ICD-10 Coordination and Maintenance Committee Meeting
September 10-11, 2024
Diagnosis Agenda

Zoom Webinar and Dial-In Information

- This meeting will be conducted via Zoom Webinar. The URL to register to join the Zoom Webinar, the password, and the call-in numbers are the same for both days of the meeting. Meeting details for each day are as follows.
- Day 1: September 10, 2024: The meeting will begin promptly at 9:00 AM ET and will end at 5:00 PM ET. Lunch will be held from 12:30 PM to 1:30 PM.
- Day 2: September 11, 2024: The meeting will begin promptly at 9:00 AM ET and will end at 5:00 PM ET. Lunch will be held from 12:30 PM to 1:30 PM.

To minimize feedback to the maximum extent possible, join the meeting using only ONE of the options listed below.

Option 1: Remote participants (attendees wishing to both view slides and ask questions during the Q&A portions of the meeting) must register to join the Zoom Webinar via the web. To register to join this Zoom Webinar conference from a PC, MAC, iPad, iPhone or Android device as well as, connect to the audio portion of the conference:

Register in advance for this webinar:
https://cms.zoomgov.com/webinar/register/WN_8hiZrGNcQYCFuH9P7LCloQ
Webinar ID: 160 744 0104
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1. From your phone, dial U.S.*: 669-254-5252 or 646-828-7666 or 833-568-8864 (Toll Free)
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Passcode: 621302

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Those participating in the Zoom Webinar may ask questions during the Q&A portions of the meeting using the "Raise Hand" feature. If time does not permit you to comment or ask a question during the Q&A session, you may submit comments and questions at any time using the "Q&A" feature.

Remaining questions and comments on proposals presented may be submitted via the ICD-10-CM mailbox at nchsicd10cm@cdc.gov.

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

Diagnosis Topics:

Contents

September 10, 2024, ICD-10 C&M Diagnosis Timed Agenda (Day 1)

**Please dial in 30mins before the topic of interest in the event ahead of schedule. Presentation times subject to change.*

Topic*	Presenter
Welcome, Diagnosis Day 1 **Time: 11:30-11:40amET**	Monica Leonard
Topic: Ectopic Pregnancies **Time: 11:40 – 11:55amET**	Traci Ramirez Dr. Nancy Fang
Topic: Utility Insecurity **Time: 11:55-12:10pmET**	Traci Ramirez Sarah DeSilvey DNP, FNP-C
Topic(s): Baked Egg Tolerance in Egg Allergy **Baked Milk Tolerance in Milk Allergy** **Time: 12:10-12:25pmET**	Cheryl Bullock
LUNCH BREAK *12:30-1:30*	
Topic: Xylazine Associated Wounds **Time: 1:30-1:45pmET**	Shannon McConnell-Lamptey Dr. Daniel Teixeira da Silva
Topic: Ledderhose Disease **Time: 1:45-2:00pmET**	Shannon McConnell-Lamptey Dr. Paul Carroll
Topic: Neovascular Glaucoma **Time: 2:00-2:15pmET**	Shannon McConnell-Lamptey Dr. Michael Repka
Topic: Topical Steroid Withdrawal **Time: 2:15-2:30pmET**	Shannon McConnell-Lamptey Dr. Peter Lio
Topic: Exposure to DES **Time: 2:30-2:45pmET**	Cheryl Bullock Karen Fernandes
Topic: Demodex Blepharitis **Time: 2:45-3:00pmET**	Shannon McConnell-Lamptey
Topic: Abnormal rheumatoid factor and anti-citrullinated protein antibody without rheumatoid arthritis **Time: 3:00-3:15pmET**	Traci Ramirez
Topic: Nipple Ischemia & Nipple Necrosis **Time: 3:15-3:30pmET**	Traci Ramirez Dr. Piltin
Topic: Encounter for Prophylactic removal of fallopian tube(s) **Time: 3:30-3:45pmET**	Traci Ramirez
Topic: Multiple Sclerosis phenotypes **Time: 3:45-4:00pmET**	Shannon McConnell-Lamptey Dr. Silvia Perez-Vilar

Topic: Inflammatory Breast Cancer (IBC) **Time: 4:00-4:15pmET**	Traci Ramirez Dr. Kathy Miller
Topic: Usher Syndrome **Time: 4:15-4:30pmET**	Shannon McConnell-Lamptey
Topic: Genetic NDDs **Time: 4:30-4:45pmET**	David Berglund, MD

September 11, 2024, ICD-10 C&M Diagnosis Timed Agenda (Day 2)

**Please dial in 30mins 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: Amyloid-related imaging abnormalities (ARIA) **Time: 9:10-9:25amET**	David Berglund, MD Danielle Abraham, PhD, MPH
Topic: Hao-Fountain Syndrome **Time: 9:25-9:40amET**	David Berglund, MD
Topic(s): Homozygous Familial Hypercholesterolemia **Time: 9:40-9:55amET**	Cheryl Bullock Patrick Lewis
Topic: Fontan circulation and related complications **Time: 9:55-10:10amET**	David Berglund, MD David Goldberg, MD
Topic: Pyrophosphate Metabolism **Time: 10:10-10:25amET**	David Berglund, MD David Goldberg, MD
Topic: Primary progressive apraxia of speech **Time: 10:25-10:40amET**	Shannon McConnell-Lamptey Dr. Rene Utianski
Topic: Lipodystrophy **Time: 10:40-10:55amET**	Shannon McConnell-Lamptey Lindsay Fourman, MD
Topic: Odontogenic Sinusitis **Time: 10:55-11:10amET**	Shannon McConnell-Lamptey Dr. John Craig
Topic: Limb Girdle Muscular Dystrophy Subtype 2I / R9 **Time: 11:10-11:25amET**	David Berglund, MD Katherine Mathews, MD
Topic: Skin Changes Due to Skin Failure **Time: 11:25-11:40amET**	Shannon McConnell-Lamptey Dr. Diane Krasner
Topic: Diabetes mellitus in remission **Time: 11:40 -11:55amET**	Shannon McConnell-Lamptey
Topic: Leukocyte Adhesion Deficiency **Time: 11:55-12:10pmET**	Cheryl Bullock
Topic: Kabuki Syndrome **Time: 12:10-12:25pmET**	Cheryl Bullock
LUNCH BREAK *12:30-1:30*	

Topic: FOXG1 Syndrome **Time: 1:30-1:45pmET**	David Berglund, MD Elli Brimble
Topic: Blast overpressure **Time: 1:45-2:00pmET**	Traci Ramirez Stephanie Panker, MD
Topic: Postprocedural open wound **Time: 2:00-2:15pmET**	Shannon McConnell-Lamptey Dr. Robert Kucejko
Topic: Lipedema **Time: 2:15-2:30pmET**	Shannon McConnell-Lamptey Dr. Karen Herbst
Topic: Lynch syndrome **Time: 2:30-2:45pmET**	David Berglund, MD Andrea Dwyer
Topic: Hypothalamic Obesity **Time: 2:45-3:00pmET**	David Berglund, MD Jennifer Miller, MD
Topic: ICMPGN **Time: 3:00-3:15pmET**	Traci Ramirez Eileen Brewer, MD
Topic: CACNA1A-NDD **Time: 3:15-3:30pmET**	David Berglund, MD Wendy Chung, MD
Topic: CTNNB1 Syndrome **Time: 3:30-3:45pmET**	David Berglund, MD Wendy Chung, MD
Topic: Addenda **Time: 3:45-4:00pmET**	Traci Ramirez

ICD-10 TIMELINE

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

September 10-11, 2024	The September 2024 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 2024	Recordings and slide presentations of the September 10-11, 2024 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, 2024	New and revised ICD-10-CM and ICD-10-PCS codes go into effect along with MS-DRG changes. Final addendum 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 11, 2024	**Deadline for receipt of public comments on proposed new codes discussed at the September 10-11, 2024 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on April 1, 2025, if applicable.**
November 2024	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, 2025 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/latest-news

November 15, 2024	**Deadline for receipt of public comments on proposed new codes and revisions discussed at the September 10-11, 2024 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2025.**
December 6, 2024	**Deadline for requestors: Those members of the public requesting that topics be discussed at the March 18-19, 2025 ICD-10 Coordination and Maintenance Committee Meeting must have their requests submitted to CMS for procedures and to NCHS for diagnosis 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. The ICD-10 Coordination and Maintenance Committee will make efforts to accommodate requested early implementation dates if the following criteria are met; new diseases or disorders and / or public health importance or emergency.
January 2025	Federal Register notice for the March 18-19, 2025 ICD-10 Coordination and Maintenance Committee Meeting will be published. This will include the tentative agenda.
February 2025	Tentative agenda for the Procedure portion of the March 18, 2025 ICD-10 Coordination and Maintenance Committee Meeting will be posted on CMS webpage at: https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html Tentative agenda for the Diagnosis portion of the March 19, 2025 ICD-10 Coordination and Maintenance Committee Meeting will be posted on NCHS homepage at: https://www.cdc.gov/nchs/icd/icd-10-maintenance/meetings.html
February 1, 2025	ICD-10 MS-DRG Grouper software and related materials posted on CMS webpage at: https://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/AcuteInpatientPPS/MS-DRG-Classifications-and-Software

February 1, 2025	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/ICD10/
February 1, 2025	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/ICD10/
March 18-19, 2025	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 2025	Recordings and slide presentations of the March 18-19, 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/ICD10/C-and-M-Meeting-Materials.html
April 1, 2025	Any new or revised ICD-10 codes or addenda previously announced will be implemented on April 1, 2025.
April 18, 2025	**Deadline for receipt of public comments on proposed new codes and revisions discussed at the March 18-19, 2025 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2025.**
April 2025	Notice of Proposed Rulemaking to be published in the Federal Register as mandated by Public Law 99-509. This notice will include references to the FY 2026 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/medicare-fee-for-service-payment/acuteinpatientpps

May 16, 2025	**Deadline for receipt of public comments on proposed new codes and revisions discussed at the March 18-19, 2025 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on April 1, 2026, if applicable.** **Deadline for receipt of public comments on proposed new diagnosis codes and revisions discussed at the March 18-19, 2025 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2026**.
May/June 2025	Final addendum 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/ICD10/index.html
June 6, 2025	**Deadline for requestors: Those members of the public requesting that topics be discussed at the September 9-10, 2025 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. The ICD-10 Coordination and Maintenance Committee will make efforts to accommodate requested early implementation dates if the following criteria are met: new diseases or disorders and/or public health importance or emergency.
July 2025	Federal Register notice for the September 9-10, 2025 ICD-10 Coordination and Maintenance Committee Meeting will be published. This will include the tentative agenda.
August 1, 2025	Hospital Inpatient Prospective Payment System final rule expected to be published in the Federal Register as mandated by Public Law 99-509. This rule will also include links to all the final codes to be implemented on October 1, 2025.

	This rule can be accessed at: https://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/AcuteInpatientPPS/index.html
August 2025	Tentative agenda for the Procedure portion of the September 9, 2025 ICD-10 Coordination and Maintenance Committee Meeting will be posted on the CMS webpage at – https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html Tentative agenda for the Diagnosis portion of the September 10, 2025 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 9-10, 2025	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.
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/ICD10/C-and-M-Meeting-Materials.html
October 1, 2025	New and revised ICD-10-CM and ICD-10-PCS codes go into effect along with MS-DRG changes. Final addendum 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/ICD10/

Contact Information
National Center for Health Statistics
ICD-10-CM Coordination and Maintenance Committee

Comments on the diagnosis proposals presented at the ICD10 Coordination and Maintenance Committee meeting should be sent to the following email address: **nchsicd10CM@cdc.gov**

Monica Leonard, Team Lead	(404)718-6443
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 virtually attend 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 meetings. 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 meeting 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.

Abnormal rheumatoid factor or anti-citrullinated protein antibody without rheumatoid arthritis

A proposal was previously presented at the September 2022, September 2023 and March 2024 ICD10 Coordination and Maintenance committee meetings. Based on public comments, revisions to the proposal have been made for reconsideration.

Rheumatoid arthritis (RA) is a well-known autoimmune condition that is characterized by the presence of inflammatory arthritis (IA)[1]. Furthermore, in up to 80% of individuals with RA there are also abnormalities of circulating biomarkers including but not limited to the autoantibodies rheumatoid factor (RF) and anti-citrullinated proteins antibody (called 'ACPA') a subset of which that is commonly tested in clinical is anti-cyclic citrullinated protein (or peptide) antibodies (called 'anti-CCP')[2].

The current paradigm for the diagnosis and treatment of RA is for a clinician to identify joint findings that are determined to be IA, diagnose that as RA based on clinical, laboratory and radiographic features, and initiate treatment. Furthermore, this is the typical clinical situation when the existing ICD-10-CM codes for RA (e.g. M06/M05) are applied. Notably, RA may be formally classified according to established criteria[3, 4]; however, in clinical practice RA is a clinical diagnosis that may or may not meet these criteria.

However, it is now well-established that RA-related immunologic tests such as RF and ACPA/anti-CCP can be present in individuals in absence of and prior to the appearance of IA, and predictive of future onset of clinical RA[5]. Furthermore, individuals who have abnormal RA-related immunologic tests without IA are identified in growing numbers in clinical care. Notably, these individuals may have symptoms such as joint pain, stiffness or swelling, but no other objective evidence of IA[6-8]. There are current recommendations for medical follow-up and lifestyle changes (e.g., smoking cessation) that can be applied to these individuals. In addition, the predictive ability of RF and ACPA for future clinical RA has underpinned multiple clinical observational studies and prevention trials in RA[9-13]. In particular, two studies have recently demonstrated that abatacept significantly reduces rate of progression from an anti-CCP positive state to clinical RA[14, 15]. Based on this, there are efforts underway to obtain approval for pharmacologic therapy in this condition to prevent or delay the future onset of clinical RA.

There are ICD-10-CM codes that can be used to designate clinical RA (e.g. M05.79), autoantibody positivity with joint symptoms/arthralgia (e.g. M25.50) as well as designations within the R category for RF and anti-CCP positivity (which are both captured under code R76.8). However, there is not a unique code for individuals who may have abnormal RA-related autoantibodies, but <u>not</u> have a diagnosis of clinical RA. As such, the introduction of a new code to accurately designate an

individual who has abnormal immunologic tests of RF and/or anti-CCP will facilitate clinical designation and care of these individuals, as well as facilitate clinical research.

This proposal was submitted by The University of Colorado, Division of Rheumatology and supported by the American College of Rheumatology (ACR).

References

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TABULAR MODIFICATIONS

M05 Rheumatoid arthritis with rheumatoid factor

New code M05.A Abnormal rheumatoid factor and anti-citrullinated protein antibody with rheumatoid arthritis

Add Code first rheumatoid arthritis with rheumatoid factor by site, if known (M05.00 to M05.8A)

R76 Other abnormal immunological findings in serum

R76.8 Other specified abnormal immunological findings in serum

Delete ~~Raised level of immunoglobulins NOS~~

New code R76.81 Abnormal rheumatoid factor and anti-citrullinated protein antibody without rheumatoid arthritis

Add Abnormal anti-cyclic citrullinated protein antibody and rheumatoid factor

Add Abnormal anti-CCP

Add Excludes1: rheumatoid arthritis with rheumatoid factor (M05.-)

New code R76.89 Other specified abnormal immunological findings in serum

Add Raised level of immunoglobulins NOS

Amyloid-Related Imaging Abnormalities

Amyloid-related imaging abnormalities (ARIA) are a finding seen with amyloid beta-directed monoclonal antibody treatment in Alzheimer's Disease (AD).

AD is a neurodegenerative disease that results in mild cognitive impairment and dementia (1). Pathological features of AD include amyloid beta plaques and tau neurofibrillary tangles (1). In 2020, AD affected an estimated 6.07 million individuals 65 years of age and older in the United States (2). Recently, the amyloid beta-directed monoclonal antibodies aducanumab (2021, accelerated approval) and lecanemab (2023, traditional approval) received U.S. Food and Drug Administration approval for the treatment of AD (3).

ARIA is thought to be the result of reduced vascular integrity due to an inflammatory response and impaired perivascular amyloid beta clearance (4-6). ARIA presents in two forms, ARIA-edema/effusion (ARIA-E) and ARIA-hemosiderin deposition (ARIA-H) (4-6). 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 (4-6). ARIA-H is a consequence of vascular leakage of blood degradation products into brain parenchyma and leptomeningeal/subpial space presenting as microhemorrhages and superficial siderosis, respectively (4-6). Both ARIA-E and ARIA-H are identified by neuroimaging (4-6). It is important to distinguish asymptomatic from symptomatic ARIA as it impacts clinical decision-making and treatment (5-7).

The term ARIA is usually reserved for neuroimaging findings in the context of amyloid beta-directed monoclonal antibody treatment of AD that may be asymptomatic or associated with a range of neurological findings and symptoms. However, similar imaging changes may occur spontaneously with diseases leading to amyloid accumulation in the brain, such as cerebral amyloid angiopathy (CAA) and AD, which may co-exist (8, 9). Although neuroimaging findings of ARIA and CAA likely share a similar pathophysiology (4-6), CAA has distinct diagnostic criteria that relies on neuroimaging and/or pathology findings (10). In addition, ARIA and CAA may have different clinical presentations when symptomatic and may be managed differently (5, 6). 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 (4-6). There is currently an ICD-10-CM code for CAA (I68.0), but there are no ICD-10-CM codes for ARIA.

The Center for Drug Evaluation and Research, U.S. Food and Drug Administration, has proposed new codes for ARIA in the same ICD-10-CM category as the code for CAA. When applicable, these codes should be used in conjunction with the ICD-10-CM code for AD. The proposed codes distinguish ARIA-E from ARIA-H and capture whether ARIA is symptomatic.

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.

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TABULAR MODIFICATIONS

G30 Alzheimer's disease

Use additional code, if applicable, to identify:

Add amyloid-related imaging abnormalities (ARIA) (I68.81-)

I68 Cerebrovascular disorders in diseases classified elsewhere

I68.8 Other cerebrovascular disorders in diseases classified elsewhere

New sub-subcategory I68.81 Amyloid-related imaging abnormalities [ARIA]

Add Use additional code, if applicable, for adverse effect of immunoglobin (T50.Z15)

New code I68.810 Amyloid-related imaging abnormalities with edema/effusion [ARIA-E], symptomatic

Add		Use additional code, if applicable, for associated symptoms
New code	I68.811	Amyloid-related imaging abnormalities with edema/effusion [ARIA-E], asymptomatic
New code	I68.812	Amyloid-related imaging abnormalities with hemosiderin deposition [ARIA-H] microhemorrhage, symptomatic
Add		Use additional code, if applicable, for associated symptoms
New code	I68.813	Amyloid-related imaging abnormalities with hemosiderin deposition [ARIA-H] microhemorrhage, asymptomatic
New code	I68.814	Amyloid-related imaging abnormalities with hemosiderin deposition [ARIA-H] superficial siderosis, symptomatic
Add		Use additional code, if applicable, for associated symptoms
New code	I68.815	Amyloid-related imaging abnormalities with hemosiderin deposition [ARIA-H] superficial siderosis, asymptomatic
New code	I68.819	Amyloid-related imaging abnormalities, unspecified
New code	I68.89	Other cerebrovascular disorders in diseases classified elsewhere

Baked Egg Tolerance in Egg Allergy

Egg allergy affects an estimated 0.5% to 2.5% of children younger than 5 years of age.[1] While 80% of children eventually outgrow egg allergy, and most were thought to develop tolerance by school age, studies indicate that some children are retaining egg allergy into adolescence.[2] It appears that the longer the egg allergy persists, the less likely tolerance will develop. Thus, it is important to understand individualized prognosis of egg allergy and develop clinical management that will improve the quality of life of egg-allergic children and, ideally, promote earlier tolerance development.

This proposal was presented at the September 2023 and March 2024 ICD-10 Coordination and Maintenance Meeting. In response to public comments, changes have been made and noted in **bold.**

It has become clear that different phenotypes of egg allergy exist, and these appear to be associated with different prognoses. There are egg-allergic patients who tolerate egg in baked products (baked egg-tolerant) but still react to stove-top cooked eggs (scrambled, fried, and boiled), and then there are patients who react to all forms of egg including well-baked products (baked egg-reactive). Baking modifies egg proteins and makes them less allergenic by destroying conformational epitopes and/or blocking epitopes through interactions with the food matrix (e.g., wheat flour).[3,4] This results in decreased IgE binding to egg proteins and increased tolerability. Clinical studies have indicated that a majority, 70-80%, of egg-allergic individuals can tolerate baked egg.[5,6]

Ovalbumin is the predominant protein in egg, making up 54% of egg white (EW), and is heat labile. Ovomucoid makes up only 11% of EW but is considered the more dominant allergen and is heat stable. In one study, higher specific IgE (sIgE) to ovomucoid was associated with reactivity to heated (well-cooked, but not baked) egg and 94% of subjects who reacted to heated egg subsequently tolerated ovomucoid-depleted heated egg.[7] Many studies have looked at using sIgE levels to total EW and components, such as ovomucoid, and/or skin prick testing to egg and components as a way to predict baked egg reactivity, however consistent cut-offs have not been found.[8,9] Additional studies are ongoing to find a biomarker for baked egg reactivity.[10]

In the meantime, baked egg tolerability is typically assessed clinically. Either the patient is already tolerating baked egg in their diet at the time of evaluation or an oral food challenge to a baked egg product (most often a muffin) is offered under supervision. Providers may use history, severity of past reactions, and testing as a guide for who to offer a baked egg challenge. Even if patients do not initially tolerate baked egg, tolerance may develop with time and regular reassessment is suggested.[11]

Egg-allergic children that tolerate baked egg appear to be more likely to outgrow their egg allergy.[12,13] Studies have shown that predominant IgE binding to ovomucoid, particularly sequential or linear epitopes, is associated with persistent egg allergy.[14,15] Those epitopes are thought to be less effected by heating or baking and matrix effects. It is possible that egg-allergic children without

predominant ovomucoid IgE binding are more likely to tolerate baked egg and be naturally predisposed to outgrowing their allergy, representing a more transient egg allergy.

There is also evidence that regular ingestion of baked egg in the diet may help children outgrow their egg allergy.[11,13,16] In one study, subjects ingesting baked egg regularly were 14.6 times more likely than subjects in the comparison group (P< .0001) to develop regular egg tolerance, and they developed tolerance earlier (median 50.0 vs 78.7 months; P< .0001). Baked egg ingestion was associated with immunologic changes, including decreasing skin prick testing to egg and egg-specific IgE levels, and increasing egg-specific IgG4 levels.[11] These immunologic changes parallel those seen in the natural resolution of egg allergy. The authors concluded that initiation of a baked egg diet accelerates the development of regular egg tolerance compared with strict avoidance. Therefore, differentiation of the different phenotypes of egg allergy can lead to different management, specifically earlier and sustained exposure to baked egg in tolerant patients as a form of possible treatment for egg allergy.

While tolerance to baked egg is associated with a better prognosis - i.e., more likely to outgrow their egg allergy, reactivity to baked egg is associated with poorer outcomes. Those that react to baked egg are less likely to outgrow their egg allergy and more likely to react to small amounts of egg. Thus, baked egg reactive patients may be considered for specific therapies due to this and a code to identify them would be beneficial.

Current ICD-10-CM codes include anaphylactic reaction due to eggs and a historical report of allergy to eggs (Z code). Neither specify tolerance of the baked form of egg, which a majority of egg-allergic patients tolerate, and which is associated with a better prognosis and increased likelihood of tolerance development. An additional ICD-10-CM code to signify baked egg tolerance would help to identify patients who may benefit from intervention and who are likely to outgrow their egg allergy, warranting close follow-up, repeat testing, and tolerance assessment.

This proposal is submitted jointly by physicians and coding professionals within the American Academy of Allery, Asthma & Immunology (AAAAI).

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TABULAR MODIFICATIONS

T78 Adverse effects, not elsewhere classified
Excludes2:complications of surgical and medical care NEC (T80-T88)
T78.0 Anaphylactic reaction due to food
Anaphylactic reaction due to adverse food reaction
Anaphylactic shock or reaction due to nonpoisonous foods
Anaphylactoid reaction due to food

T78.08 Anaphylactic reaction due to eggs

New code T78.080 Anaphylactic reaction due to egg with tolerance to baked egg

Add Excludes1: Anaphylactic reaction due to egg with reactivity to baked egg (T78.081)

New code T78.081 Anaphylactic reaction due to egg with reactivity to baked egg

Add Excludes1: Anaphylactic reaction due to egg with tolerance to baked egg (T78.080)

New code T78.089 Anaphylactic reaction due to eggs, unspecified

T78.1 Other adverse food reactions, not elsewhere classified
Use additional code to identify the type of reaction, if applicable
Excludes1: anaphylactic reaction or shock due to adverse food reaction (T78.0-)
anaphylactic reaction due to food (T78.0-)
bacterial food borne intoxications (A05.-)

Excludes2: allergic and dietetic gastroenteritis and colitis (K52.29)
allergic rhinitis due to food (J30.5)
dermatitis due to food in contact with skin (L23.6, L24.6, L25.4)
dermatitis due to ingested food (L27.2)
food protein-induced enterocolitis syndrome (K52.21)
food protein-induced enteropathy (K52.22)

New subcategory T78.12 Other adverse food reaction due to eggs

New code T87.120 Other adverse food reaction due to egg with tolerance to baked egg
Add Excludes1: Other adverse food reaction due to egg with reactivity to baked egg (T78.121)

New code T78.121 Other adverse food reaction due to egg with reactivity to baked egg
Add Excludes1: Other adverse food reaction due to egg with tolerance to baked egg (T78.120)

New code **T78.129 Other adverse food reaction due to egg with baked egg tolerance/ reactivity, unspecified**

New code T78.19 Other adverse food reactions, not elsewhere classified

Z91 Personal risk factors, not elsewhere classified
Z91.0 Allergy status, other than to drugs and biological substance
Z91.01 Food allergy status
Z91.012 Allergy to eggs

New code Z91.0120 Allergy to eggs, unspecified

New code Z91.0121 Allergy to eggs with tolerance to baked egg
Add Excludes1: Allergy to eggs with reactivity to baked egg (Z91.0122)

New code	Z91.0122	Allergy to eggs with reactivity to baked egg
Add		Excludes1: Allergy to egg with tolerance to baked egg (Z91.0121)

Baked Milk Tolerance in Milk Allergy

Cow's milk (CM) allergy affects up to 2% of children younger than 5 years of age.[1] While 80% of children eventually outgrow milk allergy, and most were thought to develop tolerance by school age, studies indicate that some children are retaining milk allergy into adolescence.[2] It appears that the longer the milk allergy persists, the less likely tolerance will develop. Thus, it is important to understand individualized prognosis of milk allergy and develop clinical management that will improve the quality of life of milk-allergic children and, ideally, promote earlier tolerance development.

This proposal was presented at the September 2023 and March 2024 ICD-10 Coordination and Maintenance Meeting. In response to public comments, changes have been made and noted in **bold.**

It has become clear that different phenotypes of milk allergy exist, and these appear to be associated with different prognoses. There are milk-allergic patients who tolerate milk in baked products (baked milk-tolerant) but still react to uncooked milk, and then there are patients who react to all forms of milk including well-baked products (baked milk-reactive). Baking modifies milk proteins and makes them less allergenic by destroying conformational epitopes and/or blocking epitopes through interactions with the food matrix (e.g., wheat flour).[3,4] This results in decreased IgE binding to milk proteins and increased tolerability. Clinical studies have indicated that a majority, 70-80%, of milk-allergic individuals can tolerate baked milk.[5,6]

The predominant protein in CM is casein, making up 80%. Casein is heat stable and is considered the more dominant allergen. Whey makes up 20% of CM protein and is heat labile. Studies have reported that higher specific IgE (sIgE) to casein is associated with reactivity to baked milk.[7,8] Many studies have looked at the using sIgE levels to total CM and components, such as casein, and/or skin prick testing to CM and components as a way to predict baked milk reactivity, however consistent cut-offs have not been found.[9]

In the meantime, baked milk tolerability is typically assessed clinically. Either the patient is already tolerating baked milk in their diet at the time of evaluation or an oral food challenge to a baked milk product (most often a muffin) is offered under supervision. Providers may use history, severity of past reactions, and testing as a guide for who to offer a baked milk challenge. Even if patients do not initially tolerate baked milk, tolerance may develop with time and regular reassessment is suggested.[10]

Milk-allergic children that tolerate baked milk appear to be more likely to outgrow their milk allergy.[10,11] Studies have shown that predominant IgE binding to casein, particularly sequential or linear epitopes, is associated with persistent milk allergy.[12,13] Those epitopes are thought to be less effected by heating or baking and matrix effects. It is possible that milk-allergic children without predominant casein IgE binding are more likely to tolerate baked milk and be naturally predisposed to outgrowing their allergy, representing a more transient milk allergy.

There is also evidence that regular ingestion of baked milk in the diet may help children outgrow their milk allergy. In one study, subjects ingesting baked milk regularly were 16 times more likely than subjects in the comparison group (P< .0001) to develop regular milk tolerance.[10] Baked milk ingestion was associated with immunologic changes, including decreasing skin prick testing to CM, and increasing casein-specific IgG4 levels.[5,10] These immunologic changes parallel those seen in the natural resolution of milk allergy. The authors concluded that initiation of a baked milk diet accelerates the development of regular CM tolerance compared with strict avoidance. Therefore, differentiation of the different phenotypes of milk allergy can lead to different management, specifically earlier and sustained exposure to baked milk in tolerant patients as a form of possible treatment for milk allergy.

While tolerance to baked milk is associated with a better prognosis - i.e., more likely to outgrow their milk allergy, reactivity to baked milk is associated with poorer outcomes. Those that react to baked milk are less likely to outgrow their milk allergy and more likely to react to small amounts of milk. Thus, baked milk reactive patients may be considered for specific therapies due to this and a code to identify them would be beneficial.

Current ICD-10-CM codes include anaphylactic reaction due to milk and dairy products, and a historical report of allergy to milk products (Z code). Neither specify tolerance of the baked form of milk, which a majority of milk-allergic patients tolerate, and which is associated with a better prognosis and increased likelihood of tolerance development.

An additional ICD-10-CM code to signify baked milk tolerance would help to identify patients who may benefit from intervention and who are likely to outgrow their milk allergy, warranting close follow-up, repeat testing, and tolerance assessment.

This proposal is submitted jointly by physicians and coding professionals within the American Academy of Allergy, Asthma & Immunology (AAAAI).

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3. Nowak-Wegrzyn A, Fiocchi A. Rare, medium, or well done? The effect of heating and food matrix on food protein allergenicity. Curr Opin Allergy Clin Immunol 2009;9:234-7.
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TABULAR MODIFICATIONS

T78 Adverse effects, not elsewhere classified
Excludes2:complications of surgical and medical care NEC (T80-T88)

T78.0 Anaphylactic reaction due to food

T78.07 Anaphylactic reaction due to milk and dairy products

New code — T78.070 Anaphylactic reaction due to milk and dairy products with tolerance to baked milk

Add — Excludes1: Anaphylactic reaction due to milk and dairy products with reactivity to baked milk (T78.071)

New code — T78.071 Anaphylactic reaction due to milk and dairy products with reactivity to baked milk

Add — Excludes1: Anaphylactic reaction due to milk and dairy products with tolerance to baked milk (T78.070)

New code — T78.079 Anaphylactic reaction due to milk and dairy products, unspecified

T78.1 Other adverse food reactions, not elsewhere classified
Use additional code to identify the type of reaction, if applicable
Excludes1: anaphylactic reaction or shock due to adverse food reaction (T78.0-)
anaphylactic reaction due to food (T78.0-)
bacterial food borne intoxications (A05.-)

Excludes2: allergic and dietetic gastroenteritis and colitis (K52.29)
allergic rhinitis due to food (J30.5)
dermatitis due to food in contact with skin (L23.6, L24.6, L25.4)
dermatitis due to ingested food (L27.2)
food protein-induced enterocolitis syndrome (K52.21)
food protein-induced enteropathy (K52.22)

New subcategory T78.11 Other adverse food reactions due to milk and dairy products

New code T78.110 Other adverse food reactions due to milk and dairy products with tolerance to baked milk

Add Excludes1: Other adverse food reaction due to milk and dairy products with reactivity to baked milk (T78.111)

New code T78.111 Other adverse food reaction due to milk and dairy products with reactivity to baked milk

Add Excludes1: Other adverse food reaction due to milk and dairy products with tolerance to baked milk (T78.110)

New code **T78.119 Other adverse food reaction due to milk and dairy products with baked milk tolerance/ reactivity, unspecified**

New code T78.19 Other adverse food reactions to other food, not elsewhere classified

Z91 Personal risk factors, not elsewhere classified
Z91.0 Allergy status, other than to drugs and biological substance
Z91.01 Food allergy status
Z91.011 Allergy to milk products

New code	Z91.0110 Allergy to milk products, unspecified
New code	Z91.0111 Allergy to milk products with tolerance to baked milk
Add	Excludes1: Allergy to milk products with reactivity to baked milk (Z91.0112)
New code	Z91.0112 Allergy to milk products with reactivity to baked milk
Add	Excludes1: Allergy to milk products with tolerance to baked milk (Z91.0111)

Blast overpressure

The term "blast" or "blast overpressure" includes both high level blast (HLB), as is seen in cases where individuals are in close proximity to exploding bombs, such as grenades or IEDs, and low-level blast (LLB), commonly associated with discharge of large caliber weapon systems. The Department of Defense (DOD) has initiated several efforts to better track service members who have been *exposed* to blast in training or operational environments, which may be repetitive in nature. DOD is requesting a unique code for blast exposures to monitor those seeking care in the medical system who have sought care as a result of these blast exposures.

Currently, there are pathophysiological features that can be used to assist in identifying blast as a unique injury, such as neuroinflammation, imaging markers, diffuse axonal injury, and autonomic nervous system activation [1, 2]. However, the clinical manifestations of these microstructural and neuroimaging changes as separate and distinct from other mechanisms of traumatic brain injury (TBI) are still unknown [3]. The DOD defines TBI as an injury event that results in a blow or jolt to the head with concurrent loss of consciousness (LOC), alteration of consciousness (AOC), or post-traumatic amnesia (PTA) around the injury event.

The Traumatic Brain Injury Center of Excellence (TBICoE) of the DOD Defense Health Agency are requesting new codes for HLB and LLB exposure. The codes should be specified to allow capture of LLB and HLB as separate subpopulations. HLB and LLB exposure codes should be used in situations after exposure to either low- or high-level blast that results in evaluation of the patient who may have symptoms requiring medical care, either resolving or persisting, that may include but not be limited to headache, ringing in the ears, hearing loss, nausea, but agnostic to reported LOC/AOC/PTA.

The current lack of blast overpressure code creates confusion and an inability to systematically and accurately survey blast overpressure exposure that may or may not result in a clinically defined injury to brain. As currently written and used, the codes for primary blast injury to brain (S06.8A) are not capturing exposure and not differentiating between HLB and LLB. The exposure to blast often does not result in diagnosable TBI per the current DOD criteria, although some alterations in neurological function have been noted in some military populations [4].

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TABULAR MODIFICATIONS

Y36	Operations of war
New subcategory	Y36.A Blast overpressure in war operations
Add	Code also: type of explosive, if known
Add	accidental detonation of onboard munitions (Y36.140)
Add	explosion of grenade (Y36.290)
Add	explosion of torpedo (Y36.040)
Add	improvised explosive device (Y36.230)
New code	Y36.A1 Low level blast overpressure blast/explosion in war operations
Add	LLB overpressure blast/explosion in war operations
New code	Y36.A2 High level blast overpressure blast/explosion in war Operations
Add	HLB overpressure blast/explosion in war operations
Y37	Military operations
New subcategory	Y37.A Blast Overpressure in military operations
Add	Code also: type of explosive, if known
Add	accidental detonation of onboard munitions (Y37.140)
Add	explosion of grenade (Y37.290)
Add	explosion of torpedo (Y37.040)
Add	improvised explosive device (Y37.230)
New code	Y37.A1 Low level blast overpressure blast/explosion in military operations
Add	LLB overpressure blast/explosion in military operations
New code	Y37.A2 High level blast overpressure blast/explosion in military operations
Add	HLB overpressure blast/explosion in military operations

CACNA1A-Related Neurodevelopmental Disorder

A proposal to create a new ICD-10-CM code for *CACNA1A*-related neurodevelopmental disorder (*CACNA1A*-NDD) was received from Pangkong Fox, PhD, Kellan Weston, PhD, Terry Jo Bichell, PhD, Laina Lusk, MMSc, LCGC, Ingo Helbig, MD, Joanna C. Jen, MD, PhD, and Wendy Chung, MD, PhD.

CACNA1A-related neurodevelopmental disorder (*CACNA1A*-NDD) is caused by disruptions to the *CACNA1A* (Calcium Voltage-Gated Channel Subunit Alpha1 A) gene at the chromosomal region 19p13. The *CACNA1A* gene encodes for the α1 subunit of the P/Q-type voltage-gated calcium channel Cav2.1.[1,2] The Cav2.1 channel is expressed broadly in the central nervous system and functions to mediate depolarization- induced calcium influx into neurons, which is essential for neurotransmitter release.[3,4] The α1 subunit determines the ion specificity and kinetics of the channel.[3]

Mutations in this gene impact the calcium current flow through the channel leading to neuronal dysfunction.[3,5] *CACNA1A* is scored as among the 2% of genes which are most intolerant to mutation in the human genome.[6]

CACNA1A-NDD has been estimated to affect 1 in 11,700 individuals,[7] indicating a global population of nearly 700,000. This frequency is comparable to other rare genetic diseases with approved ICD-10 codes, such as Angelman syndrome (1:15,000)[8] or MED13L syndrome (1:17,000).[7] *CACNA1A*-NDD is primarily caused by dominant *de novo* mutations, although there are several reports of inheritance from a carrier or mosaic parent.[1]

Initial symptoms of *CACNA1A*-NDD often present in early childhood but can begin in infancy. Spinocerebellar Ataxia Type 6 (SCA6) is a distinct adult-onset disease caused by excess trinucleotide repeat expansions and is not considered to be a neurodevelopmental disorder. Therefore, it is not included in *CACNA1A*-NDD (despite involving the same gene). Mutations in *CACNA1A* were first associated with Familial Hemiplegic Migraine Type 1 (FHM1) and Episodic Ataxia Type 2 (EA2),[9] with overlapping clinical manifestations and interictal nystagmus as a common feature. The phenotypic spectrum has been further expanded within the last 10-15 years because of the availability of unbiased genetic testing to interrogate the genome to identify spontaneous mutations in patients with complex clinical manifestations without a family history or any preconceived hypothesis of what candidate genes should be screened.[5]

CACNA1A-NDD is more broadly defined by a phenotype of developmental delay/intellectual disability, autism spectrum disorder, language delay, congenital ataxia, hypotonia, eye movement disorders such as nystagmus and paroxysmal tonic upgaze (PTU), severe hemiplegic migraine, cerebellar atrophy, a broad seizure spectrum, and epilepsies including Developmental and Epileptic Encephalopathy 42 (DEE42).[1,10–13] Seizures often correspond with subsequent speech and motor regression, and they are believed to be a trigger for further psychomotor deterioration,[1,5] partially due to the fact that over 50% of seizures in *CACNA1A*- NDD are refractory.[14] Severe hemiplegic

migraines, which can lead to cerebral edema, stroke, and even coma in some patients, are often unpredictable but can sometimes be triggered by relatively minor events such as stress or trivial head trauma.[1,5,15–18] *CACNA1A*-NDD can also be associated with neuropsychiatric disorders such as anxiety, depression, and schizophrenia.[1]

Many therapeutics that are currently in development will target the genetic mechanisms that are unique to *CACNA1A*-NDD.[10] Therefore, creation of a specific ICD-10-CM code will enable improved tracking of care and outcomes for clinical interventions specific to *CACNA1A*-NDD. An ICD-10 code will also be critical for developing and executing protocols for standards of care for *CACNA1A*-NDD. Severe seizures frequently trigger regression of development and skills; therefore, timely initiation of an effective antiepileptic treatment is crucial. Additionally, hemiplegic migraines require immediate abortive measures to prevent visits to emergency departments, hospitalization, and long-term regression. A robust treatment and rescue plan for *CACNA1A*-related hemiplegic migraine has been developed by the CACNA1A Foundation and expert clinicians in order to prevent life-threatening outcomes like cerebral edema and coma. An ICD-10-CM code will also help to avoid the use of medications contraindicated in *CACNA1A*-NDD. For example, 4-aminopyridine is commonly used to treat congenital ataxia and EA2.[19,20] However, it can also lower seizure threshold,[21] which could be detrimental to someone with both ataxia and epilepsy. There is also anecdotal evidence that Lamictal, a commonly used anti-epileptic, can exacerbate ataxia in *CACNA1A*-NDD.

The biological mechanisms by which *CACNA1A* mutations cause this broad spectrum of clinical manifestations is still unclear. An ICD-10-CM code will enable better identification and tracking of symptoms and co-morbidities across genotypes, in turn enabling development of therapies that target specific *CACNA1A* variants or groups of variants and enhancing the ability to improve care by better prediction of phenotypic outcome for a given person's mutation.

Finally, an ICD-10-CM code will be critical for tracking the progression of *CACNA1A*-NDD phenotypes with age. While it is established that pathogenic variants cause age-dependent phenotypes,[5] only small adult populations have been described thus far. For example, the prevalence of seizures is as high as 60-70% in young patients but as low as 20-25% when the cohort includes more adults; however, it is unknown whether seizures truly diminish with age or because few adults have been characterized to date.[10] Longitudinal monitoring of clinical phenotypes will enable the establishment of standards of care for these individuals across the lifespan. A unique ICD-10-CM code will serve to track the quality, safety, and efficacy of care for individuals with *CACNA1A*- NDD.

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TABULAR MODIFICATIONS

Note that the new section and category below are proposed in a separate topic, and this proposal would add a specific code for CACNA1A-NDD within this structure. Certain other proposed new codes are not shown here.

New section		Genetic disorders, not elsewhere classified (QA0-QA1)
New category	QA0	Neurodevelopmental disorders related to specific genetic pathogenic variants
Add		Code also any associated disorders, such as:
Add		attention-deficit hyperactivity disorders (F90.-)
Add		autism spectrum disorder (F84.0)
Add		epilepsy, by specific type (G40.-)
Add		intellectual disabilities (F70-F79)
Add		pervasive developmental disorders (F84.-)

New subcategory	QA0.0	Neurodevelopmental disorders related to pathogenic variants in specific genes	
New sub-subcategory	QA0.01	Neurodevelopmental disorders related to pathogenic variants in certain specific genes	
New sub-subcategory	QA0.010	Neurodevelopmental disorders, related to pathogenic variants in ion channel genes	
New code	QA0.0102	CACNA1A-related neurodevelopmental disorder	

CTNNB1 Syndrome

A proposal to create a new ICD-10-CM code for *CTNNB1* Syndrome was received from: Dr. Wendy Chung, Dr. Drew H. Scoles, Dr. Jennifer Bain, CTNNB1 Connect and Cure, Dr. Terry Jo Bichell, and Dr. Kellan Weston.

CTNNB1 syndrome is a severe neurodevelopmental disorder caused by disruption to the Beta-catenin (*CTNNB1)* gene which is located at the 3q 22.1 chromosomal region. *CTNNB1* syndrome was previously known as *CTNNB1* Neurodevelopmental Disorder, and "neurodevelopmental disorder with spastic diplegia and visual defects" (NEDSDV).

Germline disruptions in *CTNNB1* that cause pathogenic loss of function of the Beta-catenin protein are responsible for the unique phenotypes seen in this disorder.[1] Beta-catenin is a critical component of WNT signaling, a cellular growth pathway responsible for cell growth and migration, cell fate determination, and organogenesis during embryonic development.[2-5] Beta-catenin also plays a key role at synapses by anchoring cell adhesion molecules to regulate synaptic connectivity, and neuron migration.[5,6] Therefore, *CTNNB1* dysregulation is linked to aberrant neuron growth and development;[1,7] however, more accurate clinical data is needed to understand genotype-phenotype correlations.

CTNNB1 syndrome is noted for being the most frequent, recurrent monogenic cause of cerebral palsy.[8,9] Vision impairments are also common to the disorder: ~83% of individuals have ophthalmologic deficits, including strabismus, refractive errors, and exudative vitreoretinopathy.[7] Other core clinical phenotypes include moderate to severe intellectual disability, developmental delays, speech delays, behavior problems, growth abnormalities, muscle dystonia, vision impairments, craniofacial dysmorphism including microcephaly, a bulbous nasal tip and thin upper lip, and an abnormal hair pattern.[10,11] Muscle tone abnormalities are particularly prevalent in this disorder: ~86% of individuals show truncal hypotonia early in their development, while at the same time, ~78% of individuals develop peripheral hypertonia and spasticity.[10,11] There are also a rapidly growing number of reports of individuals with spinal abnormalities, specifically tethered spinal cords and cysts in the spinal cord.[7,12,13] Congenital heart deformities are also emerging as a feature of *CTNNB1,* with a recent report showing that 5 of 19 individuals had heart defects.[14]

To date there are over 400 individuals with *CTNNB1* mutations reported in the literature,[11] and prevalence has been estimated to be as high as 1:30,000.[15] The disorder almost always occurs as a *de novo* genetic event, and inheritance of the gene variant from a parent has only been observed in cases when a parent is mosaic.[7] CTNNB1 Syndrome diagnoses occur equally in all race and ethnicities and geographic regions.[16,17]

Individuals with *CTNNB1* mutations display heterogeneous phenotypes, with strong overlap with both cerebral palsy and familial exudative vitreoretinopathy (FEVR).[10] *CTNNB1* syndrome individuals need to be closely monitored from an early age for implementing treatments critical for

their care.[10] A tethered spinal cord requires early intervention to prevent debilitating nerve damage.[13] Exudative vitreoretinopathy and other ophthalmologic issues need to be treated very early in life to prevent vision loss and blindness.[10] Genetic counseling, speech therapy, feeding therapy, and applied behavior analysis are also frequently needed throughout different stages of life.[12]

At this time, ICD-10-CM coding for *CTNNB1* syndrome involves coding separate associated conditions that are present, and most often coded is cerebral palsy (G80.-). However, none of the codes enable separating patients with this syndrome as a distinct population apart from e.g. those with cerebral palsy caused by other factors. Thus, it is not possible to use this general code or others to capture epidemiological factors for this rare genetic disorder.

Creation of a specific ICD-10-CM code for *CTNNB1* syndrome would aid in: 1) providing a way to differentiate *CTNNB1* syndrome from other disorders and enable initiating early intervention in pediatric patients; 2) improving clinical care by making it possible to track outcomes from clinical interventions; and 3) identifying the clinical locations providing care to patients with the disorder.[18]

Given the involvement of multiple systems and congenital malformations that can be present, it is proposed to create a specific code for *CTNNB1* syndrome at category Q87, Other specified congenital malformation syndromes affecting multiple systems, within chapter 17.

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TABULAR MODIFICATIONS

Q87 Other specified congenital malformation syndromes affecting multiple systems

Q87.8 Other specified congenital malformation syndromes, not elsewhere classified

New code	Q87.88	CTNNB1 syndrome
Add		Code also, if applicable, associated conditions such as:
Add		cerebral palsy (G80.-)
Add		congenital heart malformations (Q20.0-Q24.9)
Add		developmental disorder of speech and language (F80.-)
Add		exudative retinopathy (H35.02-)
Add		intellectual disability (F70-F79)
Add		microcephaly (Q02)

Demodex blepharitis

This topic was presented at the March 2024 ICD10 Coordination and Maintenance meeting and based on comments received during the public comment period it is being represented for consideration. Changes are indicated in **bold**.

Blepharitis is the inflammation of the eyelids causing irritation and redness. It has classically been categorized as anterior blepharitis or posterior blepharitis with the cilium or eyelashes as the landmark. Anterior blepharitis has been further subcategorized by presumed causation into staphylococcal blepharitis, seborrheic blepharitis, and acne rosacea-associated blepharitis. Additionally, nearly 70% of all cases are due to demodex infestation, which leads to demodex blepharitis by acting as a vector for harmful bacteria [3,4,5]. Demodex blepharitis is common among all people of all ages, races, ethnicities, and genders.

Demodex, a genus of tiny parasitic mites that live in or near hair follicles of mammals, are among the smallest of arthropods with two species *Demodex folliculorum* and *Demodex brevis* typically found on humans. Symptoms of demodex blepharitis include itching and redness, ocular pain and burning, foreign body sensation, dryness, lacrimation, purulence, irritation, loss of lashes, matted or crusty lashes, and blurry vision. Research has shown that the overwhelming majority of demodex blepharitis patients have difficulty negotiating daily activities. However, it is underdiagnosed, undertreated, and often misdiagnosed despite chronically persisting signs and symptoms that often require multiple visits to an eye care practitioner.

Diagnosis is complex because demodex mites reside on the eyelids of both healthy patients and those experiencing symptoms of blepharitis. No clinical standard currently exists that determines what level of demodex infestation causes blepharitis. Additionally, the symptoms of demodex blepharitis are in alignment with those of other ocular diseases, thus making it difficult for eye care practitioners to pinpoint. Current management options include lid scrubs, tea tree oil, warm compresses, antibiotic/steroid combinations and microblepharoexfoliation. However, these treatments have low efficacy and eradication rates are low. Recently, the first and only FDA-approved treatment has been developed for the treatment of demodex blepharitis.

The American Optometric Association is requesting new codes to capture this significant condition and to facilitate research.

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TABULAR MODIFICATIONS

B88 Other infestations

B88.0 Other acariasis

Delete ~~**Acarine dermatitis**~~

Delete ~~**Dermatitis due to Demodex species**~~

Delete ~~**Dermatitis due to Dermanyssus gallinae**~~

Delete ~~**Dermatitis due to Liponyssoides sanguineus**~~

Delete ~~**Trombiculosis**~~

New code **B88.01 Infestation by Demodex mites**

Add **Demodex folliculorum infestation**

Add **Code also, if applicable eyelid inflammation (H01.8-)**

New code **B88.09 Other acariasis**

Add **Acarine dermatitis**

Add **Dermatitis due to Demodex species**

Add **Dermatitis due to Dermanyssus gallinae**

Add **Dermatitis due to Liponyssoides sanguineus**

Add **Trombiculosis**

H01 Other inflammation of eyelid

H01.8 Other specified inflammation of eyelid

Add Code also, if applicable, infestation by Demodex mites (B88.01)

New code H01.81 Other specified inflammation of right upper eyelid

New code H01.82 Other specified inflammation of right lower eyelid

New code H01.83 Other specified inflammation of right eye, unspecified eyelid

New code	H01.84	Other specified inflammation of left upper eyelid
New code	H01.85	Other specified inflammation of left lower eyelid
New code	H01.86	Other specified inflammation of left eye, unspecified eyelid
New code	H01.89	Other specified inflammation of unspecified eye, unspecified eyelid
New code	H01.8A	Other specified inflammation of right eye, upper and lower eyelids
New code	H01.8B	Other specified inflammation of left eye, upper and lower eyelids

Disorders of Pyrophosphate Metabolism

A proposal to create a new ICD-10-CM code for specific disorders of pyrophosphate metabolism was received from Dr. David Goldberg, Professor Pediatrics, Perelman School of Medicine at the University of Pennsylvania.

Pyrophosphates are molecules of phosphorous and oxygen (salts and esters of phosphoric acid) generated in cells from the breakdown of adenosine triphosphate (ATP) to Adenosine monophosphate (AMP). They play a key role in cellular metabolism, particularly in the regulation of calcium-phosphate mineral deposition and the facilitation of key signaling pathways. Abnormalities in pyrophosphate metabolism cause serious medical issues.[1,2]
Disorders of Pyrophosphate Metabolism

1. ENPP1 Deficiency- GACI and ARHR2

Ectonucleotide pyrophosphatase/phosphodiesterase 1 (ENPP1) is a protein involved in the production of pyrophosphate. Individuals with ENPP1 Deficiency, who cannot produce or have nonfunctioning ENPP1 protein, suffer from severe symptoms. ENPP1 Deficiency is rare, occurring in 1:64,000 pregnancies.[3]

In infants (age 0-6 months), ENPP1 Deficiency presents as a condition known as Generalized Arterial Calcification of Infancy (GACI) which is characterized by low serum pyrophosphate,[4] pathologic vascular calcification and cardiovascular complications (e.g., heart failure, hypertension). The overall mortality for individuals living with GACI is 55% before the age of 6 months, with almost a quarter dying in utero or being stillborn.[5,6] GACI diagnosis is typically established in infants with cardiovascular symptoms and evidence of arterial calcification or stenosis on imaging (ultrasound or CT scan) and/or genetic confirmation of ENPP1 variation.[7]

Most infants with ENPP1 Deficiency who survive GACI go on to develop Autosomal Recessive Hypophosphatemic Rickets Type 2 (ARHR2), the symptoms of which include rickets, osteomalacia, bone deformity, hearing loss, joint or ligament calcification, pain and fatigue, and impaired growth or development,[8-10] along with elevated fibroblast growth factor 23 (FGF23) levels that causes phosphate wasting.[11] While GACI is primarily managed by pediatric cardiologists and pediatric endocrinologists, ARHR2 is primarily managed by endocrinologists, geneticists and metabolic bone specialists.

2. ABCC6 Deficiency- GACI and PXE

ATP-binding cassette subfamily C member 6 (ABCC6) is a transmembrane protein that transports extracellular ATP into the intracellular space, where it is converted by ENPP1 into pyrophosphate.

ABCC6 Deficiency causes 15% of GACI cases, with similar symptomology as GACI caused by ENPP1 Deficiency.[12-14]

ABCC6 Deficiency can also cause pseudoxanthoma elasticum (PXE), an autosomal recessive disorder that leads to reduced serum pyrophosphate and ectopic mineralization of the elastic tissues in the skin, eyes, and blood vessels.[15] PXE is typified by small yellow papules on the neck and in flexural areas, leaving skin to become loose and wrinkled. The mid-dermal elastic fibers are fragmented, clumped, and calcified. The prevalence of PXE is estimated at between 1 per 100,000 and 1 per 25,000 in the general population, with slight predominance among females.[16] PXE increases risk for peripheral arterial disease, ischemic stroke, and blindness.[17]

3. CD73 Deficiency- ACDC

CD73 (ecto-5′-nucleotidase) is an enzyme that dephosphorylates AMP into adenosine in the same metabolic pathway as ABCC6 and ENPP1. Mutations in CD73 cause arterial and joint calcification manifesting in symptoms and risks similar to those of PXE. The distinct phenotype is called Arterial Calcification due to Deficiency of CD73 (ACDC). The condition has only been described in a few families but may be identified more with increased genetic testing.[2,18]

In the current ICD-10-CM code set, none of these diseases of pyrophosphate metabolism (GACI, ARHR2, PXE, or ACDC) are specifically identified—individually or collectively. When they are coded, they are placed under general categories in at least three separate locations in the ICD-10-CM classification structure in non-specific "other" codes:

- GACI is coded to Q28.8, Other specified congenital malformations of the circulatory system.
- ARHR2 is coded either to code E83.30, Disorder of phosphorus metabolism, unspecified, or to code E83.39, Other disorders of phosphorus metabolism.
- PXE is coded to Q82.8, Other specified congenital malformations of skin.
- ACDC may be coded in one or more of these "other" codes.

In some cases, these conditions may also be coded under I70.9, Other and unspecified atherosclerosis.

This absence of codes specific to these conditions can lead to inconsistent reporting of these conditions across healthcare settings. Furthermore, the paradigm is not organized in such a way that the common cause of these conditions (pyrophosphate deficiency) is recognized.

An internal analysis of these current codes revealed significant challenges in identifying the GACI, ARHR2, and PXE subpopulations from the total population of various conditions captured in these ICD-10-CM codes. The analysis revealed that the patient symptomology, diagnostic and treatment profiles of those coded under the two existing codes (E83.30 and Q82.8) have significant variation compared against expectations for individuals with GACI or ARHR2, suggesting that the true size of the population and the impact of the population on health and resource utilization is being masked by the current codes.

It is proposed to create a new fifth character subcategory for disorders of pyrophosphate metabolism, and six new codes be added to specifically name these conditions and group them together based on their common cause.

While there is an existing subcategory E83.3-, Disorders of phosphorus metabolism and phosphatases, and there are some connections to those conditions, a panel of clinical experts agreed that the pyrophosphate conditions are distinct from other conditions of phosphate metabolism and recommended that a distinct subcategory be created for classifying these disorders.

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TABULAR MODIFICATIONS

E83 Disorders of mineral metabolism

E83.3 Disorders of phosphorus metabolism and phosphatases

Add — Excludes2: disorders of pyrophosphate metabolism (E83.82-)

E83.8 Other disorders of mineral metabolism

New sub-subcategory — E83.82 Disorders of pyrophosphate metabolism

New code — E83.820 Generalized arterial calcification of infancy with unspecified genetic causality

Add — Code also, if applicable, associated conditions such as:
Add — heart failure (I50.-)
Add — other secondary hypertension (I15.8)

New code — E83.821 ENPP1 deficiency causing generalized arterial calcification of infancy

Add — Code also, if applicable, associated conditions such as:
Add — heart failure (I50.-)
Add — other secondary hypertension (I15.8)

New code — E83.822 ENPP1 deficiency causing autosomal recessive hypophosphatemic rickets type 2

New code — E83.823 ABCC6 deficiency causing generalized arterial calcification of infancy

Add — Code also, if applicable, associated conditions such as:
Add — heart failure (I50.-)
Add — other secondary hypertension (I15.8)

New code — E83.824 ABCC6 deficiency causing pseudoxanthoma elasticum

New code E83.825 CD73 deficiency causing arterial calcification

I70 Atherosclerosis

I70.9 Other and unspecified atherosclerosis
Add Excludes2: disorders of pyrophosphate metabolism (E83.82-)

Q28 Other congenital malformations of circulatory system

Q28.8 Other specified congenital malformations of circulatory system
Add Excludes2: disorders of pyrophosphate metabolism (E83.82-)

Q82 Other congenital malformations of skin

Q82.8 Other specified congenital malformations of skin
Add Excludes2: disorders of pyrophosphate metabolism (E83.82-)

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, (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] Non-tubal ectopic pregnancies include cesarean scar ectopic pregnancy, cervical ectopic pregnancy, interstitial ectopic pregnancy, and cornual ectopic pregnancy for which there is no unique ICD-10-CM code. There are unique codes for ovarian ectopic pregnancy, (O00.2-) and abdominal ectopic pregnancy, (O00.0-).

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

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 Interstitial ectopic pregnancy

New sub subcategory O00.51 Interstitial ectopic pregnancy without intrauterine pregnancy

New code O00.511 Right interstitial ectopic pregnancy without intrauterine pregnancy

New code O00.512 Left interstitial ectopic pregnancy without intrauterine pregnancy

New code	O00.519 Unspecified interstitial ectopic pregnancy without intrauterine pregnancy
New sub subcategory	O00.52 Interstitial ectopic pregnancy with intrauterine pregnancy
New code	O00.521 Right interstitial ectopic pregnancy with intrauterine pregnancy
New code	O00.522 Left interstitial ectopic pregnancy with intrauterine pregnancy
New code	O00.529 Unspecified interstitial ectopic pregnancy with intrauterine pregnancy
New subcategory	O00.6 Cornual ectopic pregnancy
New sub subcategory	O00.61 Cornual ectopic pregnancy without intrauterine pregnancy
New code	O00.611 Right cornual ectopic pregnancy without intrauterine pregnancy
New code	O00.612 Left cornual ectopic pregnancy without intrauterine pregnancy
New code	O00.619 Unspecified cornual ectopic pregnancy without intrauterine pregnancy
New sub subcategory	O00. 62 Cornual ectopic pregnancy with intrauterine pregnancy
New code	O00.621 Right cornual ectopic pregnancy with intrauterine pregnancy
New code	O00.622 Left cornual ectopic pregnancy with intrauterine pregnancy
New code	O00.629 Unspecified cornual ectopic pregnancy with intrauterine pregnancy
	O00.8 Other ectopic pregnancy
Delete	~~Cervical pregnancy~~
Delete	~~Cornual pregnancy~~

Encounter for prophylactic removal of fallopian tube(s) for persons with no known genetic/familial risk factors

This proposal was originally presented at the September 2023 and March 2024 ICD-10 Coordination and Maintenance meetings. Changes were made, and additional codes are being requested per public comments from the meetings. The changes are in **bold**.

Ovarian cancer (OC) is among the top 5 deadliest cancers in women. The American Cancer Society estimates that in 2023 about 19,710 new cases of ovarian cancer will be diagnosed; the vast majority of cases (70%) will have high grade serous histology.[1] This amounts to one woman diagnosed with high grade serous cancer every 40 minutes in the US. Despite the name "ovarian cancer," accumulating epidemiological, clinical, pathological, and molecular data over the past 20 years indicate that high grade serous carcinoma primarily originates from microscopic precancers in the fimbriated ends of fallopian tubes, rather than from the ovary itself.[2,3] Given the seemingly insurmountable obstacles to effectively screening for and treating the disease, the medical community and the patients are increasingly interested in the option of ovarian cancer prevention through fallopian tube removal (bilateral salpingectomy).

For the past decade, gynecologic surgeons have used the term *opportunistic salpingectomy* to describe the recommended practice of discussing salpingectomy for the primary prevention of ovarian cancer with post-reproductive women planning to undergo pelvic surgery for another indication (eg, hysterectomy) or as an alternative to tubal ligation for surgical sterilization.[4] While 20% of high grade serous cancer is attributable to genetic risk factors, and genetically high risk women are still advised to have both fallopian tubes and ovaries removed to reduce OC risk upon completion of child-bearing, opportunistic salpingectomy is designed to prevent the 80% of high grade serous cancer that affects women with no known risk factors. There is a lack of clear understanding of predisposing factors in this vast majority of cases diagnosed in women who are seemingly average risk for the disease.[5,6]

What is known is that bilateral salpingectomy substantially decreases ovarian cancer risk. Data from nested case-control and population-based retrospective cohort studies indicate that bilateral salpingectomy reduces the risk of ovarian cancer by at least 65 percent.[7,8] In 2022, Canadian researchers published the first prospective evidence that opportunistic salpingectomy may substantially decrease the incidence of high-grade serous carcinoma in the general population. At the time of 9 years follow-up, no high-grade serous carcinoma was observed among the 25,889 women who underwent opportunistic salpingectomy during hysterectomy or in lieu of tubal ligation for surgical sterilization. This is significantly less than the expected rate as well as the rate seen in the 32,080 women who did not undergo bilateral salpingectomy.[9] Studies that have compared the addition of opportunistic salpingectomy to a gynecological or pelvic procedure without salpingectomy have not found significant differences in ovarian endocrine function, surgical complications, operative time, or length of stay.[10,11] In the US, over a million women undergo hysterectomy or surgical sterilization annually.[12,13] By current projections, universal uptake of salpingectomy during hysterectomy and in lieu of tubal ligation could prevent nearly 2000 deaths

from ovarian cancer per year.[14] Expanding opportunistic salpingectomy to post-reproductive women undergoing non-gynecologic elective abdominal surgeries such as cholecystectomy, hernia repair, appendectomy, and gastrointestinal and urologic operations would at least double the impact of opportunistic salpingectomy on decreasing ovarian cancer incidence and mortality.

A 2020 study demonstrated feasibility of opportunistic salpingectomy at the time of elective laparoscopic cholecystectomy, with 60% of counseled patients accepting salpingectomy, a surgical success rate of 95 out of 105 (93.3%) of enrolled patients, and no attributable surgical complications. Mean additional operating time was 13 minutes.[15] Given that the morbidity of the procedure is low, it can be performed using all available approaches (open, laparoscopic, robotic, vaginal), there is no impact on ovarian function and the acceptance rate is high. Salpingectomy as a practical, population-level approach to ovarian cancer prevention.[4,16,17] Fallopian tube removal for ovarian cancer prevention was publicized in recent media coverage by the New York Times, Washington Post and Scientific American Magazine headlining the importance of empowering people to consider and choose opportunistic salpingectomy, especially when it comes to preventing a cancer for which there is neither adequate screening nor a dependable cure.[18,19,20] It is equally important that providers have the tools to offer it as a standard of care. Updating medical coding to the current standard of care is an immediate action item. Medical coding deficiencies for cancer-preventive surgeries like opportunistic salpingectomy need to be rectified because they endanger patient access and health care clinician engagement.[21] One of the most obvious coding deficiencies is that there is no ICD-10-CM code for an encounter for the purpose of opportunistic salpingectomy. The only code available is Z40.03, which requires risk factors such as an inherited genetic mutation.

Because 80% of ovarian cancers affect women with no known genetic or familial risk, the intervention should be available to all women following the completion of childbearing; it should not be restricted to those with genetic susceptibility for and/or a family history of ovarian cancer. At present, Z40.03 is the only ICD-10-CM code for prophylactic salpingectomy. Use of this singular ICD-10-CM code has been and is restricted to high risk people - people with genetic susceptibility or family history of ovarian cancer. There is no ICD-10-CM code for the risk factor of simply being a human with fallopian tubes. This is a major deficiency because having fallopian tubes is the only identifiable risk for ovarian cancer for most women. Establishing the epidemiological basis for prophylactic salpingectomy as a primary prevention strategy for ovarian cancer, both as a population-wide and as a targeted high-risk strategy, will only be feasible through the generation of separate ICD-10-CM diagnosis codes for prophylactic salpingectomy that differentiate people without known genetic/familial risk of ovarian cancer from those with known genetic/familial risk.

The Gynecologic Oncology Division of the Department of Gynecology and Obstetrics at Johns Hopkins University requested the following tabular modifications. This proposal was reviewed and is supported by the American College of Obstetricians and Gynecologists (ACOG).

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TABULAR MODIFICATIONS

Z15 Genetic susceptibility to disease
Z15.0 Genetic susceptibility to malignant neoplasm
Code first, if applicable, any current malignant neoplasm (C00-C75, C81- C96)
Use additional code, if applicable, for any personal history of malignant neoplasm (Z85.-)
New code Z15.05 Genetic susceptibility to malignant neoplasm of fallopian tube(s)

Z40 Encounter for prophylactic surgery
Excludes1: organ donations (Z52.-)
therapeutic organ removal-code to condition

Z40.0 Encounter for prophylactic surgery for risk factors related to malignant neoplasms
Admission for prophylactic organ removal

Revise
Use additional code to identify risk factor, such as genetic susceptibility to malignant neoplasm Z15.-

Z40.02 Encounter for prophylactic removal of ovary(s)

Delete
~~**Encounter for prophylactic removal of ovary(s) and fallopian tube(s)**~~

Z40.03 Encounter for prophylactic removal of fallopian tube(s)

Z40.8 Encounter for other prophylactic surgery

New code
Z40.81 Encounter for prophylactic surgery for removal of ovary(s) for persons without known genetic/familial risk factors

Add
Encounter for prophylactic oophorectomy for persons without known genetic/ familial risk factors

New code
Z40.82 Encounter for prophylactic surgery for removal of fallopian tube(s) for persons without known genetic/familial risk factors

Add
Encounter for prophylactic salpingectomy for persons without known genetic/familial risk factors

Add
Opportunistic salpingectomy

New code
Z40.89 Encounter for other prophylactic surgery

Z80 Family history of primary malignant neoplasm
Z80.4 Family history of malignant neoplasm of genital organs
Conditions classifiable to C51-C63

New code
Z80.44 Family history of malignant neoplasm of fallopian tube(s)

Z85 Personal history of malignant neoplasm
Z85.4 Personal history of malignant neoplasm of genital organs
Conditions classifiable to C51-C63

New code
Z85.4A Personal history of malignant neoplasm of fallopian tube(s)

Z86 Personal history of certain other diseases
Z86.0 Personal history of in-situ and benign neoplasms and neoplasms of uncertain behavior
Z86.00 Personal history of in-situ neoplasm
Conditions classifiable to D00-D09
New code Z86.00A Personal history of in-situ neoplasm of the fallopian tube(s)

Exposure to Diethylstilbestrol (DES)

The DES Info Association, a public advocacy support and information organization is requesting new ICD-10-CM codes to identify DES exposed individuals. Persons exposed can include females (mothers, daughters, granddaughters) and men (sons and grandsons) who have taken this drug and or exposed (intrauterine). The effects of DES continue to emerge.

The currently used ICD-10-CM code, T38.5- Poisoning by, adverse effect of and underdosing of other estrogens and progestogens lack the specificity to identify this population that is needed for possible diagnosis, treatment and hinders the ability to count the number of persons who are affected by this exposure. New ICD-10-CM codes are needed to correlate to the current medical issues facing the population of DES exposed patients.

Diethylstilbestrol (DES) is a synthetic form of the female hormone estrogen that was prescribed to pregnant women, between 1940–1971 to prevent miscarriage, premature labor, and related complications of pregnancy. Though initially it was recommended for the treatment of high-risk pregnancies, it became increasingly commonly used for routine pregnancies, (*to increase birth weight to create healthy babies*), menopausal issues, to treat vaginitis, and to halt lactation.

In 1971, researchers linked prenatal *(in utero)* DES exposure to cancer of the cervix and vagina (clear cell adenocarcinoma (CCA)) in a small group of young women. In November 1971, the Food and Drug Administration (FDA) issued a Drug Bulletin stating that Diethylstilbestrol was contraindicated for use in pregnancy. Subsequently DES exposure now is linked to additional health issues/conditions in DES exposed. DES is a known endocrine-disrupting chemical, one of a number of substances that interfere with the endocrine system to potentially cause cancer, birth defects, and other developmental abnormalities.

Unique ICD-10-CM codes are needed for the population of patients that were exposed to the drug diethylstilbestrol. Problem pregnancies were noted in DES daughters; these patients are at increased risk for breast cancer and other reproductive cancers. It is important that they are screened as high-risk patients. In addition, new codes for personal and family history of exposure to DES can enhance quality metrics for persons impacted and assist in the development of protocols to ensure appropriate screening and medical care.

The NIH/NCI have identified other health conditions in the DES exposed including cardiovascular disease, increased risk of high cholesterol, hypertension, coronary artery disease, and heart attack, pancreatic disorders, early menopause, osteoporosis, and endometriosis.

Research has demonstrated that the following genes were altered *(Including but not limited to)* during DES exposure: HOXA 9, HOXA 10, HOXA 19, HOXA 11, HOXA 13; Msx2, Wat7a, PLAP, K2.16, Lactotransferrin (LTF). The significance of the effects of DES, as a known carcinogen and teratogen, is significant as it is now having multigenerational concerns. Adverse consequences have been seen in both DES granddaughters and DES grandsons.

Proposed new codes for personal and family history would be of clinical value for data collection and possible treatment pertaining to the ongoing and complex effects of DES exposure.

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TABULAR MODIFICATIONS

Z84 Family history of other conditions

New code Z84.A Family history of exposure to diethylstilbestrol
Add Family history of DES exposure

Z91 Personal risk factors, not elsewhere classified

New code Z91.B Personal risk factor of exposure to diethylstilbestrol
Add Personal risk factor of exposure to DES

Fontan Physiology

A proposal to create a new ICD-10-CM code for Fontan physiology was received from Dr. David Goldberg, Professor Pediatrics, Perelman School of Medicine at the University of Pennsylvania.

Single ventricle congenital heart disease (SV-CHD) is comprised of a heterogenous group of anatomic and functional congenital heart lesions characterized by a native cardiac anatomy that cannot support a two-ventricle circulation. This collection of cardiac lesions may be lethal without intervention and often requires multiple palliative procedures in early infancy. While there is some variability in the early palliative procedures, the Fontan operation is the final planned palliative procedure for all forms of SV-CHD. This surgical procedure creates either an atriopulmonary or total cavopulmonary connection in which the single ventricle is assigned the role of pumping blood to the systemic circulation while the return of systemic venous blood is surgically re-routed directly to the pulmonary arteries.

While SV-CHD is estimated to occur in 1 in 10,000 live births, not all children born with these cardiac lesions will survive up to the Fontan operation. In the present era, approximately 1,100 children ultimately undergo the Fontan procedure in the United States each year, and there are thought to be about 50,000-70,000 individuals alive with the Fontan circulation in the United States today.

The relatively small number of patients living with a Fontan circulation relates to the challenges imposed by the circulation itself. Although the Fontan procedure has been successful in prolonging the life of patients with SV-CHD, it creates an imperfect cardiopulmonary physiology characterized by chronically low cardiac output and elevated central venous pressure. This “Fontan physiology” results in an increasingly well-described set of short- and long-term complications including systolic and diastolic dysfunction, cardiac dysrhythmias, peripheral venous insufficiency, liver and kidney fibrosis, and lymphatic insufficiency, all leading to a high rate of early mortality.[1-5]

Of these complications, Fontan-associated liver disease (FALD) and Fontan-associated lymphatic dysfunction warrant specific attention. These entities are unique to the circulation and result in substantially accelerated morbidity and risk of mortality.

FALD is a consequence of the unique combination of persistent exposure to central venous hypertension and low cardiac output. Manifestations include liver fibrosis / cirrhosis, regenerative arterialized liver nodules, and hepatocellular carcinoma. Numerous studies have demonstrated that the process of developing FALD begins immediately after Fontan completion, and the degree of FALD is related to both duration of exposure to the circulation and to the degree of elevation of venous pressure. In fact, many of those who develop a failed Fontan circulation and require cardiac transplantation also require liver transplantation due to the extent of liver dysfunction from FALD.[4]

Fontan-associated lymphatic dysfunction occurs in about 10% of patients and arises from the inability of the lymphatic system to adapt to the increased production of lymphatic fluid and from the

congestion of the lymphatic vessels, both caused by the high-pressure venous system. Fontan-associated lymphatic dysfunction is best described in two specific clinical scenarios: plastic bronchitis (PB) and protein losing enteropathy (PLE). The pathology leading to Fontan-associated PB involves the formation of fistulous lymphatic connections, typically between hepatic or central lymphatics and the airway. These connections lead to lymphatic leak from the interstitial vessels into the airway and subsequent development of proteinaceous casts that take the shape of the bronchial tree, hence the name plastic bronchitis. PLE is similar, but in this case the fistulous connections are to the gastrointestinal tract leading to profound lymphatic loss through the stool. Historically PLE was associated with a 5-year mortality of nearly 50%, although recent advances of care have led to improved outcomes. While PB and PLE are the most recognized of the Fontan-associated lymphatic dysfunction manifestations, abnormal fistulous lymphatic connections can also occur to other spaces in the body including the peritoneum (chyloperitoneum), the thoracic cavity (chylothorax), and the pericardial space (chylopericardium).

In the current ICD-10-CM coding system there are diagnostic codes for some anatomic variations of SV-CHD in the Q20-23 branches, but there is no ICD-10-CM code specifically for Fontan circulation. While Z-codes of medical history, such as Z87.74, Personal history of congenital malformations of heart and circulatory system, are likely to include individuals with Fontan circulation, these codes are not specific and do not accurately capture only those individuals with this specific circulation and physiology. Similarly, although many of the downstream morbidities of Fontan circulation have existing codes, conditions related to Fontan-associated lymphatic dysfunction are so unique to Fontan circulation that we propose that lymphatic dysfunction merits having its own set of related codes.

Despite the existence of the above referenced codes, there is an important ICD-10-CM coding gap defining individuals as having Fontan circulation. The codes cited above are too broad or generalized to allow for specific determinations of individuals with Fontan circulation. Rather, it mixes them into a population of others who have had congenital interventions that are distinct and separate from those with Fontan circulation. A recent 2023 report by Guo et al. found it difficult to identify individuals with Fontan circulation by using ICD-10-CM codes, requiring advanced electronic health record review using AI to adequately identify populations.[6] This distinction is important given the unique physiologic characteristics of those with a Fontan circulation. In other words, the Fontan circulation is itself a well-defined disease state, regardless of the underlying cardiac anatomy or history of other interventions.

Specific codes for Fontan circulation and for Fontan-associated lymphatic dysfunction are being requested to enable better identification and management of patients at risk for the development of the clinical sequelae unique to this physiology, and who may require unique interventions or treatment because of this physiology.

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TABULAR MODIFICATIONS

I27 Other pulmonary heart diseases
I27.8 Other specified pulmonary heart diseases

New sub-subcategory	I27.84	Fontan related circulation
New code	I27.840	Fontan-associated liver disease [FALD]
New code	I27.841	Fontan-associated lymphatic dysfunction
Add		Code also associated conditions such as:
Add		chylothorax (J94.0)
Add		Fontan associated protein-losing enteropathy (K90.89)
Add		plastic (obstructive) bronchitis (J44.89)
New code	I27.848	Other Fontan-associated condition
New code	I27.849	Fontan related circulation, unspecified

FOXG1 Syndrome

This proposal is based on a request from the FOXG1 Research Foundation, and recently updated data showing higher prevalence estimates than previously known. FOXG1 syndrome is characterized by early onset of global developmental delay which may be severe, with intellectual disability, autistic behavior, dyskinesia and hyperkinetic movements, microcephaly, hypotonia, strabismus, epilepsy, spasticity, and neurobehavioral or psychiatric manifestations (motor stereotypies, impairment of social interaction, abnormal sleep patterns, unexplained episodes of crying, restlessness, and bruxism), along with postnatal growth deficiency, feeding difficulties with poor weight gain, gastroesophageal reflux, and aspiration. Cerebral malformations may be present, as well as cortical visual impairment. Some individuals have kyphosis or scoliosis, and abnormal breathing may also occur. Speech may be absent or minimal.[1,2]

Based on a large study of severe neurodevelopmental disorder cases, it was estimated there was a prevalence of 0.6 to 2.2 FOXG1 patients per 100,000 children or ~420 – 1600 pediatric patients in the United States.[3] Based on the approach to estimating incidence using detailed assessment of mutation types and rates from López-Rivera and their supplemental data, and the approach used by Lemke to apply U.S. birth rates for finding estimated incidence in the U.S. (and 2018 reported data), and following the approach described in the March 2023 C&M proposal related to developmental and epileptic encephalopathies, applied to FOXG1 syndrome, there would be an estimate of about 3.2 (or 2.85 to 3.47) FOXG1 patients per 100,000 births, or about 121 new cases among newborns in the U.S. annually (with caveats as for other disorders that this could be an overestimate or an underestimate).[4,5]

It is proposed to add a specific code for FOXG1 syndrome, to be classified in a new separate category for genetic neurodevelopmental disorders (with further details for this category discussed separately). It is also proposed to also create a new code Neurodevelopmental disorder, related to other genes associated with transcription and gene expression, for classification.[6]

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TABULAR MODIFICATIONS

Note that the new section and category below are proposed in a separate topic, and this proposal would add a specific code for FOXG1 syndrome within this structure. Certain other proposed new codes are not shown here.

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

New category — QA0 Neurodevelopmental disorders related to specific genetic pathogenic variants

Add — Code also, if applicable, any associated conditions, such as:
Add — attention-deficit hyperactivity disorders (F90.-)
Add — autism spectrum disorder (F84.0)
Add — epilepsy, by specific type (G40.-)
Add — intellectual disabilities (F70-F79)
Add — pervasive developmental disorders (F84.-)

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

New sub-subcategory — QA0.01 Neurodevelopmental disorders related to pathogenic variants in certain specific genes

New 6th character subcategory — QA0.015 Neurodevelopmental disorders, related to genes associated with transcription and gene expression

New code — QA0.0151 FOXG1 syndrome
Add — FOXG1-related disorder
Add — FOXG1-related encephalopathy
Add — FOXG1-related neurodevelopmental disorder

New code — QA0.0159 Neurodevelopmental disorder, related to other genes associated with transcription and gene expression

Genetic Neurodevelopmental Disorders

This proposal is a repeat presentation for certain disorders that were previously presented at ICD-10 Coordination and Maintenance Committee meetings both together in March 2024, and separately in 2023. It provides an alternative option for creation of specific codes for a number of genetic neurodevelopmental disorders. There is ongoing progress in understanding of neurodevelopmental disorders, and of underlying genetic causes.

At the March 2023 ICD10 C&M meeting, there were presentations related to the ionotropic glutamate receptor (GRIN1, GRIN2A, GRIN2B, GRIN2D, GRIA1, GRIA2, GRIA3, GRIA4, and GRIK2), SCN2A-related disorders, SLC6A1-related disorders, and STXBP1-related disorders (syntaxin-binding protein 1). Also, a detailed proposal for specific codes for DLG4-related synaptopathy was presented at the September 2023 C&M meeting. It is recognized that these neurodevelopmental disorders may involve a range of associated findings and conditions, and these may include epilepsy, autism spectrum disorders, intellectual disability, and specific learning and developmental impairments, among other things. Further clinical details are available from the prior proposals and from the references included below.

A separate category for genetic neurodevelopmental disorders is proposed in chapter 17 based on public comments previously received. There have been multiple requests to consider creating a section that will enable classification of certain genetic disorders in ICD-10-CM, and this will have the potential to be more broadly applied. However, many genetic disorders are already classified in specific sections and categories of ICD-10-CM, and there is no intention of moving such conditions into this section. An alternative option is included for SLC6A1-related disorders.

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TABULAR MODIFICATIONS

New section	Genetic disorders, not elsewhere classified (QA0-QA1)	
New category	QA0	Neurodevelopmental disorders related to specific genetic pathogenic variants
Add		Code also, if applicable, any associated conditions, such as:
Add		attention-deficit hyperactivity disorders (F90.-)
Add		autism spectrum disorder (F84.0)
Add		epilepsy, by specific type (G40.-)
Add		intellectual disabilities (F70-F79)
Add		pervasive developmental disorders (F84.-)
New subcategory	QA0.0	Neurodevelopmental disorders related to pathogenic variants in specific genes
New sub-subcategory	QA0.01	Neurodevelopmental disorders related to pathogenic variants in certain specific genes
New sub-subcategory	QA0.010	Neurodevelopmental disorders, related to pathogenic variants in ion channel genes
New code	QA0.0101	SCN2A-related neurodevelopmental disorder
New code	QA0.0109	Neurodevelopmental disorder related to pathogenic variant in other ion channel gene
Add		SCN8A-related neurodevelopmental disorder

New sub-subcategory	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	GRIA1-related neurodevelopmental disorder
New code	QA0.0116	GRIA2-related neurodevelopmental disorder
New code	QA0.0117	GRIA3-related neurodevelopmental disorder
New code	QA0.0118	GRIA4-related neurodevelopmental disorder
New code	QA0.011A	GRIK2-related neurodevelopmental disorder
New code	QA0.0119	Other glutamate receptor, ionotropic, related neurodevelopmental disorder
New code	QA0.012	Neurodevelopmental disorders, related to pathogenic variants in other receptor genes

Note: for SLC6A1 related disorder, and related to the expansion for QA0.013 shown immediately below, an alternative option is also shown separately further below.

New sub-subcategory	QA0.013	Neurodevelopmental disorders, related to pathogenic variants in other transporter and solute carrier genes
New code	QA0.0131	SLC6A1-related disorder
Add		GABA transporter 1 deficiency
New code	QA0.0139	Neurodevelopmental disorder, related to pathogenic variant in other transporter or solute carrier gene
New sub-subcategory	QA0.014	Neurodevelopmental disorders, related to pathogenic variants in synapse related genes
New code	QA0.0141	Syntaxin-binding protein 1-related disorder
Add		STXBP1-related disorders
New code	QA0.0142	DLG4-related synaptopathy
New code	QA0.0149	Neurodevelopmental disorder, related to pathogenic variant in other synapse related gene
Add		Other genetic synaptopathy
New code	QA0.015	Neurodevelopmental disorders, related to genes associated with transcription and gene expression
New code	QA0.0151	FOXG1 syndrome
New code	QA0.0159	Neurodevelopmental disorder, related to other genes associated with transcription and gene expression
New code	QA0.8	Other neurodevelopmental disorders related to pathogenic variants in other specific genes

Alternative Option for SLC6A1 related disorder / GABA transporter 1 deficiency

E72 Other disorders of amino-acid metabolism

E72.8 Other specified disorders of amino-acid metabolism

New code	E72.82	Gamma aminobutyric acid transporter 1 deficiency
Add		GABA transporter 1 deficiency
Add		Gamma aminobutyric acid transporter protein type 1 deficiency
Add		GAT-1 deficiency
Add		SLC6A1-related disorder
Add		Code also any associated disorders, such as:
Add		attention-deficit hyperactivity disorders (F90.-)
Add		autism spectrum disorder (F84.0)
Add		epilepsy, by specific type (G40.-)
Add		intellectual disabilities (F70-F79)
Add		pervasive developmental disorders (F84.-)

Note: for this alternative for SLC6A1 related disorder, there would be a single new code for QA0.013, rather than the expansion shown previously.

Hao-Fountain Syndrome

A proposal to create a specific ICD-10-CM code for Hao-Fountain Syndrome has been received from Edwin Oh, MD, Christian Schaaf, MD, PhD and the Foundation for Hao-Fountain Syndrome, a patient advocacy organization representing the international Hao-Fountain Syndrome community.

Hao-fountain syndrome (HAFOUS, OMIM #616863) is a rare neurodevelopmental syndrome with an estimated prevalence of 1 in 40,000. After onset in infancy or childhood, it is characterized by developmental and intellectual disabilities, speech delay, and dysmorphic facial features (>90%) (Fountain et al., 2019). Over two-thirds of patients show abnormal MRI, hypotonia, eye anomalies, altered pain threshold, autism spectrum disorder, and delayed or impaired motor skills (Wimmer et al., 2023; Fountain et al., 2019).

It is a dominant genetic disease caused by loss of function mutations in USP7 (Ubiquitin-specific protease 7) gene (*602519) on chromosome 16p13.2 (Zheng et al., 2022). The majority of identified mutations are de novo mutations including missense, splicing, frameshift, and deletion mutations, with a majority of missense and nonsense mutations in the catalytic domain (Wimmer et al., 2023). There are currently no apparent genotype-phenotype correlations (Fountain et al., 2019). The USP7 gene encodes the deubiquitination enzyme USP7, also known as Herpesvirus Associated protease (HAUSP).

Ubiquitination is an essential process in cellular homeostasis. USP7 is involved in the MUST pathway, which regulates intracellular protein trafficking and recycling (Hao et al., 2013; Hao et al., 2015). Mutations in other proteins in the MUST pathway also cause neurodevelopmental disorders similar to Hao-Fountain Syndrome. These include Schaaf-Yang Syndrome (SYS, OMIM #615547) and Prader-Willi syndromes (PWS, OMIM #176270) (Schaaf et al., 2013; Cassidy et al., 2012). HAFOUS is unique from these disorders with a higher prevalence of seizures and the presence of white matter lesions on brain MRI.

At this time, various nonspecific ICD-10-CM codes are used for different symptoms and findings or conditions that are associated. These include code Q02, Microcephaly; codes in category K21, Gastro-esophageal reflux disease; code F82, Specific developmental disorder of motor function; and various symptom codes within subcategory R29.8, Other symptoms and signs involving the nervous and musculoskeletal systems. There is a wide spectrum of symptoms and severity amongst people with HAFOUS. However, none of these codes enable unique identification of HAFOUS.

It is proposed to classify HAFOUS in category Q87, Other specified congenital malformation syndromes affecting multiple systems. This classification for HAFOUS would place it in the same category as MED13L syndrome, a condition of similar pathology, which is coded at Q87.85.

HAFOUS is insufficiently understood, and more research is necessary to understand the spectrum of disease, potential genotype–phenotype correlations, and disease trajectories. Creation of a specific

ICD-10-CM code for HAFOUS will facilitate patient identification and participation in research studies. It will also enable collection and analyzing of disease-specific vital data. It has potential to accelerate several aspects of the drug development process. Ultimately, a unique ICD-10-CM code for HAFOUS will expedite understanding of its epidemiology and describe the broad spectrum of phenotypes, which will facilitate patient care for HAFOUS.

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TABULAR MODIFICATIONS

Q87 Other specified congenital malformation syndromes affecting multiple systems

Q87.8 Other specified congenital malformation syndromes, not elsewhere classified

New code	Q87.87	Hao-Fountain Syndrome
Add		Code also, if applicable, any associated conditions, such as:
Add		autism spectrum disorder (F84.0)
Add		developmental speech disorder (F80.-)
Add		epilepsy, by specific type (G40.-)
Add		intellectual disabilities (F70-F79)
Add		pervasive developmental disorders (F84.-)

Homozygous Familial Hypercholesterolemia

Hypercholesterolemia is a familiar diagnosis with multiple causes, most related to lifestyle issues such as diet, exercise, and smoking. In some cases, however, hypercholesterolemia is inherited.

Familial hypercholesterolemia is a common genetic disorder affecting about 30 million people worldwide. The genetic defect impairs the body's ability to remove low-density lipoprotein cholesterol (LDL-C) from the blood. LDL is known as the "bad cholesterol" because it builds up in the walls of arteries, narrowing the lumen and making it more difficult for blood to circulate, which in turn increases the risk of heart disease, acute myocardial infarction, and stroke.

There are two types of familial hypercholesterolemia:

- *Heterozygous familial hypercholesterolemia (HeFH)* - HeFH is the most common type and occurs when an individual inherits the variant (mutated) gene from one parent. HeFH is generally defined as LDL-C levels in the blood above 190 mg/dL.

- *Homozygous familial hypercholesterolemia (HoFH)* - HoFH is the rare form and occurs when an individual inherits the variant (mutated) gene from both parents. Untreated, LDL-C levels in the blood are typically in excess of 400 mg/dL.

The prevalence of heterozygous familial hypercholesterolemia is estimated to be between 1 in 250 and 1 in 300, while the prevalence of homozygous familial hypercholesterolemia is estimated to be between 1 in 250,000 and 1 in 360,000. Heterozygous familial hypercholesterolemia is more common but homozygous familial hypercholesterolemia is more severe. HoFH places individuals at extremely high risk of experiencing life-threatening cardiovascular events with substantially reduced life expectancy.

Persons with homozygous familial hypercholesterolemia are born with severely elevated levels of LDL-C, and risk increases with ongoing exposure of the vasculature to high levels of LDL-C. Symptoms of HoFH may begin during the neonatal period itself and are usually evident in early childhood. In addition to extremely elevated LDL levels in blood, excess cholesterol builds up in other tissues. Fatty yellow deposits (xanthomas) are often found under the skin and at the tendons, with interdigital xanthomas between the thumb and index finger being characteristic and specific to HoFH. Cholesterol deposits may also be found in the eyelids (xanthelasma) and inside the cornea (corneal arcus).

Chest pain due to premature coronary artery disease is common in HoFH, with acute myocardial infarction and impaired left ventricular function, including aortic valve stenosis and heart failure, sometimes seen before the age of 10. Persons with HoFH may also have renal and carotid artery stenosis early in life.

Identification of HoFH is usually based on a combination of the symptoms and physical findings as well as any family history of hypercholesterolemia. The diagnosis may be made based on clinical criteria or by genetic testing. It is imperative to identify and diagnose homozygous familial hypercholesterolemia as early in life as possible and begin intensive treatment to prevent development of premature cardiovascular disease. However, lipid screening in newborns and children is not routinely performed and most individuals are not diagnosed in a timely manner.

Treatment begins with lifestyle changes in addition to medications. In heterozygous familial hypercholesterolemia, statins and one or two additional lipid-lowering therapies are generally used. However, these are almost always insufficient for treating homozygous familial hypercholesterolemia and multiple-drug treatment is necessary to reduce LDL-C levels to treatment goals. Treatments for HeFH and HoFH may initially be similar but they have different impact on LDL-C and there are certain treatments only available for HoFH. The treatment algorithm for patients with HoFH consists of a combination of high intensity statins, additional lipid-lowering therapies, and lipoprotein apheresis.

Lipid-lowering therapies for HeFH and HoFH include oral cholesterol absorption inhibitors, such as ezetimibe, which reduces the amount of cholesterol absorbed from the diet. Subcutaneous injection of monoclonal antibodies in a class of drugs known as PCSK9 inhibitors, such as alirocumab and evolocumab, can be used for both HeFH and HoFH and can reduce LDL-C by about 20% in HoFH.

Some treatments are used only for HoFH. The microsomal triglyceride transfer protein (MTP) inhibitor lomitapide inhibits the formation of specific lipids and has been shown to reduce LDL-C by 50% in HoFH. Because of known adverse effects, particularly in the liver, physicians must enroll patients with HoFH in a risk evaluation and management program to access lomitapide and the patients must be closely monitored. Along with MTP inhibitors, angiopoietin-like 3 (ANGPTL3) inhibitors are indicated for treatment of HoFH. ANGPTL3 inhibitors use a unique pathway by blocking a specific protein, angiopoietin-like 3 (ANGPTL3), which stops the body from removing bad cholesterol.

Lipoprotein apheresis has been a mainstay treatment to remove excess LDL-C from the blood in HoFH, despite being invasive and time-consuming. It is effective in reducing LDL-C acutely by about 60% per session. However, LDL-C levels immediately begin to rise again after apheresis to about 90% of the original level in 7 days, such that apheresis is necessary on a regular basis, ideally once a week or every two weeks. It is recommended that apheresis be initiated in childhood, by the age of 6 or 7, to prevent atherosclerosis of the aortic root.

Many individuals with HoFH undergo cardiovascular interventions at a young age to treat cardiovascular disease, including aortic valve replacement, coronary artery bypass graft, and percutaneous coronary interventions. In one registry, the youngest patient to undergo CABG was 8 years old.

Without treatment, death usually occurs before the age of 20, although children have died at the age of 3 or 4. Survival depends on early diagnosis and intensive life-long treatment with a combination of lipid-lowering therapies.

Using the same ICD-10-CM code for both heterozygous and homozygous familial hypercholesterolemia muddies the data for these two very different types of familial hypercholesterolemia. HeFH and HoFH diverge in terms of the levels of LDL-C, the onset and severity of associated cardiovascular disease, and the types of treatment and monitoring needed. Data for HoFH patient gets lost in the larger HeFH group, resulting in underdiagnosis and undertreatment of this severe cholesterol disorder. Uniquely identifying HoFH in healthcare databases currently requires researchers and providers to take multiple additional steps and make multiple assumptions. A specific code for homozygous familial hypercholesterolemia will enable streamlined and more accurate study, enabling clear classification and identification of patients according to their distinct form of cholesterol disorder.

Chiesi USA, a pharmaceutical company is requesting the following tabular modifications.

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TABULAR MODIFICATIONS

E78 Disorders of lipoprotein metabolism and other lipidemias

E78.0 Pure hypercholesterolemia, unspecified

E78.00 Pure hypercholesterolemia, unspecified
Fredrickson's hyperlipoproteinemia, type IIa
Hyperbetalipoproteinemia
Low-density-lipoprotein-type [LDL] hyperlipoproteinemia
(Pure) hypercholesterolemia NOS

E78.01 Familial hypercholesterolemia

New code	E78.010	Homozygous familial hypercholesterolemia [HoFH]
New code	E78.011	Heterozygous familial hypercholesterolemia [HeFH]
New code	E78.019	Familial hypercholesterolemia, unspecified
Add		Familial hypercholesterolemia NOS

Hypothalamic obesity

This proposal is a re-presentation of a proposal originally presented at the March 2024 ICD-10 Coordination and Maintenance Committee Meeting. Changes have been made to address comments received from that proposal, and based on additional recommendations from the original submitter, Rhythm Pharmaceuticals, Inc.

Hypothalamic obesity (HO) is a highly unique form of severe obesity characterized by the rapid onset and sustained weight gain that is unresponsive to lifestyle or traditional medical interventions. HO affects approximately 5,000 to 10,000 patients in the United States.

The hypothalamus plays an important role in the body's regulation of appetite and energy balance. The term "hypothalamic obesity" describes obesity caused by injury or damage to the hypothalamus, in contrast to other forms of obesity related to the hypothalamic dysfunction such as genetic disorders. Further clinical details are available from the prior proposal.

Changes from the prior proposal are shown in **bold** in the modifications below.

TABULAR MODIFICATIONS

E23 Hypofunction and other disorders of the pituitary gland

E23.3 Hypothalamic dysfunction, not elsewhere classified

Excludes1: Prader-Willi syndrome (Q87.11)

Russell-Silver syndrome (Q87.19)

New sub-Subcategory	E23.31	Hypothalamic obesity
Add		Use additional code, if applicable, to identify associated manifestations, such as polyphagia (R63.2)
Add		Use additional code, if known, to identify body mass index (BMI) (Z68.-)
Add		**Use additional code, if applicable, to identify obesity class (E66.81-)**
Add		**Excludes1: obesity due to genetic metabolic disorder disrupting MC4R pathway (E88.82)**

New code	**E23.310**	**Hypothalamic obesity following traumatic injury to the hypothalamus**
Add		**Use additional code to identify associatedinjury, such as traumatic brain injury (S06.-)**
New code	**E23.311**	**Hypothalamic obesity following removal of neoplasm of the hypothalamus**
Add		**Use additional code to identify neoplasm**
New code	**E23.312**	**Hypothalamic obesity following treatment of neoplasm in the hypothalamus with radiation therapy**
Add		**Code first, if applicable, postprocedural complication (E89.89)**
Add		**Use additional code to identify neoplasm**
New code	**E23.313**	**Hypothalamic obesity due to neoplasm**
Add		**Hypothalamic obesity due to tumor**
Add		**Use additional code to identify neoplasm**
New code	**E23.318**	**Other hypothalamic obesity**
New code	**E23.319**	**Hypothalamic obesity, unspecified**
New code	E23.39	Other hypothalamic dysfunction

E66 Overweight and obesity

Revise **Code first, if applicable: ~~obesity complicating pregnancy, childbirth and the puerperium, if applicable (O99.21-)~~**

Add **hypothalamic obesity (E23.31-)**

Add **obesity complicating pregnancy, childbirth and the puerperium, (O99.21-)**

Add **obesity due to genetic metabolic disorder disrupting MC4R pathway (E88.82)**

Add **Prader-Willi syndrome (Q87.11)**

Excludes1: …

Delete **Prader-Willi syndrome (Q87.11)**

E88 Other and unspecified metabolic disorders

E88.8 Other specified metabolic disorders

E88.82 Obesity due to disruption of MC4R pathway

Add **Excludes1: hypothalamic obesity (acquired) (E23.31-)**

Immune Complex-mediated Membranoproliferative Glomerulonephritis (IC-MPGN)

Membranoproliferative glomerulonephritis (MPGN) is a rare cause of chronic nephritis. The term refers to a pattern of glomerular injury often characterized by mesangial expansion due to increased cellularity, increased extracellular matrix, and/or deposits of extracellular material to include immune complex and paraprotein deposits. In addition, there can be peripheral glomerular capillary wall abnormalities including thickening, duplication of basement membrane matrices, and/or deposition of immune complexes, complement, or monoclonal paraproteins. The presentation varies and changes throughout disease progression and across patient populations.

Prior to 2012, MPGN was classified into three types based on the location and characteristics of deposits identified on electron microscopy examination. Advancement in understanding the etiology of the MPGN pattern have led to a revised MPGN classification based on pathogenesis of these deposits. The revised classification uses immunofluorescence studies to distinguish three types of MPGN: 1) Complement-dominant/mediated MPGN pattern (includes C3 glomerulonephritis, C4 glomerulonephritis, and dense deposit disease (DDD)); 2) Immune-complex or immunoglobulin (Ig) mediated MPGN pattern (IC-MPGN); 3) Absence of immune complexes/minimal complement deposits with MPGN pattern (typically associated with thrombotic microangiopathy).

Immune complex-mediated MPGN (IC-MPGN) is characterized by accumulation of immunoglobulins along the capillary walls, often along with complement deposition. IC-MPGN can be idiopathic but is more commonly caused by either chronic infections (most frequently Hepatitis C and B, but also other viral and bacterial infections), monoclonal gammopathies (proliferative glomerulonephritis with monoclonal Ig (PGNMID)), or autoimmune disorders (Sjögren's syndrome, rheumatoid arthritis, and systematic lupus erythematosus (SLE)).

While evidence-based clinical guidelines reflect the updated classification of MPGN and IC-MPGN, the existing ICD-10-CM codes are insufficient for precisely defining IC-MPGN. Rather, ICD-10-CM currently includes IC-MPGN under the broad category of mesangiocapillary glomerulonephritis. This presents a potential challenge in terms of accurate documentation of the specific diagnosis and selection of the appropriate corresponding treatment. The management and treatment of IC-MPGN is dictated by its underlying pathogenesis and etiology, namely primary (idiopathic) or secondary (infection, monoclonal gammopathy, or autoimmune disease).

New IC-MPGN codes (similar to those recently approved for C3 glomerulonephritis are needed to align with the revised MPGN classification. The addition of IC-MPGN codes would be in step with current coding for C3 glomerulonephritis and DDD, which are other sub-types of MPGN. Additionally, since treatment for IC-MPGN depends on etiology, the new codes would distinguish between primary and secondary IC-MPGN.

The Renal Physicians Association has submitted the proposal for consideration.

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TABULAR MODIFICATIONS

N00 Acute nephritic syndrome

N00.5 Acute nephritic syndrome with diffuse mesangiocapillary glomerulonephritis

Delete ~~Acute nephritic syndrome with membranoproliferative glomerulonephritis, types1 and 3, or NOS~~

New subcategory N00.B Acute nephritic syndrome with immune complex membranoproliferative glomerulonephritis

New code N00.B1 Acute nephritic syndrome with idiopathic immune membranoproliferative glomerulonephritis (IC-MPGN)

New Code N00.B2 Acute nephritic syndrome with secondary immune complex membranoproliferative glomerulonephritis (IC-MPGN)

	N04	Nephrotic syndrome
	N04.5	Nephrotic syndrome with diffuse mesangiocapillary glomerulonephritis
Delete	~~Nephrotic syndrome with membranoproliferative glomerulonephritis, types 1 and 3, or NOS~~	
New subcategory	N04.B	Nephrotic syndrome with immune complex membranoproliferative glomerulonephritis (IC-MPGN)
New Code	N04.B1	Nephrotic syndrome with idiopathic immune complex membranoproliferative glomerulonephritis (IC-MPGN)
New Code	N04.B2	Nephrotic syndrome with secondary immune complex membranoproliferative glomerulonephritis (IC-MPGN)

Inflammatory Breast Cancer

Inflammatory Breast Cancer (IBC) is a distinct clinical entity of advanced breast cancer, characterized by tumor cell emboli blocking the breast lymph vessels[1]. This blockage causes inflammatory-like changes in the breast, including swelling and skin reddening, which are often mistaken for a breast infection, leading to delayed diagnosis. The current American Joint Committee on Cancer (AJCC) guidelines list established diagnostic criteria for IBC including rapid onset (less than 6 months) of erythema occupying at least 1/3 of the breast, edema, peau d' orange (orange peel-like appearance) and/or warmth (with or without an underlying mass), and pathologic confirmation of invasive carcinoma. Yet, in practice, IBC is largely a subjective, clinical diagnosis that may or may not meet these criteria. Of note, IBC can be even more difficult to recognize in women with darker skin tones. This, coupled with the increased incidence of IBC in Black or African American women[2], contributes to an even greater vulnerability for these minority populations.

Because IBC commonly lacks an underlying palpable mass and progresses rapidly, by the time symptoms manifest, the disease is already at late stage (Stage 3 or 4) with about 1/3 of women having distant metastases upon diagnosis[1]. Although IBC is a rare disease accounting for only 1-5% of breast cancers in the US[3-4], it disproportionately contributes to about 7% of breast cancer mortalities[1]; and sadly, IBC patients are faced with the dire prognosis of a five-year relative survival rate of only 39%[5]. Thus, accurate and timely diagnosis is crucial for IBC patient survival. The rarity of IBC, coupled with its diagnostic limitations, not only impacts patient outcomes but also hampers disease-related research. Thus, little is known currently about the underlying biology of IBC and no actionable biomarkers or targeted therapies have been identified to date.

Once diagnosed, IBC requires specific and intensive treatment regimens for optimal survival outcomes[6]. The National Comprehensive Cancer Network (NCCN)[7] standard of care for Stage 3 IBC (and in some cases Stage 4) is trimodality therapy consisting of: (1) Neoadjuvant therapy followed by (2) Non-skin sparing mastectomy with axillary lymph node dissection with or without delayed reconstruction followed by (3) Chest wall and regional nodal radiation.

The use of trimodality therapy for IBC patients has been associated with an increased five-year survival rate of 55.4%[5] and improved local regional recurrence rates[8]. However, only a fraction of IBC patients (with most recent reports estimating only 25%[9]) currently receive trimodality care[5].

To improve diagnosis and expedite care for IBC, the Susan G. Komen-Inflammatory Breast Cancer Research Foundation's IBC Collaborative recently developed a novel quantitative diagnostic scoring system for IBC. This IBC Scoring System,[10] available online[11], allows clinicians to assess a patient's clinical, pathologic and imaging characteristics to enable easier, quicker and more accurate diagnosis. The IBC Scoring system has recently been scientifically validated in discriminating between IBC and non-IBC patients[12]. This, combined with a new diagnosis code, will provide for more accurate and frequent diagnoses of IBC.

On behalf of the IBC Collaborative, Susan G. Komen is requesting new specific diagnosis codes for IBC to better allow physicians to identify patients more readily with this form of breast cancer, facilitate earlier intervention for patients, and raise awareness about IBC in the clinical community.

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TABULAR MODICATIONS

C50	Malignant neoplasm of breast
New subcategory	C50.A Malignant inflammatory neoplasm of breast
Add	Inflammatory breast cancer (IBC)
New code	C50.A0 Malignant inflammatory neoplasm of unspecified breast
New code	C50.A1 Malignant inflammatory neoplasm of right breast
New code	C50.A2 Malignant inflammatory neoplasm of left breast

Kabuki Syndrome

Kabuki syndrome (KS) is a rare genetic disorder affecting multiple body systems and requires comprehensive medical management[1]. A specific code for KS will assure accurate diagnoses, research, and healthcare management for individuals with KS.

A proposal was presented at the March 2024 ICD-10Coordination and Maintenance meeting. In response to public comments, a revised proposal is being submitted for reconsideration. Changes are noted in **bold.**

Kabuki syndrome is distinct and cannot be grouped with other congenital malformations. KS's unique and specific phenotypic presentation is caused by mutations in genes of histone modification, including *KMT2D* and *KDM6A*[2,3]. Patients are identified by genetic testing and characteristic phenotypes including distinct facial features, developmental delay, intellectual disability, heart defects, hypotonia, skeletal anomalies, and immune dysfunction[4]. KS requires accurate diagnosis in order for patients to receive proper treatment, particularly with precision KS therapy currently in development.

The current coding options, fails to distinguish KS from other congenital malformations, resulting in a loss of essential clinical information. It limits the ability to track and analyze KS's unique clinical characteristics, medical comorbidities, and healthcare utilization. As a result, important data related to the prevalence, treatment outcomes, and healthcare resource requirements for KS remain obscured, and medical reimbursement is delayed.

Although there are no known cures for KS, at least eight treatments specifically for KS are currently in development. One has received rare pediatric disease and orphan drug designations from the U.S. FDA[5]. Another treatment that was being developed for KS received orphan drug designation from the U.S. FDA and European Commission in 2018[6]. These designations indicate that the FDA and EMA consider KS a separate indication that cannot be grouped with others in ICD-10-CM coding.

Current best estimates for the prevalence of KS are 1:32,000-86,000 live births[7,8] (figures that were estimated in 1988 and 2004, before genetic testing for KS was available). This prevalence is similar to other rare disorders that have been provided codes, including Prader-Willi syndrome (Q87.11), SYNGAP1 Encephalopathy (F78.A1) and Angelman syndrome (Q93.51); these syndromes also have intellectual disabilities and developmental delay as hallmark symptoms. Without a specific code, KS patients cannot be tracked to corroborate prevalence estimates and document morbidity and mortality of the condition, critical for informing public health initiatives and patient care.

A unique code will improve patient care by enabling accurate diagnosis, leading to better clinical management and patient outcomes. A specific code will ensure continuation of proper treatment, aiding in the early identification of potential complications and comorbidities, as well as more easily

enabling potential retrospective studies for best practices in clinical care. In addition, a specific KS code facilitates the collection of evidence-based data, accelerating research on the syndrome's pathogenetic mechanisms, natural history, and therapeutic interventions, as well as helping discover and recruit participants for clinical studies/trials and patient registries. A dedicated code improves patient care, accelerates research and promotes health equity.

In summary, the creation of a dedicated ICD-10-CM code for KS has the potential to transform the landscape of KS research, treatment, and awareness. Accurate identification and documentation of this specific rare condition will advance public health knowledge and the well-being of affected individuals.

This proposal is submitted by and with contributions made from the following clinicians:

- Clara Tang, PhD, Director of Research at the KSF
- Bruce Bloom, JD, DDS, Chief Science Officer at the KSF
- Olaf Bodamer MD, PhD, Director of the Roya Kabuki Program at Boston Children's Hospital and Associate Professor at Harvard Medical School
- Margaret Adam, MD, Professor at University of Washington
- Jacqueline Harris, MD, Director of the Epigenetics Clinic at Kennedy Krieger Institute and Associate Professor at Johns Hopkins University School of Medicine
- Hans Bjornsson, MD, PhD, Associate Professor at Johns Hopkins University and Professor at the University of Iceland
- Brittany Simpson, MD, Assistant Professor at Cincinnati Children's Hospital Medical Center
- Ian Krantz, MD, Professor at The Children's Hospital of Philadelphia
- Drea K. Petersen, MD, Clinical Geneticist and Pediatrician at Randall Children's Hospital and Legacy Health

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TABULAR MODIFICATIONS

Q89 Other congenital malformations, not elsewhere classified

Q89.8 Other specified congenital malformations

Use additional code(s) to identify all associated manifestations

New code **Q89.81 Kabuki syndrome**
Add **Kabuki syndrome, type 1, due to KMT2D mutation**
Add **Kabuki syndrome, type 2, due to KDM6A mutation**
Add **Niikawa-Kuroki syndrome**

New code **Q89.89 Other specified congenital malformations**

Ledderhose Disease/Plantar Fibromatosis & Plantar Fasciitis

This topic was presented at the March 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. Typically, symptoms present as pain first step in the morning with improvement in ambulation.

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.

TABULAR MODIFICATIONS

	M67	Other disorders of synovium and tendon	
New subcategory	M67.A	Plantar fasciitis	
New code		M67.A0	Plantar fasciitis, unspecified site
Add			Plantar fasciitis NOS
New sub-subcategory		M67.A1	Plantar fasciitis, heel
New code			M67.A11 Plantar fasciitis, right heel
New code			M67.A12 Plantar fasciitis, left heel
New code			M67.A19 Plantar fasciitis, unspecified heel

New code	M67.A9	Other plantar fasciitis
Add		Plantar fasciitis midfoot
New code		**M67.A91 Other plantar fasciitis, right foot**
New code		**M67.A92 Other plantar fasciitis, left foot**
New code		**M67.A99 Other plantar fasciitis, unspecified foot**

	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

Limb Girdle Muscular Dystrophies (LGMD) Subtype 2I/R9

Muscular dystrophy has several major types and dozens of subtypes. Following a prior C&M proposal in March 2021, an ICD-10-CM subcategory and codes were created for one of the most common types: limb girdle muscular dystrophies (G71.03-). Within this subcategory, there are 7 codes for selected common LGMD subtypes as well as general codes for other and unspecified LGMD subtypes. It is now proposed to add a specific code for another subtype, LGMD 2I, based on prevalence and therapeutics now being developed. As in the past, the current proposal is based on a submission from and on behalf of a coalition of LGMD patient advocacy organizations and LGMD clinical experts, and reflects the input of clinicians, researchers, biopharmaceutical companies, physical therapists, coding experts, and other medical professionals familiar with LGMD.

Limb girdle muscular dystrophies are a group of genetically inherited conditions that primarily affect proximal skeletal muscle, leading to loss of muscle fibers and progressive muscle weakness. There are currently 34 identified subtypes of LGMD, each with a unique genetic cause. The subtypes also vary by the age of onset, specific symptoms, progression of the disorder, and treatment regimens.

The nomenclature for limb girdle muscular dystrophies is currently transitioning. LGMD 2I is also known as LGMD R9 FKRP-related, because the affected gene encodes instructions for making fukutin-related protein (FKRP). This enzyme is critical to muscle function and is abundantly found in skeletal muscle as well as in cardiac and respiratory muscles.

The age of onset for symptoms is quite variable but is generally between childhood and early adulthood. Initial symptoms include weakness and muscle wasting, first in the hips and legs then later in the arms and shoulders. Frequent falls are characteristic as is difficultly in lifting objects, walking, and running. Rising from the floor and climbing stairs are typically the abilities that are lost the earliest. Over time, the symptoms progressively worsen and individuals with LGMD 2I typically become wheelchair dependent. While the condition is always progressive, the rate of progression is highly variable between individuals, ranging from gradual to very rapid.

Because LGMD 2I can involve the heart and the diaphragm, cardiac and respiratory problems are seen in individuals with LGMD 2I and can occur even when the disorder is in mild stages. Some forms of muscular dystrophy exclusively affect the skeletal musculature, while others such as LGMD2I impact cardiac musculature as well. Cardiomyopathy is a prominent feature of the disease, particularly as patients enter adulthood. A significant number of individuals develop heart failure and/or require ventilatory support.

LGMD 2I is one of the most common subtypes of limb girdle muscular dystrophy. In US next-generation sequencing-based gene panel testing, the prevalence for LGMD 2I was the third highest and, notably, was higher than other subtypes such as 2L, 2D, and 2E. In the general population, prevalence of LGMD 2I is estimated at 4.52 per million, corresponding to 1,500 to 2,000 individuals in the US. Estimates of prevalence in the US vary depending on the methods. Patient registries and

natural history studies are also in use to help identify prevalence in the US but, by their nature, they do not include all patients. The Muscular Dystrophy Surveillance, Tracking and Research Network (MD STARnet) is a US population-based surveillance program funded by the Centers for Disease Control and Prevention. MD STARnet is the only program designed to collect health information on everyone with MD living in specific US areas. Using data from 6 MD STARnet sites, the preliminary estimate of the prevalence of clinically confirmed LGMD2I is 1.5 per million and the confidence interval ranges from 1.1 to 2.1 per million.

Current treatments for LGMD 2I, such as physical therapy, steroids, and pain management, are supportive and focus on management of symptoms. However, new treatments are being developed to address the underlying dysfunction.

Clinical trials are ongoing for three treatments, one small molecule drug (ribitol) and two replacement gene therapies, which address the underlying cause of the disease. The small molecule drug is currently being tested in a phase II study (NCT04800874) and a phase III study (NCT05775848). Phase I/II clinical trials are also in progress to assess the two replacement gene therapies targeted at LGMD 2I (NCT05224505, NCT05230459). The small molecule drug and one of the gene therapies were granted FDA Fast Track designation, which recognizes the need to expedite review for treatment of serious conditions with unmet needs. Of any form of LGMD, LGMD 2I has the most clinical-stage development activity in terms of number distinct therapies in development.

As with the codes created earlier for limb girdle muscular dystrophy, a unique code for LGMD 2I will enable more informed estimates of prevalence as well as identification of this condition in encoded databases consistent with the emergence of new treatments.

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TABULAR MODIFICATIONS

	G71	Primary disorders of muscles		
		G71.0	Muscular dystrophy	
			G71.03	Limb girdle muscular dystrophies
New code			G71.036	Limb girdle muscular dystrophy due to fukutin related protein dysfunction
Add				LGMD R9 FKRP-related
Add				Limb girdle muscular dystrophy due to FKRP deficiency
Add				Limb girdle muscular dystrophy type 2I
			G71.038	Other limb girdle muscular dystrophy
Delete				~~LGMD R9 FKRP-related~~
				LGMD R22 collagen 6-related
Delete				~~Limb girdle muscular dystrophy due to fukutin related protein dysfunction~~
Delete				~~Limb girdle muscular dystrophy type 2I~~
				Other autosomal recessive limb girdle muscular dystrophy

INDEX MODIFICATIONS

	Dystrophy, dystrophia
	- muscular G71.00
	- - limb-girdle G71.039
Revise	- - - R9 (autosomal recessive) <u>G71.036</u> ~~G71.038~~
Add	- - - type 2G (autosomal recessive) G71.038
Add	- - - type 2H (autosomal recessive) G71.038
Revise	- - - type 2I (autosomal recessive) <u>G71.036</u> ~~G71.038~~
Add	- - - type 2J (autosomal recessive) G71.038

Add - - - type 2K (autosomal recessive) G71.038

Add - - - type 2M (autosomal recessive) G71.038

Add - - - type 2N (autosomal recessive) G71.038

Add - - - type 2O (autosomal recessive) G71.038

Add - - - type 2P (autosomal recessive) G71.038

Add - - - type 2Q (autosomal recessive) G71.038

Add - - - type 2S (autosomal recessive) G71.038

Add - - - type 2T (autosomal recessive) G71.038

Add - - - type 2U (autosomal recessive) G71.038

Leukocyte Adhesion Deficiency Type I (LAD-I)

Leukocyte adhesion deficiency type I (LAD-I) is a rare genetic disorder caused by mutation(s) in the *ITGB2* gene that result in impaired production of CD18[1]. CD18, the beta subunit of the β2 integrins, is a glycoprotein on the surface of white blood cells that mediates their migration from inside blood vessels to sites of infection or inflammation in tissues.

This proposal was presented at the March 2024 ICD-10 Coordination and Maintenance meeting. In response to public comments, a revised proposal is being submitted for reconsideration. Changes are noted in **bold.**

Because the white blood cells with *ITGB2* mutation(s) can't get to the site of infection, patients with LAD-I have an impaired ability to fight infections, especially at sites of microbial entry, and have an abnormal hyperinflammatory response to infections. Patients are especially susceptible to recurrent atypical bacterial or fungal infections such as peri-rectal infections, as well as abnormal inflammation of tissues such as periodontitis and gingivitis. The clinical severity depends on the level of preserved CD18 on the surface of neutrophils. Mortality for patients with severe LAD-I has been reported at 60-75% by the age of 2 years[2]. Diagnosis depends on an astute clinician considering the possibility of LAD-I deficiency in a child with omphalitis, other atypical infections, or recurrent infections associated with very high neutrophil counts. However, given the rarity of LAD-I, most clinicians have never made the diagnosis or treated a patient with LAD-I.

The incidence of LAD-I is estimated to be under 10 in 1,000,000 live births[3,4]. Given the impediments to diagnosis and the greatly shortened life expectancy, the exact incidence of LAD-I is not well-established and may increase as recognition of the disease continues to grow. To quote the National Organization for Rare Diseases[5]:

> *Leukocyte adhesion deficiencies often go unrecognized and may be misdiagnosed, making it difficult to determine their true frequency in the general population.*

As of 2009, one author reported approximately 300 cases of LAD-I worldwide[6].

Frequent infections (93% of patients) and poor wound healing (86%) beginning as early as the neonatal period are the two most common presenting symptoms of LAD-I[7]. Common infections in LAD-I patients include pneumonia, gingivitis, and peritonitis, all of which can be life-threatening. Patients with severe disease (caused by <2% expression of CD18) present with frequent, severe infections which can be life-threatening[8]. Patients with severe disease have very poor prognosis without hematopoietic stem cell transplant[8]. Patients with moderate disease (2% to 30% expression of CD18) have less frequent and less severe infections but still have multiple infections affecting the skin and mucosal surfaces. Although these patients can survive to adulthood with adequate treatment[8], mortality exceeds 50% by the age of 40 years[2].

Suspicion of LAD-I may be raised by inflammation (omphalitis) and delayed separation of the umbilical stump, which occurs in 58-84% of patients[2]. The diagnosis can also be suspected in infants

with recurrent soft tissue infections, especially in those who present with abscess-like lesions without pus and in those who present with infections with an exceedingly high white blood cell count. A flow cytometry analysis demonstrating the absence of functional CD18 and the associated alpha subunit molecules (CD11) on the surface of leukocytes provides the definitive diagnoses of LAD-I[9]. This diagnosis is then confirmed with genetic testing to define the exact molecular defect.

LAD-I results from mutations in the *ITGB2* gene and is unique from the other two types of LAD (LAD-II and LAD-III) in molecular cause, diagnosis, and symptoms. LAD-II is caused by a mutation of the *SLC35C1* gene, is definitively diagnosed via flow cytometry analysis showing the absence of CD15a, and typically is associated with milder infections and no omphalitis. LAD-III is caused by a mutation in the *FERMT3* gene (also called kindlin-3 gene) and is differentially diagnosed from LAD-I via molecular testing. It commonly causes bleeding complications, bone marrow failure, and osteoporosis.

Treatment of LAD-I includes management of the repeated, prolonged infections that characterize the disease, and often involves hospitalization, long courses of antibiotic and antimicrobial medications, and preventative isolation or formation of limited social pods to minimize risk of infection. Allogeneic hematopoietic stem cell transplant (HSCT) is currently the only curative treatment. However, a high incidence of graft rejection and acute graft vs host disease (aGVHD) both pose barriers to HSCT success[6]. When HSCT is performed in LAD-I patients, 17% of patients experience graft failure and 24% experience grade II to IV acute GVHD by 100 days, with 8% of patients experiencing lethal GVHD[10].

An ICD-10-CM code would aid physicians in improving patient care by better ensuring appropriate patient treatment and making it possible to track clinical results of interventions. This is extremely important in addressing these patients' atypical serious infections and wound healing limitation. An appropriate code would intrinsically justify prolonged courses of antibiotics and would help identify a population of patients with an increased risk of complications related to otherwise standard management approaches. As an example, as a result of poor wound healing and excessive inflammation, patients with LAD-I can have significant complications resulting from standard-of-care procedures (such as routine incision and drainage) that are very well-tolerated in most other conditions.

In addition, it would promote effective communication across health care teams. Close monitoring is essential to ensuring proper care of LAD-I patients since infections typically require prolonged treatment and any medical intervention creating a wound, such as a surgery, biopsy, or circumcision, has the potential to be complicated by poor wound healing, abscess formation, and potentially fatal microbial dissemination.

An ICD-10-CM code can also help in identifying patients for clinical trials. Finally, an ICD-10-CM code will improve the ability to verify the prevalence and natural history of the disease, make genotype-phenotype correlations, and enable tailored and long-term monitoring of outcomes from future therapies.

Although no other curative therapies are currently available, an investigational genetic therapy for severe LAD-I being developed by Rocket Pharmaceuticals has demonstrated 100% 12-month survival in a Phase I/II study and has a PDUFA review date established with the FDA for March 31, 2024. An ICD-10 code for LAD-I will ensure that patients can be identified, and their outcomes tracked while receiving this new therapy as well as more conventional hematopoietic stem cell transplants.

This proposal for a specific code for LAD-I is submitted by Susan Prockop, MD, Boston Children's Hospital and Magnolia Innovation, with assistance from the medical team at Rocket Pharmaceuticals, Inc.

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TABULAR MODIFICATIONS

	D71	Functional disorders of polymorphonuclear neutrophils
Delete		~~Cell membrane receptor complex [CR3] defect~~
Delete		~~Chronic (childhood) granulomatous disease~~
Delete		~~Congenital dysphagocytosis~~
Delete		~~Progressive septic granulomatosis~~
New Code	**D71.1**	**Leukocyte Adhesion Deficiency**
Add		**LAD-I**
Add		**LAD-II**
Add		**LAD-III**
Add		**Leukocyte adhesion deficiency type I**
Add		**Leukocyte adhesion deficiency type II**
Add		**Leukocyte adhesion deficiency type III**
New Code	D71.8	Other functional disorders of polymorphonuclear neutrophils
Add		Cell membrane receptor complex [CR3] defect
Add		Chronic (childhood) granulomatous disease
Add		Congenital dysphagocytosis
Add		Progressive septic granulomatosis
New code	**D71.9**	**Functional disorders of polymorphonuclear neutrophils, unspecified**

Lipedema and lipolymphedema

This topic was originally presented at the September 2020 ICD10 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.

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. 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.[5]

Unique to lipedema is fat that is highly resistant to loss by diet, exercise, or bariatric surgery.[6-8] 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 9-11] There are three stages of lipedema.[12] Lipedema is also divided into five subtypes, or phenotypes, depending on the primary affected region:[13-15]

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

There is no cure for lipedema, but treatments aimed at reducing the lymphedema component of lipedema such as manual decongestive therapy, wrapping, exercise, compression garments and pumps, and some medical foods and medications are helpful. Expertly performed suction assisted lipectomy is the treatment of choice for suitable lipedema patients with an inadequate response to conservative and supportive measures.[14] Lipedema is thought to affect 11% of the female population.[16] Women with lipedema can develop lymphedema at any stage, called lipolymphedema, or lymphedema associated with lipedema. In some published literature, lipolymphdema is also known as Stage 4 lipedema. The use of "Stage 4" is confusing since lymphedema is not a stage of lipedema. If the natural progression of lipdema was the development of lymphedema only after Stage 3 lipedema, then the Stage 4 designation might make sense, but lymphedema can happen at any stage of lipedema.[4] The development of lymphedema with lipedema should be reported as separate ICD-10-CM codes.

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. Praecox lymphedema is currently captured in ICD-10-CM as a secondary lymphedema; it is more accurately classified under code Q82.0: Hereditary lymphedema.

Dr. Karen Herbst, with support from the American Vein & Lymphatic Society (AVLS), is submitting the following modifications to identify and track lipedema and lipolymphedema 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 (Cheek) dolorosa [Dercum]~~

New code E88.21 Dercum disease

Add Lipomatosis (Check) 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 Code also, if applicable:

Add lipedema phenotype (E88.8A-)

Add lipolymphedema (I89.A)

New code E88.831 Lipedema, Stage 1

New code E88.832 Lipedema, Stage 2

New code E88.833 Lipedema, Stage 3

New code E88.839 Lipedema, unspecified

Add Lipedema NOS

New sub-subcategory E88.8A Lipedema phenotype

Add Code first, if known, the lipedema stage (E88.83-)

New code E88.8A0 Lipedema phenotype I

Add Lipedema of buttock to hips

New code E88.8A1 Lipedema of phenotype II

Add Lipedema buttock to knees

New code E88.8A2 Lipedema phenotype III

Add Lipedema of buttock to ankles

New code E88.8A3 Lipedema phenotype IV

Add Lipedema of arms

New code E88.8A4 Lipedema phenotype V

Add Lipedema of legs

New code E88.8A8 Other lipedema phenotype

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 Lipolymphedema
Add Stage 4 lipedema

New code I89.B Idiopathic lymphedema

Q82 Other congenital malformations of skin

Q82.0 Hereditary lymphedema
Add Praecox lymphedema

Lipodystrophy

Lipodystrophy is a disorder characterized by selective, pathologic loss of adipose tissue (body fat). The loss of body fat produces a deficiency in leptin, a hormone produced by fat cells which regulates key metabolic functions, particularly the metabolism of fats and carbohydrates. Leptin's primary function is to regulate appetite and satiety, but it also has a key role in fatty acid accumulation, sensitivity to insulin, and glucose production within the body. Given the scope of leptin's effect on metabolic function, leptin deficiency results in multiple metabolic comorbidities.

Lipodystrophy can be caused by genetic mutations, ie, congenital or familial lipodystrophy, or it can be related to coexisting medical conditions, ie, acquired lipodystrophy. Importantly, lipodystrophy is also classified as either generalized or partial. Generalized refers to loss of nearly all normal fat deposits while partial refers to less complete loss of body fat. Based on these points, there are four major subtypes of lipodystrophy: congenital generalized lipodystrophy (CGL), acquired generalized lipodystrophy (AGL), familial partial lipodystrophy (FPLD), and acquired partial lipodystrophy (APL). However, for clinical management, the key distinction is between generalized lipodystrophy and partial lipodystrophy.

Individuals with generalized lipodystrophy have little or no adipose tissue. Those with partial lipodystrophy have some adipose tissue. Metabolic profiles are similar but otherwise, the disorders differ in terms of presentation, onset, severity, and treatment.

In terms of presentation, the most noticeable signs and symptoms of lipodystrophy is a very lean or muscular appearance and feeling continually hungry regardless of dietary intake. In generalized lipodystrophy, this is very striking but it is typically more subtle in partial lipodystrophy. Age of onset varies as well. In generalized lipodystrophy, the absence of body fat occurs in babies. In contrast, fat distribution is typically normal in early childhood in partial lipodystrophy, with loss of fat taking place around puberty.

Metabolic comorbidities are characteristic of lipodystrophies and place individuals with lipodystrophy at high risk of developing organ complications involving the liver, heart, and kidney. The common comorbidities include insulin resistance, diabetes mellitus which may develop in the teenage years, hypertriglyceridemia which can lead to atherosclerosis, non-alcoholic fatty liver disease (hepatic steatosis) which may result in fibrosis and cirrhosis, and polycystic ovarian syndrome.

These complications can be found in both generalized and partial lipodystrophies. However, the severity is directly related to the extent of body fat loss. Metabolic disorders occur earlier and are more severe in persons with generalized lipodystrophy. The severity is typically milder in persons with partial lipodystrophy with some subtypes at much lower risk for metabolic complications.

There is no treatment to directly reverse the loss of body fat. Instead, treatment focuses on the comorbidities that arise. In addition to diet and exercise, these include standard treatments for diabetes, such as metformin and insulin, and hypertriglyceridemia, such as fibrates and statins. Metreleptin is a recombinant leptin replacement therapy used with diet as a first line treatment for metabolic comorbidities associated with generalized lipodystrophy. It is known to improve blood

sugar levels, allowing some patients to discontinue use of insulin and to lower triglycerides. Metreleptin is not indicated for use in partial lipodystrophy in the US.

Prevalence of the lipodystrophy syndromes has been estimated at 1.3 to 4.7 cases per million in the general population, with 0.23 cases per million for generalized lipodystrophy and 2.84 cases per million for partial lipodystrophy. However, more recent data analysis suggests that generalized and partial lipodystrophies may be more prevalent than previously estimated, particularly for familial partial lipodystrophy.

It should be noted that HIV-associated lipodystrophy differs significantly from the lipodystrophy syndromes. HIV-associated lipodystrophy usually arises as a side effect of the antiretroviral therapy and is characterized by abnormal fat distribution, including both fat loss (lipoatrophy) and fat accumulation (lipohypertrophy). HIV-associated lipodystrophy is conventionally excluded from lipodystrophy studies and is the subject of separate research. Equally, localized lipodystrophy is limited to specific areas of the body and can result from various causes such as trauma, local inflammation, repeated injections, or simply natural variations. Localized lipodystrophy does not present the systemic complications seen in generalized or partial lipodystrophy syndromes.

All forms of lipodystrophy are currently classified to a single ICD-10-CM code. Lack of separate codes for generalized lipodystrophy and partial lipodystrophy has hampered efforts to establish clearer estimates of prevalence and undertake focused research on outcomes. HIV-associated lipodystrophy and localized lipodystrophy factor into this as well.

From a treatment perspective, it is essential to better identify generalized lipodystrophy and partial lipodystrophy due to the distinct therapeutic needs and responses in these conditions. In addition to distinguishing rare lipodystrophy syndromes from HIV-related lipodystrophy and local lipodystrophy, unique classification of generalized lipodystrophy and partial lipodystrophy lipodystrophy allows for tailored treatment approaches that address the specific complications associated with each subtype.

Chiesi USA, Inc., is requesting the following new ICD-10-CM codes to distinguish between the subtypes of lipodystrophy.

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TABULAR MODIFICATIONS

E88 Other and unspecified metabolic disorders

E88.1 Lipodystrophy, not elsewhere classified

Delete ~~Lipodystrophy NOS~~

Excludes1: Whipple's disease (K90.81)

New code E88.11 Partial lipodystrophy
Add Acquired partial lipodystrophy (APL)
Add Barraquer-Simons lipodystrophy
Add Familial partial lipodystrophy (FPLD)

New code E88.12 Generalized lipodystrophy
Add Acquired generalized lipodystrophy (AGL)
Add Berardinelli-Siep syndrome
Add Congenital generalized lipodystrophy (CGL)
Add Lawrence syndrome

New code E88.13 Localized lipodystrophy
Add Injection lipodystrophy
Add Insulin lipodystrophy

New code E88.14 HIV-associated lipodystrophy
Add Code first any human immunodeficiency virus [HIV] disease (B20)
Add Use additional code for adverse effect, if applicable, to identify drug (T37.5X5-)

New code E88.19 Other lipodystrophy, not elsewhere classified
Add Lipodystrophy NOS

Lynch Syndrome

This is a repeat presentation based on comments received, following a proposal to add codes for specific genetic types of Lynch syndrome that was presented in March 2024. Further clinical details are available from the prior proposal, but certain information is repeated here for convenience.

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. It is caused by pathogenic variants in genes in the DNA mismatch repair pathway that maintain fidelity during replication (MLH1, MSH2, MSH6, PMS2, EPCAM). These are inherited in an autosomal dominant fashion.

Lynch syndrome is characterized by increased risk of cancer in multiple organ systems. The cancer risks vary dramatically by gene. The lifetime risk varies by gene but 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.

The creation of new ICD-10-CM codes to specifically identify patients with Lynch syndrome has been proposed by 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. Since the cancer risks vary dramatically by gene, they have proposed ICD-10-CM codes for Lynch syndrome specifically indicating which of the five genes is responsible.

TABULAR MODIFICATIONS

Note that the new section below is proposed in separate topics also. This proposal would add a new category Genetic disorders associated with neoplasms, and codes for specific genetic causes of Lynch syndrome within that category.

New section		Genetic disorders, not elsewhere classified (QA0-QA1)
New category	QA1	Genetic disorders associated with neoplasms
Add		Code also any associated conditions, such as:
Add		malignant neoplasms (C00.0-C96.9)
Add		personal history of malignant neoplasm (Z85.-)
New subcategory		QA1.7 Genetic disorders associated with neoplasms of multiple systems

New sub-subcategory	QA1.71	Lynch syndrome	
Add		Hereditary nonpolyposis colorectal cancer susceptibility	
New code		QA1.710	MLH1 Lynch syndrome
New code		QA1.711	MSH2 Lynch syndrome
New code		QA1.712	MSH6 Lynch syndrome
New code		QA1.713	PMS2 Lynch syndrome
New code		QA1.714	EPCAM Lynch syndrome
New code		QA1.719	Lynch syndrome, unspecified

Z15 Genetic susceptibility to disease

Z15.0 Genetic susceptibility to malignant neoplasm

New sub-subcategory	Z15.06	Genetic susceptibility to malignant neoplasm of digestive system	
New code		Z15.060	Genetic susceptibility to colorectal cancer
New code		Z15.068	Genetic susceptibility to other malignant neoplasm of digestive system
Add			genetic susceptibility to biliary tract cancer
Add			genetic susceptibility to gastric cancer
Add			genetic susceptibility to pancreatic cancer
Add			genetic susceptibility to small bowel cancer
New code	Z15.07	Genetic susceptibility to malignant neoplasm of urinary tract	

Multiple Sclerosis Phenotypes

This topic was presented at the March 2023 ICD-10 Coordination and Maintenance meeting. Based on public comments, revisions to the proposal have been made for reconsideration. Changes are indicated in **bold**.

Multiple sclerosis (MS) is a chronic inflammatory disease of the central nervous system leading to demyelination and neurodegeneration. MS affects an estimated 900,000 people in the United States [1]. Diagnosis is based on a combination of signs and symptoms, radiographic findings, and laboratory findings, which are components of the 2017 McDonald Criteria [2]. The core MS phenotypes are those of relapsing and progressive disease. The pattern and course of MS is further categorized into several clinical subtypes: clinically isolated syndrome, often representing the first attack of MS, relapsing-remitting multiple sclerosis (RRMS), secondary progressive multiple sclerosis (SPMS), and primary progressive multiple sclerosis (PPMS). Clinically isolated syndrome, RRMS, and active secondary SPMS are considered to be relapsing forms of MS. The large majority of patients with MS initially follow a relapsing-remitting course, defined by acute exacerbations from which they typically completely or incompletely recover, with periods of relative clinical stability in between [3]. The transition from RRMS to SPMS usually occurs 10 to 20 years after disease onset; SPMS is associated with sustained disability progression and loss of discrete relapse events [4]. Both clinical relapses and new radiographic lesions can occur in the context of progressive MS, and establishing when the transition from relapsing to progressive MS occurs is often difficult to determine prospectively [5]. SPMS can be further characterized as either active (with clinical relapses) or non-active (without relapses). PPMS, which represents about 10 percent of adult MS cases at disease onset, is characterized by, often insidious, disease progression from onset, although occasional plateaus, temporary minor improvements, and acute relapses may occur [6]. Currently, there is only one ICD-10-CM code for MS (G35), which does not capture the MS phenotypes.

Several classes of drugs and biologics to treat MS, with varying mechanisms of action and routes of administration, are available for RRMS and active SPMS [1]. However, as of today, only one monoclonal antibody is approved for the treatment of PPMS. The proposed codes follow the recent revisions of U.S. package insert (USPI) indications performed by the U.S. Food and Drug Administration for all approved MS therapeutics and distinguishes between PPMS and relapsing forms of MS, a fundamental clinical distinction with therapeutic and prognostic implications.

This phenotype specificity would provide the ability to conduct analyses at the phenotype level, improve the interpretation of real-world data in the investigation of severe outcomes that might represent safety signals for some treatments approved to treat MS, and improve the design and interpretation of comparative safety and effectiveness studies.

The Center for Drug Evaluation and Research, U.S. Food and Drug Administration, is requesting new ICD-10-CM codes for Multiple Sclerosis (MS) to distinguish between different disease clinical

courses, evaluation of disease progression and long-term prognosis of MS in large population-based epidemiological assessments.

References

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TABULAR MODIFICATIONS

	G35	Multiple sclerosis
Delete		~~Disseminated multiple sclerosis~~
Delete		~~Generalized multiple sclerosis~~
Delete		~~Multiple sclerosis NOS~~
Delete		~~Multiple sclerosis of brain stem~~
Delete		~~Multiple sclerosis of cord~~
New code	G35.A	Relapsing-remitting multiple sclerosis
Add		Exclude1: Demyelinating disease of central nervous system, unspecified (G37.9)
New Subcategory	G35.B	Primary progressive multiple sclerosis
New code		**G35.B0 Primary progressive multiple sclerosis, unspecified**
New code		**G35.B1 Active primary progressive multiple sclerosis**
New code		**G35.B2 Non-active primary progressive multiple sclerosis**

New		
Subcategory	G35.C	Secondary progressive multiple sclerosis
New code	G35.C0	Secondary progressive multiple sclerosis, unspecified
New code	G35.C1	Active secondary progressive multiple sclerosis
New code	G35.C2	Non-active secondary progressive multiple sclerosis
New code	G35.D	Multiple sclerosis, unspecified
Add		Disseminated multiple sclerosis
Add		Generalized multiple sclerosis
Add		Multiple sclerosis NOS
Add		Multiple sclerosis of brain stem
Add		Multiple sclerosis of cord
	G37	Other demyelinating diseases of central nervous system
	G37.9	Demyelinating disease of central nervous system, unspecified
Add		Clinically isolated syndromes

Neovascular glaucoma

Neovascular glaucoma is a secondary closed angle glaucoma associated with diabetes and other retinovascular diseases, such as retinal vein occlusion and ocular ischemic syndrome.[1] The Diabetes Control Complications Trial (DCCT) noted a 24% incidence of neovascularization of the retina or optic nerve over nine years among the standard treatment group compared with 8% in the intensively treated group. Although these data are from years ago neovascular glaucoma remains an important public health issue.[2] A recent registry-based manuscript identified nearly 6000 cases in the Unites States.[2]

Neovascular glaucoma is notoriously difficult to treat. It is currently not possible to do a medical records review for the prevalence, management and outcomes of this distinct type of glaucoma as current ICD-10-CM coding uses a collective term, H40.89, Other specified glaucoma, which bundles many specific, but unrelated, conditions that share elevated intraocular pressure as a clinical sign. This bundling obscures the importance of this condition and is a key complication of diabetes mellitus often leading to severe visual loss. The use of intravitreous injected medications that inhibit vascular endothelial growth factors has greatly enhanced the ability of surgeons to treat this condition and improve the structural and functional outcomes of this disease. Having a specific ICD-10-CM code will assist in studies of the condition.

Currently, Neovascular Glaucoma in ICD-10-CM is included in a list with other specified types of glaucoma to include:

> *Right glaucoma due to vascular disorder; Right mixed mechanism glaucoma; Right neovascular glaucoma; Aphakic glaucoma, both eyes; Bilateral glaucoma due to vascular disorder; Bilateral mixed mechanism glaucoma; Bilateral neovascular glaucoma; Glaucoma associated with anterior segment anomaly; Glaucoma associated with vascular disorder; Glaucoma due to combination of mechanisms; Glaucoma with plateau iris; Left aphakic glaucoma.*

Because of the unique nature of neovascular glaucoma and it being associated with poorly controlled diabetes mellitus, the ability to identify this specific complication is important to understand the disease burden among diabetic patients in the population.

The American Academy of Ophthalmology is proposing the following tabular modifications to properly track neovascular glaucoma as an important medical complication for public health reporting, especially in the setting of diabetic retinopathy.

References:

1. Neovascular Glaucoma - EyeWiki (aao.org)
2. Diabetes control and complications trial research group. Progression of retinopathy with intensive versus conventional treatment in the Diabetes Control and Complications Trial. Ophthalmology. 1995; 102:647-661.

TABULAR MODIFICATIONS

	H40	Glaucoma
	H40.8	Other glaucoma
New sub-subcategory	H40.84	Neovascular secondary angle closure glaucoma
New code	H40.841	Neovascular secondary angle closure glaucoma, right eye
New code	H40.842	Neovascular secondary angle closure glaucoma, left eye
New code	H40.843	Neovascular secondary angle closure glaucoma, bilateral
New code	H40.849	Neovascular secondary angle closure glaucoma, unspecified eye

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.

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 so as to align clinical documentation, support effective and granular disease tracking, 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.

References

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TABULAR MODIFICATIONS

N64 Other disorders of breast

N64.5 Other signs and symptoms in breast

New Code	N64.54 Nipple ischemia
New Code	N64.55 Nipple necrosis

Odontogenic sinusitis

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 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. While a clinician could use these aforementioned codes and then separately code the infectious dental source, the infectious dental source is not always apparent at the time of evaluation by a medical doctor, so attempting to code both the dental and sinusitis codes would prove problematic.

Rhinologists, Dr. John Craig and Dr. Alberto M. Saibene are requesting new ICD-10-CM codes for ODS to facilitate both future research and clinical decision-making.

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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	J34.82	Odontogenic sinusitis
New code	J34.821	Odontogenic sinusitis, maxillary sinus
New code	J34.822	Odontogenic sinusitis, ethmoid sinus
New code	J34.823	Odontogenic sinusitis, frontal sinus
New code	J34.824	Odontogenic sinusitis, sphenoid sinus
New code	J34.829	Odontogenic sinusitis, unspecified
Add		Odontogenic sinusitis NOS

Postprocedural open deep wound without disruption

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 "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 deep wound without disruption (Z98.88)

Z98 Other postprocedural states

Z98.8 Other specified postprocedural states

New code — Z98.88 Postprocedural open deep wound without disruption
Add — Delayed abdominal closure following a procedure
Add — Postprocedural open abdomen
Add — Postprocedural temporary open surgical wound

Primary Progressive Apraxia of Speech

Primary progressive apraxia of speech (PPAOS) is the diagnosis given when apraxia of speech (AOS) is insidious, progressive, and the first or only clinical symptom associated with a neurodegenerative disease. These patients are often captured under the Primary Progressive Aphasia (PPA) consensus criteria, as AOS is one of two core features to diagnose the nonfluent/ agrammatic variant; however, this is misleading and can hide valuable information.

This imprecise labeling likely contributes to the delays to diagnosis experienced by these patients, as they do not otherwise have symptoms of aphasia. For instance, aphasia includes difficulty with comprehension (understanding spoken language and reading) and expression (writing and speaking), while AOS only impacts speaking. Over the last decade, the clinical presentation, underlying pathophysiology, and evolving neurological picture that are heralded by PPAOS have been detailed and demonstrated the distinction between PPAOS and PPA. The lack of widespread distinction between them has stifled progress in understanding the similarities and differences of these distinct but related disorders. While it is difficult to estimate, PPAOS likely has a prevalence of more than 4/100,000 and is estimated to reflect 20% of patients diagnosed with PPA. Some studies have shown that more patients with nonfluent/agrammatic of PPA have AOS than aphasia and, in fact, AOS has been recognized as the most prevalent feature of that variant.

Additional neurological signs and symptoms typically occur approximately 5 years from time of onset of speech problems. The most common signs and symptoms that occur later in patients with PPAOS include aphasia and those linked to the extrapyramidal system. Patients tend to develop features of corticobasal syndrome and progressive supranuclear palsy but do not meet strict criteria for either when AOS is the sole feature. Significantly, patients with PPAOS have been found to have better survival compared to those with PPA. PPAOS patients ultimately end up mute (unable to speak or verbally communicate). It is also common to observe changes in balance and gait, ultimately resulting in patients being wheelchair bound and being in an akinetic rigid state close to time of death. They often die from physical difficulties associated with disease progression (e.g., aspiration pneumonia or falls).

Currently, there are no available curative or disease modifying treatments for PPAOS or the evolving neurologic syndromes. However, symptomatic behavioral treatment is recommended. Research has demonstrated the potential of working with a speech-language pathologist to maintain speech function or slow the progression of deterioration. Recent studies have looked at transcranial direct current stimulation (tDCS) and transcranial magnetic stimulation (TMS) and shown some promise in improving speed and accuracy of speech, although more work is needed. Referrals to speech-language pathologists may facilitate identifying augmentative and alternative means of communication, and referrals to occupational therapy may support alternative means of access in the cases where limb rigidity, spasticity, or apraxia preclude typing on speech generating devices. Meeting with a speech-language pathologist regularly also allows for identification and management of later emerging speech, language, cognitive, and/or swallowing difficulties.

This proposal is submitted by the Neurodegenerative Research Group at Mayo Clinic, a multidisciplinary research group comprised of clinician scientists in speech-language pathology, neurology, neuropsychology, and neuroradiology. A new ICD-10-CM diagnosis code is being requested to specifically identify individuals with PPAOS to better track the incidence, prevalence, and progression.

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TABULAR MODIFICATIONS

G31 Other degenerative diseases of nervous system, not elsewhere classified

G31.0 Frontotemporal dementia

G31.01 Pick's disease
Primary progressive aphasia
Progressive isolated aphasia

G31.8 Other specified degenerative diseases of nervous system

New code G31.87 Primary progressive apraxia of speech

Skin changes due to skin failure

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]

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:

1. Langemo DK, Brown G. Skin fails too: Acute, chronic, and end-stage skin failure. Advances in Skin & Wound Care, 2006, 19(4), 206-211.

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 sub-subcategory	L98.81	Acute skin changes due to skin failure
Add		Code also underlying condition
New code	L98.810	Acute skin changes due to skin failure, skin intact
New code	L98.811	Acute skin changes due to skin failure, partial thickness
New code	L98.812	Acute skin changes due to skin failure, full thickness
New sub-subcategory	L98.82	Chronic skin changes due to skin failure
Add		Code also underlying condition
New code	L98.820	Chronic skin changes due to skin failure, skin intact
New code	L98.821	Chronic skin changes due to skin failure, partial thickness
New code	L98.822	Chronic skin changes due to skin failure, full thickness
New sub-subcategory	L98.83	End-stage skin changes due to skin failure
Add		Code also underlying condition
New code	L98.830	End-stage skin changes due to skin failure, skin intact
New code	L98.831	End-stage skin changes due to skin failure, partial thickness
New code	L98.832	End-stage skin changes due to skin failure, full thickness
New code	L98.89	Other specified disorders of the skin and subcutaneous tissue

Topical steroid withdrawal

This topic was presented at the March 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**.

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 “Steroid Withdrawal Syndrome and Steroid-induced Dermatitis” among others. The National Eczema Association, additionally, acknowledges TSW as a separate clinical entity from AD and highlights the absence of a formally recognized diagnostic criteria for TSW [4] (two independent, yet congruent, sets of diagnostic criteria are under consideration. [7] This lack of standardized criteria and inconsistency in naming create constraints in conducting population studies.

The most up-to-date literature on TSW indicate the onset of symptoms arise from potencies of TCS which were 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]. 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][9].

The management and treatment of TSW focus on total cessation of steroids, 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, steroid-free wet wrap therapy, dupilumab, psychological support and lifestyle adjustments. However, no current therapy appears to offer more than symptomatic control.

Currently, there is no 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 reported 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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TABULAR MODIFICATIONS

L30 Other and unspecified dermatitis

L30.8 Other specified dermatitis

New code **L30.81 Topical steroid-induced chronic dermatitis**
Add **Topical steroid withdrawal syndrome**

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

New code L30.89 Other specified dermatitis

Type 2 diabetes mellitus in remission

This topic was presented at the March 2024 ICD10 Coordination and Maintenance meeting. Based on public comments, revisions to the proposal have been made to 1.) address concerns of code selection in addition to diabetic complications with the addition of an Excludes 1 note, and 2.) emphasize the importance of adding this code as a means of embracing remission as a key therapeutic goal in T2DM.

People with type 2 diabetes mellitus (T2DM) should be considered in remission after sustaining normal blood glucose (sugar) levels for three months or more, according to a new consensus statement from the American Diabetes Association® (ADA), the Endocrine Society, the European Association for the Study of Diabetes and Diabetes UK jointly published in 2021 in Diabetes Care, the Journal of Clinical Endocrinology & Metabolism, Diabetologia, and Diabetic Medicine, respectively.

Traditional T2DM management, based on the belief that T2DM is irreversible, has concentrated on glycemic control through diabetes education and glucose-lowering medication intensification. With this approach, however, adequate glycemic control is not achieved in a considerable proportion of patients. Thus, weight management is increasingly taking center stage in T2DM, and there is accumulating evidence that T2DM remission can be achieved through weight loss interventions. Moreover, patients in remission should thereafter be kept under regular review with annual testing and intervention management as a means of achieving long term remission and preventing macro or microvascular complications.

Individuals with complications like retinopathy, nephropathy, or neuropathy may find it more challenging to achieve remission. Managing these complications often requires a multidisciplinary approach and focusing on both glycemic control and specific treatments for complications. Therefore, this proposal focuses on remission only in the context of uncomplicated T2DM.

Permanente Medicine is requesting the following tabular modification to report remission status when achieved in uncomplicated type 2 diabetics. This new code reflects the goal to achieve clinical regression along the diabetes continuum in alignment with the current clinical guidelines.

References

1. Buse JB, Caprio S, Cefalu WT, et al. How do we define cure of diabetes? Diabetes Care 2009;32:2133–2135
2. Shibib L, Al-Qaisi M, Ahmed A, Miras AD, Nott D, Pelling M, Greenwald SE, Guess N. Reversal and Remission of T2DM - An Update for Practitioners. Vasc Health Risk Manag. 2022 Jun 14;18:417-443. doi: 10.2147/VHRM.S345810. PMID: 35726218; PMCID: PMC9206440.

TABULAR MODIFICATIONS

E11 Type 2 diabetes mellitus

E11.9 Type 2 diabetes mellitus without complications

New code	E11.A Type 2 diabetes mellitus without complications in remission
Add	Excludes1: Type 2 diabetes mellitus, with complications (E11.0-E11.8)
Add	Excludes1: Type 2 diabetes mellitus, without complications not in remission (E11.9)

Usher Syndrome

This topic was presented at the September 2023 and March 2024 ICD10 Coordination and Maintenance meeting. Based on public comments, revisions to the proposal have been made for reconsideration. Usher syndrome (USH) is a hereditary disorder with well-defined genetic causation that results in impairment of both hearing and vision. USH is a recessive genetic disorder that is responsible for 50% of those with hereditary deafblindness[1]. Hearing loss varies in age of onset and severity as described further below. Retinitis pigmentosa, the visual component of USH, is a progressive and untreatable retinal degeneration that initially causes nightblindness followed by loss of peripheral vision and finally impaired central vision. USH also affects balance in some patients. Variants in at least 12 genes have been identified as causing USH with a combined estimated incidence of 4-17:100,000[1]. Three clinically identifiable categories of USH account for the majority of patient presentations and are characterized by age of onset and severity of symptoms[2]:

• Type 1: Children have profound hearing loss or deafness at birth and may have severe balance problems that may lead to delayed motor milestones. Many obtain little or no benefit from hearing aids, but early use of cochlear implants may allow for development of speech. Decreased night vision by age 10, progressing to severe vision loss by midlife.

• Type 2: Moderate to severe hearing loss at birth. Normal balance. Decreased night vision by adolescence, progressing to severe vision loss by midlife.

• Type 3: Progressive hearing loss in childhood or early teens. Normal to near-normal balance in childhood. Chance of later problems. Vision loss varies in severity and age of onset; night vision problems often begin in teens and progress to severe vision loss by midlife.

Given the current absence of a unique ICD-10-CM code that simultaneously captures the auditory and visual manifestations of USH, clinicians are forced to choose among several non-specific codes including: H35.5 (Hereditary retinal dystrophy), H35.53 (Other dystrophies primarily involving the sensory retina), H35.52 (Pigmentary retinal dystrophy), H91.93 (Unspecified hearing loss, bilateral), Q87.89 (Other specified congenital malformation syndromes, not elsewhere classified). The proposed new codes are important at a systems level as data about patients with USH cannot currently be reliably extracted from general medical databases or disease-specific registries such as the American Academy of Ophthalmology's IRIS Registry.

Usher 1F Collaborative, the Usher Syndrome Coalition, and the Usher Syndrome Society, which are all nonprofit patient advocacy organizations, are requesting the following new codes to enable better tracking of these cases and treatment outcomes.

References
1. https://www.nidcd.nih.gov/health/usher-syndrome#3
2. Nolen RM, Hufnagel RB, Friedman TB, Turriff AE, Brewer CC, Zalewski CK, King KA, Wafa TT, Griffith AJ, Brooks BP, Zein WM. Atypical and ultra-rare Usher syndrome: a review. Ophthalmic Genet. 2020 Oct;41(5):401-412. doi: 10.1080/13816810.2020.1747090. Epub 2020 May 6. PMID: 32372680.

TABULAR MODIFICATIONS

	Q99	Other chromosome abnormalities, not elsewhere classified
	Q99.8	Other specified chromosome abnormalities
New sub-subcategory	Q99.81	Usher syndrome
New code	Q99.811	Usher syndrome, type 1
New code	Q99.812	Usher syndrome, type 2
New code	Q99.813	Usher syndrome, type 3
New code	Q99.818	Other Usher syndrome
New code	Q99.819	Usher syndrome, unspecified
New code	Q99.89	Other specified chromosome abnormalities

Utility Insecurity

This proposal represents a formal Gravity Project (GP) ICD-10-CM submission representing needed terms from the project's consensus, evidence-based analysis of the utility insecurity domain. Utility insecurity covers the experience of difficulty paying for utilities, burdensome utility costs due to housing inadequacies, and at its most severe, not having utility service because it was disconnected due to inability to pay for services.

At the September 2022, ICD10 Coordination and Maintenance Committee meeting a new code was requested, Z59.12 Inadequate housing utilities. As a subset of inadequate housing, it is bound to the elements of utility insecurity that intersect with the quality of housing. This can reasonably include both burdensome costs related to housing inadequacies and the disconnection of services.

Given that difficulty paying for utilities is not a problem of housing, but one of finances, this aspect of utility insecurity is not well represented by Z59.12. In the past, Gravity has built novel concepts to cover the breadth of the other core socioeconomic insecurities (food insecurity, housing instability, transportation insecurity, etc). Gravity recognizes that Z59.12 is currently implemented and part of HHS guidance. Given this, the GPis reluctant to change this term. However, to support consistent documentation of the full breadth of utility insecurity, Gravity recommends adapting its prior concept of financial insecurity to include a specific utility insecurity subtype to represent the "difficulty paying" concept, as detailed below.

The GP also requests the revision of utility insecurity inclusion term, (unable to obtain adequate utilities due to limited financial resources) from Z59.87 Material hardship, due to limited financial resources, NEC to focus utility insecurity documentation between Z59.12 and the proposed new financial insecurity inclusion terms.

TABULAR MODIFICATION

Z59 Problems related to housing and economic circumstances
Z59.1 Inadequate housing
Z59.12 Inadequate housing utilities
Lack of electricity services
Lack of gas services
Lack of oil services
Lack of water services
Add Excludes2: financial insecurity, difficulty paying for utilities (Z59.861)
Z59.19 Other inadequate housing
Add Poor housing weatherization

Z59.8 Other problems related to housing and economic circumstances

New subcategory Z59.86 Financial Insecurity

Delete ~~Bankruptcy~~

New code Z59.861 Financial insecurity, difficulty paying for utilities

Add Difficulty paying for electricity

Add Difficulty paying for heat

Add Difficulty paying for oil

Add Difficulty paying water bill

Add Utility disconnect notice due to inability to pay

Add Excludes2: inadequate housing utilities (Z59.12)

New code Z59.868 Other specified financial insecurity

Add Bankruptcy

New code Z59.869 Financial insecurity, unspecified

Z59.87 Material hardship, due to limited financial resources, NEC

Delete ~~Unable to obtain adequate utilities due to limited financial resources~~

Xylazine-associated wounds

This topic was originally presented at the March 2024 ICD10 Coordination and Maintenance meeting. Based on comments received during the public comments period, a revised proposal is being presented for consideration. Changes are indicated in **bold**.

On April 12, 2023, the White House Office of National Drug Control Policy (ONDCP) declared fentanyl adulterated or associated with xylazine an emerging drug threat. In their response plan, the ONDCP urged the creation of ICD-10-CM codes specific to xylazine and its health consequences.[1] This proposal addresses the need for standardized diagnostic coding for one distinct and ulcerating consequence: that of xylazine-associated wounds.

Xylazine is an α-2 receptor agonist approved by the Food and Drug Administration (FDA) for sedation and analgesia in veterinary medicine.[2] Xylazine is not approved for use in humans due to unsafe hypotension and central nervous system depression. Xylazine first emerged as an illicit drug in the early 2000s in Puerto Rico and 2006 in Philadelphia, PA.[3] In 2009, Rodríguez et al. reported a higher prevalence of skin wounds among people who injected xylazine-containing drugs compared to people who injected drugs without xylazine.[4] As xylazine has become pervasive in Philadelphia's illicit opioid supply, wounds and skin and soft tissue infections have come to dominate the needs of people who use drugs. In 2021, Xylazine was present in 90% of illicit fentanyl samples tested by the Philadelphia Department of Public Health.[5] Between 2020 and 2021, there was a 39% increase in hospitalizations for skin and soft tissue infections related to injection drug use in Philadelphia.[6] Wound care providers across community, outpatient, and hospital settings in Philadelphia have developed an expertise in identifying, evaluating, and treating xylazine-associated wounds. However, the lack of standardized ICD-10-CM codes has hindered continuity of care, as well as the evaluation and monitoring of xylazine-wounds and associated sequelae.

Xylazine-associated wounds are distinctly recognizable, yet their etiology is poorly understood.[7] Xylazine-associated wounds are consistently described in peer-reviewed literature as partial to full thickness skin defects with progressive necrosis of the skin, muscle, tendon, and bone.[8,9,10,11,12,13,14,15,16] Although xylazine-associated wounds commonly develop at sites of injection, they can appear anywhere on the body irrespective of the route of xylazine administration.[7,12,17,18] Xylazine-associated wounds may initially appear as areas of blistered skin, often over reddish-purple discolored tissue which evolve into a thick layer of eschar overlying a partial or full thickness ulcer that progressively increases in size and depth.[11,19] Indeed, xylazine-associated wounds are often typified by the presence of necrotic tissue (eschar, and slough) and wound diameters greater than 10 cm.[19] Xylazine-associated wounds increase a person's risk of bacteremia, endocarditis, sepsis, limb amputation, and death.[12,18] Treatment of xylazine-associated wounds is often complicated by patients' experiences of opioid and xylazine withdrawal, co-occurring mental illness, local and systemic infections, physical disability, and health-related social needs such as housing instability and food insecurity.[9,10,14]

Catherine Tomson, Daniel Teixeira da Silva, MD, MSHP, and Rachel Neuschatz, RN, MSN of the Substance Use Prevention and Harm Reduction Division in the Philadelphia Department of Public Health are requesting the following tabular modifications for reporting, evaluation, and monitoring of xylazine-associated wounds.

References:

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TABULAR MODIFICATIONS

T65 Toxic effect of other and unspecified substances

T65.8 Toxic effect of other specified substances

New sub-subcategory T65.84 Toxic effect of xylazine

Add		Use additional code(s) for all associated manifestations, such as:
Add		cellulitis and acute lymphangitis (L03.-)
Add		cutaneous abscess, furuncle and carbuncle (L02.-)
Add		non-pressure chronic ulcer of lower limb, not elsewhere classified (L97.-)
Add		**non-pressure chronic ulcer of skin, not elsewhere classified (L98.4-)**
Add		**other psychoactive substance dependence with withdrawal (F19.23-)**
New code	T65.841	Toxic effect of xylazine, accidental (unintentional)
Add		Toxic effect of xylazine NOS
New code	T65.842	Toxic effect of xylazine, intentional self-harm
New code	T65.843	Toxic effect of xylazine, assault
New code	T65.844	Toxic effect of xylazine, undetermined

	L97	Non-pressure chronic ulcer of lower limb, not elsewhere classified
	L97.2	Non-pressure chronic ulcer of calf
Add		Non-pressure chronic ulcer of shin
	L98	Other disorders of skin and subcutaneous tissue, not elsewhere classified
	L98.4	Non-pressure chronic ulcer of skin, not elsewhere classified
New sub-subcategory	L98.43	Non-pressure chronic ulcer of abdomen
New code	L98.431	Non-pressure chronic ulcer of abdomen limited to breakdown of skin
New code	L98.432	Non-pressure chronic ulcer of abdomen with fat layer exposed

New code	L98.433	Non-pressure chronic ulcer of abdomen with necrosis of muscle
New code	L98.434	Non-pressure chronic ulcer of abdomen with necrosis of bone
New code	L98.435	Non-pressure chronic ulcer of abdomen with muscle involvement without evidence of necrosis
New code	L98.436	Non-pressure chronic ulcer of abdomen with bone involvement without evidence of necrosis
New code	L98.438	Non-pressure chronic ulcer of abdomen with other specified severity
New code	L98.439	Non-pressure chronic ulcer of abdomen with unspecified severity
New sub-subcategory	L98.44	Non-pressure chronic ulcer of chest
New code	L98.441	Non-pressure chronic ulcer of chest limited to breakdown of skin
New code	L98.442	Non-pressure chronic ulcer of chest with fat layer exposed
New code	L98.443	Non-pressure chronic ulcer of chest with necrosis of muscle
New code	L98.444	Non-pressure chronic ulcer of chest with necrosis of bone
New code	L98.445	Non-pressure chronic ulcer of chest with muscle involvement without evidence of necrosis
New code	L98.446	Non-pressure chronic ulcer of chest with bone involvement without evidence of necrosis
New code	L98.448	Non-pressure chronic ulcer of chest with other specified severity
New code	L98.449	Non-pressure chronic ulcer of chest with unspecified severity

New sub-subcategory	L98.45	Non-pressure chronic ulcer of neck
New code	L98.451	Non-pressure chronic ulcer of neck limited to breakdown of skin
New code	L98.452	Non-pressure chronic ulcer of neck with fat layer exposed
New code	L98.453	Non-pressure chronic ulcer of neck with necrosis of muscle
New code	L98.454	Non-pressure chronic ulcer of neck with necrosis of bone
New code	L98.455	Non-pressure chronic ulcer of neck with muscle involvement without evidence of necrosis
New code	L98.456	Non-pressure chronic ulcer of neck with bone involvement without evidence of necrosis
New code	L98.458	Non-pressure chronic ulcer of neck with other specified severity
New code	L98.459	Non-pressure chronic ulcer of neck with unspecified severity
New sub-subcategory	L98.46	Non-pressure chronic ulcer of face
New code	L98.461	Non-pressure chronic ulcer of face limited to breakdown of skin
New code	L98.462	Non-pressure chronic ulcer of face with fat layer exposed
New code	L98.463	Non-pressure chronic ulcer of face with necrosis of muscle
New code	L98.464	Non-pressure chronic ulcer of face with necrosis of bone
New code	L98.465	Non-pressure chronic ulcer of face with muscle involvement without evidence of necrosis
New code	L98.466	Non-pressure chronic ulcer of face with bone involvement without evidence of necrosis

New code L98.468 Non-pressure chronic ulcer of face with other specified severity

New code L98.469 Non-pressure chronic ulcer of face with unspecified severity

New sub-subcategory L98.47 Non-pressure chronic ulcer of groin

New code L98.471 Non-pressure chronic ulcer of groin limited to breakdown of skin

New code L98.472 Non-pressure chronic ulcer of groin with fat layer exposed

New code L98.473 Non-pressure chronic ulcer of groin with necrosis of muscle

New code L98.474 Non-pressure chronic ulcer of groin with necrosis of bone

New code L98.475 Non-pressure chronic ulcer of groin with muscle involvement without evidence of necrosis

New code L98.476 Non-pressure chronic ulcer of groin with bone involvement without evidence of necrosis

New code L98.478 Non-pressure chronic ulcer of groin with other specified severity

New code L98.479 Non-pressure chronic ulcer of groin with unspecified severity

New code L98.A Non-pressure chronic ulcer of upper limb, not elsewhere classified
Add Includes: chronic ulcer of upper limb NOS
Add non-healing ulcer of skin
Add ulcer of skin NOS

Add Exclude2: pressure ulcer (pressure area) (L89.-)
Add gangrene (I96)
Add skin infections (L00-L08)
Add specific infections classified to A00-B99
Add ulcer of lower limb NEC (L97.-)
Add varicose ulcer (I83.0-I83.93)

New sub-subcategory	L98.A1	Non-pressure chronic ulcer of upper arm
Add		Non-pressure chronic ulcer of axilla
New sub-subcategory	L98.A11	Non-pressure chronic ulcer of right upper arm
New code	L98.A111	Non-pressure chronic ulcer of right upper arm limited to breakdown of skin
New code	L98.A112	Non-pressure chronic ulcer of right upper arm with fat layer exposed
New code	L98.A113	Non-pressure chronic ulcer of right upper arm with necrosis of muscle
New code	L98.A114	Non-pressure chronic ulcer of right upper arm with necrosis of bone
New code	L98.A115	Non-pressure chronic ulcer of right upper arm with muscle involvement without evidence of necrosis
New code	L98.A116	Non-pressure chronic ulcer of right upper arm with bone involvement without evidence of necrosis
New code	L98.A118	Non-pressure chronic ulcer of right upper arm with other specified severity
New code	L98.A119	Non-pressure chronic ulcer of right upper arm with unspecified severity
New sub-subcategory	L98.A12	Non-pressure chronic ulcer of left upper arm
New code	L98.A121	Non-pressure chronic ulcer of left upper arm limited to breakdown of skin
New code	L98.A122	Non-pressure chronic ulcer of left upper arm with fat layer exposed
New code	L98.A123	Non-pressure chronic ulcer of left upper arm with necrosis of muscle
New code	L98.A124	Non-pressure chronic ulcer of left upper arm with necrosis of bone

New code	L98.A125	Non-pressure chronic ulcer of left upper arm with muscle involvement without evidence of necrosis
New code	L98.A126	Non-pressure chronic ulcer of left upper arm with bone involvement without evidence of necrosis
New code	L98.A128	Non-pressure chronic ulcer of left upper arm with other specified severity
New code	L98.A129	Non-pressure chronic ulcer of left upper arm with unspecified severity
New sub-subcategory	L98.A19	Non-pressure chronic ulcer of unspecified upper arm
New code	L98.A191	Non-pressure chronic ulcer of unspecified upper arm limited to breakdown of skin
New code	L98.A192	Non-pressure chronic ulcer of unspecified upper arm with fat layer exposed
New code	L98.A193	Non-pressure chronic ulcer of unspecified upper arm with necrosis of muscle
New code	L98.A194	Non-pressure chronic ulcer of unspecified upper arm with necrosis of bone
New code	L98.A195	Non-pressure chronic ulcer of unspecified upper arm with muscle involvement without evidence of necrosis
New code	L98.A196	Non-pressure chronic ulcer of unspecified upper arm with bone involvement without evidence of necrosis
New code	L98.A198	Non-pressure chronic ulcer of unspecified upper arm with other specified severity
New code	L98.A199	Non-pressure chronic ulcer of unspecified upper arm with unspecified severity

New sub-subcategory	L98.A2	Non-pressure chronic ulcer of forearm
New sub-subcategory	L98.A21	Non-pressure chronic ulcer of right forearm
New code	L98.A211	Non-pressure chronic ulcer of right forearm limited to breakdown of skin
New code	L98.A212	Non-pressure chronic ulcer of right forearm with fat layer exposed
New code	L98.A213	Non-pressure chronic ulcer of right forearm with necrosis of muscle
New code	L98.A214	Non-pressure chronic ulcer of right forearm with necrosis of bone
New code	L98.A215	Non-pressure chronic ulcer of right forearm with muscle involvement without evidence of necrosis
New code	L98.A216	Non-pressure chronic ulcer of right forearm with bone involvement without evidence of necrosis
New code	L98.A218	Non-pressure chronic ulcer of right forearm with other specified severity
New code	L98.A219	Non-pressure chronic ulcer of right forearm with unspecified severity
New sub-subcategory	L98.A22	Non-pressure chronic ulcer of left forearm
New code	L98.A221	Non-pressure chronic ulcer of left forearm limited to breakdown of skin
New code	L98.A222	Non-pressure chronic ulcer of left forearm with fat layer exposed
New code	L98.A223	Non-pressure chronic ulcer of left forearm with necrosis of muscle
New code	L98.A224	Non-pressure chronic ulcer of left forearm with necrosis of bone

New code	L98.A225	Non-pressure chronic ulcer of left forearm with muscle involvement without evidence of necrosis
New code	L98.A226	Non-pressure chronic ulcer of left forearm with bone involvement without evidence of necrosis
New code	L98.A228	Non-pressure chronic ulcer of left forearm with other specified severity
New code	L98.A229	Non-pressure chronic ulcer of left forearm with unspecified severity
New sub-subcategory	L98.A29	Non-pressure chronic ulcer of unspecified forearm
New code	L98.A291	Non-pressure chronic ulcer of unspecified forearm limited to breakdown of skin
New code	L98.A292	Non-pressure chronic ulcer of unspecified forearm with fat layer exposed
New code	L98.A293	Non-pressure chronic ulcer of unspecified forearm with necrosis of muscle
New code	L98.A294	Non-pressure chronic ulcer of unspecified forearm with necrosis of bone
New code	L98.A295	Non-pressure chronic ulcer of unspecified forearm with muscle involvement without evidence of necrosis
New code	L98.A296	Non-pressure chronic ulcer of unspecified forearm with bone involvement without evidence of necrosis
New code	L98.A298	Non-pressure chronic ulcer of unspecified forearm with other specified severity
New code	L98.A299	Non-pressure chronic ulcer of unspecified forearm with unspecified severity

New sub-subcategory	L98.A3	Non-pressure chronic ulcer of hand
New sub-subcategory	L98.A31	Non-pressure chronic ulcer of right hand
New code	L98.A311	Non-pressure chronic ulcer of right hand limited to breakdown of skin
New code	L98.A312	Non-pressure chronic ulcer of right hand with fat layer exposed
New code	L98.A313	Non-pressure chronic ulcer of right hand with necrosis of muscle
New code	L98.A314	Non-pressure chronic ulcer of right hand with necrosis of bone
New code	L98.A315	Non-pressure chronic ulcer of right hand with muscle involvement without evidence of necrosis
New code	L98.A316	Non-pressure chronic ulcer of right hand with bone involvement without evidence of necrosis
New code	L98.A318	Non-pressure chronic ulcer of right hand with other specified severity
New code	L98.A319	Non-pressure chronic ulcer of right hand with unspecified severity
New sub-subcategory	L98.A32	Non-pressure chronic ulcer of left hand
New code	L98.A321	Non-pressure chronic ulcer of left hand limited to breakdown of skin
New code	L98.A322	Non-pressure chronic ulcer of left hand with fat layer exposed
New code	L98.A323	Non-pressure chronic ulcer of left hand with necrosis of muscle
New code	L98.A324	Non-pressure chronic ulcer of left hand with necrosis of bone

New code	L98.A325	Non-pressure chronic ulcer of left hand with muscle involvement without evidence of necrosis
New code	L98.A326	Non-pressure chronic ulcer of left hand with bone involvement without evidence of necrosis
New code	L98.A328	Non-pressure chronic ulcer of left hand with other specified severity
New code	L98.A329	Non-pressure chronic ulcer of left hand with unspecified severity
New sub-subcategory	L98.A39	Non-pressure chronic ulcer of unspecified hand
New code	L98.A391	Non-pressure chronic ulcer of unspecified hand limited to breakdown of skin
New code	L98.A392	Non-pressure chronic ulcer of unspecified hand with fat layer exposed
New code	L98.A393	Non-pressure chronic ulcer of unspecified hand with necrosis of muscle
New code	L98.A394	Non-pressure chronic ulcer of unspecified hand with necrosis of bone
New code	L98.A395	Non-pressure chronic ulcer of unspecified hand with muscle involvement without evidence of necrosis
New code	L98.A396	Non-pressure chronic ulcer of unspecified hand with bone involvement without evidence of Necrosis
New code	L98.A398	Non-pressure chronic ulcer of unspecified hand with other specified severity
New code	L98.A399	Non-pressure chronic ulcer of unspecified hand with unspecified severity

TABULAR MODIFICATIONS PROPOSED ADDENDA

All approved modifications will be effective April 1, 2025

Tuberculosis (A15-A19)

Add — Use additional code, if applicable, for associated cachexia (E88.A)

B59 Pneumocystosis

Revise — Pneumonia due to Pneumocystis ~~jiroveci~~ jirovecii

E23 Hypofunction and other disorders of the pituitary gland

E23.0 Hypopituitarism

Add — Use additional code, if applicable, for associated cachexia (E88.A)

J65 Pneumoconiosis associated with tuberculosis

Add — Use additional code, if applicable, for associated cachexia (E88.A)

K56 Paralytic ileus and intestinal obstruction without hernia

K56.4 Other impaction of intestine

K56.41 Fecal impaction

Delete — ~~Excludes1: constipation (K59.0-)~~

Add — Excludes2: constipation (K59.0-)

Delete — ~~Excludes2: incomplete defecation (R15.0)~~

K59 Other functional intestinal disorders

K59.0 Constipation

Delete — ~~Excludes1: fecal impaction (K56.41)~~

Add — Excludes2: fecal impaction (K56.41)

Delete — ~~Excludes2: incomplete defecation (R15.0)~~

K72 Hepatic failure, not elsewhere classified

Add — Use additional code, if applicable, for ascites (R18.8)

K73 Chronic hepatitis, not elsewhere classified

Add — Use additional code, if applicable, for ascites (R18.8)

K90 Intestinal malabsorption
K90.8 Other intestinal malabsorption
K90.82 Short bowel syndrome
Add Excludes1: Postsurgical malabsorption, not elsewhere classified (K91.2)

K90.822 Short bowel syndrome without colon in continuity
Short bowel syndrome without colonic continuity
Add Use additional code, if applicable, for colostomy status (Z93.3)

N18 Chronic kidney disease (CKD)
Revise Use additional code, if applicable, to identify: ~~kidney transplant status, if applicable,~~ ~~(Z94.0)~~
Add associated cachexia (E88.A)
Add kidney transplant status, (Z94.0)

O04 Complications following (induced) termination of pregnancy
Revise Excludes2: encounter for elective termination of pregnancy~~, uncomplicated~~ (Z33.2)

O26 Maternal care for other conditions predominantly related to pregnancy
O26.6 Liver and biliary tract disorders in pregnancy, childbirth and the puerperium
O26.64 Intrahepatic cholestasis of pregnancy
Add Use additional code for hepatic obstruction (K76.89)

Injury, poisoning and certain other consequences of external causes (S00-T88)

Note: Use secondary code(s) from Chapter 20, External causes of morbidity, to indicate cause of injury. Codes within the T section that include the external cause do not require an additional external cause code

Delete ~~Excludes1: birth trauma (P10-P15)~~
Delete ~~obstetric trauma (O70-O71)~~

Add Excludes2: birth trauma (P10-P15)
Add obstetric trauma (O70-O71)

Revise The appropriate 7th character is to be added to each code from category S06, except as noted below

Z3A Weeks of gestation

Note: Codes from category Z3A are for use, only on the maternal record, to indicate the weeks of gestation of the pregnancy, if known.

Revise Code first obstetric condition or encounter for delivery (O09-O60, O80-O82 O94-O9A)

INDEX MODIFICATIONS PROPOSED ADDENDA

All approved modifications will be effective April 1, 2025

Anomaly, anomalous (congenital) (unspecified type) Q89.9
Revise - ~~Rieger's~~ Rieger Q13.81

Decrease(d)
- blood
Revise - - glucose E16.2
Revise - - - level 1 E16.A1
Revise - - - level 2 E16.A2
Revise - - - level 3 E16.A3
Revise - glucose ~~R73.09~~ E16.2

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

Dissection
- precerebral artery, congenital (nonruptured) Q28.1
Delete ~~- - Heartland A93.8~~

Glaucoma H40.9
- in (due to)
Revise - - ~~Rieger's~~ Rieger anomaly Q13.81 [H42]

Infection, infected, infective (opportunistic) B99.9
Revise - Cronobacter (sakazakii) ~~B96.89~~ A49.8
Revise - Pneumocystis ~~jiroveci~~ jirovecii (pneumonia) B59

Myocarditis
Revise - virus, viral ~~I40.0~~ B33.22

Revise Pneumocystis ~~jiroveci~~ jirovecii (pneumonia) B59

Pneumonia (acute) (double) (migratory) (purulent) (septic) (unresolved) J18.9
- in (due to)
Revise - - pneumocystosis (Pneumocystis carinii) (Pneumocystis ~~jiroveci~~ jirovecii) B59
Revise - Pneumocystis (carinii) (~~jiroveci~~ jirovecii) B59

Revise ~~Rieger's~~ Rieger anomaly or syndrome Q13.81

TABULAR MODIFICATIONS PROPOSED ADDENDA

All approved modifications will be effective October 1, 2025

B18 Chronic viral hepatitis
Add Use additional code, if applicable, for ascites (R18.8)

D53 Other nutritional anemias
D53.8 Other specified nutritional anemias
Anemia associated with deficiency of copper
Anemia associated with deficiency of molybdenum
Anemia associated with deficiency of zinc
Delete Excludes1: ~~nutritional deficiencies without anemia, such as~~:
Delete ~~copper deficiency NOS (E61.0)~~
Delete ~~molybdenum deficiency NOS (E61.5)~~
Delete ~~zinc deficiency NOS (E60)~~
Add Excludes2: nutritional deficiencies without anemia, such as:
Add copper deficiency NOS (E61.0)
Add molybdenum deficiency NOS (E61.5)
Add zinc deficiency NOS (E60)

D68 Other coagulation defects
D68.6 Other thrombophilia
D68.61 Antiphospholipid syndrome
Delete Excludes1: ~~anti-phospholipid antibody, finding without diagnosis (R76.0)~~
Delete ~~anti-phospholipid antibody with hemorrhagic disorder (D68.312)~~
Delete ~~lupus anticoagulant syndrome (D68.62)~~
Add Excludes2: anti-phospholipid antibody with hemorrhagic disorder (D68.312)
Add lupus anticoagulant syndrome (D68.62)

D68.62 Lupus anticoagulant syndrome
Delete Excludes1: ~~anticardiolipin syndrome (D68.61)~~
Delete ~~antiphospholipid syndrome (D68.61)~~
Delete ~~lupus anticoagulant (LAC) finding without diagnosis (R76.0)~~
Delete ~~lupus anticoagulant (LAC) with hemorrhagic disorder (D68.312)~~
Add Excludes2: anticardiolipin syndrome (D68.61)
Add antiphospholipid syndrome (D68.61)
Add lupus anticoagulant (LAC) with hemorrhagic disorder (D68.312)

E27 Other disorders of adrenal gland

E27.5 Adrenomedullary hyperfunction
Add Code also, if applicable:
Add malignant pheochromocytoma (C74.1-)
Add pheochromocytoma (benign) (D35.0-)
Add secondary hypertension (I15.2)

E35 Disorders of endocrine glands in diseases classified elsewhere
Code first underlying disease, such as:
late congenital syphilis of thymus gland [Dubois disease] (A50.59)
Add sequelae of tuberculosis of other organs (B90.8)
Delete ~~Use additional code, if applicable, to identify:~~
~~sequelae of tuberculosis of other organs (B90.8)~~

E71 Disorders of branched-chain amino-acid metabolism and fatty-acid metabolism
E71.5 Peroxisomal disorders
E71.54 Other peroxisomal disorders
E71.540 Rhizomelic chondrodysplasia punctata
Delete ~~Excludes1: chondrodysplasia punctata NOS (Q77.3)~~

E88 Other and unspecified metabolic disorders
E88.0 Disorders of plasma-protein metabolism, not elsewhere classified
Revise Code also, if applicable, ligneous conjunctivitis (H10.51-)

H33 Retinal detachments and breaks
H33.3 Retinal breaks without detachment
Delete ~~Excludes1: peripheral retinal degeneration without break (H35.4-)~~
Add Excludes2: peripheral retinal degeneration without break (H35.4-)

H35 Other retinal disorders
H35.4 Peripheral retinal degeneration
Delete ~~Excludes1: peripheral retinal degeneration with retinal break (H33.3-)~~
Add Excludes2: peripheral retinal degeneration with retinal break (H33.3-)

H53 Visual disturbances
H53.0 Amblyopia ex anopsia
H53.03 Strabismic amblyopia
Delete ~~Excludes1: strabismus (H50.-)~~
Add Excludes1: strabismus (H50.-)

I51 Complications and ill-defined descriptions of heart disease
Delete ~~Excludes1: any condition in I51.4-I51.9 due to hypertension (I11.-)~~
Delete ~~any condition in I51.4-I51.9 due to hypertension and chronic kidney disease (I13.-)~~
Delete ~~heart disease specified as rheumatic (I00-I09)~~

Add — Excludes2: heart disease specified as rheumatic (I00-I09)

I51.5 Myocardial degeneration
Add — Excludes1: myocardial degeneration due to hypertension (I11.-)
Add — myocardial degeneration due to hypertension and chronic kidney disease (I13.-)

I51.7 Cardiomegaly
Add — Excludes1: cardiomegaly due to hypertension (I11.-)
Add — cardiomegaly due to hypertension and chronic kidney disease (I13.-)

I80 Phlebitis and thrombophlebitis
Revise — Code first, if applicable:

J40 Bronchitis, not specified as acute or chronic
Revise — Excludes1: allergic bronchitis NOS (J45.909-) (removing hyphen)

J43 Emphysema
Delete — Excludes1: ~~emphysema due to inhalation of chemicals, gases, fumes or vapors (J68.4)~~
Add — Excludes2: emphysema due to inhalation of chemicals, gases, fumes or vapors (J68.4)

J44 Other chronic obstructive pulmonary disease
Delete — Excludes1: ~~chronic bronchitis NOS (J42)~~
Delete — ~~chronic simple and mucopurulent bronchitis (J41.-)~~
Delete — ~~chronic tracheitis (J42)~~
Delete — ~~chronic tracheobronchitis (J42)~~

Add — Excludes2: chronic bronchitis NOS (J42)
Add — chronic simple and mucopurulent bronchitis (J41.-)
Add — chronic tracheitis (J42)
Add — chronic tracheobronchitis (J42)

M21 Other acquired deformities of limbs
M21.1 Varus deformity, not elsewhere classified
M21.15 Varus deformity, not elsewhere classified, hip
Revise — M21.159 Varus deformity, not elsewhere classified, unspecified hip

M24 Other specific joint derangements
M24.0 Loose body in joint
M24.07 Loose body in ankle and toe joints
Revise — M24.076 Loose body in unspecified toe ~~joints~~ joint(s)

M36 Systemic disorders of connective tissue in diseases classified elsewhere
M36.3 Arthropathy in other blood disorders
Add Code first underlying disease, such as:
Add delta-beta thalassemia (D56.2)
Add disease of blood and blood-forming organs, unspecified (D75.9)
Add other hemoglobinopathies (D58.2)
Add thalassemia (D56.-)

M61 Calcification and ossification of muscle
M61.1 Myositis ossificans progressiva
M61.12 Myositis ossificans progressiva, upper arm
Revise M61.129 Myositis ossificans progressiva, unspecified upper arm

M92 Other juvenile osteochondrosis
M92.8 Other specified juvenile osteochondrosis
Delete ~~Calcaneal apophysitis~~

Q75 Other congenital malformations of skull and face bones
Q75.0 Craniosynostosis
Q75.02 Coronal craniosynostosis
Revise Q75.029 Coronal craniosynostosis, unspecified

Q77 Osteochondrodysplasia with defects of growth of tubular bones and spine
Q77.3 Chondrodysplasia punctata
Revise Excludes1: Rhizomelic chondrodysplasia punctata (~~E71.43~~ E71.540)

O90 Complications of the puerperium, not elsewhere classified
O90.4 Postpartum acute kidney failure
Revise Excludes1: ~~non-~~anuria and oliguria (R34)

R40 Somnolence, stupor and coma
R40.2 Coma
R40.2A Nontraumatic coma due to underlying condition
Add Excludes1: coma scale, best motor response (R40.24.-)
Add coma scale, best verbal response (R40.22.-)
Add coma scale, eyes open (R40.21.-)
Add Glasgow coma scale, total score (R40.23.-)

R65 Symptoms and signs specifically associated with systemic inflammation and infection
R65.2 Severe sepsis
R65.21 Severe sepsis with septic shock

Add — Excludes1: postprocedural septic shock (T81.12-)

S62 Fracture at wrist and hand level
Revise — S62.9 Unspecified fracture of ~~wrist and~~ hand
Revise — S62.90 Unspecified fracture of unspecified ~~wrist and~~ hand
Revise — S62.91 Unspecified fracture of right ~~wrist and~~ hand
Revise — S62.92 Unspecified fracture of left ~~wrist and~~ hand

S74 Injury of nerves at hip and thigh level
S74.2 Injury of cutaneous sensory nerve at hip and thigh level
Revise — S74.21 Injury of cutaneous sensory nerve at hip and ~~high~~ thigh level, right leg

Z56 Problems related to employment and unemployment
Z56.6 Other physical and mental strain related to work
Add — Workplace stress

Z56.8 Other problems related to employment
Z56.89 Other problems related to employment
Add — Furloughed
Add — Underemployed

Z59 Problems related to housing and economic circumstances
Z59.02 Unsheltered homelessness
Add — Lives in a homeless encampment

INDEX MODIFICATION PROPOSED ADDENDA

All approved modifications will be effective October 1, 2025

Aneurysm (anastomotic) (artery) (cirsoid) (diffuse) (false) (fusiform) (multiple) (saccular) I72.9
- brain I67.1
- - mycotic I67.1

Add Note: if applicable, see also: Sequelae, infectious disease
- - - with endocarditis - see also Endocarditis

Ascites (abdominal) R18.8
- due to

Revise - - hepatitis – see also Hepatitis
Revise - - - chronic active (see also Hepatitis, chronic active) ~~K71.51~~K73.2
Add - - - - with toxic liver disease K71.51

Apophysitis (bone) -see also Osteochondropathy

Revise - calcaneus ~~M92.8~~ M92.6

Cholestasis NEC K83.1

Add - intrahepatic K76.89

Dactylitis

Revise - sickle-cell ~~D57.00~~ D57.04
Add - - Beta plus D57.454
Add - - Beta zero D57.434
Revise - - Hb C ~~D57.219~~ D57.214
Add - - Hb SD D57.814
Add - - Hb SE D57.814
Revise - - Hb SS ~~D57.00~~ D57.04
Revise - - specified NEC ~~D57.819~~ D57.814
Add - - thalassemia D57.414

Revise Desmoid (extra-abdominal) (tumor)~~ - see Neoplasm, connective tissue, uncertain behavior~~

Diabetes, diabetic (mellitus) (sugar) E11.9
- due to drug or chemical E09.9
- - with

Add - - - retina, hemorrhage E09.39

- due to underlying condition E08.9
- - with

Add - - - retina, hemorrhage E08.39

- specified type NEC E13.9
- - with
Add - - - retina, hemorrhage E13.39

- type 1 E10.9
- - with
Add - - - retina, hemorrhage E10.39
- type 2 E11.9
- - with
Add - - - retina, hemorrhage E11.39

Disease
Add - Heartland A93.8
- hemoglobin or Hb

- - Hb-SD D57.80
- - - with
Add - - - - priapism D57.818

- - Hb-SE D57.80
- - - with
Add - - - - priapism D57.818

- - specified NEC D57.80
- - - with
Add - - - - priapism D57.218

- - spherocytosis D57.80
- - - with
Add - - - - priapism D57.818

- - Hb-C D57.20
- - thalassemia D57.40
- - - with
Revise - - - - priapism ~~D57.458~~ D57.418

- - - beta plus D57.44
- - - - with
Add - - - - priapism D57.458

- - - beta zero D57.42
- - - - with
Add - - - - - priapism D57.438
- - S or SS D57.1
- - - with

- - - - acute chest syndrome D57.01
- - - - crisis (painful) D57.04
Revise - - - - - with ~~complication specified NEC D57.09~~
Add - - - - - - dactylitis D57.04
Add - - - - - - specified complication D57.09

- - - beta plus D57.44
- - - - with
- - - - - crisis D57.459
Revise - - - - - - with ~~specified complication NEC D57.458~~
Add - - - - - - - dactylitis D57.454
Add - - - - - - - specified complication D57.458

- - - beta zero D57.42
- - - - with
- - - - - crisis D57.439
Revise - - - - - - with ~~specified complication NEC D57.438~~
Add - - - - - - - dactylitis D57.434
Add - - - - - - - specified complication D57.438

- sickle cell
- - Hb-SD D57.80
- - - with
Add - - - - - priapism D57.818

- - Hb-SE D57.80
- - - with
Add - - - - priapism D57.818

- - specified NEC D57.80
- - - with
Add - - - - priapism D57.818

- - spherocytosis D57.80
- - - with
Add - - - - priapism D57.818

- - thalassemia D57.40
- - - with
Add - - - - priapism D57.438

- - - beta plus D57.44
- - - - with
Add - - - - - priapism D57.458

- - - beta zero D57.42
- - - - with
Add - - - - - priapism D57.438

Dysfunction
- sexual (due to) R37
- - alcohol F10.981
- - - in
Revise - - - - abuse ~~F15.181~~ F10.181
Revise - - - - dependence ~~F15.281~~ F10.281

Epilepsy, epileptic, epilepsia (attack) (cerebral) (convulsion) (fit) (seizure) G40.909
Revise - Lafora progressive myoclonus (see also Epilepsy, myoclonic, progressive, Lafora) G40.C09
- progressive (familial) myoclonic - see Epilepsy, myoclonus, progressive
Revise - - Lafora (see also Epilepsy, myoclonic, progressive, Lafora) G40.C09

Frostbite (superficial) T33.90
Revise - leg ~~T33.9-~~ T33.99
Revise - - with tissue necrosis (see also Frostbite, by specific site on leg, if specified) ~~T34.9-~~ T34.99

Revise GERD (gastroesophageal reflux disease) (see also Disease, gastroesophageal reflux) K21.9

Hemoglobinopathy (mixed) D58.2
- sickle-cell D57.1
- - with thalassemia D57.40
- - - with
- - - - acute chest syndrome D57.411
Revise - - - - - with ~~specified complication NEC D57.418~~
Add - - - - - - dactylitis D57.414
Add - - - - - - specified complication D57.418

Hypertrophy, hypertrophic
- bone M89.30
Revise - - pubic ramus [pubis] M89.38

Injury -see also specified injury type T14.90
- intracranial (traumatic) (see also, if applicable, Compression, brain, traumatic) S06.9-
Revise - - cerebellar ~~hemorrhage, traumatic~~ – see also Injury, intracranial, focal brain injury
Add - - - hemorrhage, traumatic – see Injury, intracranial, intracerebral hemorrhage,

traumatic

Add - thalamic, thalamus – see also Injury, intracranial, focal brain injury
Add - - hemorrhage, traumatic – see Injury, intracranial, intracerebral hemorrhage, traumatic

Osteitis - see also Osteomyelitis
- deformans M88.9
- - in (due to)
- - - neoplastic disease (see also Neoplasm, by type and site) D49.9 [M90.60]
Revise - - - - pubic ramus [pubis] D49.2 [M90.68]

Osteoarthropathy (hypertrophic) M19.90
- specified type NEC M89.40
Revise - - pubic ramus [pubis] M89.48

Osteolysis M89.50
Revise - pubic ramus [pubis] M89.58

Osteomalacia M83.9
- vitamin-D-resistant in adults E83.31 [M90.8-]
Revise - - pubic ramus [pubis] E83.31 [M90.88]

Osteomyelitis (general) (infective) (localized) (neonatal) (purulent) (septic) (staphylococcal) (streptococcal) (suppurative) (with periostitis) M86.9
- chronic (or old) M86.60
- - with draining sinus M86.40
Revise - - - pubic ramus [pubis] M86.48
- - hematogenous NEC M86.50
- - - multifocal M86.30
Revise - - - - pubic ramus [pubis] M86.38
Revise - - - pubic ramus [pubis] M86.58

Osteonecrosis M87.9
- idiopathic aseptic M87.00
Revise - - pubic ramus [pubis] M87.050
- secondary NEC M87.30
- - due to
- - - drugs M87.10
Revise - - - - pubic ramus [pubis] M87.150
- - - hemoglobinopathy NEC D58.2 [M90.50]
Revise - - - - pubic ramus [pubis] D58.2 [M90.58]
- - - trauma (previous) M87.20
Revise - - - - pubic ramus [pubis] M87.250
- - in
- - - caisson disease T70.3 [M90.50]

Revise - - - - pubic ramus [pubis] T70.3 [M90.58]
Revise - - pubic ramus [pubis] M87.350
- specified type NEC M87.80
Revise - - pubic ramus [pubis] M87.850

Osteopathy - see also Osteomyelitis, Osteonecrosis, Osteoporosis
- after poliomyelitis M89.60
Revise - - pubic ramus [pubis] M89.68

Osteoporosis (female) (male) M81.0
- age-related M81.0
- - with current pathologic fracture M80.00
Revise - - - pubic ~~pubis~~ ramus [pubis] M80.0A
- disuse M81.8
- - with current pathological fracture M80.80
Revise - - - pubic ~~pubis~~ ramus [pubis] M80.0A
- postmenopausal M81.0
- - with pathological fracture M80.00
Revise - - - pubic ~~pubis~~ ramus [pubis] M80.0A
- specified type NEC M81.8
- - with pathological fracture M80.80
Revise - - - pubic ~~pubis~~ ramus [pubis] M80.0A

Paget's disease
- bone M88.9
Revise - - pubic ramus [pubis] M88.88

Priapism N48.30
- due to
Add - - sickle cell disease – see Disease, sickle cell by type, with priapism

Schizophrenia, schizophrenic F20.9
- undifferentiated (type) F20.3
Revise - - chronic ~~F20.5~~ F20.9

Short, shortening, shortness
Revise - bowel syndrome ~~K91.2~~ see Syndrome, short bowel

Syndrome -see also Disease
Revise - cardiorenal ~~-see Hypertension, cardiorenal~~ Note: see Failure, heart; also see Failure, renal
Revise - cardiovascular renal ~~-see Hypertension, cardiorenal~~ Note: see Failure, heart; also see Failure, renal
- myeloproliferative (chronic) D47.1
Add - - pancytopenia (acquired), D47.1 [D61.818]

	Tabacism, tabacosis, tabagism - see also Poisoning, tobacco
Revise	- meaning dependence (without remission) – see Dependence, drug, nicotine ~~F17.200~~
Delete	~~- - with~~
Delete	~~- - - disorder F17.299~~
Delete	~~- - - - in remission F17.211~~
Delete	~~- - - - specified disorder NEC F17.298~~
Revise	Tower skull ~~Q75.009~~ Q75.058
Revise	Xanthoma(s), xanthomatosis (primary) (familial) (hereditary) ~~E75.5~~ E78.2

ICD-10-CM External Cause of Injuries Index

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	Accident (to) X58
Revise	- ~~bare foot~~ barefoot water ~~skiier~~ skier V94.4
	Activity
Add	- pickleball Y93.73
Add	- protesting (political, social) Y93.89
Revise	Environmental pollution related condition- see category ~~Z57-~~Z77
Revise	Hanged herself or himself ~~-- see Hanging, self-inflicted~~ T71.162