NEC

Complication(s) (from) (of)
- eye H57.9
Add - - postprocedural (H59.-)

Confusion, confused R41.0
Add - acute R41.0
Add - - with dementia F05

Dehiscence (of)
- closure of
- - fascia (muscular) (superficial) T81.328
Add - - - abdominal T81.321
- - muscle or muscle flap T81.328
Add - - - abdominal T81.321

Delivery
- - by
- - - malposition, malpresentation
Revise - - - - without obstruction (see also Delivery, complicated by, ~~obstruction~~ obstructed labor) O32.9

Revise De Morgan's spots (senile angiomas) ~~I78.1~~D18.-

Depression (acute) (mental) F32.A
Revise - central nervous system ~~R09.2~~ G98.8

Dependence
- opioid use
- - moderate or severe F11.20
- - - with
- - - - opioid-induced
- - - - - anxiety disorder F11.288
Delete ~~- - - - - anxiety disorder F11.988~~
- - - - - depressive disorder F11.24
Delete ~~- - - - - depressive disorder F11.94~~
- - - - - sexual dysfunction F11.281

Delete	~~- - - - - sexual dysfunction F11.981~~
	Diabetes, diabetic (mellitus) (sugar) E11.9
Add	- double - see Diabetes, specified type NEC
Add	- hybrid - see Diabetes, specified type NEC
	- type 1 E10.9
	- - with
Add	- - - diabetes type 2 - see Diabetes, specified type NEC
	- type 2 E11.9
	- - with
Add	- - - diabetes type 1- see Diabetes, specified type NEC
	Diarrhea, diarrheal (disease) (infantile) (inflammatory) R19.7
Add	- with
Add	- - melena K92.1
Revise	Dog bite - see ~~Bite~~ External Cause of Injuries Index bite, bitten, dog
	Dysfunction
	- sexual (due to) R37
	- - alcohol F10.981
Add	- - - in
Add	- - - - abuse F10.181
Add	- - - - dependence F10.281
	Encounter (with health service) (for) Z76.89
Add	- comfort Z51.5
Add	- hospice Z51.5
	Foreign body
Revise	- feeling of, in throat ~~R09.89~~ R09.A2
	Granuloma
Revise	- lung (infectious) (see also Fibrosis, lung) J98.4
Delete	~~Homocystinemia R79.83~~
Revise	Homocystinuria (Homocystinemia) E72.11
	Myeloma (multiple) C90.0-
Add	- smoldering D47.2
Add	Neurogenic stunned myocardium I5A

	Opioid(s)
	- induced, without use disorder
Revise	- - anxiety disorder ~~F11.988~~ F11.288
Revise	- - depressive disorder ~~F11.94~~ F11.24
Revise	- - sexual dysfunction ~~F11.981~~ F11.281
Add	PANS (Pediatric Acute-onset Neuropsychiatric Syndrome) D89.89
	Pharyngitis (acute) (catarrhal)(gangrenous) (infective) (malignant) (membranous)
Delete	(phlegmonous) (pseudomembranous) (simple) (subacute) (suppurative) (ulcerative) ~~(viral)~~ J02.9
	Polyp
Revise	- cecum ~~D12.0~~ K63.5
Add	- - adenomatous D12.6
	Pregnancy (single) (uterine) - see also Delivery and Puerperal Z33.1
	- complicated by (care of) (management affected by)
	- - fetal (maternal care for)
	- - - chromosomal abnormality (conditions in Q90-Q99) O35.10
Add	- - - - Down Syndrome O35.13
Add	- - - - Edwards' Syndrome O35.12
Add	- - - - Klinefelter Syndrome O35.15
Add	- - - - Patau Syndrome O35.11
Add	- - - - Trisomy X O35.15
Add	- - - - XYY Syndrome O35.15
	Problem (with) (related to)
Revise	- new step-parent affecting child ~~Z62.898~~ Z62.823
	Protrusion, protrusio
	- device, implant or graft (see also Complications, by site and type, mechanical) T85.698
Revise	- - arterial graft NEC - see Complication, graft ~~cardiovascular device,~~ ~~mechanical,~~ vascular, mechanical protrusion
	Schizophrenia, schizophrenic F20.9
	- catatonic (type) (excited) (withdrawn) F20.2
Add	- - catatonic NOS F06.1
	Shift
Add	- midline, brain G93.5
Add	- - traumatic S06.A0
Add	- - - with herniation S06.A1

	Spots, spotting (in) (of)
Revise	- De Morgan's (senile angiomas) ~~I78.1~~ D18.-
Add	Stunned myocardium, neurogenic I5A
	Syndrome -see also Disease
Add	- Alpha-gal Z91.014
Revise	- cardiovascular renal ~~-see Hypertension, cardiorenal~~ – see Syndrome, cardiorenal
	- Korsakoff (-Wernicke) (nonalcoholic) F04
Revise	- - alcoholic ~~F10.26~~ F10.9-
Add	- - - with dependence F10.2-
Add	- pediatric acute-onset neuropsychiatric (PANS) D89.89
Add	- post -polypectomy coagulation K91.89
	- Usher Q99.819
Revise	- - specified NEC ~~Q99.814~~ Q99.818
	- - type 1 Q99.811
	- - type 2 Q99.812
	- - type 3 Q99.813
Revise	- - type 4 ~~Q99.814~~ Q99.818
	- Wernicke-Korsakoff (nonalcoholic) F04
Revise	- - alcoholic ~~F10.26~~ F10.9-
Add	- - - with dependence F10.2-
	Thrombophlebitis I80.9
Revise	- leg – see Phlebitis, leg ~~I80.3~~
Delete	- - ~~superficial I80.0~~
Revise	- lower extremity - see Phlebitis, leg ~~I80.299~~
Add	- - ~~deep (see also Phlebitis, leg, deep) I80.29~~
	Thrombosis, thrombotic (bland) (multiple) (progressive) (silent) (vessel) I82.90
	- intracranial (arterial) I66.9
Revise	- - venous sinus (any) ~~G08~~ I67.6
Add	Transaminasemia R74.01
	Transection
Delete	~~Transaminasemia R74.01~~
Revise	Wernicke-Korsakoff's syndrome or psychosis (alcoholic) ~~F10.96~~ F10.9-
Add	- with dependence F10.2-

	Use (of) - opioid F11.90 - - with
Revise	- - - opioid-associated amnestic syndrome ~~F11.988~~ F11.288

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ICD-10-CM External Cause of Injuries Index
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Inhalation
Revise - food (any type) (into respiratory tract) (with asphyxia, obstruction respiratory tract, suffocation) ~~- see categories T17 and T18~~ T17.92-

Interruption of respiration (by)
Revise - food (lodged in esophagus) ~~- see categories T17 and T18~~ T18.120

Suffocation (accidental) (by external means) (by pressure) (mechanical) - see also category T71
- due to, by
Revise - - food, any type (aspiration) (ingestion) (inhalation) ~~- see categories T17 and T18~~ T17.92-

ICD-10-CM TABLE of NEOPLASMS

All approved modifications will be effective October 1, 2026

Neoplasm	Malignant Primary	Malignant Secondary	Ca in situ	Benign	Uncertain Behavior	Unspecified Behavior
Neoplasm, neoplastic						
Add **femur (any part)**	C40.2-	C79.51				