CENTERS FOR DISEASE™
CONTROL AND PREVENTION

ICD-10 Coordination and Maintenance Committee Meeting
September 13-14, 2022
Diagnosis Agenda

Zoom Webinar and Dial-In Information

• This meeting will be conducted via Zoom Webinar. The URL 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 13, 2022: 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 14, 2022: 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 join the Zoom Webinar via the web. 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:
Click the following URL:

https://cms.zoomgov.com/j/1618774054?pwd=RzlGenRwL09TbHZKVEcrNnJDRi90QT09
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Option 2: Dial-in access is available for listen-only participants. Listen-only participants are participants who wish to only listen to the meeting and do not wish to comment or ask questions during the Q&A portions of the meeting.

1. From your phone, dial U.S.*: 669-254-5252 or 646-828-7666 or 833-568-8864 (Toll Free)
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   SIP: 1618774054@sip.zoomgov.com
   Passcode: 703819

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Those participating in the Zoom Webinar may ask questions during the Q&A portions of the meeting using the "Raise Your 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. All comments and questions submitted using the "Q&A" feature, along with CDC's responses to them, will be posted as soon as possible after the meeting on the CDC web page located at: ICD - ICD-10-CM - Coordination and Maintenance Committee (cdc.gov). Remaining questions may be submitted via the NCHS mailbox at nchsicd10CM@cdc.gov .

*Please note that registration is not required to attend the Zoom Webinar.

Welcome and announcements

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

Diagnosis Topics:

Contents

ICD-10 TIMELINE

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

September 13-14, 2022	The September 2022 ICD-10 Coordination and Maintenance Committee Meeting.
September 2022	Recordings and slide presentations of the September 13-14, 2022 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/icd10cm_maintenance.htm **Procedure code portion of the recording and related materials–** https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html
October 1, 2022	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/Comprehensive-Listing-of-ICD-10-CM-Files.htm **Procedure addendum –** https://www.cms.gov/medicare/coding/icd10
October 14, 2022	**Deadline for receipt of public comments on proposed new codes discussed at the September 13-14, 2022 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on April 1, 2023.**
November 2022	Any new ICD-10 codes required to capture new diseases or technology that will be implemented on the following April 1 will be announced. Information on any new codes to be implemented April 1, 2023, will be posted on the following websites:

	https://www.cdc.gov/nchs/icd/Comprehensive-Listing-of-ICD-10-CM-Files.htm
November 14, 2022	**Deadline for receipt of public comments on proposed new codes and revisions discussed at the September 13-14, 2022 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2023.**
December 2, 2022	**Deadline for requestors: Those members of the public requesting that topics be discussed at the March 7-8, 2023 ICD-10 Coordination and Maintenance Committee Meeting must have their requests submitted to CMS for procedures and to NCHS for diagnoses by this date.** Procedure code requests should be directed to CMS at https://mearis.cms.gov. Diagnosis code requests should be directed to NCHS at nchsicd10cm@cdc.gov. Requestors should indicate if they are submitting their code request for consideration for an October 1, 2023, implementation date or an April 1, 2024, implementation date.
January 2023	The ICD-10 Coordination and Maintenance Committee will make efforts to accommodate the requested implementation date for each request submitted, however, the Committee will determine which requests will be presented for consideration for an October 1, 2023 implementation date or an April 1, 2024 implementation date. Federal Register notice for the March 7-8, 2023 ICD-10 Coordination and Maintenance Committee Meeting will be published. This will include the tentative agenda.
February 2023	Tentative agenda for the Procedure portion of the March 7, 2023 ICD-10 Coordination and Maintenance Committee Meeting posted on CMS webpage as follows: https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html

	Tentative agenda for the Diagnosis portion of the March 8, 2023, ICD-10 Coordination and Maintenance Committee Meeting posted on NCHS homepage as follows: https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm
February 1, 2023	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, 2023	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/Comprehensive-Listing-of-ICD-10-CM-Files.htm https://www.cms.gov/medicare/coding/icd10
February 1, 2023	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/Comprehensive-Listing-of-ICD-10-CM-Files.htm https://www.cms.gov/medicare/coding/icd10
March 7-8, 2023	ICD-10 Coordination and Maintenance Committee Meeting.
March 2023	Recordings and slide presentations of the March 7-8, 2023 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/icd10cm_maintenance.htm **Procedure code portion of the recording and related materials–** https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html

April 1, 2023	Any new ICD-10 codes will be implemented on April 1, 2023.
April 7, 2023	**Deadline for receipt of public comments on proposed new codes and revisions discussed at the March 7-8, 2023 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2023**.
April 2023	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 2024 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 5, 2023	**Deadline for receipt of public comments on proposed new codes and revisions discussed at the March 7-8, 2023 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on April 1, 2024.** **Deadline for receipt of public comments on proposed new diagnosis codes and revisions discussed at the March 7-8, 2023 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2024**.
May/June 2023	Final addendum posted on web pages as follows: **Diagnosis addendum -** https://www.cdc.gov/nchs/icd/Comprehensive-Listing-of-ICD-10-CM-Files.htm **Procedure addendum -** https://www.cms.gov/medicare/coding/icd10

June 9, 2023	**Deadline for requestors: Those members of the public requesting that topics be discussed at the September 12-13, 2023 ICD-10 Coordination and Maintenance Committee Meeting, must have their requests submitted to CMS for procedures and NCHS for diagnoses.** Requestors should indicate if they are submitting their code request for consideration for an April 1, 2024, implementation date or an October 1, 2024, implementation date.
July 2023	Federal Register notice for the September 12-13, 2023, ICD-10 Coordination and Maintenance Committee Meeting will be published. This will include the tentative agenda.
August 1, 2023	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, 2023. This rule can be accessed at: https://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/AcuteInpatientPPS/index.html
August 2023	Tentative agenda for the Procedure portion of the September 12, 2023, 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 13, 2023, ICD-10 Coordination and Maintenance Committee Meeting will be posted on the NCHS webpage at – https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm
September 12-13, 2023	The September 2023 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 2023	Recordings and slide presentations of the September 12-13, 2023 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/icd10cm_maintenance.htm **Procedure code portion of the recording and related materials–** https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html
October 1, 2023	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/Comprehensive-Listing-of-ICD-10-CM-Files.htm **Procedure addendum –** https://www.cms.gov/medicare/coding/icd10
October 13, 2023	**Deadline for receipt of public comments on proposed new codes discussed at the September 12-13, 2023 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on April 1, 2024.**
November 2023	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, 2024 will be posted on the following websites: https://www.cdc.gov/nchs/icd/Comprehensive-Listing-of-ICD-10-CM-Files.htm https://www.cms.gov/medicare/coding/icd10
November 15, 2023	**Deadline for receipt of public comments on proposed new codes and revisions discussed at the September 12-13, 2023 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2024.**

Contact Information
Mailing address:

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

Donna Pickett	(301) 458-4434
David Berglund	(301) 458-4095
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Continuing Education Credits

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

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

If you plan to attend or participate via telephone the ICD-10 Coordination and Maintenance (C&M) Committee Meeting, you should be aware that CMS /NCHS do not provide certificates of attendance for these calls. Instead, the AAPC will accept your printed topic packet as proof of participation. Please retain your topic packet copy as the AAPC may request them for any conference call you entered 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 arthritis-related immunological findings without current or prior diagnosis of clinically-apparent inflammatory arthritis

Rheumatoid arthritis (RA) is a well-known autoimmune condition that is characterized by the presence of inflammatory arthritis (IA). 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 antibodies to cyclic citrullinated proteins (anti-CCP).

There are established diagnostic classification criteria for RA that incorporate several features including the presence and distribution of joint tenderness and swelling, autoantibodies and inflammatory markers, duration of symptoms and in some criteria the presence of imaging evidence of joint damage. However, while these criteria are commonly used in clinical research of RA, in routine clinical practice RA is commonly diagnosed and treated even if the classification criteria are not met. Indeed, in most current clinical practices, ICD-10-CM for RA are applied based on a clinical diagnosis and not necessarily fulfillment of established classification criteria.

The current paradigm for the diagnosis and treatment of RA is for a clinician to identify joint findings that are determined to be clinically-apparent IA, diagnosis 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., M60, M50.) are applied.

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. Furthermore, individuals who have abnormal RA-related immunologic tests without IA are identified in growing numbers in clinical care. 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.

While there are ICD-10-CM codes that can be used to designate clinical RA, RF and anti-CCP positivity, there is not currently a clear way in the existing ICD-10-CM to code individuals who may exhibit RA-related biomarkers but not have clinically-apparent IA. As such, the introduction of a new code to accurately designate an individual who has abnormal RA-related immunologic test 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 is supported by the American College of Rheumatology (ACR).

References

1. Smolen JS, Aletaha D, Barton A, Burmester GR, Emery P, Firestein GS, Kavanaugh A, McInnes IB, Solomon DH, Strand V, Yamamoto K. Rheumatoid arthritis. Nat Rev Dis Primers. 2018;4:18001. Epub 2018/02/09. doi: 10.1038/nrdp.2018.1. PubMed PMID: 29417936.

2. Whiting PF, Smidt N, Sterne JA, Harbord R, Burton A, Burke M, Beynon R, Ben-Shlomo Y, Axford J, Dieppe P. Systematic review: accuracy of anti-citrullinated Peptide antibodies for diagnosing rheumatoid arthritis. Ann Intern Med. 2010;152(7):456-64; W155-66. Epub 2010/04/07. doi: 10.7326/0003-4819-152-7-201004060-00010. PubMed PMID: 20368651.

3. Arnett FC, Edworthy SM, Bloch DA, McShane DJ, Fries JF, Cooper NS, Healey LA, Kaplan SR, Liang MH, Luthra HS, et al. The American Rheumatism Association 1987 revised criteria for the classification of rheumatoid arthritis. Arthritis Rheum. 1988;31(3):315-24. Epub 1988/03/01. doi: 10.1002/art.1780310302. PubMed PMID: 3358796.

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5. Deane KD, Holers VM. Rheumatoid arthritis pathogenesis, prediction, and prevention: an emerging paradigm shift. Arth Rheum. 2021;73:181-93.

6. Gizinski AM, Mascolo M, Loucks JL, Kervitsky A, Meehan RT, Brown KK, Holers VM, Deane KD. Rheumatoid arthritis (RA)-specific autoantibodies in patients with interstitial lung disease and absence of clinically apparent articular RA. Clin Rheumatol. 2009;28(5):611-3. Epub 2009/03/03. doi: 10.1007/s10067-009-1128-9. PubMed PMID: 19252818; PMCID: PMC4084723.

7. Fischer A, Solomon JJ, du Bois RM, Deane KD, Olson AL, Fernandez-Perez ER, Huie TJ, Stevens AD, Gill MB, Rabinovitch AM, Lynch DA, Burns DA, Pineiro IS, Groshong SD, Duarte Achcar RD, Brown KK, Martin RJ, Swigris JJ. Lung disease with anti-CCP antibodies but not rheumatoid arthritis or connective tissue disease. Respir Med. 2012;106(7):1040-7. Epub 2012/04/17. doi: 10.1016/j.rmed.2012.03.006. PubMed PMID: 22503074; PMCID: PMC3753791.

TABULAR MODIFICATIONS

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 arthritis-related immunological findings without current or prior diagnosis of clinically-apparent inflammatory arthritis

New code R76.89 Other specified abnormal immunological findings in serum
Add Raised level of immunoglobulins NOS

Acinetobacter Baumannii Infections

Acinetobacter are commonly found in the environment, like in soil and water. Acinetobacter baumannii accounts for most Acinetobacter infections in humans. It is a gram-negative bacteria which can cause infections in the blood, urinary tract, lungs, or in wounds in different parts of the body.[1,2] It can also "colonize" or live in a patient without causing infections or symptoms, especially in respiratory secretions or open wounds.[1]

Antibiotic resistance is common in Acinetobacter baumannii, and carbapenem resistance is a challenging threat to hospitalized patients. In 2017, carbapenem-resistant Acinetobacter caused an estimated 8,500 infections in hospitalized patients and 700 estimated deaths in the United States.[1,3]

This proposal is based on internal CDC review.

References
1. CDC. Acinetobacter in Healthcare Settings. Centers for Disease Control and Prevention, National Center for Emerging and Zoonotic Infectious Diseases (NCEZID), Division of Healthcare Quality Promotion (DHQP). https://www.cdc.gov/hai/organisms/acinetobacter.html

2. CDC. Multi-site Gram-negative Surveillance Initiative. Centers for Disease Control and Prevention, National Center for Emerging and Zoonotic Infectious Diseases (NCEZID), Division of Healthcare Quality Promotion (DHQP). https://www.cdc.gov/hai/eip/mugsi.html

3. CDC. Antibiotic Resistance Threats in the United States, 2019. Atlanta, GA: U.S. Department of Health and Human Services, CDC; 2019. http://www.cdc.gov/DrugResistance/Biggest-Threats.html

TABULAR MODIFICATIONS

A41 Other sepsis

A41.5 Sepsis due to other Gram-negative organisms

New code A41.54 Sepsis due to Acinetobacter baumannii
Add Use additional code, if applicable, for resistance to carbapenem (Z16.13)

B96 Other bacterial agents as the cause of diseases classified elsewhere

B96.8 Other specified bacterial agents as the cause of diseases classified elsewhere

New code B96.83 Acinetobacter baumannii as the cause of diseases classified elsewhere
Add Use additional code, if applicable, for resistance to carbapenem (Z16.13)

J15 Bacterial pneumonia, not elsewhere classified

J15.6 Pneumonia due to other Gram-negative bacteria

Delete			~~Pneumonia due to other aerobic Gram-negative bacteria~~	
Delete			~~Pneumonia due to Serratia marcescens~~	
New code		J15.61	Pneumonia due to Acinetobacter baumannii	
Add			Use additional code, if applicable, for resistance to carbapenem (Z16.13)	
New code		J15.69	Pneumonia due to other Gram-negative bacteria	
Add			Pneumonia due to other aerobic Gram-negative bacteria	
Add			Pneumonia due to Serratia marcescens	
	Z16	Resistance to antimicrobial drugs		
		Z16.1	Resistance to beta lactam antibiotics	
New code		Z16.13	Resistance to carbapenem	
	Z22	Carrier of infectious disease		
		Z22.3	Carrier of other specified bacterial diseases	
New sub-subcategory		Z22.34	Carrier of Acinetobacter baumannii	
New code			Z22.340	Carrier of carbapenem-resistant Acinetobacter baumannii
New code			Z22.341	Carrier of carbapenem-sensitive Acinetobacter baumannii
New code			Z22.349	Carrier of Acinetobacter baumannii, unspecified
New sub-subcategory		Z22.35	Carrier of Enterobacterales	
Add			Carrier of E. coli	
Add			Carrier of K. pneumoniae	
New code			Z22.350	Carrier of carbapenem-resistant Enterobacterales
New code			Z22.358	Carrier of other Enterobacterales
Add				Carrier of carbapenem-sensitive Enterobacterales
Add				Carrier of extended-spectrum beta-lactamase producing Enterobacterales
Add				Carrier of ESBL-producing Enterobacterales
New code			Z22.359	Carrier of Enterobacterales, unspecified

Acute HIV Infection Syndrome and HIV Pre-Exposure Prophylaxis (PrEP)

HIV disease has been a significant public health issue for many years, and despite improvements, there is a need for continuing efforts to reduce the burden of HIV disease.[1] Acute HIV infection syndrome is the initial infection with HIV, which involves symptoms, but is appropriate to differentiate from AIDS, or later HIV disease.

Pre-exposure prophylaxis (PrEP) for HIV is recommended for people at risk of HIV to prevent them from getting HIV from sex or injection drug use.[2,3] There are needs to increase awareness of pre-exposure prophylaxis among some at risk groups, to enable wider use, and reduce the burden of HIV disease.[4]

A request for a specific code for pre-exposure prophylaxis (PrEP) for HIV was received from a private individual, referencing the needs for tracking this. Acute HIV infection syndrome is a code in the WHO ICD-10, and this proposal would add it to ICD-10-CM. This proposal is based on internal NCHS review of these issues, and CDC HIV expert review is supportive.

References

1. CDC. HIV-Related Death Rate in U.S. Fell by Half From 2010 to 2017. National Center for HIV/AIDS, Viral Hepatitis, STD, and TB Prevention; Centers for Disease Control and Prevention. https://www.cdc.gov/nchhstp/newsroom/2020/hiv-related-death-rate-press-release.html
2. HIV.gov. Pre-Exposure Prophylaxis. U.S. Department of Health & Human Services. https://www.hiv.gov/hiv-basics/hiv-prevention/using-hiv-medication-to-reduce-risk/pre-exposure-prophylaxis
3. CDC. PrEP (Pre-Exposure Prophylaxis). Division of HIV Prevention, National Center for HIV/AIDS, Viral Hepatitis, STD, and TB Prevention, Centers for Disease Control and Prevention. https://www.cdc.gov/hiv/basics/prep.html
4. Baugher AR, Trujillo L, Kanny D, et al. Racial, Ethnic, and Gender Disparities in Awareness of Preexposure Prophylaxis Among HIV-Negative Heterosexually Active Adults at Increased Risk for HIV Infection — 23 Urban Areas, United States, 2019. MMWR Morb Mortal Wkly Rep 2021;70:1635–1639. https://doi.org/10.15585/mmwr.mm7047a3.

TABULAR MODIFICATIONS

	B20	Human immunodeficiency virus [HIV] disease		
Add		Excludes1: acute HIV infection syndrome (B23.0)		
New Category	B23	Human immunodeficiency virus [HIV] disease resulting in other conditions		
New code		B23.0	Acute HIV infection syndrome	
	Z29	Encounter for other prophylactic measures		
		Z29.8	Encounter for other specified prophylactic measures	
New code			Z29.81	Encounter for HIV pre-exposure prophylaxis
Add				Code also, if applicable, risk factors for HIV, such as:
Add				contact with and (suspected) exposure to human immunodeficiency virus [HIV] (Z20.6)
Add				high risk sexual behavior (Z72.5-)
New code			Z29.89	Encounter for other specified prophylactic measures

Age-Related Osteoporosis with Current Pathological Fracture, Pelvis

Autologous bone graft is used in a variety of orthopedic and maxillofacial procedures. The iliac crest of the pelvis is the most common site of autologous bone graft harvesting. In patients with osteoporosis, pathological fracture of the iliac crest can occur, either during or after the bone graft harvesting. Published series and reviews have reported incidence rates of up to 1%. This complication may be associated with significant pain and delayed postoperative mobility, although it typically does not require surgical intervention.

Osteoporosis with pathological fracture of pelvis is currently coded as “osteoporosis with current pathological fracture, femur”. This is anatomically incorrect. Codes specific to pelvis are needed to accurately identify this condition. Clinically, treatment of an osteoporotic fracture of the pelvis would differ significantly from treatment of an osteoporotic fracture of the femur. Since ICD-10-CM codes are used for a variety of purposes, including clinical decision making, statistical analysis, patient safety measures, etc., it is important that codes accurately identify the clinical condition they describe.

The Agency for Healthcare Research and Quality (AHRQ) is requesting creation of new codes for “age-related osteoporosis with current pathological fracture of pelvis”, and “other osteoporosis with current pathological fracture of pelvis”. These would allow more accurate reporting of the location of osteoporotic fractures.

References

Dimitriou R, Mataliotakis GI, Angoules AG, Kanakaris NK, Giannoudis PV. Complications following autologous bone graft harvesting from the iliac crest and using the RIA: a systematic review. Injury. 2011 Sep;42 Suppl 2:S3-15. doi: 10.1016/j.injury.2011.06.015.

Migliorini F, Cuozzo F, Torsiello E, Spiezia F, Oliva F, Maffulli N. Autologous Bone Grafting in Trauma and Orthopaedic Surgery: An Evidence-Based Narrative Review. J Clin Med. 2021 Sep 24;10(19):4347. doi: 10.3390/jcm10194347.
Almaiman M, Al-Bargi HH, Manson P. Complication of anterior iliac bone graft harvesting in 372 adult patients from may 2006 to may 2011 and a literature review. Craniomaxillofac Trauma Reconstr. 2013 Dec;6(4):257-66. doi: 10.1055/s-0033-1357510.

TABULAR MODIFICATIONS

M80 Osteoporosis with current pathological fracture

M80.0 Age-related osteoporosis with current pathological fracture

New subcategory M80.0B Age-related osteoporosis with current pathological fracture, pelvis

New code M80.0B1 Age-related osteoporosis with current pathological fracture, right pelvis

New code M80.0B2 Age-related osteoporosis with current pathological fracture, left pelvis

New code M80.0B9 Age-related osteoporosis with current pathological fracture, unspecified pelvis

M80.8 Other osteoporosis with current pathological fracture

New subcategory M80.8B Other osteoporosis with current pathological fracture, pelvis

New code M80.8B1 Other osteoporosis with current pathological fracture, right pelvis

New code M80.8B2 Other osteoporosis with current pathological fracture, left pelvis

New code M80.8B9 Other osteoporosis with current pathological fracture, unspecified pelvis

Alagille Syndrome

Alagille syndrome is a rare genetic disorder that primarily affects the liver. However, this syndrome can affect multiple organ systems of the body including the cardiovascular system, skeletal system, eyes, and kidneys.

The specific symptoms and severity of Alagille syndrome can vary greatly from one person to another, even within the same family. Some individuals may have mild forms of the disorder while others may have more serious forms.

Common symptoms, which often develop during the first three months of life, include blockage of the flow of bile from the liver (cholestasis), yellowing of the skin and mucous membranes (jaundice), poor weight gain and growth, severe itching (pruritus) and pale, loose stools. Additional symptoms include heart murmurs, congenital heart defects, vertebral (back bone) differences, thickening of the ring that normally lines the cornea in the eye (posterior embryotoxon) and distinctive facial features.

Most people with Alagille syndrome have mutations in one copy of the JAG1 gene. A small percentage (less than 1 percent) of patients have mutations of the NOTCH2 gene. These mutations are inherited as autosomal dominant traits, however in about half of cases the mutation occurs as a new change in the individual and was not inherited from a parent. The current estimated incidence of ALGS is between 1:30,000 and 1:45,000 with no difference in gender (https://www.ncbi.nlm.nih.gov/books/NBK1273/).

This condition is currently classified (an inclusion term) under code, Q44.7 Other congenital malfunctions of the liver, which also includes accessory liver, congenital absence of liver congenital hepatomegaly and congenital malformation of liver NOS.

A specific code for Alagille syndrome will effectively enable meeting the needs of clinical practice, patient and provider education, and epidemiology research for a condition for which the medical and scientific information and public health implications have been rapidly evolving. This request will allow for clearer identification of patients with Alagille Syndrome.

This proposal was submitted from the Global Liver Institute (GLI).

TABULAR MODIFICATIONS

Q44 Congenital malformations of gallbladder, bile ducts and liver

Q44.7 Other congenital malformations of liver

Add — Code also, if applicable, associated malformations affecting other systems

Delete — ~~Accessory liver~~
Delete — ~~Alagille's syndrome~~
Delete — ~~Congenital absence of liver~~
Delete — ~~Congenital hepatomegaly~~
Delete — ~~Congenital malformation of liver NOS~~

New code — Q44.70 Other congenital malformation of liver, unspecified
Add — Congenital malformation of liver, NOS

New code — Q44.71 Alagille syndrome
Add — Alagille-Watson syndrome

New code — Q44.79 Other congenital malformations of liver
Add — Accessory liver
Add — Congenital absence of liver
Add — Congenital hepatomegaly

Anal Fistula

This topic was presented at the March 2022 Coordination and Maintenance meeting. Based on public comments, revisions to the proposal have been made for reconsideration. An anal fistula is an inflammatory tract or connection between the surface of the anal canal and, most frequently, the perianal skin or perineum, typically evolving from an anal abscess.[1] The disease has significant implications for a patient's quality of life, as symptoms range from pain and discharge to fecal incontinence.

Anal fistulas are typically classified using the Parks classification system, which considers the external sphincter as a central point of reference to describe five distinct types of fistulas: superficial, intersphincteric, transsphincteric, suprasphincteric, and extrasphincteric.[2] The classification system describes the anatomic location of the fistula and facilitates the identification of a treatment pathway. The system is also useful in describing the complexity of the condition and related treatment protocols.

While clinical definitions of complex anal fistula can vary, clinicians are aligned on a consistent definition of simple fistula. According to several clinical guidelines, an anal fistula is considered to be "simple" when the tract is intersphincteric or low intersphincteric (crossing <30% of the external anal sphincter).[3] In addition, simple fistulas have a single external and internal opening, are associated with no pain or fluctuation to suggest presence of perianal abscess and have no evidence of a rectovaginal fistula or anorectal stricture.[2]

The management of patients with anal fistulas varies depending on severity of disease and underlying comorbidities (such as Crohn's disease).[4] Treatment and management of simple fistulas are relatively straightforward compared with complex anal fistulas. Complex anal fistulas can be much more challenging to manage, resulting in high disease burden, diminished health-related quality of life, and increased healthcare resource use and costs.[5] Treatments vary by location and fistula type, and include fistulotomies, endoanal advancement flap or ligation of the intersphincteric fistula tract (LIFT), proctectomies, and fecal diversions.[6]

A common complication of anal fistula surgery is recurrence of fistulas after surgery, which represents a challenging problem as these fistulas are usually associated with higher risk of re-recurrence and fecal incontinence.[7]

Current ICD-10-CM coding, K60.3 Anal fistula, does not differentiate between simple versus complex fistulas, nor does it distinguish between persistent, and recurrent fistulas. This lack of specificity decreases the opportunity to use ICD-10-CM codes for accurate disease tracking.

Takeda Pharmaceuticals America, Incorporated is proposing the following tabular modifications to enable better tracking of complex fistulas, facilitating greater understanding of anal fistula epidemiology, and improving treatment paradigms. The American Gastroenterological Association (AGA) has reviewed and supports this proposal.

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TABULAR MODIFICATIONS

K50 Crohn's disease [regional enteritis]
Add — Use additional code to identify any associated anal fistula (K60.3-) or rectal fistula (K60.4-) or anorectal fistula (K60.5-)

K51 Ulcerative colitis
Add — Use additional code to identify any associated anal fistula (K60.3-) or rectal fistula (K60.4-) or anorectal fistula (K60.5-)

K60 Fissure and fistula of anal and rectal regions

New subcategory — K60.3 Anal fistula
Add — Code first, if applicable:
Crohn's disease (K50.-)
ulcerative colitis (K51.-)
Add — Excludes1: congenital fistula (Q43.6)
New sub-subcategory — K60.31 Anal fistula, simple
Add — Low intersphincteric anal fistula
Add — Superficial anal fistula

New code — K60.311 Anal fistula, simple, persistent
Add — Anal fistula, simple, chronic

New code — K60.312 Anal fistula, simple, recurrent
New code — K60.319 Anal fistula, simple, unspecified

New sub-subcategory		K60.32	Anal fistula, complex
Add			Extrasphincteric anal fistula
Add			Intersphincteric anal fistula
Add			Suprasphincteric anal fistula
Add			Transsphincteric anal fistula
Add			Code also, if applicable: perianal abscess (K61.0) rectovaginal fistula (N82.3) stenosis of anus and rectum (K62.4)
New code			K60.321 Anal fistula, complex, persistent
Add			Anal fistula, complex, chronic
New code			K60.322 Anal fistula, complex, recurrent
New code			K60.329 Anal fistula, complex, unspecified
New code		K60.39	Other anal fistula
Add			Anal fistula NOS
New subcategory	K60.4	Rectal fistula	
Add		Excludes1: congenital fistula (Q43.6)	
New sub-subcategory		K60.41	Rectal fistula, simple
Add			Low intersphincteric rectal fistula
Add			Superficial rectal fistula
New code			K60.411 Rectal fistula, simple, persistent
Add			Rectal fistula, simple, chronic
New code			K60.412 Rectal fistula, simple, recurrent
New code			K60.419 Rectal fistula, simple, unspecified
New sub-subcategory		K60.42	Rectal fistula, complex
Add			Extrasphincteric rectal fistula
Add			Intersphincteric rectal fistula
Add			Suprasphincteric rectal fistula
Add			Transsphincteric rectal fistula
New code			K60.421 Rectal fistula, complex, persistent
Add			Rectal fistula, complex, chronic

New code		K60.422	Rectal fistula, complex, recurrent
New code		K60.429	Rectal fistula, complex, unspecified
New code	K60.49	Other rectal fistula	
Add		Rectal fistula NOS	
New subcategory	K60.5	Anorectal fistula	
Add		Excludes1: congenital fistula (Q43.6)	
New sub-subcategory	K60.51	Anorectal fistula, simple	
Add		Low intersphincteric anorectal fistula	
Add		Superficial anorectal fistula	
New code		K60.511	Anorectal fistula, simple, persistent
Add			Anorectal fistula, simple, chronic
New code		K60.512	Anorectal fistula, simple, recurrent
New code		K60.519	Anorectal fistula, simple, unspecified
New sub-subcategory	K60.52	Anorectal fistula, complex	
Add		Extrasphincteric anorectal fistula	
Add		Intersphincteric anorectal fistula	
Add		Suprasphincteric anorectal fistula	
Add		Transsphincteric anorectal fistula	
New code		K60.521	Anorectal fistula, complex, persistent
Add			Anorectal fistula, complex, chronic
New code		K60.522	Anorectal fistula, complex, recurrent
New code		K60.529	Anorectal fistula, complex, unspecified
New code	K60.59	Other anorectal fistula	
Add		Anorectal fistula NOS	

INDEX MODIFICATIONS

	Fistula (cutaneous) L98.8
Add	- anorectal (infectional) K60.5-
Revise	- anus, anal ~~(recurrent)~~ (infectional) K60.3-
Add	- rectal (infectional) K60.4-

Anuria, Oliguria and Hepatorenal Syndrome in Complicating the Puerperium

The National Center for Health Statistics (NCHS) received a request to clarify coding related to the current code of O90.4, postpartum acute kidney failure. Currently, anuria, oliguria and hepatorenal syndrome complicating the puerperium are classified together under code O90.4.

The NCHS determined it was best to create separate codes for hepatorenal syndrome complicating the puerperium and create a new code for other postpartum acute kidney failure to include puerperal anuria and oliguria.

Anuria, complicating the puerperium, refers to the lack or no production of urine. Oliguria, complicating the puerperium, refers to low urine output. Hepatorenal syndrome (HRS) is a kidney impairment in decompensated liver disease characterized by decreased renal perfusion caused by severe splanchnic vasodilatation in the absence of shock, renal disease, or nephrotoxic effects of medications or other substances.

The American College of Obstetricians and Gynecologists (ACOG) has reviewed and supports the proposal.

TABULAR MODIFICATIONS

O90 Complications of the puerperium, not elsewhere classified

O90.4 Postpartum acute kidney failure

Delete ~~Hepatorenal syndrome following labor and delivery~~

Add Excludes1: anuria and oliguria (R34)

New code O90.41 Hepatorenal syndrome following labor and delivery

New code O90.49 Other postpartum acute kidney failure
Add Postpartum acute kidney failure
Add Puerperal anuria
Add Puerperal oliguria

R34 Anuria and oliguria
Excludes1: anuria and oliguria complicating abortion or ectopic or molar pregnancy (O00-O07, O08.4)
anuria and oliguria complicating pregnancy (O26.83-)
Revise anuria and oliguria complicating the puerperium (O90.4-)

INDEX MODIFICATIONS

	Anuria R34
Add	- puerperal O90.49
	Failure, failed
	- renal N19
	- - following
Revise	- - - labor and delivery (acute) O90.49
Revise	Hepatorenal syndrome following labor and delivery O90.41
	Oliguria R34
Add	- puerperal O90.49
	Puerperal, puerperium (complicated by, complications)
	- failure
Revise	- - renal, acute O90.49
Revise	- hepatorenal syndrome O90.41
	- necrosis, liver (acute) (subacute) (conditions in subcategory K72.0) O26.63
Revise	- - with renal failure O90.49
	- renal
Revise	- - failure O90.49
Revise	- uremia (due to renal failure) O90.49
	Syndrome
	- hepatorenal K76.7
Revise	- - following delivery O90.41
Revise	- - postpartum, puerperal O90.41

Autosomal Dominant Hypocalcemia

The National Center for Health Statistics received a proposal requesting new ICD-10-CM codes for autosomal dominant hypocalcemia.

Autosomal dominant hypocalcemia, commonly abbreviated as ADH, is a genetic disorder of calcium metabolism mediated by hypoparathyroidism associated with impaired secretion of parathyroid hormone.

There are two types of ADH. ADH type1 is caused by mutations in the gene *CASR*. This gene codes for a protein called calcium-sensing receptor (CaSR). CaSR plays a key role in controlling parathyroid hormone, which in turn controls calcium metabolism. In ADH, the genetic mutation causes the body to sense and interpret even very low levels of calcium as normal and then prevent any increase to the amount of calcium in the blood. ADH type 2, which is rarer, is caused by mutations in the *GNA11* gene, with similar effect.

Individuals with ADH may have no family history of the condition; in other cases the genetic mutation is familial. The disorder has an autosomal dominance pattern, meaning that one copy of the mutated gene is sufficient to cause the disorder. So, the child of a person with an autosomal dominant condition has a 50% chance of inheriting the condition.

Individuals with ADH may remain asymptomatic. Otherwise, the disorder causes a variety of symptoms which vary from mild, to severe, to debilitating. Symptoms and complications may impact multiple organ systems, particularly the musculoskeletal, nervous, and renal systems.

Acute symptoms commonly involve the muscles because calcium is essential to muscle contraction. For example, people with ADH often suffer from muscle spasms, especially of the hands and feet, muscle cramping and twitching, and paresthesia such as prickling and tingling. More severely, muscle symptoms include bronchospasm, which can lead to respiratory distress, as well as impaired cardiac contractility and arrhythmias.

In the nervous system, manifestations include seizures and abnormal movement, such as gait and coordination disorders. Calcium build-up and formation of calculi in the basal ganglia of the brain, the area which helps to control movement, is a known complication of ADH. Cognitive impairment, sometimes referred to as brain fog, can also arise.

People with ADH are at significantly increased risk of developing renal disorders, notably nephrolithiasis (kidney stones), nephrocalcinosis (permanent calcifications of the kidney tissue itself), and chronic kidney disease up to and including kidney failure. Hypercalciuria, abnormally high concentration of calcium in urine, is a frequent finding and contributes to formation of the kidney stones and calcifications and the impairment of kidney function. Abnormal blood levels of other minerals, such as hyperphosphatemia and hypomagnesemia, are common findings as well.

After identification of the hypocalcemia and related findings, the diagnosis of ADH is made by genetic testing. Median age at diagnosis is about 25 years old, although individuals with the most

severe symptoms tend to present earlier in life, including in infancy, while the disorder may be identified much later in others. ADH is seen equally in men and women. Prevalence of ADH in the US population is not precisely known but a recent analysis of genetic sequencing data coupled with laboratory data from electronic health records arrived at an estimated prevalence of 3.9 per 100,000.

Treatment of ADH focuses on addressing hypocalcemia and the associated symptoms. Individuals routinely take oral calcium supplements, though they may require IV calcium infusions on an urgent basis when acute symptoms are present. Prescription active vitamin D is also commonly used to improve calcium absorption, because hypoparathyroidism impairs the kidneys' ability to convert the precursor form of vitamin D into its activated form.

Unfortunately, chronic use of calcium and active vitamin D can paradoxically cause worsening hypercalciuria, leading to calcifications in the kidneys and basal ganglia. This requires an ongoing balancing act in the level of treatment to address the symptoms without causing the complications. Due to the narrow range of parameters, patients often remain symptomatic despite currently available treatments. With the associated and significant risks of renal complications, treatment may be avoided in completely asymptomatic individuals.

People with ADH require life-long monitoring. In stable individuals, serum calcium is usually measured twice a year. However, this can be much more frequent when dosages of calcium and active vitamin D are being adjusted, as happens when patients are experiencing symptoms. A 24-hour urine is collected once a year to monitor creatinine, calcium excretion, and other indicators of renal function. As indicated, renal and basal ganglia imaging is periodically performed for calcifications.

In addition to ADH, new codes are being proposed for other types of hypoparathyroidism due to impaired parathyroid hormone secretion. These include secondary hypoparathyroidism, for example associated with neoplasm or thalassemia, and autoimmune hypoparathyroidism in which individuals produce anti-parathyroid gland antibodies.

New ICD-10-CM codes will provide coding specificity to identify individuals with autosomal dominant hypocalcemia for treatment, tracking, and research.

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TABULAR MODIFICATIONS

E20 Hypoparathyroidism

E20.8 Other hypoparathyroidism

New subcategory

E20.81 Hypoparathyroidism due to impaired parathyroid hormone secretion

New code
E20.810 Autosomal dominant hypocalcemia
Add
Autosomal dominant hypocalcemia type 1 (ADH1)
Add
Autosomal dominant hypocalcemia type 2 (ADH2)

Add
Code also, if applicable, any associated conditions, such as:
Add
calculus of kidney (N20.0)
Add
chronic kidney disease (N18.-)
Add
respiratory distress (J80, R06.-)
Add
seizure disorder (G40.-, R56.9)

New code
E20.811 Secondary hypoparathyroidism in diseases classified elsewhere
Add
Code first underlying condition if known

New code
E20.812 Autoimmune hypoparathyroidism
Add
Code first, if applicable, underlying condition such as:
Add
autoimmune polyglandular failure (E31.0)
Add
Schmidt's syndrome (E31.0)

New code
E20.818 Other specified hypoparathyroidism due to impaired parathyroid hormone secretion
Add
Familial isolated hypoparathyroidism

New code
E20.819 Hypoparathyroidism due to impaired parathyroid hormone secretion, unspecified

New code
E20.89 Other specified hypoparathyroidism
Add
Familial hypoparathyroidism

E83 Disorders of mineral metabolism
Excludes1: dietary mineral deficiency (E58-E61)
parathyroid disorders (E20-E21)
vitamin D deficiency (E55.-)
E83.5 Disorders of calcium metabolism
Add Excludes1: autoimmune hypoparathyroidism (E20.812)
Add autosomal dominant hypocalcemia (E20.810)
chondrocalcinosis (M11.1-M11.2)
hungry bone syndrome (E83.31)
hyperparathyroidism (E21.0-E21.3)
Add secondary hypoparathyroidism in diseases classified elsewhere (E20.811)

Bronchiolitis Obliterans and Bronchiolitis Obliterans Syndrome

Bronchiolitis obliterans and bronchiolitis obliterans syndrome (BOS) together with chronic lung allograft dysfunction had a prior proposal for ICD-10-CM code expansion, September 2021. Based on comments from that time, a revised proposal is re-presented. Please see the prior proposal for further clinical details.

In brief, bronchiolitis obliterans or obliterative bronchiolitis may occur for a number of reasons, in a clinical syndrome characterized by airflow limitation not reversible with inhaled bronchodilators which may be associated with progressive dyspnea. BOS may occur following lung transplant, with fibrosis involving terminal and respiratory bronchioles; it may also follow stem cell transplant with chronic graft-versus-host-disease. Chronic lung allograft dysfunction may involve BOS, or restrictive allograft syndrome (RAS), or a mix of these, and potentially other clinical issues.

TABULAR MODIFICATIONS

Revise	Chronic lower respiratory diseases ~~(J40-J47)~~ (J40-J4A)			
	J42	Unspecified chronic bronchitis		
Add		Excludes1: bronchiolitis obliterans (J44.81)		
	J44	Other chronic obstructive pulmonary disease		
New subcategory		J44.8	Other specified chronic obstructive pulmonary disease	
New code			J44.81	Bronchiolitis obliterans and bronchiolitis obliterans syndrome
Add				Code first, if applicable:
Add				complication of bone marrow transplant (T86.09)
Add				complication of stem cell transplant (T86.5)
Add				heart-lung transplant rejection (T86.31)
Add				lung transplant rejection (T86.810)
Add				other complications of heart-lung transplant (T86.39)
Add				other complications of lung transplant (T86.818)
Add				Code also, if applicable, associated conditions, such as:
Add				chronic graft-versus-host disease (D89.811)
Add				chronic lung allograft dysfunction (J4A.-)
Add				chronic respiratory conditions due to chemicals, gases, fumes and vapors (J68.4)

New code J44.89 Other specified chronic obstructive pulmonary disease
Add Chronic asthmatic (obstructive) bronchitis
Add Chronic emphysematous bronchitis

J4A Chronic Lung Allograft Dysfunction
Add Code first, if applicable:
Add heart-lung transplant rejection (T86.31)
Add lung transplant rejection (T86.810)
Add other complications of heart-lung transplant (T86.39)
Add other complications of lung transplant (T86.818)
Add Code also, if applicable: bronchiolitis obliterans syndrome (J44.81)

New code J4A.0 Restrictive allograft syndrome
Add Note: for mixed chronic lung allograft dysfunction, code both restrictive allograft syndrome, J4A.0, and bronchiolitis obliterans syndrome, J44.81.

New code J4A.8 Other chronic lung allograft dysfunction

New code J4A.9 Chronic lung allograft dysfunction, unspecified

J68 Respiratory conditions due to inhalation of chemicals, gases, fumes and vapors

J68.4 Chronic respiratory conditions due to chemicals, gases, fumes and vapors
Delete ~~Emphysema (diffuse) (chronic) due to inhalation of chemicals, gases, fumes and vapors~~
Delete ~~Obliterative bronchiolitis (chronic) (subacute) due to inhalation of chemicals, gases, fumes and vapors~~
Delete ~~Pulmonary fibrosis (chronic) due to inhalation of chemicals, gases, fumes and vapors~~

Add Use additional code, if applicable, for chronic conditions, such as:
Add emphysema (J43.-)
Add obliterative bronchiolitis (J44.81)
Add pulmonary fibrosis (J84.10)

INDEX MODIFICATIONS

Asthma, asthmatic (bronchial) (catarrh) (spasmodic) J45.909
Revise - chronic obstructive J44.89
Revise - with chronic obstructive bronchitis J44.89
Revise - with chronic obstructive pulmonary disease J44.89

Bronchiolitis (acute) (infective) (subacute) J21.9
Revise - chronic (fibrosing) ~~(obliterative)~~ J44.89
Add - - obliterative J44.81
Revise - fibrosa obliterans ~~J44.9~~ J44.81
Revise - obliterans ~~J42~~ (see also Bronchiolitis, obliterative) J44.81
Add - - syndrome J44.81
Revise - obliterative (chronic) (subacute) (see also Bronchiolitis, obliterans) ~~J44.9~~ J44.81

Bronchitis (diffuse) (fibrinous) (hypostatic) (infective) (membranous) J40
- asthmatic J45.9
Revise - - chronic J44.89
- chronic J42
Revise - - asthmatic (obstructive) J44.89
Revise - - emphysematous J44.89
Revise - - obliterans ~~J44.9~~ see Bronchiolitis, obliterans
Revise - - obstructive J44.89
Revise - - with airways obstruction J44.89

Revise - emphysematous (obstructive) J44.89
Revise - obliterans (chronic) ~~J44.9~~ see Bronchiolitis, obliterans
Revise - obstructive (chronic) (diffuse) J44.89
Revise - with obstruction (airway) (lung) J44.89

Disease, diseased - see also Syndrome
- lung J98.4
- - obstructive (chronic) J44.9
Revise - - - with bronchitis J44.89

Dyspnea (nocturnal) (paroxysmal) R06.00
- asthmatic (bronchial) J45.909
- - with
- - - bronchitis J45.909
Revise - - - - chronic J44.89

Obstruction, obstructed, obstructive
- airway J98.8
Revise - - with bronchitis (chronic) J44.89

Pneumatocele (lung) J98.4

Revise - tension ~~J44.9~~ J98.8

Revise Vanishing lung J44.89

Chronic Migraine with Aura

The National Center for Health Statistics received a proposal from Wisconsin Physicians Service Corporation requesting a new ICD-10-CM code for chronic migraine with aura.

"Migraine is a common, multifactorial brain disorder with recurring disabling attacks of headache and associated features."[2] These features include migraine with and without auras.[2] The international Classification of Headache Disorders, third edition defines Chronic Migraine as a headache occurring on 15 or more days a month for more than three months, which, on at least eight days a month has the features of migraine headache. Chronic headache criteria includes both migraine with and without aura.[1]

A new ICD-10-CM code will provide coding specificity for individuals with chronic migraines with aura for treatment and research.

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TABULAR MODIFICATIONS

	G43	Migraine	
New subcategory	G43.E	Chronic migraine with aura	
Add		Excludes1: migraine with aura (G43.1-)	
New sub-subcategory	G43.E0	Chronic migraine with aura, not intractable	
Add		Chronic migraine with aura, without refractory migraine	
New code		G43.E01	Chronic migraine with aura, not intractable, with status migrainosus
New code		G43.E09	Chronic migraine with aura, not intractable, without status migrainosus
Add			Chronic migraine with aura NOS

New sub-subcategory	G43.E1	Chronic migraine with aura, intractable	
Add		Chronic migraine with aura, with refractory migraine	
New code		G43.E11	Chronic migraine with aura, intractable, with status migrainosus
New code		G43.E19	Chronic migraine with aura, intractable, without status migrainosus

Coronary Microvascular Dysfunction

Approximately 112 million people globally are affected by angina[1] and a significant proportion of these patients experience ischemia with non-obstructive coronary arteries (INOCA) and myocardial infarction with non-obstructive coronary arteries (MINOCA) due to pathologies in the microvasculature.[2] The microcirculatory system is largely responsible for the regulation and distribution of blood flow to the myocardium and is composed of an extensive network of narrow vessels downstream from the epicardial arteries.[2] Coronary microvascular dysfunction (CMD) is a condition that impacts the microvasculature by restricting microvascular flow and increasing microvascular resistance. Manifestations of CMD widely range in severity and presentation from angina to heart failure.[2,3]

Due to the growing body of evidence on CMD and its clinical impact on patients, the 2021 Guideline for the Evaluation and Diagnosis of Chest Pain indicated that patients with INOCA benefit from an assessment of functional significance to help guide management of the condition.[4] The guidelines also recommend testing practices that can aid in the diagnosis of microvascular flow abnormalities to support risk stratification.[4] Additionally, studies have indicated that the presence of CMD is not benign, as it is associated with significantly higher rates of major adverse cardiac events.[5] There is higher risk of CMD in women, those with hypertension, diabetes, and other insulin-resistant states.[4]

A proposal to create specific ICD-10-CM codes for CMD was received from Abbott Laboratories, Inc., to ensure afflicted patients receive appropriate diagnosis and care.

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TABULAR MODIFICATIONS

I20 Angina pectoris

I20.8 Other forms of angina pectoris

Delete ~~Angina equivalent~~

Delete ~~Angina of effort~~

Delete ~~Coronary slow flow syndrome~~
Delete ~~Stenocardia~~
Delete ~~Stable angina~~

New code I20.81 Angina pectoris with coronary microvascular dysfunction
Add Angina pectoris with coronary microvascular disease

New code I20.89 Other forms of angina pectoris
Add Angina equivalent
Add Angina of effort
Add Coronary slow flow syndrome
Add Stenocardia
Add Stable angina

I21 Acute myocardial infarction

New code I21.B Myocardial infarction with coronary microvascular dysfunction
Add Myocardial infarction with coronary microvascular disease
Add Myocardial infarction with nonobstructive coronary arteries [MINOCA] with microvascular disease

I24 Other acute ischemic heart diseases

I24.8 Other forms of acute ischemic heart disease

New code I24.81 Acute presentation of coronary microvascular dysfunction
Add Acute (presentation of) coronary microvascular disease

New code I20.89 Other forms of acute ischemic heart disease

I25 Chronic ischemic heart disease

I25.8 Other forms of chronic ischemic heart disease

New code I25.85 Chronic presentation of coronary microvascular dysfunction
Add Chronic (presentation of) coronary microvascular disease

Dense Breast(s) on Mammography

The National Center for Health Statistics received a request for a new ICD-10-CM codes to identify dense breast(s) on mammography. Breasts are made up of lobules, ducts, fatty and fibrous connective tissue. Lobules are the small glands that produce milk, while ducts are the tiny tubes that carry the milk from the lobules to the nipple. Together, the lobules and ducts are referred to as glandular tissue. Fibrous tissue and fat give breasts their size and shape and hold the other structures in place.

Fibrous and glandular tissue are harder to see through on a mammogram, so the breast tissue may be called 'dense'. Having dense breast tissue is common. Some women have more dense breast tissue. For most women, breasts become less dense with age. For others, there's little change.

The Breast Imaging Reporting and Data System, called BI-RADS, is used to group different types of breast density. This system, developed by the American College of Radiology, helps clinicians to interpret and report specific mammogram findings. BI-RADS classifies breast density into four categories, as follows:

(a) Almost entirely fatty breast tissue, found in about 10% of women
(b) Scattered areas of dense glandular tissue and fibrous connective tissue (scattered fibroglandular breast tissue) found in about 40% of women
(c) Heterogeneously dense breast tissue with many areas of glandular tissue and fibrous connective tissue, found in about 40% of women
(d) Extremely dense breast tissue, found in about 10% of women

It is proposed to create new codes to allow code assignment to capture both the screening mammogram and the finding of dense breasts on the same encounter.

References
https://www.cancer.org/cancer/breast-cancer/screening-tests-and-early-detection/mammograms/breast-density-and-your-mammogram-report.html
https://www.cdc.gov/cancer/breast/basic_info/dense-breasts.htm
https://www.cancer.gov/types/breast/breast-changes/dense-breasts

TABULAR MODIFICATIONS

R92 Abnormal and inconclusive findings on diagnostic imaging of breast

R92.2 Inconclusive mammogram

Delete ~~Dense breasts NOS~~

Inconclusive mammogram NEC

Delete ~~Inconclusive mammography due to dense breasts~~

Inconclusive mammography NEC

Add Code also, if applicable, density of breast (R92.3-)

New subcategory R92.3 Mammographic density found on imaging of breast

Add Code also, if applicable, inconclusive mammogram (R92.2)

New code R92.30 Dense breasts, unspecified

Add Dense breasts NOS

Add Low density

New subcategory R92.31 Mammographic fatty tissue density of breast

Add Breast Imaging Reporting and Data System (BI-RADS): A

New code R92.311 Mammographic fatty tissue density of right breast

New code R92.312 Mammographic fatty tissue density of left breast

New code R92.313 Mammographic fatty tissue density of bilateral breasts

New subcategory R92.32 Mammographic fibroglandular density of breast

Add Breast Imaging Reporting and Data System (BI-RADS): B

New code R92.321 Mammographic fibroglandular density of right breast

New code R92.322 Mammographic fibroglandular density of left breast

New code R92.323 Mammographic fibroglandular density of bilateral breasts

New subcategory R92.33 Mammographic heterogeneous density of breast

Add Breast Imaging Reporting and Data System (BI-RADS): C

New code R92.331 Mammographic heterogeneous density of right breast

New code	R92.332 Mammographic heterogeneous density of left breast
New code	R92.333 Mammographic heterogeneous density of bilateral breasts
New subcategory	R92.34 Mammographic extreme density of breast
Add	Breast Imaging Reporting and Data System (BI-RADS): D
New code	R92.341 Mammographic extreme density of right breast
New code	R92.342 Mammographic extreme density of left breast
New code	R92.343 Mammographic extreme density of bilateral breasts

INDEX MODIFICATIONS

	Breast - see also condition
Revise	- dense ~~R92.2~~ R92.3-
Revise	Dense breasts ~~R92.2~~ R92.3-
	Density
Add	- breast R92.3-
	Findings, abnormal, inconclusive, without diagnosis - see also Abnormal
	- mammogram NEC R92.8
Delete	- - inconclusive result (due to dense breasts) R92.2
	Inconclusive
Delete	- mammogram (due to dense breasts) R92.2

Eating Disorders

The Fifth Edition of the Diagnostic and Statistical Manual of Mental Disorders (DSM-5), published by the American Psychiatric Association (APA), is a clinical classification with relevant terminology designed to facilitate reliable and consistent diagnosing of mental and behavioral health conditions. The classification subcategorizes many of its clinical diagnoses by severity, course, and other descriptive feature specifiers.

Most of these specifiers and defining features are reflected in the ICD-10-CM today as unique codes or inclusion terms resulting from proposals such as this (e.g., Bipolar Disorders, Major Depressive Disorder, Substance Use Disorders, and, more recently, Neurocognitive Disorders).

Kaiser Permanente Medicine request the following tabular modifications to further the alignment of the two publications by updating the ICD-10-CM to recognize the diagnostic subcategories and other descriptive features of the DSM-5 for feeding and eating disorders (e.g., severity and remission).

The American Psychiatric Association has reviewed and supports this proposal.

TABULAR MODIFICATIONS

F50 Eating disorders
Excludes1: anorexia NOS (R63.0)
feeding problems of newborn (P92.-)
polyphagia (R63.2)

Excludes2: feeding difficulties (R63.3)
feeding disorder in infancy or childhood (F98.2-)

F50.0 Anorexia nervosa
Excludes1: loss of appetite (R63.0)
psychogenic loss of appetite (F50.89)
F50.00 Anorexia nervosa, unspecified

F50.01 Anorexia nervosa, restricting type
Add Anorexia nervosa, restricting type, extreme
Add Anorexia nervosa, restricting type, in full remission
Add Anorexia nervosa, restricting type, in partial remission
Add Anorexia nervosa, restricting type, mild

Add Anorexia nervosa, restricting type, moderate
Add Anorexia nervosa, restricting type, severe

F50.02 Anorexia nervosa, binge eating/purging type
Excludes1: bulimia nervosa (F50.2)
Add Anorexia nervosa, binge eating/purging type, extreme
Add Anorexia nervosa, binge eating/purging type, in full remission
Add Anorexia nervosa, binge eating/purging type, in partial remission
Add Anorexia nervosa, binge eating/purging type, mild
Add Anorexia nervosa, binge eating/purging type, moderate
Add Anorexia nervosa, binge eating/purging type, severe

F50.2 Bulimia nervosa
Bulimia NOS
Hyperorexia nervosa
Excludes1: anorexia nervosa, binge eating/purging type (F50.02)
Add Bulimia nervosa, restricting type, extreme
Add Bulimia nervosa, restricting type, in full remission
Add Bulimia nervosa, restricting type, in partial remission
Add Bulimia nervosa, restricting type, mild
Add Bulimia nervosa, restricting type, moderate
Add Bulimia nervosa, restricting type, severe

F50.8 Other eating disorders
Delete ~~Excludes2: pica of infancy and childhood (F98.3)~~
F50.81 Binge eating disorder
Add Binge eating disorder, extreme
Add Binge eating disorder in full remission
Add Binge eating disorder in partial remission
Add Binge eating disorder, mild
Add Binge eating disorder, moderate
Add Binge eating disorder, severe

F50.82 Avoidant/restrictive food intake disorder
Add Avoidant/restrictive food intake disorder, in remission

New code F50.83 Pica in adults
Add Pica in adults, in remission
Add Excludes1: pica of infancy and childhood (F98.3)

New F50.84 Rumination disorder in adults
Add Rumination in adults, in remission
Excludes1: rumination disorder in infancy and childhood (F98.21)

F50.89 Other specified eating disorder
Delete ~~Pica in adults~~
Psychogenic loss of appetite

F98.2 Other feeding disorders of infancy and childhood
Excludes2: anorexia nervosa and other eating disorders (F50.-)
feeding difficulties (R63.3)
feeding problems of newborn (P92.-)
pica in infancy and childhood (F98.3)

Revise F98.21 Rumination disorder ~~of~~ in infancy and childhood
Add Excludes1: rumination disorder in adults (F50.84)

F98.3 Pica of infancy and childhood
Add Excludes1: pica in adults (F50.83)
Add Pica in infancy and childhood, in remission

Extraocular Muscle Entrapment

This is a representation from the September 2021 and March 2022 ICD-10 Coordination and Maintenance Committee Meetings of the Extraocular Muscle Entrapment proposal with the recommended modification to add "unspecified" to the sub-subcategory H50.68, Extraocular muscle entrapment. The modifications are in **bold.**

Extraocular muscle entrapment in a nondisplaced orbital fracture, although a well-known entity in pediatric trauma, is atypical in adults. It can present with a triad of bradycardia, nausea, and in rare cases, syncope, and result in severe fibrosis of damaged and incarcerated muscle.[1]

An article published by AO Surgery Reference, "The inferior rectus muscle is the most common ocular muscle to become entrapped with an orbital floor fracture (trap-door phenomenon) and this may not be visible on conventional x-rays. Entrapment requires urgent freeing of the muscle to prevent necrosis of the incarcerated muscle. Clinical examination should give evidence on impaired ocular muscle function. Entrapment is often associated with severe ocular pain on attempted range of motion, as well as nausea and vomiting, especially in children".[2]

The National Center for Health Statistics received a proposal requesting for the creation of ICD-10-CM codes for extraocular muscle entrapment for coding specificity and research.

The American Academy of Ophthalmology (AAO) and American Association of Oral and Maxillofacial Surgeons (AAOMS) supports this proposal.

References

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2. Cornelius, C.-P., Gellrich, N., Hillerup, S., Kusumoto, K., & Schubert, W. (n.d.). Emergency treatment. Retrieved from Surgery Reference: https://surgeryreference.aofoundation.org/cmf/trauma/midface/further-reading/emergency-treatment#

TABULAR MODIFICATIONS

	H50	Other strabismus	
	H50.6	Mechanical strabismus	
New subcategory	H50.62	Inferior oblique muscle entrapment	
New code		H50.621	Inferior oblique muscle entrapment, right eye
New code		H50.622	Inferior oblique muscle entrapment, left eye
New code		H50.629	Inferior oblique muscle entrapment, unspecified eye
New sub-subcategory	H50.63	Inferior rectus muscle entrapment	
New code		H50.631	Inferior rectus muscle entrapment, right eye
New code		H50.632	Inferior rectus muscle entrapment, left eye
New code		H50.639	Inferior rectus muscle entrapment, unspecified eye

New sub-subcategory	H50.64	Lateral rectus muscle entrapment	
New code		H50.641	Lateral rectus muscle entrapment, right eye
New code		H50.642	Lateral rectus muscle entrapment, left eye
New code		H50.649	Lateral rectus muscle entrapment, unspecified eye
New sub-subcategory	H50.65	Medial rectus muscle entrapment	
New code		H50.651	Medial rectus muscle entrapment, right eye
New code		H50.652	Medial rectus muscle entrapment, left eye
New code		H50.659	Medial rectus muscle entrapment, unspecified eye
New sub-subcategory	H50.66	Superior oblique muscle entrapment	
New code		H50.661	Superior oblique muscle entrapment, right eye
New code		H50.662	Superior oblique muscle entrapment, left eye
New code		H50.669	Superior oblique muscle entrapment, unspecified eye
New sub-subcategory	H50.67	Superior rectus muscle entrapment	
New code		H50.671	Superior rectus muscle entrapment, right eye
New code		H50.672	Superior rectus muscle entrapment, left eye
New code		H50.679	Superior rectus muscle entrapment, unspecified eye
New sub-subcategory	H50.68	Extraocular muscle entrapment**, unspecified**	
New code		H50.681	Extraocular muscle entrapment**,** unspecified, right eye
New code		H50.682	Extraocular muscle entrapment**,** unspecified, left eye
New code		H50.689	Extraocular muscle entrapment**, unspecified, unspecified eye**

Familial Adenomatous Polyposis

Familial adenomatous polyposis (FAP) is an inherited disorder which predisposes to colon cancer, and also is associated with development of large numbers of colon polyps. There are three types of FAP, classic, attenuated, and autosomal recessive. Both the classic and attenuated types are cause by mutations in the APC gene, while the autosomal recessive type is caused by mutations in the MUTYH gene (also called MUTYH-associated polyposis).

The autosomal recessive FAP is a milder type of familial adenomatous polyposis. People with the autosomal recessive type have fewer polyps than those with the classic type. Fewer than 100 polyps typically develop, rather than hundreds or thousands.

The genetic mutations affect cell ability to maintain normal growth and function. Cell overgrowth resulting from mutation leads to the colon polyps seen in familial adenomatous polyposis. While it is expected that most people with mutations in the APC gene will develop colorectal cancer, the number of polyps and the time frame in malignancy develops depend on the particular mutation in the gene.

People with the classic type of familial adenomatous polyposis may develop multiple colon polyps in their teenage years. Without colectomy, polyps will be expected to become malignant. The average age for colon cancer to develop in classic familial adenomatous polyposis is 39 years. In attenuated FAP, polyp growth is delayed, and the average age of colorectal cancer development is 55 years.

In people with classic FAP, the number of polyps increases with age and hundreds to thousands of polyps can develop in the colon. This may also be development of desmoid tumors, noncancerous or uncertain behavior growths. These are fibrous tumors, that may be provoked by colectomy, and may tend to recur after being surgically removed. In both classic and attenuated FAP, benign and malignant tumors may sometimes be found in other places in the body, including the duodenum, stomach, bones, skin, and other tissues; the combination of colon polyps and growths outside the colon may be referred to as Gardner syndrome.

The autosomal recessive FAP is a milder type of familial adenomatous polyposis. People with the autosomal recessive type have fewer polyps than those with the classic type. Fewer than 100 polyps typically develop, rather than hundreds or thousands.

Reference
MedlinePlus. Familial adenomatous polyposis. National Library of Medicine, NIH.
https://medlineplus.gov/genetics/condition/familial-adenomatous-polyposis/

TABULAR MODIFICATIONS

Option #1

Z15 Genetic susceptibility to disease
Z15.8 Genetic susceptibility to other disease

New code	Z15.82	Familial adenomatous polyposis
Add		Code also associated clinical findings, such as:
Add		benign neoplasm of colon (D12.6)
Add		malignant neoplasm of colon (C18.-)

Option #2

D13 Benign neoplasm of other and ill-defined parts of digestive system
D13.9 Benign neoplasm of ill-defined sites within the digestive system

New code	D13.91	Familial adenomatous polyposis
Add		Code also associated clinical findings, such as:
Add		benign neoplasm of colon (D12.6)
Add		malignant neoplasm of colon (C18.-)
New code	D13.99	Benign neoplasm of ill-defined sites within the digestive system

Family History of Adenomatous Polyps

The National Center for Health Statistics received a proposal requesting new ICD-10-CM codes for family history of adenomatous polyps.

It is important to identify adenomatous/serrated polyps (D12.-), or adenoma, current or by history, as they carry a high risk of cancer and are considered premalignant. About 70% colorectal cancers originate from adenomatous adenomas and 25% - 30% arise from (sessile) serrated polyps. [1] This is relevant to colorectal cancer screening and surveillance. The addition of the new code will provide more specific data for research and screening protocols.

New ICD-10-CM codes will provide coding specificity for family history of adenomatous polyps for individuals with this familial risk factor.

References:
[1]ACG Clinical Guidelines: Colorectal Cancer Screening 2021: Official journal of the American College of Gastroenterology | ACG (lww.com)
[2]Task Force on Colorectal Cancer Recommends Colon Cancer Screening at 45 - Gastroenterologist San Antonio (gastroconsa.com)
[3]Understanding Your Pathology Report: Colon Polyps (Sessile or Traditional Serrated Adenomas) (cancer.org)

TABULAR MODIFICATIONS

Z83 Family history of other specific disorders

Z83.7 Family history of diseases of the digestive system

Z83.71 Family history of colonic polyps

Add — Conditions classifiable to K00-K93, or unspecified

Add — Excludes2: family history of adenomatous polyps (Z83.71-.)

New code — Z83.710 Family history of adenomatous and serrated polyps

Add — Conditions classifiable to D12.-

Add — Family history of tubular adenoma polyps

Add — Family history of tubulovillous adenoma polyps

Add — Family history of villous adenoma polyps

New code — Z83.711 Family history of hyperplastic colon polyps

New code — Z83.718 Other family colon polyps

Add — Family history of inflammatory colon polyps

New code — Z83.719 Family history of colon polyps, unspecified

Add — Family history of colon polyps NOS

Flank Anatomical Specificity

The "flank" (also known as "latus" or "lumbar region") of the thorax is a unique area of the body that lies between on the lateral aspect of the thorax between the rib cage and the iliac bone of the hip (below the rib cage and above the ilium). [Alberts, D; et al. (2012). Dorland's illustrated medical dictionary (32nd ed.). Philadelphia, PA: Saunders/Elsevier. p. 714]. Simply is it "the fleshy part of the side between the ribs and the hip" [https://www.merriam-webster.com/dictionary/flank].

This proposal was presented at the March 2021 Coordination and Maintenance meeting. In response to public comments, a revised proposal is being submitted for reconsideration. Changes are noted in **bold.**

There are times when a patient will seek medical care because of "flank pain" as opposed to abdominal or back pain. Pathology specific to flank pain can include kidney stones, pyelonephritis, gall bladder or liver disease, or muscle spasm to name a few. In addition, injuries to this area can lead to different muscle or intra-abdominal pathology.

The specific anatomical locale helps determine the clinician's evaluation process as well as resource utilization. The division of the frontal and lateral aspects of the abdomen allows for greater specificity in evaluating the patient. Currently, ICD-10-CM directs the term "flank" to the abdomen.

The American College of Emergency Physicians (ACEP) requests specific codes be added to the ICD-10-CM code set to better capture this specific anatomic region. This proposal is supported by the American Academy of Pediatrics.

TABULAR MODIFICATIONS

L02 Cutaneous abscess, furuncle and carbuncle
L02.2 Cutaneous abscess, furuncle and carbuncle of trunk
Excludes1: non-newborn omphalitis (L08.82)
omphalitis of newborn (P38.-)
Excludes2: abscess of breast (N61.1)
abscess of buttocks (L02.3)
abscess of female external genital organs (N76.4)
abscess of male external genital organs (N48.2, N49.-)
abscess of hip (L02.4)

L02.21 Cutaneous abscess of trunk

Revise **L02.212 Cutaneous abscess of back [any part, except buttock <u>and flank</u>]**

New code L02.217 Cutaneous abscess of flank

L02.22 Furuncle of trunk
Boil of trunk
Folliculitis of trunk
Revised **L02.222 Furuncle of back [any part, except buttock <u>and flank</u>]**
New code L02.227 Furuncle of flank

L02.23 Carbuncle of trunk
Revise **L02.232 Carbuncle of back [any part, except buttock <u>and flank</u>]**
New code L02.237 Carbuncle of flank

L03 Cellulitis and acute lymphangitis
L03.3 Cellulitis and acute lymphangitis of trunk
L03.31 Cellulitis of trunk
New code **L03.31A Cellulitis of flank**

L03.32 Acute lymphangitis of trunk
New code **L03.32A Acute lymphangitis of flank**

R10 Abdominal and pelvic pain
Excludes1: renal colic (N23)
Add Excludes2: **costovertebral (angle) tenderness (R39.85)**
dorsalgia (M54.-)
Add flatulence and related conditions (R14.-)

R10.1 Pain localized to upper abdomen
Add **Excludes2: pain localized to lateral abdomen (R10.A-)**
Add **pelvic and perineal pain (R10.2-)**

R10.2 Pelvic and perineal pain
Add **Excludes2: pain localized to other parts of lower abdomen (R10.3-)**
Add **pain localized to upper abdomen (R10.1-)**

New code **R10.20 Pelvic and perineal pain unspecified side**
New code **R10.21 Pelvic and perineal pain right side**
New code **R10.22 Pelvic and perineal pain left side**
New code **R10.23 Pelvic and perineal pain bilateral**

New code **R10.24 Suprapubic pain**

R10.3 Pain localized to other parts of lower abdomen
Add **Excludes2: pain localized to lateral abdomen R10.4-**
Add **pelvic and perineal pain (R10.2-)**

New subcategory **R10.A Pain localized to lateral flank abdomen**
Add **Latus pain**
Add **Excludes2: pain localized to other parts of lower abdomen (R10.3-)**
Add **pain localized to upper abdomen (R10.1-)**
New code **R10.A0 Flank pain, unspecified side**
New code **R10.A1 Flank pain, right side**
New code **R10.A2 Flank pain, left side**
New code **R10.A3 Flank pain, bilateral**

R10.8 Other abdominal pain

New subcategory **R10.8A Flank tenderness**
New code **Rl0.8A1 Right flank tenderness**
New code **Rl0.8A2 Left flank tenderness**
New code **R10.8A3 Suprapubic tenderness**

New code **R10.85 Abdominal pain of multiple sites**
Add **Excludes1: abdominal rigidity NOS (Rl9.3)**
Add **generalized abdominal pain associated with acute abdomen (R10.0)**
Add **generalized abdominal pain NOS (Rl0.84)**
Add **localized abdominal pain (Rl0.l-Rl0.4-)**

R39 Other and unspecified symptoms and signs involving the genitourinary system
R39.8 Other symptoms and signs involving the genitourinary system
New code **R39.85 Costovertebral (angle) tenderness**
Add **Excludes2: abdominal and pelvic pain (Rl0.-)**

S30 Superficial injury of abdomen, lower back, pelvis and external genitals
S30.1 Contusion of abdominal wall and latus region
Delete ~~Contusion of flank~~
Delete ~~Contusion of groin~~

New code S30.10 Contusion of abdominal wall and latus region, unspecified
New code S30.11 Contusion of abdominal wall and latus region
New code S30.12 Contusion of flank
New code S30.13 Contusion of groin

S30.8 Other superficial injuries of abdomen, lower back, pelvis, and external genitals

S30.81 Abrasion of abdomen, lower back, pelvis, and external genitals
New code S30.81A Abrasion of flank

S30.82 Blister (nonthermal) of abdomen, lower back, pelvis, and external genitals
New code S30.82A Blister (nonthermal) of flank

S30.84 External constriction of abdomen, lower back, pelvis and external genitals
New code S30.84A External constriction of flank

S30.85 Superficial foreign body of abdomen, lower back, pelvis, and external genitals
New code S30.85A Superficial foreign body of flank

S30.86 Insect bite (nonvenomous) of abdomen, lower back, pelvis, and external genitals
New code S30.86A Insect bite (nonvenomous) of flank

S30.87 Other superficial bite of abdomen, lower back, pelvis, and external genitals
New code S30.87A Other superficial bite of flank

S30.9 Unspecified superficial injury of abdomen, lower back, pelvis, and external genitals
New code S30.9A Unspecified superficial injury of flank

S31 Open wound of abdomen, lower back, pelvis and external genitals

S31.1 Open wound of abdominal wall without penetration into peritoneal cavity

S31.10 Unspecified open wound of abdominal wall without penetration into peritoneal cavity

New code S31.106 Unspecified open wound of abdominal wall, right flank without penetration into peritoneal cavity

New code S31.107 Unspecified open wound of abdominal wall, left flank without penetration into peritoneal cavity

New code S31.10A Unspecified open wound of abdominal wall, unspecified flank without penetration into peritoneal cavity

Add Open wound of abdominal wall of flank NOS without penetration into peritoneal cavity

S31.11 Laceration without foreign body of abdominal wall without penetration into peritoneal cavity

New code S31.116 Laceration without foreign body of abdominal wall, right flank without penetration into peritoneal cavity

New code S31.117 Laceration without foreign body of abdominal wall, left flank without penetration into peritoneal cavity

New code S31.11A Laceration without foreign body of abdominal wall, unspecified flank without penetration into peritoneal cavity

Add Laceration without foreign body of flank NOS without penetration into peritoneal cavity

S31.12 Laceration with foreign body of abdominal wall without penetration into peritoneal cavity

New code S31.126 Laceration with foreign body of abdominal wall, right flank without penetration into peritoneal cavity

New code S31.127 Laceration with foreign body of abdominal wall, left flank without penetration into peritoneal cavity

New code S31.12A Laceration with foreign body of abdominal wall unspecified flank without penetration into peritoneal cavity

Add Laceration with foreign body of abdominal wall of flank NOS without penetration into peritoneal cavity

S31.13 Puncture wound of abdominal wall without foreign body without penetration into peritoneal cavity

New code S31.136 Puncture wound of abdominal wall without foreign body, right flank without penetration into peritoneal cavity

New code S31.137 Puncture wound of abdominal wall without foreign body, left flank without penetration into peritoneal cavity

New code S31.13A Puncture wound of abdominal wall without foreign body, unspecified flank without penetration into peritoneal cavity

Add Puncture wound of abdominal wall of flank NOS without foreign body

S31.14 Puncture wound of abdominal wall with foreign body without penetration into peritoneal cavity

New code S31.146 Puncture wound of abdominal wall with foreign body, right flank without penetration into peritoneal cavity

New code S31.147 Puncture wound of abdominal wall with foreign body, left flank without penetration into peritoneal cavity

New code S31.14A Puncture wound of abdominal wall with foreign body, unspecified flank without penetration into peritoneal cavity

Add Puncture wound of abdominal wall with foreign body of flank NOS without penetration into peritoneal cavity

S31.15 Open bite of abdominal wall without penetration into peritoneal cavity

New code S31.156 Open bite of abdominal wall, right flank without penetration into peritoneal cavity

New code S31.157 Open bite of abdominal wall, left flank without penetration into peritoneal cavity

New code S31.15A Open bite of abdominal wall, unspecified flank without penetration into peritoneal cavity

Add Open bite of abdominal wall of flank NOS without penetration into peritoneal cavity

S31.6 Open wound of abdominal wall with penetration into peritoneal cavity

S31.60 Unspecified open wound of abdominal wall with penetration into peritoneal cavity

New code S31.606 Unspecified open wound of abdominal wall, right flank with penetration into peritoneal cavity

New code S31.607 Unspecified open wound of abdominal wall, left flank with penetration into peritoneal cavity

New code S31.60A Unspecified open wound of abdominal wall, unspecified flank with penetration into peritoneal cavity

Add Unspecified open wound of abdominal wall of flank NOS, with penetration into peritoneal cavity

S31.61 Laceration without foreign body of abdominal wall with penetration into peritoneal cavity

New code — S31.616 Laceration without foreign body of abdominal wall, right flank with penetration into peritoneal cavity

New code — S31.617 Laceration without foreign body of abdominal wall, left flank with penetration into peritoneal cavity

New code — S31.61A Laceration without foreign body of abdominal wall, unspecified flank with penetration into peritoneal cavity

Add — Laceration without foreign body of abdominal wall of flank NOS, with penetration into peritoneal cavity

S31.62 Laceration with foreign body of abdominal wall with penetration into peritoneal cavity

New code — S31.626 Laceration with foreign body of abdominal wall, right flank with penetration into peritoneal cavity

New code — S31.627 Laceration with foreign body of abdominal wall, left flank with penetration into peritoneal cavity

New code — S31.62A Laceration with foreign body of abdominal wall, unspecified flank with penetration into peritoneal cavity

Add — Laceration with foreign body of abdominal wall, flank NOS, with penetration into peritoneal cavity

S31.63 Puncture wound without foreign body of abdominal wall with penetration into peritoneal cavity

New code S31.636 Puncture wound of abdominal wall without foreign body, right flank with penetration into peritoneal cavity

New code S31.637 Puncture wound of abdominal wall without foreign body, left flank with penetration into peritoneal cavity

New code S31.63A Puncture wound of abdominal wall without foreign body, unspecified flank with penetration into peritoneal cavity

Add Puncture wound of abdominal wall without foreign body, flank NOS, with penetration into peritoneal cavity

S31.64 Puncture wound with foreign body of abdominal wall with penetration into peritoneal cavity

New code S31.646 Puncture wound of abdominal wall with foreign body, right flank with penetration into peritoneal cavity

New code S31.647 Puncture wound of abdominal wall with foreign body, left flank with penetration into peritoneal cavity

New code S31.64A Puncture wound of abdominal wall with foreign body, unspecified flank with penetration into peritoneal cavity

Add Puncture wound of abdominal wall with foreign body, flank NOS, with penetration into peritoneal cavity

S31.65 Open bite of abdominal wall with penetration into peritoneal cavity

New code S31.656 Open bite of abdominal wall, right flank with penetration into peritoneal cavity

New code	S31.657 Open bite of abdominal wall, left flank with penetration into peritoneal cavity
New code	S31.65A Open bite of abdominal wall, unspecified flank with penetration into peritoneal cavity
Add	Open bite of abdominal wall, flank NOS, with penetration into peritoneal cavity

Foreign Body Entering Into or Through a Natural Orifice

Foreign bodies can enter through natural body orifices. Some of which are benign and cause irritation (i.e. bead in the ear or nose). However other types of foreign bodies can have significant morbidity or mortality.

Button batteries can result in rapid caustic tissue injury with both acute and chronic complications. An extensive analysis of 8648 battery ingestion cases, 8161 were button batteries and 487 were cylindrical cells (AA, AAA). Of the button battery ingestions, 62.5% were in children under 6 years of age and 15.9% involved adults over 60 years old[1]. It has been reported that 12.6% of children under age 6 who ingested a 20 mm button battery suffered a major complication[2]. Ingestion of multiple magnets can cause serious conditions such as pinch the intestine walls quickly resulting in tissue necrosis and bowel perforation[3].

Currently in ICD-10-CM, there is not a specific way to identify these more serious types of foreign bodies entering into or through a natural orifice. Instead of adding each of these types of foreign bodies to the existing codes in T16-T19, the American Academy of Pediatrics is requesting that the WHO ICD-10 code category W44, Foreign body entering into or through eye or natural orifice be incorporated in the ICD-10-CM classification structure.

The American Academy of Pediatrics are requesting the following tabular modifications.

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TABULAR MODIFICATIONS

Effects of foreign body entering through natural orifice (T15-T19)

Excludes2: foreign body accidentally left in operation wound (T81.5-)

foreign body in penetrating wound - See open wound by body region residual foreign body in soft tissue (M79.5)

splinter, without open wound - See superficial injury by body region

Add Code also, if known, foreign body entering into or through a natural orifice (W44.-)

New category	W44 Foreign body entering into or through a natural orifice
Add	Excludes2: contact with sharp glass (W25) contact with other sharp objects (W26) foreign body or object entering through skin (W45)
Add	The appropriate 7th character is to be added to each code from category W44
Add	A - initial encounter
Add	D - subsequent encounter
Add	S - sequela
New subcategory	W44.A Battery entering into or through a natural orifice
New code	W44.A0 Battery unspecified, entering into or through a natural orifice
New code	W44.A1 Button battery entering into or through a natural orifice
New code	W44.A9 Other batteries entering into or through a natural orifice
New subcategory	W44.B Plastic entering into or through a natural orifice
New code	W44.B0 Plastic object unspecified, entering into or through a natural orifice
New code	W44.B1 Plastic bead entering into or through a natural orifice
New code	W44.B2 Plastic coin entering into or through a natural orifice
New code	W44.B3 Plastic toy and toy part entering into or through a natural orifice
New code	W44.B4 Plastic jewelry entering into or through a natural orifice
New code	W44.B5 Plastic bottle entering into or through a natural orifice

New code	W44.B9 Other plastic object entering into or through a natural orifice
New subcategory	W44.C Glass entering into or through a natural orifice
New code	W44.C0 Glass unspecified, entering into or through a natural orifice
New code	W44.C1 Sharp glass entering into or through a natural orifice
Add	Glass shard entering into or through a natural orifice
New code	W44.C2 Intact glass entering into or through a natural orifice
Add	Intact glass bottle entering into or through a natural orifice
New subcategory	W44.D Magnetic metal entering into or through a natural orifice
New code	W44.D0 Magnetic metal object unspecified, entering into or through a natural orifice
New code	W44.D1 Magnetic metal bead entering into or through a natural orifice
New code	W44.D2 Magnetic metal coin entering into or through a natural orifice
New code	W44.D3 Magnetic metal toy entering into or through a natural orifice
New code	W44.D4 Magnetic metal jewelry entering into or through a natural orifice
New code	W44.D9 Other magnetic metal objects entering into or through a natural orifice
New subcategory	W44.E Non-magnetic metal entering into or through a natural orifice
New code	W44.E0 Non-magnetic metal object unspecified, entering into or through a natural orifice

New code	W44.E1 Non-magnetic metal bead entering into or through a natural orifice
New code	W44.E2 Non-magnetic metal coin entering into or through a natural orifice
New code	W44.E3 Non-magnetic metal toy entering into or through a natural orifice
New code	W44.E4 Non-magnetic metal jewelry entering into or through a natural orifice
New code	W44.E9 Other non-magnetic metal objects entering into or through a natural orifice
Add	Bottle cap entering into or through a natural orifice
Add	Can lid entering into or through a natural orifice
Add	Pull tab entering into or through a natural orifice
New subcategory	W44.F Objects of natural or organic material entering into or through a natural orifice
New code	W44.F0 Objects of natural or organic material unspecified entering into or through a natural orifice
New code	W44.F1 Bezoar entering into or through a natural orifice
New code	W44.F2 Rubber band entering into or through a natural orifice
New code	W44.F3 Food entering into or through a natural orifice
New code	W44.F4 Insect entering into or through a natural orifice
New code	W44.F9 Other object of natural or organic material, entering into or through a natural orifice
New subcategory	W44.G Other non-organic objects entering into or through a natural orifice
New code	W44.G0 Other non-organic objects unspecified, entering into or through a natural orifice
New code	W44.G1 Audio device entering into or through a natural orifice

Add	Ear buds
Add	Hearing aids
New code	W44.G2 Combination metal and plastic toy and toy part entering into or through natural orifice
New code	W44.G3 Combination metal and plastic jewelry entering into or through a natural orifice
New code	W44.G9 Other non-organic objects entering into or through a natural orifice
New subcategory	W44.H Other sharp object entering into or through a natural orifice
New code	W44.H0 Other sharp object unspecified, entering into or through a natural orifice
New code	W44.H1 Needle entering into or through a natural orifice
Add	Dart entering into or through a natural orifice
Add	Hypodermic needle entering into or through a natural orifice
Add	Safety pin entering into or through a natural orifice
Add	Sewing needle entering into or through a natural orifice
New code	W44.H2 Knife, sword or dagger entering into or through a natural orifice
New code	W44.8 Other foreign body entering into or through a natural orifice
Add	Foreign body NOS entering into or through a natural orifice
New code	W44.9 Unspecified foreign body entering into or through a natural orifice

Frailty Risk Analysis Index

As the US population ages, the number of operations performed on elderly patients will increase. Frailty predicts postoperative mortality and morbidity more than age alone, thus presenting opportunities to identify the highest-risk surgical patients and improve their outcomes.

Studies have been done to determine if surgical outcomes of frail patient can be improved by facility-wide frailty screening and subsequent administrative review of perioperative surgical decision making. The Frailty Analysis Index can be performed during the pre-surgical care visit. Patients with high frailty scores (a Risk Analysis Index of 42 or more on a linear scale from zero to 81) must have a documented care plan that reflects their frailty-associated risks. Surgeons have two options: They can refer the selected patients for further evaluation by their primary care physician or they can use evidence-based protocols to prepare patients for surgery. Additionally, surgeons can document that the patient has been informed about his or her risk score and has engaged with the physician in a shared decision-making process. The result of that process is documented in a plan which might include deciding not to proceed with surgery.

Study findings revealed that this Frailty Screening Initiative (FSI) using Risk Analysis Index scores had a 3-fold survival benefit after controlling for age, frailty, and predicted mortality.

New specific ICD-10-CM codes will promote a more accurate assessment of the patient's severity of illness and needs for care. The Regulatory Committee of the Association of Clinical Documentation Integrity Specialists (ACDIS) propose the following revisions:

References
https://jamanetwork.com/journals/jamasurgery/fullarticle/2587479
https://achp.org/prevent-unnecessary-surgery/

TABULAR MODIFICATIONS

R54 Age-related physical debility
Delete ~~Frailty~~
Delete ~~Old age~~
Delete ~~Senescence~~
Delete ~~Senile asthenia~~
Delete ~~Senile debility~~
Excludes1: age-related cognitive decline (R41.81)
sarcopenia (M62.84)
senile psychosis (F03)
senility NOS (R41.81)

New code R54.8 Other age-related physical debility
Add Frailty
Add Old age
Add Senescence
Add Senile asthenia
Add Senile debility

New subcategory R54.A Frailty risk
Add Code first, underlying condition
New code R54.A0 Frailty risk analysis index score unspecified
New code R54.A1 Frailty risk analysis index score 0-41
New code R54.A2 Frailty risk analysis index score 42-81

Immunoglobulin A Nephropathy (IgAN)

The Renal Physicians Association (RPA) is requesting a new ICD-10-CM code for Immunoglobulin A Nephropathy (IgAN), the most common form of glomerulonephropathy.[i] The proposal was presented at the March and September 2021 ICD-10 Coordination and Maintenance meetings. Based on public comments from September 2021 C&M meeting, revised code titles are being presented for consideration and noted in bold.

IgAN affects approximately 2.5 per 100,000 persons worldwide. In the U.S., approximately 130 thousand patients have IgAN (incidence of 20-45 patients per million/year). In approximately 25% of patients with the condition, the nephropathy may progress to end-stage renal disease (ESRD) within 10-15 years.[ii] It is estimated that IgAN accounts for up to 10% of all patients in need of renal replacement therapy for ESRD in western countries.[iii] IgAN represents a particularly significant burden on the health care system because patients are usually relatively young when they reach ESRD. Also, the disease recurs in up to 60% of the patients who have received renal transplantation, though not all will develop clinically significant disease.[iv]

IgAN is characterized by deposition of immune complexes containing Immunoglobulin A in the glomerulus and proliferation of mesangial cells.[v,vi] The course of disease progression in IgAN can usually be predicted by clinical signs (hypertension, proteinuria, impaired renal function) and histologic lesions (extent of sclerosis and tubulointerstitial damage). Higher levels and longer duration of proteinuria are the strongest prognostic risk factors for disease progression.[vii,viii] There are a number of specific therapies that are used in the treatment of IgAN patients.[ix, x]

IgAN is diagnosed by renal biopsy.[xi] Immuno-fluorescence shows abundant deposition of IgA in the glomeruli, mainly in the mesangial region. The histological changes are variable but are dominated by mesangial proliferation and matrix expansion.[xii] It is commonly diagnosed between the ages of 16 and 35 years, usually due to the discovery of micro- or macrohematuria not attributable to other causes, with or without proteinuria.

Specific coding for IgAN is critical for accurately identifying cases, allowing for etiology-related research, patient segmentation, and therapeutic selection. A recommendation for a revision to the ICD-10-CM coding for IgAN is in line with the consensus of a group of experts in renal pathology, nephrology, and complement biology and therapeutics, as well as IgAN patients. Feedback from this group suggests that current coding for IgAN is neither sufficient nor adequate for identifying and differentiating IgAN patients because:

1. Current codes do not distinguish IgAN from other glomerular lesions that may have different treatment pathways, and do not enable a clear understanding of the epidemiology of the disease.

2. The distinctions between the different types of glomerular lesions in current codes may not be precise enough to indicate the severity or course of IgA nephropathy.

Currently, IgAN cases are commonly coded as N02.8, Recurrent and persistent hematuria with other morphologic changes, however IgAN is a well-defined condition. This expansion is recommended for further specificity. The RPA supports these revisions.

References

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TABULAR MODIFICATIONS

N02 Recurrent and Persistent Hematuria

N02.B Recurrent and persistent immunoglobulin A nephropathy

New code	**N02.B1 Recurrent and persistent immunoglobulin A nephropathy with glomerular lesion**
New code	**N02.B2 Recurrent and persistent immunoglobulin A nephropathy with focal and segmental glomerular lesion**
Add	**Recurrent and persistent immunoglobulin A nephropathy with focal and segmental hyalinosis or sclerosis**
New code	**N02.B3 Recurrent and persistent immunoglobulin A nephropathy with diffuse membranoproliferative glomerulonephritis**
New code	**N02.B4 Recurrent and persistent immunoglobulin A nephropathy with diffuse membranous glomerulonephritis**
New code	**N02.B5 Recurrent and persistent immunoglobulin A nephropathy with diffuse mesangial proliferative glomerulonephritis**
New code	**N02.B6 Recurrent and persistent immunoglobulin A nephropathy with diffuse mesangiocapillary glomerulonephritis**
New code	**N02.B9 Other recurrent and persistent immunoglobulin A nephropathy**

INDEX MODIFICATIONS

	Nephropathy
Revise	- IgA ~~N02.8~~ N02.B-
Revise	- - with glomerular lesion ~~N02.9~~ N02.B1
Revise	- - - focal and segmental hyalinosis or sclerosis ~~N02.1~~ N02.B2
Revise	- - - membranoproliferative (diffuse) ~~N02.5~~ N02.B3
Revise	- - - membranous (diffuse) ~~N02.2~~ N02.B4
Revise	- - - mesangial proliferative (diffuse) ~~N02.3~~ N02.B5
Revise	- - - mesangiocapillary (diffuse) ~~N02.5~~ N02.B6
Revise	- - - proliferative NEC ~~N02.8~~ N02.B9
Revise	- - - specified pathology NEC ~~N02.8~~ N02.B9

Inappropriate Sinus Tachycardia (IST)

This is a representation from the March 2022 ICD-10 Coordination and Maintenance Committee Meeting of the Inappropriate Sinus Tachycardia (IST) proposal with the recommended title modification adding "as state" to the code title at I47.11, Inappropriate sinus tachycardia. The modification is in **bold.**

Inappropriate sinus tachycardia's definition is sinus heart rate >100 bpm at rest (with a mean 24-hour heart rate >90 bpm not due to primary causes) and is associated with distressing symptoms of palpitations.

The prevalence of IST was estimated in a middle-aged population of people with and without hypertension. Using a definition of a resting heart rate >100 bpm and an average heart rate of >90 bpm on 24-hour Holter monitoring, the IST prevalence was 1.2% (7 of 604 patients), including both symptomatic and asymptomatic patients.[2]

The mechanisms leading to IST are not completely understood, but there are several underlying diseases that can result in this syndrome, including increased sinus node automaticity, beta-adrenergic hypersensitivity, decreased parasympathetic activity, and impaired neurohumoral modulation. β-Adrenergic receptor antibodies can sensitize β- adrenergic receptors in some patients, while other patients might have increased sympathetic activity and sensitivity, with or without inherent impaired sinus node automaticity.

National Center for Health Statistics received a request to create an ICD-10-CM code for inappropriate sinus tachycardia for coding specificity to accurately track cases, allowing for etiology related research, patient segmentation, and therapeutic selection.

References

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TABULAR MODIFICATIONS

I47 Paroxysmal tachycardia

I47.1 Supraventricular tachycardia

Delete ~~Atrial (paroxysmal) tachycardia~~
Delete ~~Atrioventricular [AV] (paroxysmal) tachycardia~~
Delete ~~Atrioventricular re-entrant (nodal) tachycardia [AVNRT] [AVRT]~~
Delete ~~Junctional (paroxysmal) tachycardia~~
Delete ~~Nodal (paroxysmal) tachycardia~~

New code	I47.10	Supraventricular tachycardia, unspecified
New code	I47.11	Inappropriate sinus tachycardia**, as stated**
New code	I47.19	Other supraventricular tachycardia
Add		Atrial (paroxysmal) tachycardia
Add		Atrioventricular [AV] (paroxysmal) tachycardia
Add		Atrioventricular re-entrant (nodal) tachycardia [AVNRT] [AVRT]
Add		Junctional (paroxysmal) tachycardia
Add		Nodal (paroxysmal) tachycardia
Add		Supraventricular tachycardia

Insulin Resistant Syndrome

This is a representation from the September 2021 and March 2022 ICD-10 Coordination and Maintenance Committee Meetings of the Insulin Resistant Syndrome proposal with the recommended modification to add "syndrome" to the inclusion term Insulin resistance, Type B at E88.818, Other insulin resistance. The modification is in **bold.**

The National Institute of Health defines metabolic syndrome as the presence of at least 3 of the following traits (including the ones that are controlled by medication): large waist, elevated triglyceride level, reduced HDL cholesterol, increased blood pressure and elevated fasting blood glucose. Other names for metabolic syndrome are: Dysmetabolic syndrome, Hypertriglyceridemic waist, Insulin resistance syndrome, Obesity syndrome or Syndrome X.

The National Heart, Lung and Blood Institute states the following: Insulin Resistance also may increase your risk for metabolic syndrome. Insulin resistance is a condition in which the body cannot use its insulin properly. Insulin is a hormone that helps move blood sugar into cells where it is used for energy. Insulin resistance can lead to high blood sugar levels, and it is intricately linked to overweight and obesity. Genetics and aging may also contribute to the development of this syndrome.

Type A and B insulin-resistance syndrome belongs to the group of extreme insulin-resistance syndromes (which includes leprechaunism, the lipodystrophies, Rabson-Mendenhall syndrome) (characterized by the triad of hyperinsulinemia, acanthosis nigricans (skin lesions associated with insulin resistance), and signs of hyperandrogenism in females without lipodystrophy and who are not overweight.[1]

This proposal is supported by the Office of Genomics Precision Public Health and American College of Medical Genetics and Genomics.

References

1. Orphanet: Insulin resistance syndrome type A. INSERM US14 -- ALL RIGHTS RESERVED
https://www.orpha.net/consor/cgi-bin/OC_Exp.php?Expert=2297

TABULAR MODIFICATIONS

E88 Other and unspecified metabolic disorders
Use additional codes for associated conditions
Excludes1: histiocytosis X (chronic) (C96.6)
E88.8 Other specified metabolic disorders
Revise E88.81 Metabolic syndrome and other insulin
Delete ~~Dysmetabolic syndrome X~~
Use additional codes for associated manifestations, such as:
obesity (E66.-)

New code Add	E88.810	Metabolic syndrome Dysmetabolic syndrome
New code	E88.811	Insulin resistance syndrome, Type A
New code Add	E88.818	Other insulin resistance Insulin resistance **syndrome,** Type B
New code	E88.819	Insulin resistance, unspecified

Intestinal Microbial Overgrowth

Intestinal microbial overgrowth includes several diseases including small intestinal bacterial overgrowth (SIBO), intestinal methanogen overgrowth (IMO) and small intestinal fungal overgrowth (SIFO). These diseases result from the overpopulation of bacteria, methanogenic archaea or fungi in the intestines and can lead to debilitating symptoms with significant effect on quality of life. Over the past 2 decades, there has been significant progress in the diagnosis and treatment of these diseases specially SIBO and IMO with various published practice guidelines including the American College of Gastroenterology (ACG).[1-3]

Small intestinal bacterial overgrowth (SIBO): Common symptoms of SIBO include abdominal distention, abdominal bloating, diarrhea, and abdominal pain/discomfort. Malnutrition, weight loss, and anemia are seen in more severe cases.[4] SIBO is estimated to affect at least 33% of patients with unexplained GI symptoms.[1] The gold standard for a diagnosis of SIBO is the presence of $\geq 10^3$ CFU/mL of jejunal aspirate by culture.[2] Alternatively, SIBO can be diagnosed by a rise in exhaled hydrogen levels from baseline after consumption of a fermentable sugar substrate such as glucose or lactulose.[2] Antibiotics remain the core treatment modality for SIBO. A systematic review of the literature found 23 trials addressing the role of antibiotics in SIBO.[5] The most commonly used antibiotics were clindamycin, metronidazole, neomycin, rifaximin and tetracycline.[1] Short term courses of elemental diets can also be a safe and effective alternative to antibiotics. Elemental diets are believed to be fully absorbed within the first few feet of small intestine, limiting the delivery of nutrients to the microbes in mid/distal small bowel and colon.[6]

Intestinal methanogen overgrowth (IMO): Methanogens are classified under the kingdom of archaea and are not bacteria, hence, excessive archaea cannot be classified under bacterial overgrowth. Symptoms of IMO include abdominal bloating, distention, constipation and abdominal discomfort.[7] Severity of symptoms directly correlates with the amount of methane produced by the gut methanogens.[1] IMO is diagnosed by detection of exhaled methane levels >10 ppm during breath testing.[1, 2, 7] In patients with IMO, single antibiotic therapy may not have the expected effectiveness. *Methanobrevibacter smithii*, which is the main archaeon responsible for methane production in humans, is resistant to several antibiotics. In a retrospective study [8], subjects with IMO who received either neomycin or rifaximin alone did not have a substantial improvement. However, combination of neomycin with rifaximin decreased methane production in more than 80 % of subjects, with a similar response in constipation-related symptoms. Similar findings were seen in prospective trials.[9]

Small intestinal fungal overgrowth (SIFO) symptoms include abdominal pain, gas, bloating, fullness, belching, indigestion, and diarrhea. SIFO is diagnosed via small bowel aspiration and fungal cultures, and the choice of antifungal therapy is mainly driven by the culture and sensitivity results. Irrespective of geographic distribution, *Candida albicans* and *Candida glabrata* are the most common species isolated from the small bowel.[10] Overgrowth of H_2S-producing bacteria are also associated with abdominal pain and diarrhea.[11]

Overall, intestinal microbial overgrowth has multiple objectively-defined subtypes which are diagnosed and treated differently. Currently, there are no codes dedicated to intestinal microbial overgrowth or its subtypes. Given this deficiency, physicians and coders commonly use: "A04.9: Bacterial intestinal infection, unspecified" (however this code relates to intestinal infections and not a microbial overgrowth), or "K63.8: Other specified diseases of intestine" which has an unacceptable specificity for microbial overgrowth.

This lack of specificity decreases the opportunity to use ICD-10-CM codes for accurate disease tracking. The following revisions are proposed to achieve sufficient clinical granularity for diagnosis of intestinal microbial overgrowth and further improve international classification, tracking, and surveillance.

Specific codes for intestinal microbial overgrowth are requested by Drs. Rezaie and Pimentel who are the lead authors of the North American consensus on breath testing to diagnose SIBO and IMO[2], and the American College of Gastroenterology (ACG) clinical guidelines for diagnosis and treatment of microbial overgrowth.[1]

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TABULAR MODIFICATIONS

K63 Other diseases of intestine

K63.8 Other specified diseases of intestine

New sub-subcategory — K63.82 Intestinal microbial overgrowth

New code — K63.821 Intestinal methanogen overgrowth

New sub-subcategory — K63.822 Small intestinal bacterial overgrowth

New code — K63.8221 Small intestinal bacterial overgrowth, hydrogen-subtype

New code — K63.8222 Small intestinal bacterial overgrowth, hydrogen sulfide-subtype

New code — K63.8229 Small intestinal bacterial overgrowth, unspecified

New code — K63.823 Small intestinal fungal overgrowth

Intrahepatic Cholestasis in Pregnancy

The American College of Obstetricians and Gynecologists (ACOG) is proposing to add new codes that allow for the reporting of Intrahepatic Cholestasis in Pregnancy (ICP) for each trimester.

ICP is a pregnancy-specific disorder where bile acid transport in the liver is altered. This leads to a build-up of bile acid in both the maternal circulation as well as the fetal amniotic fluid. There are both maternal and fetal risks of intrahepatic cholestasis of pregnancy. Maternal risks include an increased risk of pre-eclampsia and gestational diabetes.[1] Fetal risks include preterm birth, increased respiratory distress after birth when matched to the same gestational age non ICP pregnancies, meconium staining of the amniotic fluid and stillbirth.[1]

Intrahepatic cholestasis of pregnancy affects about 0.5% of pregnancies in the United States.[1] The exact cause of ICP is unknown but is thought to be related to the increased hormones of pregnancy altering bile acid transport. About 15% of cases are linked with a genetic mutation, most commonly ABCB4.[2] ICP most often occurs in the 2nd and 3rd trimesters of pregnancy, but cases have also been diagnosed as early as 5 weeks.[1]

In the United States, the Society for Maternal-Fetal Medicine (SMFM) has a set of guidelines for the diagnosis and treatment of intrahepatic cholestasis of pregnancy.[1] The main symptom of ICP is pruritus which is often on the hands and feet but can become generalized and severe. The syndrome is diagnosed by a total bile acid blood test which is elevated in ICP. The most often agreed-upon diagnosis level in the United States is 10 μmol/L.[1] Other pre-pregnancy liver conditions should also be screened for with a thorough personal and family history as well as necessary laboratory evaluation prior to diagnosis. SMFM has outlined a specific set of treatment guidelines including fetal monitoring, medication treatment with ursodeoxycholic acid and appropriately timed infant delivery to prevent complications such as stillbirth. This disorder is a unique disease requiring its own specific management in pregnancy.[1,3]

Intrahepatic cholestasis of pregnancy resolves after the peripartum period is over. Some patients might have underlying liver disorders that predisposed them to the development of intrahepatic cholestasis of pregnancy, but the disorder itself is pregnancy-specific.

There is a need for a specific code for intrahepatic cholestasis of pregnancy both to allow for the proper identification and treatment of these patients as well as being able to gather more information collectively about the outcomes of these pregnancies.

It has also been shown that some patients with intrahepatic cholestasis of pregnancy are at risk for later diseases. In looking at the long-term health of these patients, they are more likely to have other hepatobiliary diseases in the future including cholelithiasis, pancreatitis and need for cholecystectomy.[5,6,7,8] There is also an increased risk of hypothyroidism. Given that there are some

life-long health concerns with the disorder, it would be beneficial to be able to track these patients in order to hopefully modify their lifestyle factors to possibly prevent further disease.

In summary, intrahepatic cholestasis of pregnancy is a pregnancy specific liver disorder with its own set of diagnostic criteria and implications for pregnancy as well as future health. Patients with this disorder would benefit by the creation of an ICD-10-CM code as the disorder differs from other liver disorders with which it is currently classified.

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TABULAR MODIFICATIONS

O26 Maternal care for other conditions predominantly related to pregnancy

O26.6 Liver and biliary tract disorders in pregnancy, childbirth and the puerperium

Use additional code to identify the specific disorder

Excludes2: hepatorenal syndrome following labor and delivery (O90.4)

O26.64 Intrahepatic cholestasis of pregnancy

New code O26.641 Intrahepatic cholestasis of pregnancy, first trimester

New code O26.642 Intrahepatic cholestasis of pregnancy, second trimester

New code O26.643 Intrahepatic cholestasis of pregnancy, third trimester

New code O26.649 Intrahepatic cholestasis of pregnancy, unspecified trimester

Leukodystrophies

Leukodystrophies, or more specifically inherited leukodystrophies, are a group of diseases affecting the white matter of the brain, that cause significant morbidities and death in 1 of 3 patients by age 8 years.[1] The Global Leukodystrophy Alliance (GLIA), an NIH-funded consortium composed of clinicians, scientists, patients, and patient advocacy groups, is requesting new ICD-10-CM codes for certain separate and genetically distinct leukodystrophy diseases. NCHS has received letters of support from various professional and patient groups.

This proposal was presented at the March 2022 Coordination and Maintenance Meeting. Based on public comment, changes have been made and resubmitted for reconsideration. Changes are noted in **bold**.

Leukodystrophies may present at any age from preterm infants and neonates to late adulthood and have been reported across all ethnicities and regions of the world. 30 years ago only a few leukodystrophies were recognized as distinct disease entities, but in the past 10 years over 400 genetically unique leukodystrophies have been reported.[2] Even though most leukodystrophies are individually rare, as a group leukodystrophies affect close to 1 in 4,000 live births.[3,4] Further, consensus work in the community has defined a group of leukodystrophies with unique genetic causes and well-studied, distinct clinical and pathophysiological features.

Currently there are only specific ICD-10-CM codes for six of the primary leukodystrophies (X-linked Adrenoleukodystrophy, ALD- E71.52x; Metachromatic leukodystrophy, MLD- E75.25; Krabbe disease- E75.23, Refsum's disease- G60.1; Zellweger syndrome- E71.510; and E71.511 Neonatal Adrenoleukodystrophy). Otherwise, many leukodystrophies are indexed under a single ICD-10-CM code E75.29, Other sphingolipidosis.
Prior to ICD-10-CM, there were not specific ICD codes for ALD, MLD, or Krabbe. The advent of specific ICD-10-CM codes for ALD, Krabbe, and MLD enabled clinical trials, newborn screening, and studies of racial disparities.[6,7]

The leukodystrophies proposed for unique ICD-10-CM codes all have unique genetic causes; distinct clinical courses and morbidities; and have different treatments- either currently available or in clinical trials. For example, VWM has a sputtering clinical course[8] and has a clinical trial with the α-agonist guanabenz.[9] In contrast, Canavan's disease has an early rapid progression[10] and potential treatment with antisense oligonucleotides.[11]

Creation of specific ICD-10-CM codes for this heterogenous, complex group of disorders known as leukodystrophies is critical for patient care, clinical trials, and research. The diversity in causes should be reflected by a diversity of codes to best represent these disorders. The importance of and difference between these leukodystrophy disorders can be seen in the codes already created for ICD-11. In ICD-11, Leukodystrophy has its own ICD-11 category, there are five new leukodystrophy codes, and there are also fourteen new leukodystrophy indices.

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TABULAR MODIFICATIONS

E74 Other disorders of carbohydrate metabolism

E74.0 Glycogen storage disease

E74.09 Other glycogen storage disease

Andersen disease

Delete ~~Hers disease~~

Delete ~~Tauri disease~~

Glycogen storage disease, types 0, IV, VI-XI

Add Hers disease

Liver phosphorylase deficiency

Muscle phosphofructokinase deficiency

Add Tauri disease

E75 Disorders of sphingolipid metabolism and other lipid storage disorders
E75.2 Other sphingolipidosis
Excludes1:adrenoleukodystrophy [Addison-Schilder] (E71.528)
New code **E75.27 Pelizaeus-Merzbacher disease**
New code **E75.28 Canavan disease**

E79 Disorders of purine and pyrimidine metabolism
E79.8 Other disorders of purine and pyrimidine metabolism
Delete ~~**Hereditary xanthinuria**~~
New code **E79.81 Aicardi-Goutières syndrome**
New code **E79.82 Hereditary xanthinuria**
New code **E79.89 Other specified disorders of purine and pyrimidine metabolism**

E88 Other and unspecified metabolic disorders
E88.4 Mitochondrial metabolism disorders
New code **E88.43 Disorders of mitochondrial tRNA synthetases**

G11 Hereditary ataxia
New code G11.5 Hypomyelination - hypogonadotropic hypogonadism - hypodontia
Add 4H syndrome
Add Pol III-related leukodystrophy
New code G11.6 Leukodystrophy with vanishing white matter disease

G23 Other degenerative diseases of basal ganglia
New code G23.3 Hypomyelination with atrophy of the basal ganglia and cerebellum
Add H-ABC

G31 Other degenerative diseases of nervous system, not elsewhere classified
G31.8 Other specified degenerative diseases of nervous system
New code **G31.80 Leukodystrophy, unspecified**
New code **G31.86 Alexander Disease**

G90 Disorders of autonomic nervous system
New code **G90.A LMNB1-related autosomal dominant leukodystrophy**

G93 Other Disorders of the Brain

G93.4 Other and unspecified encephalopathy

New code	**G93.42 Megaloencephalic leukoencephalopathy with subcortical cysts**
New code	**G93.43 Leukoencephalopathy with calcifications and cysts**
New code	**G93.44 Adult-onset leukodystrophy with axonal spheroids**
Add	**Adult-onset leukoencephalopathy with axonal spheroids and pigmented glia**

Lymphoma in Remission

Lymphoma is a cancer of the lymphatic system, which is part of the body's germ-fighting network. The lymphatic system includes the lymph nodes (lymph glands), spleen, thymus gland and bone marrow. Lymphoma can affect all those areas as well as other organs throughout the body.[1]

In general, the goal of treatment is to destroy as many lymphoma cells as possible and to induce a complete remission. Complete remission means that all evidence of disease is eliminated. Patients who go into remission are sometimes cured of their disease. Treatment can also keep non-Hodgkin lymphoma (NHL) in check for many years, even though imaging or other studies show remaining sites of disease.[2]

A new ICD-10-CM code will provide coding specificity for the distinct types of lymphoma in remission. The absence of lymphoma in remission codes will hinder the ability to make meaningful comparisons to assess and evaluate differences in patient care, statistical data, resource consumption (i.e., overall length of stay, additional drugs, etc.), and accurate clinical outcomes of lymphoma cases.

The National Center of Health Statistics received a proposal requesting new ICD-10-CM codes for lymphoma in remission from Alliance Dedicated Cancer Centers (ADCC).

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TABULAR MODIFICATIONS

C81 Hodgkin lymphoma

New subcategory

C81.0 Nodular lymphocyte predominant Hodgkin lymphoma

New code

C81.0A Nodular lymphocyte predominant Hodgkin lymphoma, in remission

Add

Nodular sclerosis classical Hodgkin lymphoma, in remission

C81.2 Mixed cellularity Hodgkin lymphoma
Mixed cellularity classical Hodgkin lymphoma

New code C81.2A Mixed cellularity Hodgkin lymphoma, in remission
Add Mixed cellularity classical Hodgkin lymphoma, in remission

C81.3 Lymphocyte depleted Hodgkin lymphoma
Lymphocyte depleted classical Hodgkin lymphoma

New code C81.3A Lymphocyte depleted Hodgkin lymphoma, in remission
Add Lymphocyte depleted classical Hodgkin lymphoma, in remission

C81.4 Lymphocyte-rich Hodgkin lymphoma
Lymphocyte-rich classical Hodgkin lymphoma
New code C81.4A Lymphocyte-rich Hodgkin lymphoma, in remission
Add Lymphocyte-rich classical Hodgkin lymphoma, in remission

C81.7 Other Hodgkin lymphoma
Classical Hodgkin lymphoma NOS
Other classical Hodgkin lymphoma

New code C81.7A Other Hodgkin lymphoma, in remission
Add Classical Hodgkin lymphoma NOS, in remission
Add Other classical Hodgkin lymphoma, in remission

C81.9 Hodgkin lymphoma, unspecified
New code C81.9A Hodgkin lymphoma, unspecified, in remission

C82 Follicular lymphoma
C82.0 Follicular lymphoma grade I
New code C82.0A Follicular lymphoma grade I, in remission

C82.1 Follicular lymphoma grade II
New code C82.1A Follicular lymphoma grade II, in remission

C82.2 Follicular lymphoma grade III, unspecified
New code C82.2A Follicular lymphoma grade III, unspecified, in remission

C82.3 Follicular lymphoma grade IIIa
New code C82.3A Follicular lymphoma grade IIIa, in remission

C82.4 Follicular lymphoma grade IIIb
New code C82.4A Follicular lymphoma grade IIIb, in remission

C82.5 Diffuse follicle center lymphoma

New code C82.5A Diffuse follicle center lymphoma, in remission

C82.6 Cutaneous follicle center lymphoma

New code C82.6A Cutaneous follicle center lymphoma, in remission

C82.8 Other types of follicular lymphoma

New code C82.8A Other types of follicular lymphoma, in remission

C82.9 Follicular lymphoma, unspecified

New code C82.9A Follicular lymphoma, unspecified, in remission

C83 Non-follicular lymphoma

C83.0 Small cell B-cell lymphoma
Lymphoplasmacytic lymphoma
Nodal marginal zone lymphoma
Non-leukemic variant of B-CLL
Splenic marginal zone lymphoma

New code C83.0A Small cell B-cell lymphoma, in remission
Add Lymphoplasmacytic lymphoma, in remission
Add Nodal marginal zone lymphoma, in remission
Add Non-leukemic variant of B-CLL, in remission
Add Splenic marginal zone lymphoma, in remission

C83.1 Mantle cell lymphoma
Centrocytic lymphoma

New code C83.1A Mantle cell lymphoma, in remission
Add Centrocytic lymphoma, in remission

C83.3 Diffuse large B-cell lymphoma

New code C83.3A Diffuse large B-cell lymphoma, in remission
Add Anaplastic diffuse large B-cell lymphoma, in remission
Add CD30-positive diffuse large B-cell lymphoma, in remission
Add Centroblastic diffuse large B-cell lymphoma, in remission
Add Diffuse large B-cell lymphoma, subtype not specified, in remission
Add Immunoblastic diffuse large B-cell lymphoma, in remission
Add Plasmablastic diffuse large B-cell lymphoma, in remission
Add T-cell rich diffuse large B-cell lymphoma, in remission

C83.5 Lymphoblastic (diffuse) lymphoma

New code	C83.5A	Lymphoblastic (diffuse) lymphoma, in remission
Add		B-precursor lymphoma, in remission
Add		Lymphoblastic B-cell lymphoma, in remission
Add		Lymphoblastic lymphoma NOS, in remission
Add		Lymphoblastic T-cell lymphoma, in remission
Add		T-precursor lymphoma, in remission

C83.7 Burkitt lymphoma

New code	C83.7A	Burkitt lymphoma, in remission
Add		Atypical Burkitt lymphoma, in remission
Add		Burkitt-like lymphoma, in remission

C83.8 Other non-follicular lymphoma

New code	C83.8A	Other non-follicular lymphoma, in remission
Add		Intravascular large B-cell lymphoma, in remission
Add		Lymphoid granulomatosis, in remission
Add		Primary effusion B-cell lymphoma, in remission

C83.9 Non-follicular (diffuse) lymphoma, unspecified

New code	C83.9A	Non-follicular (diffuse) lymphoma, unspecified, in remission

C84 Mature T/NK-cell lymphomas

C84.0 Mycosis fungoides

New code	C84.0A	Mycosis fungoides, in remission

C84.1 Sézary disease

New code	C84.1A	Sézary disease, in remission

C84.4 Peripheral T-cell lymphoma, not classified

New code	C84.4A	Peripheral T-cell lymphoma, not classified, in remission
Add		Lennert's lymphoma, in remission
Add		Lymphoepithelioid lymphoma, in remission
Add		Mature T-cell lymphoma, not elsewhere classified, in remission

C84.6 Anaplastic large cell lymphoma, ALK-positive
Anaplastic large cell lymphoma, CD30-positive

New code C84.6A Anaplastic large cell lymphoma, ALK-positive, in remission
Add Anaplastic large cell lymphoma, CD30-positive, in remission

C84.7 Anaplastic large cell lymphoma, ALK-negative
New code C84.7B Anaplastic large cell lymphoma, ALK-negative, in remission

C84.A Cutaneous T-cell lymphoma, unspecified
New code C84.AA Cutaneous T-cell lymphoma, unspecified, in remission

C84.Z Other mature T/NK-cell lymphomas
New code C84.ZA Other mature T/NK-cell lymphomas, in remission

C84.9 Mature T/NK-cell lymphomas, unspecified
NK/T cell lymphoma NOS

New code C84.9A Mature T/NK-cell lymphomas, unspecified, in remission
Add NK/T cell lymphoma NOS, in remission

C85 Other specified and unspecified types of non-Hodgkin lymphoma

C85.1 Unspecified B-cell lymphoma
New code C85.1A Unspecified B-cell lymphoma, in remission

C85.2 Mediastinal (thymic) large B-cell lymphoma
New code C85.2A Mediastinal (thymic) large B-cell lymphoma, in remission

C85.8 Other specified types of non-Hodgkin lymphoma
New code C85.8A Other specified types of non-Hodgkin lymphoma, in remission

C85.9 Non-Hodgkin lymphoma, unspecified
New code C85.9A Non-Hodgkin lymphoma, unspecified, in remission
Add Lymphoma NOS, in remission
Add Malignant lymphoma NOS, in remission
Add Non-Hodgkin lymphoma NOS, in remission

C86 Other specified types of T/NK-cell lymphoma
C86.0 Extranodal NK/T-cell lymphoma, nasal type

New code C86.00 Extranodal NK/T-cell lymphoma, nasal type not having achieved remission
Add Extranodal NK/T-cell lymphoma, nasal type NOS

Add			Extranodal NK/T-cell lymphoma, nasal type with failed remission
New code		C86.01	Extranodal NK/T-cell lymphoma, nasal type, in remission
	C86.1	Hepatosplenic T-cell lymphoma Alpha-beta and gamma delta types	
New code		C86.10	Hepatosplenic T-cell lymphoma not having achieved remission
Add			Hepatosplenic T-cell lymphoma NOS
Add			Hepatosplenic T-cell lymphoma with failed remission
New code		C86.11	Hepatosplenic T-cell lymphoma, in remission
	C86.2	Enteropathy-type (intestinal) T-cell lymphoma Enteropathy associated T-cell lymphoma	
New code		C86.20	Enteropathy-type (intestinal) T-cell lymphoma not having achieved remission
Add			Enteropathy associated T-cell lymphoma NOS
Add			Enteropathy associated T-cell lymphoma not having achieved remission
Add			Enteropathy associated T-cell lymphoma with failed remission
Add			Enteropathy-type (intestinal) T-cell lymphoma NOS
Add			Enteropathy-type (intestinal) T-cell lymphoma with failed remission
New code		C86.21	Enteropathy-type (intestinal) T-cell lymphoma, in remission
Add			Enteropathy associated T-cell lymphoma, in remission
	C86.3	Subcutaneous panniculitis-like T-cell lymphoma	
New code		C86.30	Subcutaneous panniculitis-like T-cell lymphoma not having achieved remission
Add			Subcutaneous panniculitis-like T-cell lymphoma NOS
Add			Subcutaneous panniculitis-like T-cell lymphoma with failed remission
New code		C86.31	Subcutaneous panniculitis-like T-cell lymphoma, in remission
	C86.4	Blastic NK-cell lymphoma Blastic plasmacytoid dendritic cell neoplasm (BPDCN)	

New code	C86.40	Blastic NK-cell lymphoma not having achieved remission
Add		Blastic NK-cell lymphoma NOS
Add		Blastic NK-cell lymphoma with failed remission
Add		Blastic plasmacytoid dendritic cell neoplasm (BPDCN) NOS
Add		Blastic plasmacytoid dendritic cell neoplasm (BPDCN) not having achieved remission
Add		Blastic plasmacytoid dendritic cell neoplasm (BPDCN) with failed remission
New code	C86.41	Blastic NK-cell lymphoma, in remission
Add		Blastic plasmacytoid dendritic cell neoplasm (BPDCN), in remission

C86.5 Angioimmunoblastic T-cell lymphoma
Angioimmunoblastic lymphadenopathy with dysproteinemia (AILD)

New code	C86.50	Angioimmunoblastic T-cell lymphoma not having achieved remission
Add		Angioimmunoblastic lymphadenopathy with dysproteinemia (AILD) NOS
Add		Angioimmunoblastic lymphadenopathy with dysproteinemia (AILD) not having achieved remission
Add		Angioimmunoblastic lymphadenopathy with dysproteinemia (AILD) with failed remission
Add		Angioimmunoblastic T-cell lymphoma NOS
Add		Angioimmunoblastic T-cell lymphoma with failed remission
New code	C86.51	Angioimmunoblastic T-cell lymphoma, in remission
Add		Angioimmunoblastic lymphadenopathy with dysproteinemia (AILD), in remission

C86.6 Primary cutaneous CD30-positive T-cell proliferations
Lymphomatoid papulosis
Primary cutaneous anaplastic large cell lymphoma
Primary cutaneous CD30-positive large T-cell lymphoma

New code	C86.60	Primary cutaneous CD30-positive T-cell proliferations not having achieved remission
Add		Lymphomatoid papulosis NOS
Add		Lymphomatoid papulosis not having achieved remission
Add		Lymphomatoid papulosis with failed remission
Add		Primary cutaneous anaplastic large cell lymphoma NOS
Add		Primary cutaneous anaplastic large cell lymphoma not having achieved remission

Add		Primary cutaneous anaplastic large cell lymphoma with failed remission
Add		Primary cutaneous CD30-positive large T-cell lymphoma NOS
Add		Primary cutaneous CD30-positive large T-cell lymphoma not having achieved remission
Add		Primary cutaneous CD30-positive T-cell proliferations with failed remission
New code	C86.61	Primary cutaneous CD30-positive T-cell proliferations, in remission
Add		Lymphomatoid papulosis, in remission
Add		Primary cutaneous anaplastic large cell lymphoma, in remission
Add		Primary cutaneous CD30-positive large T-cell lymphoma, in remission

C88 Malignant immunoproliferative diseases and certain other B-cell lymphomas

C88.0 Waldenström macroglobulinemia
Lymphoplasmacytic lymphoma with IgM-production
Macroglobulinemia (idiopathic) (primary)

New code	C88.00	Waldenström macroglobulinemia not having achieved remission
Add		Lymphoplasmacytic lymphoma with IgM-production, NOS
Add		Lymphoplasmacytic lymphoma with IgM-production not having achieved remission
Add		Lymphoplasmacytic lymphoma with IgM-production with failed remission
Add		Macroglobulinemia (idiopathic) (primary) NOS
Add		Macroglobulinemia (idiopathic) (primary) not having achieved remission
Add		Macroglobulinemia (idiopathic) (primary) with failed remission
Add		Waldenström macroglobulinemia NOS
Add		Waldenström macroglobulinemia with failed remission
New code	C88.01	Waldenström macroglobulinemia, in remission
Add		Lymphoplasmacytic lymphoma with IgM-production, in remission
Add		Macroglobulinemia (idiopathic) (primary), in remission

C88.2 Heavy chain disease
Franklin disease
Gamma heavy chain disease
Mu heavy chain disease

New code	C88.20	Heavy chain disease not having achieved remission
Add		Franklin disease NOS
Add		Franklin disease not having achieved remission
Add		Franklin disease with failed remission
Add		Gamma heavy chain disease NOS
Add		Gamma heavy chain disease not having achieved remission
Add		Gamma heavy chain disease with failed remission
Add		Heavy chain disease NOS
Add		Heavy chain disease with failed remission
Add		Mu heavy chain disease NOS
Add		Mu heavy chain disease not having achieved remission
Add		Mu heavy chain disease with failed remission
New code	C88.21	Heavy chain disease, in remission
Add		Franklin disease, in remission
Add		Gamma heavy chain disease, in remission
Add		Mu heavy chain disease, in remission

C88.3 Immunoproliferative small intestinal disease
Alpha heavy chain disease
Mediterranean lymphoma

New code	C88.30	Immunoproliferative small intestinal disease not having achieved remission
Add		Alpha heavy chain disease immunoproliferative small intestinal disease not having achieved remission
Add		Alpha heavy chain disease NOS
Add		Alpha heavy chain disease with failed remission
Add		Immunoproliferative small intestinal disease NOS
Add		Immunoproliferative small intestinal disease with failed remission
Add		Mediterranean lymphoma NOS
Add		Mediterranean lymphoma not having achieved remission
Add		Mediterranean lymphoma with failed remission
New code	C88.31	Immunoproliferative small intestinal disease, in remission
Add		Alpha heavy chain disease, in remission
Add		Mediterranean lymphoma, in remission

	C88.4	Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue [MALT-lymphoma]	
Add		Lymphoma of bronchial-associated lymphoid tissue [BALT-lymphoma]	
		Lymphoma of skin-associated lymphoid tissue [SALT-lymphoma]	
Delete		~~Lymphoma of bronchial-associated lymphoid tissue [BALT-lymphoma]~~	
New code	C88.40	Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue [MALT-lymphoma] not having achieved remission	
Add		Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue [MALT-lymphoma] NOS	
Add		Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue [MALT-lymphoma] with failed remission	
Add		Lymphoma of bronchial-associated lymphoid tissue [BALT-lymphoma] NOS	
Add		Lymphoma of bronchial-associated lymphoid tissue [BALT-lymphoma] not having achieved remission	
Add		Lymphoma of bronchial-associated lymphoid tissue [BALT-lymphoma] with failed remission	
Add		Lymphoma of skin-associated lymphoid tissue [SALT-lymphoma] NOS	
Add		Lymphoma of skin-associated lymphoid tissue [SALT-lymphoma] not having achieved remission	
Add		Lymphoma of skin-associated lymphoid tissue [SALT-lymphoma] with failed remission	
New code	C88.41	Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue [MALT-lymphoma], in remission	
Add		Lymphoma of bronchial-associated lymphoid tissue [BALT-lymphoma], in remission	
Add		Lymphoma of skin-associated lymphoid tissue [SALT-lymphoma], in remission	
	C88.8	Other malignant immunoproliferative diseases	
New code	C88.80	Other malignant immunoproliferative diseases not having achieved remission	
Add		Other malignant immunoproliferative diseases NOS	
Add		Other malignant immunoproliferative diseases with failed remission	

New code		C88.01	Other malignant immunoproliferative diseases, in remission
	C88.9	Malignant immunoproliferative disease, unspecified Immunoproliferative disease NOS	
		C88.90	Malignant immunoproliferative disease, unspecified not having achieved remission
Add			Immunoproliferative disease NOS
Add			Immunoproliferative disease NOS not having achieved remission
Add			Immunoproliferative disease NOS with failed remission
Add			Malignant immunoproliferative disease, unspecified NOS
Add			Malignant immunoproliferative disease, unspecified with failed remission
		C88.91	Malignant immunoproliferative disease, unspecified, in remission
			Immunoproliferative disease NOS, in remission

Lysosome-Associated Membrane Protein 2 (LAMP2) Deficiency (Danon Disease)

Danon disease is an X-linked dominant metabolic disorder originally described as "lysosomal glycogen storage disease with normal acid maltase"[1]. Subsequently, a causative gene (*LAMP2*) was identified. Mutations in this gene result in a primary deficiency of lysosome-associated membrane protein 2, an intracellular protein that mediates endosomal-lysosomal fusion, an integral component of autophagy. The absence of intracellular LAMP2 protein in tissues/organs is associated with impaired autophagic and mitochondrial function and accumulation of intracellular debris resulting in functional impairment[2].

Danon disease is a multi-systemic disorder and represents one of the most aggressive cardiomyopathies ever characterized, especially for male patients. There currently are no available disease-modifying therapies that mitigate progression to end stage heart failure and death. As a result, most male Danon patients do not live beyond adolescence or early adulthood in the absence of heart transplantation.

Danon disease involves the heart, skeletal muscles, central nervous system, and retina, although the non-cardiac manifestations are most frequently reported in males. Given the multisystemic presentation and rarity of Danon disease, proper identification and management of patients has been challenging. Disease heterogeneity is also apparent between males and females, who manifest significant clinical differences due to the X-linked inheritance of the disease[3]. The average age of initial symptoms in males is 12 years and the average age of death is 19 years[4]. The average age of initial symptoms in females is 28 and of death is 35 years[4]. Within each gender, there exist considerable variability of presentation, organ involvement, and rapidity of cardiac deterioration[4].

Danon disease is diagnosed via genetic testing for pathogenic variations of the *LAMP2* gene[5]. As of 2019, at least 146 cases had been reported in the literature with molecular confirmation indicating Danon disease[6]. The true prevalence of Danon disease is not known. A crude estimate of prevalence can be based on extrapolation from studies of pediatric and adult hypertrophic cardiomyopathy (for which there is an overall prevalence of approximately 1:500 in the overall population); in these series, LAMP2 pathogenic variants have been identified as underlying approximately 1% - 4% of hypertrophic cardiomyopathy cases[4,7,8] although there is still no consensus on these estimates[6,9]. These data yield an estimate of 10,000 – 25,000 total cases in the US. The volume of documented cases is rising with wider availability of genetic testing[10] and genetically-targeted therapies in development, which may motivate more widespread diagnostic efforts.

Danon disease was originally classified as a glycogen storage disease (GSD), but is distinct from other GSDs, which are caused by a deficient enzyme of glycogen metabolism. Danon disease differs from other GSDs in terms of genetic cause, pathophysiology (i.e., it is a transport disorder, not a storage disorder), prevalence, symptoms, and treatment; this distinction also applies to Danon disease compared with other disorders in the E74.0 subcategory, as shown in the comparisons noted below.

Von Gierke disease is GSD type I, and involves gene GSDIA or GSDIB, and is autosomal recessive with incidence 1:100,000 births. Symptoms include low blood sugar, enlarged liver and kidneys, and treatment involves diet. The ICD-10-CM code is E74.01.

Pompe disease is GSD type II, involving gene GAA, autosomal recessive, incidence 1:40,000 births. Symptoms involve respiratory issues, muscle weakness, and hypertrophic cardiomyopathy (HCM); treatment involves enzyme replacement. The ICD-10-CM code is E74.02.

Danon disease is GSD type IIB, involving the gene LAMP2, X-linked dominant, prevalence 1:12,000 – 1:50,000 individuals. Symptoms include HCM, dilated cardiomyopathy (DCM), and musculoskeletal weakness. Treatment includes heart transplant.

Cori disease is GSD III, involving the gene PYGL, autosomal recessive, with incidence 1:100,000 births. Symptoms include low blood sugar, and enlarged liver; treatment involves diet. It is coded to E74.03.

Andersen disease is GSD type IV, involving the gene GBE, autosomal recessive, incidence 1:600,000 – 1:800,000 births. Symptoms include enlarged liver, fibrosis, and cirrhosis; treatment includes liver transplant. It is coded to E74.09 (not specific).

McArdle disease is GSD type V, involving gene PYGM, autosomal recessive, with prevalence 1:100,000 individuals. Symptoms include cramps and fatigue, and treatment involves diet and exercise. It is coded to E74.04.

No specific treatment is currently available for Danon disease[11]. Current management focuses on surveillance and symptom palliation, with heart transplantation provided when clinically indicated. There is currently a Danon disease-directed gene therapy treatment under investigation; a registrational trial is anticipated over the coming year.

Danon disease has unique monitoring and management needed to provide optimal patient care (e.g., alcohol septal ablation for HCM, which is not recommended for Danon disease per the 2020 AHA/ACC HCM Guidelines, or a patient being evaluated for heart transplant for exercise intolerance because skeletal myopathy has not yet been diagnosed).

A specific ICD-10-CM code for Danon disease will facilitate patient identification for appropriate treatments as well as for clinical trials, and will facilitate better understanding of prevalence, onset, and disease progression, as well as supporting further research in the evaluation and treatment of Danon disease.

A proposal to create a specific code for Danon disease was received from University of Colorado, Adult Medical Genetics Program.

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TABULAR MODIFICATIONS

E74 Metabolic Disorders

E74.0 Glycogen Storage Disorders

New Code	E74.05	Lysosome-associated membrane protein 2 [LAMP2] deficiency
Add		Danon disease

MED13L Syndrome

The MED13L Foundation is requesting a unique International Classification of Disease, 10th Revision, Clinical Modification (ICD-10-CM) code specific for MED13LSyndrome.

MED13L Syndrome is a genetic disorder with a world-wide prevalence estimated at over 1 million individuals.[1] The syndrome is "characterized by [moderate to severe] intellectual disability, heart malformation, and hypotonia."[2] Notably, MED13L Syndrome may present with severe cyanotic forms of congenital heart diseases and other congenital cardiac defects. In addition, MED13L Syndrome has been associated with mitochondrial dysfunction.[2] The condition is also characterized by significant speech impairment and dysmorphic facial features. "Most children with the syndrome [also] . . . take longer to learn to sit and walk independently (delayed motor skills)." [3] "Other features may include short stature, cleft palate, problems with coordination (ataxia), and recurrent seizures (epilepsy)."[3]

MED13L Syndrome is an autosomal dominant monogenic disorder diagnosed in children associated with the mutation and disruption of function of the MED13L(Mediator Complex Subunit 13L) gene, which is located on chromosome 12.[2] The MED13 gene is a component of the CDK8- kinase module, which can reversibly bind the Mediator complex, a multi-protein complex required for assembly and stabilization of the pre-initiation complex. Congenital malformations have been associated with thirty-two sub-units of the Mediator complex,[4] and the complex is "essential for transcription initiation….The core function of Mediator is to transmit signals from various transcription factors to RNA polymerase II (Pol II) . . . . Binding of the CDK8-module to Mediator has been reported to prevent the association of Mediator with the Pol II pre-initiation complex, thus preventing transcription initiation and/or re-initiation."[5] As such, MED13L "plays a role in the control of cell growth, repression of cell cycle target genes, and cell cycle inhibition."[6] Reported cases of MED13L Syndrome are overwhelmingly de novo (i.e., spontaneous) mutations.[4, 7, 14] Nevertheless, there are also reports of parental germline mosaicism (presence of mutation in some of the sperms/eggs) leading to more than one affected individual in the families.[7, 8] Approximately 92% of known cases involve de novo mutations, 3% involve hereditary mutations, and 5% of cases are of unknown inheritances.[9] "Although [different] MED13L mutations have been associated with this syndrome, a causative mechanism(s) for the multiple facets of [the clinical presentation of] this disorder has not [yet] been established."[2, 10]

A specific diagnosis code for MED13L Syndrome would provide research and public health institutions a more reliable way to track the epidemiology associated with MED13L Syndrome. Currently, clinicians cannot specifically characterize and report on MED13L Syndrome through existing diagnosis codes, which inhibits accurate data collection and

systematic tracking of incidence and outcomes. The result is MED13L Syndrome being categorized with other disorders that are phenotypically distinct.

Further, lack of a specific code can cause inaccurate coding. Indeed, a unique code is vital to allow epidemiologists and researchers to understand and reliably track the full prevalence of the condition, as well as associated rates of morbidity and mortality. This inhibits accurate reporting and can delay treatment—ultimately contributing to increased costs and burdens on the health system as a whole which impacts physicians, patients, and their families.

Due to deficits in identification and diagnosis—which are compounded by the absence of a unique diagnosis code—there are significant limitations on the current data about the incidence, prevalence, and epidemiology of MED13L Syndrome.[1] Further, as noted, the estimated worldwide prevalence of MED13L Syndrome is over 1 million individuals.[1] As such, although rare, MED13L Syndrome is significantly represented among birth cohorts relative to many other rare genetic conditions. It also presents in birth cohorts at comparable incidence rates as other rare conditions assigned ICD-10-CM codes, such as SYNGAP1.

Currently, there is no cure or pharmacologic treatment for MED13L Syndrome. Management of the syndrome generally involves speech, behavioral, and occupational therapy. Depending on the presentation of the syndrome, therapy could also involve treatment for epilepsy and management of congenital anomalies (e.g., congenital heart defects, cleft palates, etc.) if present.

Ongoing surveillance is typically part of effective management, including assessments to exclude for development of congenital heart defects, ophthalmologic examinations for eye problems, and monitoring for other clinical anomalies.[12] The MED13L Foundation is currently leading an effort to develop therapeutics for MED13L Syndrome, including the repurposing of small molecule drugs, de novo small molecule and biologic drugs, and genetic medicines such as anti-sense oligonucleotides, gene therapy, and gene-editing constructs. An ICD-10-CM code is critical to the continued development of these programs, in addition to facilitating more reliable public health tracking for the condition.

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TABULAR MODIFICATIONS

Q87 Other specified congenital malformation syndromes affecting multiple systems

Use additional code(s) to identify all associated manifestations

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

Excludes1:Zellweger syndrome (E71.510)

New code Q87.85 MED13L Syndrome
Add Asadollahi-Rauch Syndrome
Add Mediator complex subunit 13L syndrome

Add Code also, if applicable, any associated manifestations such as:
autism spectrum disorder (F84.0-)
congenital malformations of cardiac septa (Q21-)
epilepsy and recurrent seizures (G40.-)
intellectual disability (F70-F79)

Membranous Nephropathy

The National Kidney Foundation has requested new ICD-10-CM codes for membranous nephropathy to simplify and clarify diagnostic coding.

Membranous nephropathy (MN), a common cause of nephrotic syndrome in adults,[1] is characterized by thickening of the renal glomerular basement membrane (GBM), resulting from the accumulation of immune reactants in this structure. MN has a global incidence of 8-10 per million and approximately 12 cases per million in the U.S.[2]

MN patients typically present with overt nephrotic syndrome or non-nephrotic proteinuria, commonly with edema and/or hypertension.[3] Progression of nephrotic syndrome predicts higher risk of end-stage kidney disease (ESKD) and thrombo-embolic events.[3] Half of MN patients who present with nephrotic syndrome have a worsening disease progression, and about 30% of MN patients may develop ESKD. In untreated patients, 40-50% progress to ESKD and will require dialysis or renal transplantation.[2] These treatments are particularly burdensome on the patient and health care system, especially regarding transplantation, as the likelihood of MN recurrence following transplantation is common, leading to an increased risk of kidney failure and additional invasive therapies.[4]

MN is caused by autoantibodies directed against the M-type phospholipase A2 receptor (PLA2R)[5] that resides on glomerular podocytes, the cells which overlay the GBM. These antibodies react with podocyte PLA2R and initiate processes that lead to GBM thickening and glomerular injury in MN. Thus, MN is diagnosed by structural (renal biopsy) and/or serological (the presence in the blood or glomerulus of anti-PLA2R antibodies) criteria.

MN has been historically classified into two subtypes: "primary" and "secondary" forms of MN.[6]

1) Primary MN comprises approximately 75-80% of cases. Light microscopy typically shows uniform thickening of the GBM in primary MN. The Jones silver methenamine stain shows "spikes" of GBM material extending between "holes" of unstained immune deposits in the GBM,[7,8] which correspond to subepithelial deposits of IgG and C3 revealed by immunofluorescence microscopy.[7] There are significant correlations between the anti-PLA2R antibody found in primary MN and disease outcomes. High titers of these antibodies have been associated with severe disease progression while low titers have been found to increase the likelihood of remission after kidney transplantation.[7]

2) Other forms of MN comprise the remaining 20-25% of MN cases. Those cases with a recognizable underlying etiology (e.g., autoimmune diseases, diabetes, cancer, infections, non-steroidal anti-inflammatory drugs) are categorized as "secondary" MN.[3,9] Light microscopy shows endocapillary hypercellularity in secondary MN, especially in cases of malignancy.[7] Immunofluorescence reveals subepithelial deposits of IgG, including IgG1, IgG2, or -IgG3 in secondary MN cases.[7] Deposits of C3 and C1q are also present in secondary MN.[10] As the presence of anti-PLA2R is often low in these secondary forms of MN, disease outcomes often vary, and typically follow the outcomes associated with the etiologic cause in this form.[11]

Primary and secondary forms of MN have distinct treatment pathways. Initial treatment for primary MN involves antibody-targeted therapies, immunosuppressive agents, and supportive care for blood pressure control and proteinuria reduction. Initial treatment for secondary MN follows the treatment pathways of the underlying condition.[12] Since treatment is often targeted at the etiologic cause in secondary MN, differential diagnosis is critical to distinguish between the two forms.[11] To reflect the process and time it takes to achieve a specific diagnosis in MN, a code for unspecified MN is also necessary.

An advisory panel of expert stakeholders (including clinicians, researchers, and patient educator/advocates) have indicated that the term "membranous nephropathy" (MN) is generally used in clinical guidelines that describe this condition as well as being used in clinical practice (rather than the term "diffuse membranous glomerulonephritis").[12] This condition will generally present with either nephrotic syndrome or isolated proteinuria, and will be further classified as primary or secondary MN. Specific codes to identify these would enable tracking to be more clinically accurate, useful, and up-to-date with scientific advances. This proposal builds on a previous proposal submitted by the National Kidney Foundation in December 2021.

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TABULAR MODIFICATIONS

N04 Nephrotic syndrome

N04.2 Nephrotic syndrome with diffuse membranous glomerulonephritis

New code	N04.20	Nephrotic syndrome with diffuse membranous glomerulonephritis, unspecified
Add		Membranous nephropathy NOS with nephrotic syndrome
New code	N04.21	Primary membranous nephropathy with nephrotic syndrome
Add		Idiopathic membranous nephropathy with nephrotic syndrome
New code	N04.22	Secondary membranous nephropathy with nephrotic syndrome
Add		Code first, if applicable, other disease or disorder or poisoning causing membranous nephropathy
Add		Use additional code, if applicable, for adverse effect of drug causing membranous nephropathy
New code	N04.29	Other nephrotic syndrome with diffuse membranous glomerulonephritis

N06 Isolated proteinuria with specified morphological lesion

N06.2 Isolated proteinuria with diffuse membranous glomerulonephritis

New code	N06.20	Isolated proteinuria with diffuse membranous glomerulonephritis, unspecified
Add		Membranous nephropathy, NOS
Add		Excludes1: membranous nephropathy with nephrotic syndrome (N04.20)

New code	N06.21	Primary membranous nephropathy with isolated proteinuria
Add		Idiopathic membranous nephropathy (with isolated proteinuria)
Add		Primary membranous nephropathy, NOS
Add		Excludes1: primary membranous nephropathy with nephrotic syndrome (N04.21)
New code	N06.22	Secondary membranous nephropathy with isolated proteinuria
Add		Secondary membranous nephropathy, NOS
Add		Code first, if applicable, other disease or disorder or poisoning causing membranous nephropathy
Add		Use additional code, if applicable, for adverse effect of drug causing membranous nephropathy
Add		Excludes1: secondary membranous nephropathy with nephrotic syndrome (N04.22)
New code	N06.29	Other isolated proteinuria with diffuse membranous glomerulonephritis

Myelin Oligodendrocyte Glycoprotein Antibody Disease

Myelin oligodendrocyte glycoprotein (MOG) antibody disease (MOG-AD) is an inflammatory demyelinating condition of the central nervous system (CNS) characterized by a monophasic or relapsing course of neurological dysfunction, which does not meet the typical criteria for multiple sclerosis (MS) or other known neuroinflammatory conditions, and occurs in the presence of serum MOG antibodies detected using specific cell-based assays.[1] MOG-AD is often associated with neuromyelitis optica spectrum disorder (NMOSD). Although the clinical course and presentation for these diseases can be similar, there are some critical differences in pathophysiology, prognoses and outcomes. Recognition and identification of MOG-AD versus other neuroinflammatory conditions, such as NMOSD, is critical to ensure appropriate clinical evaluation, treatment plans and optimal outcomes for MOG-AD patients. It has been proposed that a specific ICD-10-CM code for MOG-AD will facilitate research and data collection as well as improve diagnosis and patient care. This proposal is based on a submission from Jonathan D. Santoro, MD, Division of Neurology, Department of Pediatrics, Children's Hospital Los Angeles, Keck School of Medicine at USC and The MOG Project.

MOG-AD can occur in all decades of life, with a slight predominance in women and with median age of onset in the early to mid-thirties. The most common presenting feature is optic neuritis (ON), occurring in 54–61% of patients, followed by myelitis, acute disseminated encephalomyelitis (ADEM) or an ADEM-like presentation (e.g., brainstem attack).[1] A relapsing course has been reported in 44–83% of patients and more commonly involves the optic nerve. MOG-positive ON is frequently bilateral and associated with optic nerve head swelling.[1]

A large body of immunologic and clinical evidence has made clear that MOG-AD is a distinct entity.[2] The autoantibodies in MOG-AD have a unique target and the disease has unique immunologic mechanisms that distinguish it from other conditions, such as aquaporin-4 (AQP-4) - positive NMOSD and MS.[3-4] While MOG-AD was initially believed to represent a subset of patients with neuromyelitis optica (NMO), it is now clear that this is not the case. MOG is located on the surface of myelin sheaths and oligodendrocyte processes, whereas AQP-4 water channels, the target of seropositive NMO, are located on astrocytic foot processes.[5] It has also been shown that about one-third of demyelinating lesions in patients with MOG-AD have MOG-dominant myelin loss, but relatively preserved oligodendrocytes. This differs from AQP-4-positive NMOSD lesions, which exhibit myelin-associated glycoprotein-dominant oligodendrogliopathy.[6] In addition, complement activation plays a significant role in AQP-4-IgG-positive NMOSD and is a potential treatment target,[7-8] but appears to be less important in MOG-AD.[6] Most recently, a new neutrophil granulocyte-specific biomarker has been shown to have high sensitivity and specificity for rapid differentiation among patients with NMOSD, MOG-AD, and relapsing-remitting MS.[9]

Differences in pathology of MOG-AD, MS, and AQP-4-positive NMOSD result in distinct clinical symptoms and prognoses. MOG-IgG optic neuritis has a higher likelihood of being recurrent, bilateral, associated with intervertebral disc edema, and have perineural enhancement around the optic nerve on magnetic resonance imaging in contrast to demyelinating optic neuritis arising from MS.[4] In addition, the overall course of optic neuritis in MOG-AD differs from that in MS patients. MS patients with optic neuritis generally have unilateral involvement and occurrence only once

during the course of their disease, while involvement in MOG-AD is bilateral and relapsing.[2,10-12] Recovery from attacks in MOG-AD is typically better than that seen in AQP-4-positive NMOSD.[2] In addition, ADEM is a relatively common presentation in pediatric patients with MOG-AD, but is not seen in either MS or AQP-4-IgG-positive NMOSD.[4,13] The prognoses for MOG-AD and AQP-4-IgG-positive NMOSD also differ, with much higher risk for fatality and permanent disability with the latter condition.[4,14]

There is a very clear consensus in the clinical literature that MOG-AD is a distinct pathologic and clinical entity with a unique disease course and management requirements.[6, 14-18] Patients with MOG-AD exhibit unique characteristics. Although various data points such as clinical picture, severity and antibody titers are established, prognostic factors for MOG-AD are still lacking. For example, physicians have difficulty predicting whether a patient will relapse. For patients who do relapse, the consensus is to provide maintenance therapy, but there is currently no clinical consensus on what treatments are the most effective.

Inflammatory demyelinating CNS diseases are a heterogeneous group, and although the sensitivity and specificity of diagnostic criteria have improved, misdiagnosis is not infrequent and occurs in up to 10% of cases.[19] Results from one study showed that 9% of patients with ADEM according to clinical criteria were misdiagnosed, since the pathology was similar to that of MS.[20] This is particularly concerning, given that certain treatment protocols used for other similar disorders like MS have been found to exacerbate MOG-AD symptoms.

A study in the UK found an incidence rate for MOG-AD of about 3.4/1 000 000 person-years.[21] This compared to a nationwide incidence rate of about 1.6/1 000 000 persons in Holland (from 2014 to 2017), with a higher rate in children (of 3.1/1 000 000, vs 1.3/1 000 000 in adults).[21]

An individual code for MOG-AD will enable better identification and tracking of this distinct set of patients, which will advance the clinical understanding of MOG-AD, and subsequently enable improvement in the diagnostic and treatment paradigms and facilitate research and data collection. Increased awareness of MOG-AD will help to decrease diagnostic delay in these patients, which may potentially decrease the likelihood of permanent disability, such as blindness and paralysis. Furthermore, accurate identification is critical to facilitate research that will further elucidate the marked needs and characteristics of MOG-AD, as well as the development of effective treatment for MOG-AD.

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TABULAR MODIFICATIONS

G37 Other demyelinating diseases of central nervous system

G37.8 Other specified demyelinating diseases of central nervous system

New code	G37.81	Myelin Oligodendrocyte Glycoprotein Antibody Disease
Add		Code also associated manifestations, such as:
Add		neuromyelitis optica (G36.0)
Add		noninfectious acute disseminated encephalomyelitis (G04.81)

Nontraumatic Coma Due to Underlying Condition

This is a representation from the September 2021 and March 2022 ICD-10 Coordination and Maintenance Committee Meetings of the Coma Due to Underlying Condition proposal with the recommended modification to add "nontraumatic" to the proposed code title R40.2A, Coma due to underlying condition. The modification is in **bold.**

After the recent coding guideline changes which limits Glasgow coma scale codes to traumatic brain injury (TBI), the National Center for Health Statistics received a proposal for the creation of a new ICD-10-CM code for "Coma NEC."

R40.20, Unspecified Coma, is the only code available for coma in patients who do not have TBI but have conditions without combination codes describing the coma; for example, coma secondary to spontaneous brain hemorrhage.

TABULAR MODIFICATIONS

R40 Somnolence, stupor and coma

Excludes1: neonatal coma (P91.5)
somnolence, stupor and coma in diabetes (E08-E13)
somnolence, stupor and coma in hepatic failure (K72.-)
somnolence, stupor and coma in
hypoglycemia (nondiabetic) (E15)

R40.2 Coma
Code first any associated:
fracture of skull (S02.-)
intracranial injury (S06.-)

Note: One code from each subcategory, R40.21-R40.23, is required to complete the coma scale

New code R40.2A **Nontraumatic** coma due to underlying condition
Add Secondary coma

Add Code first underlying condition

Obesity in Children, Adolescents, and Adults

Obesity is a highly prevalent chronic disease with complex inflammatory and endocrinological pathophysiology.[1] The American Medical Association has recognized obesity as a disease since 2013.[2] Obesity is associated with serious health and social consequences.[3] Childhood obesity is defined by a body mass index (BMI = body weight in kilograms divided by height in meters squared (kg/m^2)) $\geq 95^{th}$ percentile for age and sex.[4] Currently, approximately 1 in 5 U.S. children have obesity.[5] Furthermore, the proportion of children and adolescents 2-19 years with severe obesity (BMI $\geq$120% above the 95^{th} percentile) in 2015-2018 was 7.6%.[6] Obesity in childhood predisposes to obesity in both adolescence and adulthood.[7]

Obesity puts children and adolescents at risk for serious short- and long-term adverse health outcomes later in life, including cardiovascular disease (CVD), hypertension (HTN), dyslipidemia, insulin resistance, type 2 diabetes mellitus (T2DM), obstructive sleep apnea (OSA), obesity-related glomerulopathy (ORG), and non-alcoholic fatty liver disease (NAFLD).[8-12] In addition to physical and metabolic consequences, obesity in childhood and adolescence is associated with poor psychological and emotional health, stigmatization, bullying, increased stress, depressive symptoms, and low self-esteem.[13] Significantly, the severity of comorbidities increases with increasing adiposity.[8] Furthermore, stigmatization, depression and low self-esteem contribute to binge eating, social isolation, avoidance of health care services, and decreased physical activity.[14]

Obesity in childhood and adolescence is associated with increased health care utilization and costs.[15] In an analysis of the National Inpatient Sample database from 2006-2016, the most common conditions that co-occur with a diagnosis of obesity and increase costs and utilization included mood disorders, asthma, and diabetes.[15]

Interventions to manage and treat childhood obesity consist of *family-based*, intensive and comprehensive lifestyle interventions.[16] The United States Preventive Services Task Force (USPSTF) recognizes that weight management or lifestyle interventions that include nutritional and physical activity counseling and deliver $\geq$26 hours in a 6-month period as being effective in improving weight status among children and adolescents as well as cardiovascular risk factors.[16] These interventions have been shown to be cost-effective with important health benefits to caregivers as well.[16, 17]

Obesity in children and adolescents is determined by age- and gender-specific percentiles. Therefore, a child or adolescent may suffer from obesity at a lower BMI than an adult. For adults, the overweight range is from a BMI of 25.0 to <30. Obesity in adults is subdivided into the following: Class 1: BMI of 30 to < 35; Class 2: BMI of 35 to < 40; Class 3: BMI of 40 or higher (sometimes categorized as "severe" obesity).[18] Obesity in children uses a classification system recognizing BMI ≥95th percentile as class I obesity, BMI ≥120% of the 95th percentile as class II obesity, and BMI ≥140% of the 95th percentile as class III obesity.[19]

Previous etiological hypotheses around obesity suggested an imbalance in caloric intake. This understanding is no longer accepted, and obesity is understood to be a complex, inflammatory

disease.[1] Therefore, identifying excess calories as the cause of obesity does not reflect current medical understanding.

Providers and medical associations recognize that children and adolescents with obesity, and their families, experience stigmatization that can negatively affect health-seeking behavior and outcomes.[14] This stigmatization may come directly from providers and includes terminology such as "morbid" and assumptions that obesity is due to personal choices relating to caloric consumption. Using people-first language, e.g., "children *with obesity"* rather than "obese children," is an important written and verbal communication strategy to avoid stigmatization.[14] Use of language that may cause stigmatization, such as "morbid" or "excess calories," or the use of non-people-first language may impact how providers document their services.

Accordingly, these difficulties may impact how obesity is diagnosed, classified, managed, and tracked over time in a clinical setting. Several studies have determined that current ICD codes have low sensitivity in identifying obesity and may impact clinical care by, for example, lowering referral rates to specialists and weight management programs.[20] These shortcomings may also have serious implications for current and future health of the growing population of children and adolescents suffering from obesity.[5,15]

Proposed changes to the ICD-10-CM obesity codes have been received from CDC, the Division of Nutrition, Physical Activity and Obesity of the National Center for Chronic Disease Prevention and Health Promotion with further input from additional obesity experts.

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TABULAR MODIFICATIONS

E66 Overweight and obesity

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

Use additional code to identify body mass index (BMI), if known (Z68.-)

Delete ~~Excludes1: adiposogenital dystrophy (E23.6)~~
~~lipomatosis NOS (E88.2)~~
~~lipomatosis dolorosa [Dercum] (E88.2)~~
~~Prader-Willi syndrome (Q87.11)~~

Add Excludes2: adiposogenital dystrophy (E23.6)
Add lipomatosis NOS (E88.2)
Add lipomatosis dolorosa [Dercum] (E88.2)
Add Prader-Willi syndrome (Q87.11)

E66.0 Obesity due to excess calories

Revise E66.01 Extreme ~~Morbid (severe)~~ obesity due to excess calories

Revise Excludes1: extreme ~~morbid (severe)~~ obesity with alveolar hypoventilation (E66.2)

Revise E66.2 Extreme ~~Morbid (severe)~~ obesity with alveolar hypoventilation
Obesity hypoventilation syndrome (OHS)
Pickwickian syndrome

E66.8 Other obesity

New sub-subcategory	E66.81	Obesity in children and adolescents	
Add		Use additional code to identify body mass index (BMI), pediatric, if known (Z68.5-)	
New code		E66.811	Obesity in children and adolescents, class 1
New code		E66.812	Obesity in children and adolescents, class 2
New code		E66.813	Obesity in children and adolescents, class 3
Add			Severe obesity in children and adolescents
New code		E66.819	Obesity in children and adolescents, unspecified
New sub-subcategory	E66.82	Obesity in adults	
Add		Use additional code to identify body mass index (BMI), adult, if known (Z68.1-Z68.45)	
New code		E66.821	Obesity in adults, class 1
New code		E66.822	Obesity in adults, class 2
New code		E66.823	Obesity in adults, class 3
Add			Severe obesity (in adults)
New code		E66.829	Obesity in adults, unspecified
New code	E66.89	Other obesity	

INDEX MODIFICATIONS

	Obesity E66.9
Add	- extreme (see also Obesity, severe) E66.823
Add	- - with
Add	- - - alveolar hypoventilation E66.2
Add	- - - obesity hypoventilation syndrome (OHS) E66.2
Add	- - due to excess calories E66.01
Revise	- morbid (see also Obesity, severe) E66.823 ~~E66.01~~
Revise	- severe E66.823 ~~E66.01~~
Add	- - in adolescents E66.813
Add	- - in adults E66.823
Add	- - in children E66.813
Revise	- specified type NEC E66.89

Phelan-McDermid Syndrome

Phelan-McDermid syndrome was recognized as a distinct diagnosis in the 1990's with the first case report published in the medical literature in 1992. Often abbreviated as PMS, it is a genetic neurodevelopmental condition with multi-system manifestations.

The genetic cause of Phelan-McDermid syndrome has been established to be defects in a specific portion of the long arm of chromosome 22. In about 80% of individuals, the cause is a deletion in a segment of chromosome 22 known as q13.3. For this reason, Phelan-McDermid syndrome was initially named 22q13.3 deletion syndrome. Less commonly, in about 20% of cases, Phelan-McDermid syndrome is caused by pathogenic variants in the *SHANK3* gene located at the distal long arm of chromosome 22.

Signs and symptoms of Phelan-McDermid syndrome are often seen in infancy and early childhood. Classically, these include low muscle tone in infants, delays in reaching developmental milestones, delayed speech, or inability to develop functional speech, and global developmental delay leading to varying degrees of intellectual disability. Certain minor dysmorphic features are also characteristic of Phelan-McDermid syndrome, such as elongation of the head, full brow, deep-set eyes, wide nasal bridge and bulbous nose, prominent ears, and large, fleshy hands.

As children with Phelan-McDermid syndrome age, other disorders and symptoms are often seen, although the specific symptoms vary with the individual and may occur only in certain subgroups or at specific stages in life. Common disorders and symptoms include seizures, gastrointestinal disorders such as cyclic vomiting and constipation, sleep difficulties, lymphedema, and neuropsychiatric illness. A significant minority have kidney abnormalities such as multicystic kidneys and vesicoureteral reflux. Of note, people with Phelan-McDermid syndrome often have decreased perception of pain as well as low levels of sweating, putting them at risk for unrecognized injury and overheating.

Phelan-McDermid syndrome is also known to have a strong association with autism. It is estimated that approximately 75-80% of people with Phelan-McDermid syndrome also have autism spectrum disorder. Conversely, it is estimated that approximately 1% of people with autism have Phelan-McDermid syndrome.

From its association with autism, it is believed that the prevalence of Phelan-McDermid syndrome is approximately 1 in 10,000 to 15,000 people. However, this is an extrapolation and may be understated because of the challenges in identifying the diagnosis distinctly in healthcare databases.

The diagnosis of Phelan-McDermid syndrome is made definitively by genetic testing. Chromosomal microarray analysis is the first line of testing and is required for diagnosis in most cases to detect a deletion. Whole exome/genome sequencing is required for detecting sequence variants in *SHANK3*. As technologies in genetic sequencing continue to become less costly and more widely used, the known prevalence of Phelan-McDermid syndrome is likely to continue to increase.

There is currently no treatment for the underlying genetic basis of Phelan-McDermid syndrome. Care is directed at managing the symptoms and risks for each affected individual. Families often see teams of specialists, including a neurologist, pediatrician or primary care physician, gastroenterologist, nephrologist, endocrinologist, and psychiatrist, as well as a speech-language pathologist and behavioral, occupational, and physical therapists.

There are currently three clinical trials related to Phelan-McDermid syndrome at various stages in the U.S. Three other treatment trials were recently completed in the U.S.

Given that the main cause of Phelan-McDermid syndrome is a deletion in chromosome 22, subcategory Q93.5 is an appropriate location for a new code. This is also consistent with WHO ICD-11, which categorizes Phelan-McDermid syndrome under code LD44, Deletions of the autosomes.

In addition to aiding in identifying the true prevalence of Phelan-McDermid syndrome, a specific ICD-10-CM code would enable tracking the disorder, calculating the public health impact, assist in clinical trials and retrospective research.

This proposal is submitted by the Phelan-McDermid Syndrome foundation.

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See also: https://pmsf.org/about-pms/

TABULAR MODIFICATIONS

Q93 Monosomies and deletions from the autosomes, not elsewhere classified

Q93.5 Other deletions of part of a chromosome

Q93.51 Angelman syndrome

New code Q93.52 Phelan-McDermid syndrome
Add 22q13.3 deletion syndrome

Add Use additional code(s) to identify any associated conditions, such as:
Add autism spectrum disorder (F84.0)
Add degree of intellectual disabilities (F70-F79)
Add epilepsy and recurrent seizures (G40.-)
Add lymphedema (I89.0)

Q93.59 Other deletions of part of a chromosome

Short Bowel Syndrome and Intestinal Failure

This topic was presented at the September 2020 Coordination and Maintenance Meeting. This proposal has been revised to divide short bowel syndrome (SBS) into those with and without colon in continuity with the residual small bowel. SBS with colon in continuity is when the colon has been anastomosed to residual small bowel. This includes ileocolonic and jejunocolonic anastomoses. SBS with no colon in continuity is when all colon has been resected, or otherwise is not in continuity with the residual small bowel. This includes mucus fistula, ileostomy, jejunostomy, duodenostomy patients and jejuno/ileo-rectal anastomosis that meet the definition for SBS.

Short bowel syndrome (SBS) is a condition that occurs when your body is unable to absorb enough nutrients from the foods you eat because you do not have enough small intestine. Short bowel syndrome is caused by the physical absence or loss of massive portions of intestine (typically to < 200 cm of residual intestine). Many, but not all individuals with SBS may also develop intestinal failure (IF), which is the inability to absorb enough nutrients and/or fluid necessary to maintain nutritional autonomy. Conversely, not all patients with IF suffer from SBS, but may have a myriad of different malabsorptive disorders. Treatment of patients with SBS and IF is complex, including the management of intravenous nutrition and fluids, and the prevention and treatment of nutrient deficiencies and dehydration. Additional complications of the disease can affect the liver, kidney, brain, and bones. It is likely that lack of knowledge and understanding of both SBS and IF has led to misclassification of these diseases under various identifiers, thereby commonly reported prevalence numbers may be either over- or under-representations of actual disease prevalence.

In patients with short bowel syndrome, the colon assumes a substantially greater role than normal with regard to both fluid and nutrient absorption than in individuals with a fully intact digestive tract. The colon absorbs fluid, electrolytes, some amino acids and medium chain triglycerides (MCT), but most importantly, through fermentation of unabsorbed carbohydrates by what is termed “carbohydrate salvage,” the colon becomes a factory for energy production through the production of short chain fatty acids. The amount of energy produced may amount to as much as 1000 kcal daily.

Individuals with short bowel syndrome who have no residual colon after resection have a poorer prognosis, respond to a lesser degree to dietary therapy, and are more likely to require long-term or permanent intravenous feeding than those individuals who have colon in continuity. This may relate largely to a decrease in glucagon-like peptide 2 (GLP-2) production in the former.

Creating a unique ICD-10-CM codes for SBS would facilitate better care via (a) exposing regional variations and areas for improvements in care, and (b) ultimately, enabling continuity of care by tracking patients across systems and optimizing standards of care.

Alan Buchman, MD, MSPH is proposing the following tabular modifications for better delineation in these conditions. **Changes in bold.**

TABULAR MODIFICATIONS

K90 Intestinal malabsorption

K90.8 Other intestinal malabsorption

New subcategory — K90.82 Short bowel syndrome

New code — K90.821 Short bowel syndrome with colon in continuity
New code — K90.822 Short bowel syndrome without colon in continuity
New code — K90.829 Short bowel syndrome, unspecified

New code — K90.83 Intestinal failure

Sickle-Cell Retinopathy

Sickle-cell disease is the most common inherited blood disorder. Sickle cell retinopathy is characterized by the blockage of outer retinal vessels, resulting in both non-proliferative and proliferative retinopathy, that can lead to complications such as vision impairment and blindness. Treatment of sickle cell retinopathy includes observation, retinal ablation, advanced retinal-vitreal surgery and the intravitreal injection of anti-VEGF biologic agents to control the proliferative retinopathy. The expanding use of intravitreal biologic agents is changing the prognosis for sickle-cell retinal disease.

Bevacizumab has been found to allow patients to maintain vision without the need for more invasive interventions such as vitrectomy to clear vitreous hemorrhage or to repair retinal detachment. At present there is no specific ICD-10-CM diagnostic code for proliferative sickle cell retinopathy. This absence makes it difficult to determine medical necessity of management. Specific reporting is also important from a public health perspective to be able to study the prevalence of this specific blinding disease in the US population. Retinal issues occur in the same eye or in fellow eyes of sickle-cell patients, either synchronously or asynchronously. Currently physicians and facilities use the following non-specific code, H35.2-, Other nondiabetic proliferative retinopathy.

There is no synonym identifying sickle cell retinal disease in the current ICD-10-CM. While including synonym(s) would be an improvement in ICD-10-CM there are several limitations to this approach. First, it would not allow the public health evaluation and study of this specific retinal disease. Second, it does not specify the proliferative form of the disease as compared with background retinopathy. Third, non-diabetic proliferative retinopathies have differing treatments based on ocular or systemic diagnosis. To that goal new diagnostic codes are being requested for ICD-10-CM.

The American Academy of Ophthalmology is proposing the following tabular modifications to report treatment accurately and to support the need for disease-specific reporting of sickle cell proliferative retinopathy.

TABULAR MODIFICATIONS

H35 Other retinal disorders

H35.2 Other non-diabetic proliferative retinopathy
Proliferative vitreo-retinopathy

Add Thalassemia proliferative retinopathy

Add Excludes2: Proliferative sickle-cell retinopathy (H36.82-)

H36 Retinal disorders in diseases classified elsewhere
Code first underlying disease, such as:
lipid storage disorders (E75.-)
sickle-cell disorders (D57.-)

Excludes1: arteriosclerotic retinopathy (H35.0-)
Diabetic retinopathy (E08.3-, E09.3-, E10.3-, E11.3-, E13.3-)

New subcategory	H36.8	Other retinal disorders in diseases classified elsewhere
New sub-subcategory	H36.81	Non proliferative sickle-cell retinopathy
New code		H36.810 Non proliferative sickle cell retinopathy, unspecified eye
New code		H36.811 Non proliferative sickle cell retinopathy, right eye
New code		H36.812 Non proliferative sickle cell retinopathy, left eye
New code		H36.813 Non proliferative sickle cell retinopathy, bilateral
New sub-subcategory	H36.82	Proliferative sickle-cell retinopathy
New code		H36.820 Proliferative sickle cell retinopathy, unspecified eye
New code		H36.821 Proliferative sickle cell retinopathy, right eye
New code		H36.822 Proliferative sickle cell retinopathy, left eye
New code		H36.823 Proliferative sickle cell retinopathy, bilateral
New code	H36.89	Other retinal disorders in diseases classified elsewhere
Add		Retinal dystrophy in lipid storage disorders

Social Determinants of Health

This proposal was originally submitted by the Gravity Project (GP) and presented at the March 2021 and September 2021 and March 2022 ICD-10 Coordination and Maintenance (C&M) meetings. The American Academy of Pediatrics (AAP) also had several code requests that were presented at the September 2021and March 2022 C&M meeting. Parts of the proposal were previously approved and will be implemented on October 1, 2022. The files are posted on the CDC webpage Comprehensive Listing ICD-10-CM Files (cdc.gov). **This proposal will be considered for April 1, 2023, implementation.**

Subsequently, we have received additional code requests from the GP and CMS, as well as modifications to AAP's previously presented code request for Z91.1Patient's noncompliance with medical treatment and regimen expansion that have been incorporated in the proposal and are **bold**.

One of the five overarching goals of Healthy People 2030 is 'attaining health literacy to improve the health and well-being of all". Included in this goal is the adoption of the following definition. Personal health literacy is the degree to which individuals have the ability to find, understand, and use information and services to inform health-related decisions and actions for themselves and others

New ICD-10-CM codes are being requested at the T74.A- and the T76.A- previously expanded codes to include Child financial abuse, confirmed and Child financial abuse, suspected. There is also a request for inclusion terms under codes T74.3 and T76.3 to describe patients who identify that they have been threatened with harm. Receiving threats is a subtype of psychological abuse within the American Psychological Association.

The proposed Y07.0 Spouse or partner, perpetrator of maltreatment and neglect is being expanded as requested by comments from the March 2022 meeting.

A request for a new term under Z58 to cover basic necessities unavailable in the environment, Z58.81 Services unavailable in physical environment. Additional code requests were received to expand code Z59.1 Inadequate housing to further describe housing inadequacy.

Health Insurance Coverage, Healthy People 2030 includes several objectives that relate to improving the proportion of people with some form of health or dental insurance or reducing the proportion of people under 65 who are uninsured. Earlier this year, the Centers for Medicare and Medicaid Services (CMS) unveiled an initiative to reduce the uninsured rate among children and increase Medicaid enrollment for parents and pregnant people. Lack of insurance affects access to care and preventative services as well as an increase in mortality. Although one could say that the current code Z59.7 Insufficient social insurance and welfare support applies, our consensus statement is that it is not specific enough to support this

important use case. Social insurance is a broad-based term and welfare support can include different types of assistance programs outside of insurance coverage. The Gravity Project community determined that a specific code to identify insufficient health insurance coverage is needed to properly identify this situation.

New codes and revisions have been made to further describe circumstances related to food insecurity, housing instability, transportation needs, utility difficulties, and interpersonal safety to illuminate their impact on health outcomes. These health-related social needs (HRSNs), defined as individual-level, adverse social conditions that negatively impact a person's health or healthcare, are significant risk factors associated with worse health outcomes as well as increased healthcare utilization. Assessment of HRSNs is an essential mechanism for capturing the interaction between social, community, and environmental factors associated with health status and health outcomes. While widespread interest in addressing HRSNs exists, action is inconsistent, with 92 percent of hospitals screening for one or more of the five HRSNs, but only 24 percent of hospitals screening for all five HRSNs. Utilization of screening tools to identify the burden of unmet HRSNs can be a helpful first step in identifying necessary community partners and connecting individuals to resources in their communities. For data collection of responses to screening tools, there is a need to better track data elements of the HRSN domains.

References

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TK Fraze, AL Brewster, VA Lewis, LB Beidler, GF Murray, CH Colla. Prevalence of screening for food insecurity, housing instability, utility needs, transportation needs, and interpersonal violence by US physician practices and hospitals. JAMA Network Open 2019; 2:e1911514.10.1001/jamanetworkopen.2019.11514.31532515.

TABULAR MODIFICATIONS

T74 Adult and child abuse, neglect, and other maltreatment, confirmed

T74.3 Psychological abuse, confirmed
Bullying and intimidation, confirmed
Intimidation through social media, confirmed
Add **Target of threatened harm, confirmed**
Add **Target of threatened physical violence, confirmed**
Add **Target of threatened sexual abuse, confirmed**

New subcategory T74.A Financial abuse, confirmed
New code T74.A1 Adult financial abuse, confirmed
New code **T74.A2 Child financial abuse, confirmed**

T76 Adult and child abuse, neglect, and other maltreatment, suspected
T76.3 Psychological abuse, suspected
Bullying and intimidation, suspected
Intimidation through social media, suspected
Add **Target of threatened harm, suspected**
Add **Target of threatened physical violence, suspected**
Add **Target of threatened sexual abuse, suspected**

New subcategory T76.A Financial abuse, suspected
New code T76.A1 Adult financial abuse, suspected
New code **T76.A2 Child financial abuse, suspected**

Y07 Perpetrator of assault, maltreatment and neglect
Note: Codes from this category are for use only in cases of confirmed abuse (T74.-)
Selection of the correct perpetrator code is based on the relationship between the perpetrator and the victim

Add Includes: **perpetrator of verbal abuse**

Y07.0 Spouse or partner, perpetrator of maltreatment and neglect
Spouse or partner, perpetrator of maltreatment and neglect against spouse or partner

Y07.01 Husband, perpetrator of maltreatment and neglect
New code **Y07.010 Husband, current, perpetrator of maltreatment and neglect**

New code **Y07.011 Husband, former, perpetrator of maltreatment and neglect**

Y07.02 Wife, perpetrator of maltreatment and neglect
New code **Y07.020 Wife, current, perpetrator of maltreatment and neglect**
New code **Y07.021 Wife, former, perpetrator of maltreatment and neglect**

Y07.03 Male partner, perpetrator of maltreatment and neglect
Add **Intimate or dating partner, perpetrator of maltreatment and neglect**

New code		**Y07.030 Male partner, current, perpetrator of maltreatment and neglect**
New code		**Y03.031 Male partner, former, perpetrator of maltreatment and neglect**
	Y07.04	**Female partner, perpetrator of maltreatment and neglect**
Add		**Intimate or dating partner, perpetrator of maltreatment and neglect**
New code		**Y07.040 Female partner, current, perpetrator of maltreatment and neglect**
New code		**Y07.041 Female partner, former, perpetrator of maltreatment and neglect**
New code	**Y07.05**	**Non-binary partner, perpetrator of maltreatment and neglect**
Add		**Gender non-conforming partner, perpetrator of maltreatment and neglect**
New code		**Y07.050 Non-binary partner, current, perpetrator of maltreatment and neglect**
New code		**Y07.051 Non-binary partner, former, perpetrator of maltreatment and neglect**
	Y07.4	**Other family member, perpetrator of maltreatment or neglect**
New Code	**Y07.44**	**Child (biological, step, in-law, foster, adopted), perpetrator ofmaltreatment, and neglect**
Add		**Daughter, perpetrator of maltreatment, and neglect**
Add		**Non-binary child, perpetrator of maltreatment, and neglect**
Add		**Son, perpetrator of maltreatment, and neglect**
New Code	**Y07.45**	**Grandchild (biological, step, in law, foster, adopted), perpetrator of maltreatment, and neglect**
Add		**Granddaughter, perpetrator of maltreatment, and neglect**
Add		**Grandson, perpetrator of maltreatment, and neglect**

Add **Non-binary grandchild, perpetrator of maltreatment, and neglect**

New Code **Y07.46 Grandparent, perpetrator of maltreatment, and neglect**
Add **Grandfather, perpetrator of maltreatment, and neglect**
Add **Grandmother, perpetrator of maltreatment, and neglect**
Add **Non-binary grandparent, perpetrator of maltreatment, and neglect**

New Code **Y07.47 Parental sibling, perpetrator of maltreatment and neglect**
Add **Aunt, perpetrator of maltreatment and neglect**
Add **Non-binary parental sibling, perpetrator of maltreatment or neglect**
Add **Uncle, perpetrator of maltreatment and neglect**

Y07.49 Other family member, perpetrator of maltreatment and neglect
Y07. 499 Other family member, perpetrator of maltreatment and neglect
Add **Excludes2: parental sibling, perpetrator of maltreatment and neglect (Y07.47)**

Y07.5 Non-family member, perpetrator of maltreatment and neglect

New Code **Y07.54 Acquaintance or friend, perpetrator of maltreatment and neglect**

Z55 Problems related to education and literacy

New code **Z55.6 Problems related to health literacy**
Add **Difficulty understanding health related information**
Add **Difficulty understanding medication instructions**
Add **Problem completing medical forms**

Z58 Problems related to physical environment
New subcategory Z58.8 Other problems related to physical environment
New code Z58.81 **Basic services unavailable in** physical environment

Add — Unable to obtain **utilities**, due to inadequate physical environment
Add — Unable to obtain internet service, due to **unavailability in geographic area**
Add — **Unable to obtain telephone service, due to unavailability in geographic area**

New code — Z58.89 Other problems related to physical environment

Z59 Problems related to housing and economic circumstances
Z59.1 Inadequate Housing
Delete — ~~**Lack of heating**~~
Delete — ~~**Restriction of space**~~
Delete — ~~**Technical defects in home preventing adequate care**~~
Delete — ~~**Unsatisfactory surroundings**~~

New code — **Z59.10 Inadequate housing, unspecified**
Add — **Inadequate housing, NOS**

New code — **Z59.11 Inadequate housing**
Add — **Pest infestation**
Add — **Restriction of space**
Add — **Technical defects in home preventing adequate care**
Add — **Unsatisfactory surroundings**

New code — **Z59.12 Inadequate environmental temperature**
Add — **Lack of air conditioning**
Add — **Lack of heating**

New code — **Z59.13 Inadequate utilities**
Add — **Lack of electricity services**
Add — **Lack of gas services**
Add — **Lack of oil services**
Add — **Lack of water services**

Add — **Excludes2: lack of adequate food (Z59.4-)**
Add — **other problems related to housing and economic circumstances (Z59.8-)**

Z59.7 Insufficient social insurance and welfare support
Add — **Inadequate social and welfare insurance**
Add — **Insufficient social and welfare insurance**

New code — **Z59.70 Insufficient social insurance and welfare support, unspecified**

New code **Z59.71 Insufficient health insurance coverage**
Add **No health insurance coverage**

New code **Z59.79 Other insufficient social insurance and welfare support**

Z59.8 Other problems related to housing and economic circumstances

Z59.81 Housing instability, housed
Foreclosure on home loan
Past due on rent or mortgage
Unwanted multiple moves in the last 12 months

Z59.811 Housing instability, housed, with risk of homelessness
Imminent risk of homelessness

Z59.812 Housing instability, housed, homelessness in past 12 months

Z59.819 Housing instability, housed unspecified

Add **Excludes2: extreme poverty (Z59.5)**
Add **financial insecurity (Z59.86)**
Add **low income (Z59.6)**
Add **material hardship, not elsewhere classified (Z59.87)**

Z59.82 Transportation insecurity
Add **Excludes2: unavailability and inaccessibility of health-care facilities (Z75.3)**

Revise Z59.87 Material hardship, **due to limited financial resources, not elsewhere classified**
Revise Material deprivation **due to limited financial resources**
Revise Unable to obtain adequate childcare **due to limited financial resources**
Revise Unable to obtain adequate clothing **due to limited financial resources**
Revise Unable to obtain adequate utilities **due to limited financial resources**
Revise Unable to obtain basic needs, **due to limited financial resources**

Z60 Problems related to social environment
Z60.4 Social exclusion and rejection
Exclusion and rejection on the basis of personal characteristics, such as unusual physical appearance, illness or behavior
Add **Social isolation**

Z62 Problems related to upbringing
Z62.8 Other specified problems related to upbringing
Z62.81 Personal history of abuse in childhood

New code **Z62.814 Personal history of child financial abuse**

New code **Z62.815 Personal history of intimate partner abuse in childhood**

Z91 Personal risk factors, not elsewhere classified
Z91.1 Patient's noncompliance with medical treatment and regimen
Add **Code also, if applicable, to identify underdosing of specific drug (T36-T50 with final character 6)**

New sub-category Z91.14 Patient's other noncompliance with medication regimen
Patient's underdosing of medication

New code Z91.141 Patient’s other noncompliance with medication regimen due to financial hardship

New code Z91.148 Patient’s other noncompliance with medication regimen for other reason

New sub-category Z91.15 Patient's noncompliance with renal dialysis
New code Z91.151 Patient's noncompliance with renal dialysis due to financial hardship

New code Z91.158 Patient's noncompliance with renal dialysis for other reason

Z91.4 Personal history of psychological trauma, not elsewhere classified
Z91.41 Personal history of adult abuse

New code Z91.413 Personal history of adult financial abuse

New code **Z91.414 Personal history of adult intimate partner abuse**

INDEX MODIFICATIONS

	Maltreatment
	- adult
Add	- -threatened abuse (harm)
Add	- - - confirmed T74.31
Add	- - - suspected T76.31
	-child
Add	- -threatened abuse (harm)
Add	- - - confirmed T74.32
Add	- - - suspected T76.32
	Threatened
Add	- abuse (harm) – *see* Maltreatment

Wasting Disease (Syndrome) Due to Underlying Condition

This is a representation from the September 2021 and March 2022 ICD-10 Coordination and Maintenance Committee Meetings of the Wasting Disease (Syndrome) Due to Underlying Condition proposal with recommended modifications. The modifications are in **bold.**

Wasting disease (syndrome) is an involuntary, on-going loss of more than 10% of body weight with reduction in muscle mass, with or without loss of fat due to underlying condition. The manifestations of the disease occur in multiple conditions as an indicator of end-stage progression and complicate those concurrent conditions.

Wasting disease (syndrome) is a metabolic-catabolic syndrome that is a severe complication of a chronic, primary disease. It has a constellation of signs and symptoms and is a manifestation signaling the later end- stage or morbidity of an underlying condition and is typically irreversible.

The National Center for Health Statistics received a request to create an ICD-10-CM code for wasting disease (syndrome) due to underlying condition for coding specificity to aid in capturing severity of illness for mortality of the underlying conditions and to interrupt the physiological progress or pathways of the condition in hopes of supplying a better clinical outcome for the patients.

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TABULAR MODIFICATIONS

E88 Other and unspecified metabolic disorders

New code	E88.A	Wasting disease (syndrome) due to underlying condition
Add		**Cachexia due to underlying condition**
Add		Code first underlying condition
Add		**Excludes1: cachexia NOS (R64)**
Add		**Excludes2: failure to thrive (R62.51, R62.7)**

	R64	**Cachexia**
Delete		**~~Wasting syndrome~~**
Delete		**~~Code first underlying condition, if known~~**
Add		**Exclude1: cachexia due to underlying condition (E88.A)**

TABULAR MODIFICATIONS PROPOSED ADDENDA
All proposed effective October 1, 2024

A40 Streptococcal sepsis

Revise — Code first, if applicable, postprocedural ~~streptococcal~~ sepsis (T81.44-)
Add — sepsis due to central venous catheter (T80.211-)
streptococcal sepsis during labor (O75.3)
Revise — streptococcal sepsis following abortion or ectopic or molar pregnancy (O03.37, O03.87, O04.87, O07.37, ~~O08.0~~O08.82)
Revise — streptococcal sepsis following immunization (T88.0-)
Revise — streptococcal sepsis following infusion, transfusion, or therapeutic injection (~~T80.211-,~~ T80.22-, T80.29-)

A41 Other sepsis

Revise — Code first, if applicable, postprocedural sepsis (T81.44-)
Add — sepsis due to central venous catheter (T80.211-)
sepsis during labor (O75.3)
Revise — sepsis following abortion, ectopic or molar pregnancy (O03.37, O03.87, O04.87, O07.37, ~~O08.0~~ O08.82)
sepsis following immunization (T88.0-)
Revise — sepsis following infusion, transfusion, or therapeutic injection (~~T80.211-,~~ T80.22-, T80.29-)

A41.5 Sepsis due to other Gram-negative organisms
A41.52 Sepsis due to Pseudomonas
Revise — Pseudomonas ~~aeroginosa~~ aeruginosa

C61 Malignant neoplasm of prostate

Revise — Use additional, if applicable, code to identify:
hormone sensitivity status (Z19.1-Z19.2)
rising PSA following treatment for malignant neoplasm of prostate (R97.21)

C92 Myeloid leukemia
Add — Code also, if applicable, pancytopenia (acquired) (D61.818)

C94 Other leukemias of specified cell type
C94.8 Other specified leukemias
Add — Code also, if applicable, eosinophilia (D72.18)

D12 Benign neoplasm of colon, rectum, anus and anal canal

Delete ~~Excludes1: benign carcinoid tumors of the large intestine, and rectum (D3A.02-)~~
Delete ~~polyp of colon NOS (K63.5)~~

Add Excludes2: benign carcinoid tumors of the large intestine, and rectum (D3A.02-)
Add polyp of colon NOS (K63.5)

E13 Other specified diabetes mellitus
E13.0 Other specified diabetes mellitus with hyperosmolarity
E13.00 Other specified diabetes mellitus with hyperosmolarity without nonketotic hyperglycemic-hyperosmolar coma (NKHHC)
Delete ~~Excludes2: type 2 diabetes mellitus (E11.-)~~

E71 Disorders of branched-chain amino-acid metabolism and fatty-acid metabolism
E71.3 Disorders of fatty-acid metabolism
E71.31 Disorders of fatty-acid oxidation
E71.312 Short chain acyl CoA dehydrogenase deficiency
Revise SCAD <u>deficiency</u>

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

E87 Other disorders of fluid, electrolyte and acid-base balance
E87.0 Hyperosmolality and hypernatremia
Add Excludes1: diabetes with hyperosmolarity (E08, E09, E11, E13 with ending .00 or .01)

F64 Gender identity disorders
F64.0 Transsexualism
Delete ~~Gender identity disorder in adolescence and adulthood~~
Gender dysphoria in adolescents and adults
Add Gender identity disorder in adolescence and adulthood
Add Gender incongruence in adolescents and adults
Add Transgender
Add Exlcudes1: gender identity disorder of childhood (F64.2)

F64.2 Gender identity disorder of childhood
Gender dysphoria in children
Add Gender incongruence of childhood

F64.9 Gender identity disorder, unspecified
Add Gender incongruence, unspecified
Gender dysphoria, unspecified
Gender-role disorder NOS

G32 Other degenerative disorders of nervous system in diseases classified elsewhere
G32.0 Subacute combined degeneration of spinal cord in diseases classified elsewhere

Code first underlying disease, such as:
Revise vitamin B12 deficiency anemia, unspecified ~~anemia~~ (D51.9)
Revise other dietary vitamin B12 deficiency anemia ~~dietary~~ (D51.3)
Revise vitamin B12 deficiency anemia due to intrinsic factor deficiency ~~pernicious~~ (D51.0)

H49 Paralytic strabismus
H49.8 Other paralytic strabismus
H49.81 Kearns-Sayre syndrome

Revise ~~Use additional code~~ Code also, if applicable, ~~for~~ other manifestations, such as:
heart block (I45.9)

Infections with a predominantly sexual mode of transmission (A50-A64)

Revise Excludes1: ~~human immunodeficiency virus [HIV] disease (B20)~~ nonspecific and nongonococcal urethritis (N34.1)
Delete ~~nonspecific and nongonococcal urethritis (N34.1)~~
Reiter's disease (M02.3-)

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

I25 Chronic ischemic heart disease
I25.1 Atherosclerotic heart disease of native coronary artery
I25.11 Atherosclerotic heart disease of native coronary artery with angina pectoris

Revise I25.112 ~~Atherosclerosic~~ Atherosclerotic heart disease of native coronary artery with refractory angina pectoris

I43 Cardiomyopathy in diseases classified elsewhere

Code first underlying disease, such as:
amyloidosis (E85.-)
Revise glycogen storage disease (E74.0-)

I71 Aortic aneurysm and dissection
I71.5 Thoracoabdominal aortic aneurysm, ruptured

Revise I71.51 Supraceliac aneurysm of the thoracoabdominal aorta, ruptured
Revise I71.52 Paravisceral aneurysm of the thoracoabdominal aorta, ruptured

I71.6 Thoracoabdominal aortic aneurysm, without rupture

Revise I71.61 Supraceliac aneurysm of the thoracoabdominal aorta, without rupture
Revise I71.62 Paravisceral aneurysm of the thoracoabdominal aorta, without rupture

J41 Simple and mucopurulent chronic bronchitis
Delete ~~Excludes1: chronic bronchitis NOS (J42)~~
Delete ~~chronic obstructive bronchitis (J44.-)~~

Add Excludes2: chronic bronchitis NOS (J42)
Add chronic obstructive bronchitis (J44.-)

J43 Emphysema
Excludes1: compensatory emphysema (J98.3)
Delete ~~emphysema with chronic (obstructive) bronchitis (J44.-)~~
Delete ~~emphysematous (obstructive) bronchitis (J44.-)~~

Add Excludes2: emphysema with chronic (obstructive) bronchitis (J44.-)
Add emphysematous (obstructive) bronchitis (J44.-)

J44 Other chronic obstructive pulmonary disease
Delete ~~Excludes1: bronchiectasis (J47.-)~~
Delete ~~emphysema without chronic bronchitis (J43.-)~~

Add Excludes2: bronchiectasis (J47.-)
Add emphysema without chronic bronchitis (J43.-)

K51 Ulcerative colitis
K51.4 Inflammatory polyps of colon

Delete ~~Excludes1: adenomatous polyp of colon (D12.6)~~
Delete ~~polyposis of colon (D12.6)~~
Delete ~~polyps of colon NOS (K63.5)~~

Add Excludes2: adenomatous polyp of colon (D12.6)
Add polyposis of colon (D12.6)
Add polyps of colon NOS (K63.5)

K63 Other diseases of intestine
K63.5 Polyp of colon

Delete ~~Excludes1: adenomatous polyp of colon (D12.-)~~
Delete ~~inflammatory polyp of colon (K51.4-)~~
Delete ~~polyposis of colon (D12.6)~~

Add Exculdes2: adenomatous polyp of colon (D12.-)
Add inflammatory polyp of colon (K51.4-)
Add polyposis of colon (D12.6)

K80 Cholelithiasis
K80.4 Calculus of bile duct with cholecystitis

Any condition listed in K80.5 with cholecystitis (with cholangitis)

Revise Codes also, if applicable, fistula of bile duct (K83.3)

M24 Other specific joint derangements
M24.1 Other articular cartilage disorders

Revise Excludes2: chondrocalcinosis (M11.1-, M11.2-)
internal derangement of knee (M23.-)
metastatic calcification (E83.5)
Revise ochronosis (E70.29)

M32 Systemic lupus erythematosus (SLE)
M32.1 Systemic lupus erythematosus with organ or system involvement
M32.19 Other organ or system involvement in systemic lupus erythematosus

Add Use additional code(s) to identify organ or system involvement, such as encephalitis (G05.3)

N16 Renal tubulo-interstitial disorders in diseases classified elsewhere

Code first underlying disease, such as:
brucellosis (A23.0-A23.9)
cryoglobulinemia (D89.1)
Revise glycogen storage disease (E74.0-)

N35 Urethral stricture
N35.8 Other urethral stricture
N35.81 Other urethral stricture, male
Revise N35.812 Other ~~urethral~~ bulbous urethral stricture, male

N81 Female genital prolapse
N81.6 Rectocele

Add Excludes1: rectocele with prolapse of uterus (N81.2-N81.4)

Excludes2: perineocele (N81.81)
rectal prolapse (K62.3)
Delete ~~rectocele with prolapse of uterus (N81.2-N81.4)~~

O11 Pre-existing hypertension with pre-eclampsia

Revise Includes: conditions in ~~O10~~ O10 complicated by pre-eclampsia
pre-eclampsia superimposed pre-existing hypertension

O26 Maternal care for other conditions predominantly related to pregnancy
O26.8 Other specified pregnancy related conditions
O26.89 Other specified pregnancy related conditions

Add Use additional code, if applicable, to identify specific condition such as insulin resistance (E88.81)

Pregnancy, childbirth and the puerperium (O00-O9A)

Revise Use additional code, if applicable, from category Z3A, Weeks of gestation, to identify the specific week of the pregnancy, if known.

P29 Cardiovascular disorders originating in the perinatal period
P29.0 Neonatal cardiac failure
Add Code also associated underlying condition

Q90 Down syndrome
Add Code also associated physical condition(s), such as atrioventricular septal defect (Q21.2)

Revise Use additional code(s) to identify any associated ~~physical conditions and~~ degree of intellectual disabilities (F70-F79)

S06 Intracranial injury
S06.3 Focal traumatic brain injury

Delete ~~Excludes1: any condition classifiable to S06.4-S06.6~~

Revise Excludes2: ~~focal cerebral edema (S06.1)~~ any condition classifiable to S06.4-S06.6
Add focal cerebral edema (S06.1)

T75 Other and unspecified effects of other external causes
T75.3 Motion sickness
Airsickness
Seasickness
Travel sickness

Revise Use additional external cause code to identify vehicle or type of motion (Y92.81~~-, Y93.5 -~~)

W62 Contact with nonvenomous amphibians

Revise Excludes1: contact with venomous amphibians ~~(T63.81-R63.83)~~ (T63.81-T63.83).

Z79 Long term (current) drug therapy

Z79.4 Long term (current) use of insulin

Excludes2: long-term (current) use of injectable non-insulin antidiabetic drugs (Z79.85)
long term (current) use of oral antidiabetic drugs (Z79.84)
long term (current) use of oral hypoglycemic drugs (Z79.84)
Add long term (current) use of noninsulin injectable drug (Z79.84)

Z79.8 Other long term (current) drug therapy
Z79.84 Long term (current) use of oral hypoglycemic drugs
Long term (current) use of oral antidiabetic drugs

Excludes2: long-term (current) use of injectable non-insulin antidiabetic drugs (Z79.85)

	long term (current) use of insulin (Z79.4)
Add	long term (current) use of noninsulin injectable drug (Z79.85)

INDEX MODIFICATION PROPOSED ADDENDA
All proposed effective October 1, 2024

Add Burkholderia
Add - cepacia A49.8
Add - mallei A24.0
Add - pseudomallei – see Melioidosis

Cachexia R64
Revise - due to malnutrition ~~R64~~ E43

Colibacillosis A49.8
Revise - generalized – see also, Sepsis, Escherichia coli ~~A41.50~~ A41.51

Complication(s) (from) (of)
- catheter (device) NEC -see also Complications, prosthetic device or implant
- - epidural infusion T85.9
- - - mechanical
Revise - - - - malfunction ~~T85.610~~ T85.690

- - subdural infusion T85.9
- - - mechanical
Revise - - - - malfunction ~~T85.610~~ T85.690

Dependence (on) (syndrome) F19.20
- drug NEC F19.20
- - psychoactive NEC F19.20
Add - - - in remission F19.21

Diabetes, diabetic (mellitus) (sugar) E11.9
- with
Revise - - ~~Kimmelsteil~~ Kimmelstiel -Wilson disease E11.21

- due to drug or chemical E09.9
- - with
Revise - - - ~~Kimmelsteil~~ Kimmelstiel -Wilson disease E09.21

- due to underlying condition E08.9
- - with
Revise - - - ~~Kimmelsteil~~ Kimmelstiel ~~-WIlson~~ Wilson disease E08.21

- specified type NEC E13.9
- - with
Revise - - - ~~Kimmelsteil~~ Kimmelstiel -Wilson disease E13.21

- type 1 E10.9
- - with
Revise - - - ~~Kimmelsteil~~ Kimmelstiel -Wilson disease E10.21

- type 2 E11.9
- - with
Revise - - - ~~Kimmelsteil~~ Kimmelstiel -Wilson disease E11.21

Disorder (of) -see also Disease
- bone M89.9
- - development and growth NEC M89.20
Revise - - - ilium ~~M89.259~~ M89.28
Revise - - - ischium ~~M89.259~~ M89.28
- eating (adult) (psychogenic) F50.9
Add - - specified NEC F50.89

Edema, edematous (infectious) (pitting) (toxic) R60.9
- lung J81.1
- - with heart condition or failure -see Failure, ventricular, left
Add - - - newborn P29.0

Encephalitis (chronic) (hemorrhagic) (idiopathic) (nonepidemic) (spurious) (subacute) G04.90
Revise - lupus erythematosus, systemic M32.19 [G05.3]

Gastropathy K31.9
Add - specified NEC K31.89

Grief F43.21
Revise - prolonged ~~F43.29~~ F43.81

Add Homocystinemia R79.83
Revise ~~Homocystinemia, homocystinuria~~ Homocystinuria E72.11

Revise Hyperosmolality – (see also, Diabetes, by type, with hyperosmolarity) E87.0

Hypertrophy, hypertrophic
- bone M89.30
Revise - - ilium ~~M89.359~~ M89.38
Revise - - ischium ~~M89.359~~ M89.38
Add - - pubic ramus M89.38

Infection, infected, infective (opportunistic) B99.9
- bacterial NOS A49.9
- - as cause of disease classified elsewhere B96.89
Add - - - Cronobacter (sakazakii) B96.89

Add - Cronobacter (sakazakii) B96.89
Add - - as cause of disease classified elsewhere B96.89
Add - - generalized A41.59

- Pseudomonas NEC A49.8
Add - - generalized A41.52
Add - Serratia NEC A49.8
Add - - as cause of disease classified elsewhere B96.89
Add - - generalized A41.53

Intolerance
- milk NEC K90.49
Revise ~~—~~ - orthostatic, chronic G90.A

Leukodystrophy E75.29
Add - metachromatic E75.25

Revise Lipodermatosclerosis (see also Insufficiency, venous) M79.3 -see Varix, leg, with, inflammation
Add - with
Add - - varicose veins -see Varix, leg, with, inflammation
Add - - - ulcerated -see Varix, leg, with, ulcer, with inflammation by site
Revise - ulcerated -see also Ulcer, by site -see Varix, leg, with, ulcer, with inflammation by site

Malfunction -see also Dysfunction
- catheter device NEC T85.618
- - infusion NEC T82.514
Revise - - - cranial ~~T85.610~~ (see also Complication(s), catheter, cranial infusion, mechanical) T85.690
Revise - - - epidural ~~T85.610~~ (see also Complication(s), catheter, cranial infusion, mechanical) T85.690
Revise - - - intrathecal (see also Complication(s), catheter, cranial infusion, mechanical) ~~T85.610~~ T85.690
Revise - - - spinal (see also Complication(s), catheter, cranial infusion, mechanical) ~~T85.610~~ T85.690
Revise - - - subarachnoid (see also Complication(s), catheter, cranial infusion, mechanical) ~~T85.610~~ T85.690
Revise - - - subdural (see also Complication(s), catheter, cranial infusion, mechanical) ~~T85.610~~ T85.690

Neuromyopathy G70.9
Revise - paraneoplastic (see also, Neoplasm, by site, if known) D49.9 [G13.0]

Neuropathy, neuropathic G62.9
Revise - paraneoplastic (sensorial) (Denny Brown) (see also, Neoplasm, by site, if known) D49.9 [G13.0]

Neutropenia, neutropenic (chronic) (genetic) (idiopathic) (immune) (infantile) (malignant) (pernicious) (splenic)
Add - specified NEC D70.8

Osteitis -see also Osteomyelitis
Revise - deformans (see also Paget's disease, bone) M88.9
- - in (due to)
Revise - - - malignant neoplasm of bone (see also, Neoplasm, malignant, by site) C41.9 [M90.60]
Revise - - - neoplastic disease –(see also Neoplasm, by type and site) D49.9 [M90.60]
Revise - - - - carpus ~~D49.9~~ D49.2 [M90.64-]
Revise - - - - clavicle ~~D49.9~~ D49.2 [M90.61-]
Revise - - - - femur ~~D49.9~~ D49.2 [M90.65-]
Revise - - - - fibula ~~D49.9~~ D49.2 ~~[M90.65-]~~ [M90.66-]
Revise - - - - finger ~~D49.9~~ D49.2 [M90.64-]
Revise - - - - humerus ~~D49.9~~ D49.2 [M90.62-]
Revise - - - - ilium ~~D49.9~~ D49.2 ~~[M90.65-]~~ [M90.68]
Revise - - - - ischium ~~D49.9~~ D49.2 ~~[M90.65-]~~ [M90.68]
Revise - - - - metacarpus ~~D49.9~~ D49.2 [M90.64-]
Revise - - - - metatarsus ~~D49.9~~ D49.2 [M90.67-]
Revise - - - - multiple sites ~~D49.9~~ D49.89 [M90.69]
Revise - - - - neck ~~D49.9~~ D49.2 [M90.68]
Add - - - - pubic ramus D49.2 [M90.68]
Revise - - - - radius ~~D49.9~~ D49.2 [M90.63-]
Revise - - - - rib ~~D49.9~~ D49.2 [M90.68]
Revise - - - - scapula ~~D49.9~~ D49.2 [M90.61-]
Revise - - - - skull ~~D49.9~~ D49.2 [M90.68]
Revise - - - - tarsus ~~D49.9~~ D49.2 [M90.67-]
Revise - - - - tibia ~~D49.9~~ D49.2 [M90.66-]
Revise - - - - toe ~~D49.9~~ D49.2 [M90.67-]
Revise - - - - ulna ~~D49.9~~ D49.2 [M90.63-]
Revise - - - - vertebra ~~D49.9~~ D49.2 [M90.68]

Osteoarthropathy (hypertrophic) M19.90
- specified type NEC M89.40
Revise - - ilium ~~M89.459~~M89.48
Revise - - ischium ~~M89.459~~M89.48
Add - - pubic ramus M89.48

	Osteodystrophy Q78.9
Revise	- ilium ~~M89.559~~ M89.58
Revise	- ischium ~~M89.559~~ M89.58
Add	- pubic ramus M89.58
	Osteolysis M89.50
Revise	- ilium ~~M89.559~~ M89.58
Revise	- ischium ~~M89.559~~ M89.58
Add	- pubic ramus M89.58
	Osteomalacia M83.9
	- vitamin-D-resistant in adults E83.31 [M90.8-]
Revise	- - ilium E83.31 ~~[M90.859]~~ [M90.88]
Revise	- - ischium E83.31 ~~[M90.859]~~ [M90.88]
Add	- - pubic ramus 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	- - - ilium ~~M86.459~~ M86.48
Revise	- - - ischium ~~M86.459~~ M86.48
Add	- - - pubic ramus M86.48
	- - hematogenous NEC M86.50
Revise	- - - ilium ~~M86.559~~ M86.58
Revise	- - - ischium ~~M86.559~~ M86.58
Add	- - - pubic ramus M86.58
	- - - multifocal M86.30
Revise	- - - - ilium ~~M86.359~~ M86.38
Revise	- - - - ischium ~~M86.359~~ M86.38
Add	- - - pubic ramus M86.38
	Osteonecrosis M87.9
	- idiopathic aseptic M87.00
Add	- - pubic ramus M87.050
	- secondary NEC M87.30
	- - due to
	- - - drugs M87.10
Revise	- - - - ilium ~~M87.159~~ M87.150
Revise	- - - - ischium ~~M87.159~~ M87.150
Add	- - - - pubic ramus M87.150

- - - hemoglobinopathy NEC D58.2 [M90.50]
Revise - - - - ilium D58.2 ~~[M90.55]~~ [M90.58]
Revise - - - - ischium D58.2 ~~[M90.55]~~ [M90.58]
Add - - - - pubic ramus D58.2 [M90.58]

- - - trauma (previous) M87.20
Revise - - - - ilium ~~M87.25 -~~ M87.250
Revise - - - - ischium ~~M87.25 -~~ M87.250
Add - - - - pubic ramus M87.250

- - in
- - - caisson disease T70.3 [M90.50]
Revise - - - - ilium T70.3 ~~[M90.55]~~ [M90.58]
Revise - - - - ischium T70.3 ~~[M90.55]~~ [M90.58]
Add - - - - pubic ramus T70.3 [M90.58]

Add - - pubic ramus M87.350

- specified type NEC M87.80
Revise - - ilium ~~M87.85 -~~ M87.850
Revise - - ischium ~~M87.85 -~~ M87.850
Add - - pubic ramus M87.850

Osteopathy -see also Osteomyelitis, Osteonecrosis, Osteoporosis
- after poliomyelitis M89.60
Revise - - ilium ~~M89.659~~ M89.68
Revise - - ischium ~~M89.659~~ M89.68
Add - - pubic ramus M89.68

Paget's disease
- bone M88.9
Revise - - ilium ~~M88.85 -~~ M88.88
Revise - - ischium ~~M88.85 -~~ M88.88
Add - - pubic ramus M88.88

Revise Pancolitis, ~~ulcerative (chronic)~~ (see also, Colitis) ~~K51.00~~
Add - ulcerative (chronic) K51.00
Revise - - with
Revise - - - complication K51.019
Revise - - - abscess K51.014
Revise - - - fistula K51.013
Revise - - - obstruction K51.012
Revise - - - rectal bleeding K51.011
Revise - - - specified complication NEC K51.018

Revise	Pellagra (alcoholic) ~~(with polyneuropathy)~~ E52
Add	- with
Add	- - polyneuropathy E52 [G63]
	Rectocele
	- female (without uterine prolapse) N81.6
	- - with uterine prolapse N81.4
Add	- - - complete N81.3
	Resistance, resistant (to)
Delete	~~- complicating pregnancy O26.89~~
	- insulin E88.81
Add	- - complicating pregnancy O26.89-
	Schizophrenia, schizophrenic F20.9
	- undifferentiated (type) F20.3
Revise	- - chronic ~~F20.9~~ F20.5
	Sepsis (generalized) (unspecified organism) A41.9
Add	- Cronobacter A41.59
Add	- MRSA (Methicillin resistant Staphylococcus aureus) A41.02
	Short, shortening, shortness
	- stature (child) (hereditary) (idiopathic) NEC R62.52
	- - due to
	- - - genetic causes E34.329
Revise	- - - - ACAN gene variant ~~E34.828~~ E34.328
Revise	- - - - aggrecan deficiency ~~E34.828~~ E34.328
Revise	- - - - NPR-2 gene variant ~~E34.828~~ E34.328
Revise	- - - - specified genetic cause NEC ~~E34.828~~ E34.328
	Syndrome -see also Disease
Revise	- hyperosmolality (see also, Diabetes, by type, with hyperosmolarity) E87.0
Revise	- ~~Kimmelsteil~~ Kimmelstiel -Wilson -see Diabetes, specified type, with ~~Kimmelsteil~~ Kimmelstiel -Wilson disease