



**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023
Diagnosis Agenda**

Welcome and announcements

Carolyn Greene, MD

David Berglund, MD

Diagnosis Topics:

Contents

Abnormal antibody without a diagnosis of rheumatoid arthritis	14
Traci Ramirez	
Kevin D. Deane, MD/PhD	
Professor of Medicine, William P. Arend Endowed Chair in Rheumatology Research The University of Colorado, Division of Rheumatology	
Adverse Effects of Immune Checkpoint Inhibitor (ICI) Immunotherapy	17
Cheryl Bullock	
Kerry Reynolds, MD	
Clinical Director, Inpatient Oncology	
Director, Severe Immunotherapy Complications Service	
Massachusetts General Hospital	
Anal Fistula.....	22
Shannon McConnell-Lamphey	
Pradeep P. Nazarey, MD	
US Medical Affairs, Gastroenterology Business Unit	
Takeda Pharmaceuticals USA Inc.	
Anosognosia.....	28
Cheryl Bullock	
Baked Egg Tolerance in Egg Allergy	30
Cheryl Bullock	
Julia Upton, MD, FRCP(C), FAAAAI	
Staff Physician The Hospital for Sick Kids	

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Baked Milk Tolerance in Milk Allergy	33
Cheryl Bullock Julia Upton, MD, FRCP(C), FAAAAI Staff Physician The Hospital for Sick Kids	
Bicuspid Aortic Valve.....	36
David Berglund, MD Simon C. Body, MBChB, MPH, FAHA Boston University School of Medicine Director, International Bicuspid Aortic Valve Consortium	
Cement Pulmonary Embolism and Fat Pulmonary Embolism	39
David Berglund, MD Richard H. White, MD Professor of Medicine, UC Davis	
Cholestatic Pruritus.....	42
Traci Ramirez	
Coding of Firearm Injuries Default.....	43
Traci Ramirez Cathy Barber Senior Researcher, Harvard Injury Control Research Center Harvard Chan School of Public Health	
DLG4-related synaptopathy.....	45
David Berglund, MD Zeynep Tümer, MD, PhD, DMSc Professor of Applied Human Molecular Genetics with special focus on neurodevelopmental disorders University of Copenhagen, Denmark	
Duffy Phenotype	49
David Berglund, MD Maureen Okam Achebe, MD MPH Brigham and Women’s Hospital Harvard Medical School	
Early-stage type 1 diabetes	53
Shannon McConnell-Lamprey Brigitte I Frohnert MD PhD Associate Professor of Pediatrics Barbara Davis Center for Diabetes	

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

University of Colorado Anschutz Medical Campus Encounter for prophylactic removal of fallopian tube(s) for persons with no known genetic/familial risk factors.....	55
Traci Ramirez Rebecca Stone, MD Associate Professor and Director of Gynecologic Oncology Department of Gynecology and Obstetrics Johns Hopkins School of Medicine	
Estrogen, and other hormones and factors receptor status.....	59
Traci Ramirez Elizabeth Valencia, MD, JD, FCLM Assistant Professor, Department of Radiology, Division of Breast Imaging & Intervention, Mayo Clinic	
Family History of Familial Adenomatous Polyposis.....	63
David Berglund, MD	
Flank Anatomical Specificity	64
Cheryl Bullock	
Gulf War Illness.....	74
Shannon McConnell-Lamprey Beatrice A. Golomb, MD, PhD Professor of Medicine UC San Diego School of Medicine	
Hyperoxaluria	76
Shannon McConnell-Lamprey Cristin Kaspar, MD, MS Biostatistics, MS Physiology & Biophysics Medical Director Clinical Development Alnylam Pharmaceuticals Pediatric Nephrologist, Children’s Wisconsin Pediatric Nephrology	
Hypoglycemia level	79
Shannon McConnell-Lamprey Daniel Chernavsky, MD Senior Director Medical Affairs at Dexcom	
Injection Drug Use.....	82
Cheryl Bullock	

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Lymphoma in Remission	85
Traci Ramirez	
Multifidus Muscle Dysfunction	97
David Berglund, MD	
Chris Gilligan, MD, MBA	
Associate Chief Medical Officer, Brigham and Women’s Hospital	
Nasal Valve Collapse	100
Shannon McConnell-Lampzey	
Thomas Higgins, MD, MSPH	
Fellowship-Trained Rhinologist	
Clinical Associate Professor with the Department of Otolaryngology, University of Louisville School of Medicine	
Pediatric Obesity Body Mass Index.....	103
David Berglund, MD	
Personal History of Colon Polyps.....	105
David Berglund, MD	
Personal History of Immune Checkpoint Inhibitor (ICI) Immunotherapy	107
Cheryl Bullock	
Kerry Reynolds, MD	
Clinical Director, Inpatient Oncology	
Director, Severe Immunotherapy Complications Service	
Massachusetts General Hospital	
Post-exertional malaise/post-exertional symptom exacerbation.....	109
Traci Ramirez	
Lisa McCorkell, MPP	
Co-Founder of Patient-Led Research Collaborative	
Serotonin Syndrome.....	112
Traci Ramirez	
Usher Syndrome.....	114
Shannon McConnell-Lampzey	
Eliot Shearer, MD, PhD	
Associate, Department of Otolaryngology and Communication Enhancement	
Boston Children’s Hospital	
Assistant Professor of Otolaryngology Head and Neck Surgery	

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Harvard Medical School
TABULAR MODIFICATIONS PROPOSED ADDENDA..... 116
Traci Ramirez

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

ICD-10 TIMELINE

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

September 12-13, 2023	The September 2023 ICD-10 Coordination and Maintenance Committee Meeting will be held virtually by Zoom Webinar.
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 and revisions 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 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/Latest_News
November 15, 2023	Deadline for receipt of public comments on proposed new codes and revisions discussed at the September 12-13, 2023 ICD-10

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2024.

December 1, 2023

Deadline for requestors: Those members of the public requesting that topics be discussed at the March 19-20, 2024 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, 2024 implementation date, or an April 1, 2025 implementation date.

The ICD-10 Coordination and Maintenance Committee will make efforts to accommodate the requested implementation date for each request submitted, however, the Committee will determine which requests will be presented for consideration for an October 1, 2024 implementation date or an April 1, 2025 implementation date.

January 2024

Federal Register notice for the March 19-20, 2024 ICD-10 Coordination and Maintenance Committee Meeting will be published. This will include the tentative agenda.

February 2024

Tentative agenda for the Procedure portion of the March 19, 2024 ICD-10 Coordination and Maintenance Committee Meeting posted on CMS webpage at:

<https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html>

Tentative agenda for the Diagnosis portion of the March 20, 2024 ICD-10 Coordination and Maintenance Committee Meeting posted on NCHS homepage at:

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

February 1, 2024

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>

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

- February 1, 2024** 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, 2024** 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 19-20, 2024 The ICD-10 Coordination and Maintenance Committee Meeting is anticipated to be fully virtual by zoom and dial-in. Those who wish to attend must participate via Zoom Webinar or by dialing in.
- March 2024 Recordings and slide presentations of the March 19-20, 2024 ICD-10 Coordination and Maintenance Committee Meeting will be posted on the following web pages:
Diagnosis code portion of the recording and related materials–
https://www.cdc.gov/nchs/icd/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, 2024 Any new or revised ICD-10 codes will be implemented on April 1, 2024.
- April 19, 2024** **Deadline for receipt of public comments on proposed new codes and revisions discussed at the March 19-20, 2024 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2024.**
- April 2024 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 2025 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:

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

<https://www.cms.gov/medicare/medicare-fee-for-service-payment/acuteinpatientpps>

May 17, 2024

Deadline for receipt of public comments on proposed new codes and revisions discussed at the March 19-20, 2024 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on April 1, 2025.

Deadline for receipt of public comments on proposed new diagnosis codes and revisions discussed at the March 19-20, 2024 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2025.

May/June 2024

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/index.html>

June 7, 2024

Deadline for requestors: Those members of the public requesting that topics be discussed at the September 10-11, 2024 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, 2025 implementation date or an October 1, 2025 implementation date.

The ICD-10 Coordination and Maintenance Committee will make efforts to accommodate the requested implementation date for each request submitted, however, the Committee will determine which requests will be presented for consideration for an April 1, 2025 implementation date or an October 1, 2025 implementation date.

July 2024

Federal Register notice for the September 10-11, 2024 ICD-10 Coordination and Maintenance Committee Meeting will be published. This will include the tentative agenda.

August 1, 2024

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, 2024.

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

This rule can be accessed at:

<https://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/AcuteInpatientPPS/index.html>

August 2024

Tentative agenda for the Procedure portion of the September 10, 2024, 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 11, 2024, 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 10-11, 2024

The September 2024 ICD-10 Coordination and Maintenance Committee Meeting is anticipated to be fully virtual by zoom and dial-in. Those who wish to attend must participate via Zoom Webinar or by dialing in.

September 2024

Recordings and slide presentations of the September 10-11, 2024 ICD-10 Coordination and Maintenance Committee Meeting will be posted on the following web pages:

Diagnosis code portion of the recording and related materials–
https://www.cdc.gov/nchs/icd/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, 2024

New and revised ICD-10-CM and ICD-10-PCS codes go into effect along with MS-DRG changes. Final addendum available on web pages as follows:

Diagnosis addendum –
<https://www.cdc.gov/nchs/icd/Comprehensive-Listing-of-ICD-10-CM-Files.htm>

Procedure addendum –
<https://www.cms.gov/Medicare/Coding/ICD10/>

October 11, 2024

Deadline for receipt of public comments on proposed new codes discussed at the September 10-11, 2024 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on April 1, 2025.

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

November 2024

Any new ICD-10 codes that will be implemented on the following April 1 will be announced. Information on any new codes to be implemented April 1, 2025, will be posted on the following websites:

<https://www.cdc.gov/nchs/icd/Comprehensive-Listing-of-ICD-10-CM-Files.htm>

https://www.cms.gov/Medicare/Coding/ICD10/Latest_News

November 13, 2024

Deadline for receipt of public comments on proposed new codes and revisions discussed at the September 10-11, 2024 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2025.

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Contact Information

Mailing address:

National Center for Health Statistics
ICD-10-CM Coordination and Maintenance Committee
3311 Toledo Road
Hyattsville, Maryland 20782

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

David Berglund	(301) 458-4095
Cheryl Bullock	(301) 458-4297
Shannon McConnell-Lamprey	(301) 458-4612
Traci Ramirez	(301) 458-4454

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Continuing Education Credits

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

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

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

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

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

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

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

Abnormal antibody without a diagnosis of rheumatoid arthritis

The proposal was previously presented at the September 2022 ICD10 Coordination and Maintenance meeting and is being represented today to address comments received.

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

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

However, it is now well-established that RA-related immunologic tests such as RF and ACPA/anti-CCP can be present in individuals in absence of and prior to the appearance of IA, and predictive of future onset of clinical RA⁵. Furthermore, individuals who have abnormal RA-related immunologic tests without IA are identified in growing numbers in clinical care. Notably, these individuals may have symptoms such as joint pain, stiffness or swelling, but no other objective evidence of IA⁶⁻⁸. There are current recommendations for medical follow-up and lifestyle changes (e.g., smoking cessation) that can be applied to these individuals. In addition, the predictive ability of RF and ACPA for future clinical RA has underpinned multiple clinical observational studies and prevention trials in RA⁹⁻¹⁵, and it is expected that there soon will be approved pharmacologic for RA prevention. Indeed, some companies are in the process of requesting FDA approval for therapeutic agents in this condition to prevent or delay the future onset of clinical RA.

Importantly, while there are ICD-10-CM codes that be used to designate clinical RA (e.g. M05.79), autoantibody positivity with joint symptoms/arthritis (e.g. M25.50), as well as designations within the R category for RF and anti-CCP positivity (e.g. R76.8), there is not currently a clear way in the existing ICD-10-CM system to designate individuals who may exhibit RA-related biomarkers/immunologic findings, but not have clinical RA. As such, the introduction of a new code to accurately designate an individual who has abnormal 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.

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

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. Aletaha D, Neogi T, Silman AJ, Funovits J, Felson DT, Bingham CO, Birnbaum NS, Burmester GR, Bykerk VP, Cohen MD, Combe B, Costenbader KH, Dougados M, Emery P, Ferraccioli G, Hazes JM, Hobbs K, Huizinga TW, Kavanaugh A, Kay J, Kvien TK, Laing T, Mease P, Ménard HA, Moreland LW, Naden RL, Pincus T, Smolen JS, Stanislawska-Biernat E, Symmons D, Tak PP, Upchurch KS, Vencovský J, Wolfe F, Hawker G. 2010 Rheumatoid arthritis classification criteria: an American College of Rheumatology/European League Against Rheumatism collaborative initiative. *Arthritis Rheum*. 2010;62(9):2569-81. doi: 10.1002/art.27584. PubMed PMID: 20872595.
4. 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.
5. Deane KD, Holers VM. Rheumatoid Arthritis Pathogenesis, Prediction, and Prevention: An Emerging Paradigm Shift. *Arthritis Rheumatol*. 2021;73(2):181-93. Epub 2020/07/01. doi: 10.1002/art.41417. PubMed PMID: 32602263; PMCID: PMC7772259.
6. Stack RJ, van Tuyl LH, Sloots M, van de Stadt LA, Hoogland W, Maat B, Mallen CD, Tiwana R, Raza K, van Schaardenburg D. Symptom complexes in patients with seropositive arthralgia and in patients newly diagnosed with rheumatoid arthritis: a qualitative exploration of symptom development. *Rheumatology (Oxford)*. 2014;53(9):1646-53. Epub 2014/04/15. doi: 10.1093/rheumatology/keu159. PubMed PMID: 24729397.
7. van Steenberg HW, Aletaha D, Beaart-van de Voorde LJ, Brouwer E, Codreanu C, Combe B, Fonseca JE, Hetland ML, Humby F, Kvien TK, Niedermann K, Nuno L, Oliver S, Rantapaa-Dahlqvist S, Raza K, van Schaardenburg D, Schett G, De Smet L, Szucs G, Vencovsky J, Wiland P, de Wit M, Landewe RL, van der Helm-van Mil AH. EULAR definition of arthralgia suspicious for progression to rheumatoid arthritis. *Ann Rheum Dis*. 2017;76(3):491-6. Epub 2016/12/20. doi: 10.1136/annrhumdis-2016-209846. PubMed PMID: 27991858.
8. Ten Brinck RM, van Steenberg HW, van Delft MAM, Verheul MK, Toes REM, Trouw LA, van der Helm-van Mil AHM. The risk of individual autoantibodies, autoantibody combinations and levels for arthritis development in clinically suspect arthralgia. *Rheumatology (Oxford)*. 2017;56(12):2145-53. doi: 10.1093/rheumatology/kex340. PubMed PMID: 28968865; PMCID: PMC6703997.
9. Deane KD, Holers VM. Rheumatoid arthritis pathogenesis, prediction, and prevention: an emerging paradigm shift. *Arth Rheum*. 2021;73:181-93.
10. Gerlag DM, Safy M, Maijer KI, Tang MW, Tas SW, Starmans-Kool MJF, van Tubergen A, Janssen M, de Hair M, Hansson M, de Vries N, Zwinderman AH, Tak PP. Effects of B-cell directed therapy on the preclinical stage of rheumatoid arthritis: the PRAIRI study. *Ann Rheum Dis*. 2019;78(2):179-85. Epub 2018/12/07. doi: 10.1136/annrhumdis-2017-212763. PubMed PMID: 30504445; PMCID: PMC6352407.
11. Krijbolder DI, Verstappen M, van Dijk BT, Dakkak YJ, Burgers LE, Boer AC, Park YJ, de Witt-Luth ME, Visser K, Kok MR, Molenaar ETH, de Jong PHP, Böhringer S, Huizinga TWJ, Allaart CF, Niemantsverdriet E, van der Helm-van Mil AHM. Intervention with methotrexate in patients with arthralgia at risk of rheumatoid arthritis to reduce the development of persistent arthritis and its disease burden (TREAT EARLIER): a randomised, double-blind, placebo-controlled, proof-of-concept trial. *Lancet*. 2022;400(10348):283-94. doi: 10.1016/S0140-6736(22)01193-X. PubMed PMID: 35871815.
12. van Boheemen L, Turk S, Beers-Tas MV, Bos W, Marsman D, Griep EN, Starmans-Kool M, Popa CD, van Sijl A, Boers M, Nurmohamed MT, van Schaardenburg D. Atorvastatin is unlikely to prevent rheumatoid arthritis in high risk individuals: results from the prematurely stopped STATins to Prevent Rheumatoid Arthritis (STAPRA) trial. *RMD Open*. 2021;7(1). doi: 10.1136/rmdopen-2021-001591. PubMed PMID: 33685928; PMCID: PMC7942258.
13. Deane K, Striebich C, Feser M, Demourelle K, Moss L, Bemis E, Frazer-Abel A, Fleischer C, Sparks J, Solow E, James J, Guthridge J, Davis J, Graf J, Kay J, Danila M, Bridges L, Forbess L, O'Dell J, McMahon M, Grossman J, Horowitz D, Tiliakos A, Schiopu E, Fox D, Carlin J, Arriens C, Bykerk V, Jan R, Pioro M, Husni E, Fernandez-Pokorny A, Walker S, Booher S, Greenleaf M, Byron M, Keyes-Elstein L, Goldmuntz E, Holers M. Hydroxychloroquine Does Not Prevent the Future Development of Rheumatoid Arthritis in a Population with Baseline High Levels of Antibodies to Citrullinated Protein Antigens and Absence of Inflammatory Arthritis: Interim Analysis of the StopRA Trial (Abstract 1604). *Arthritis Rheumatol*. 74 (Suppl 9)2022.
14. Rech J, Kleyer A, Østergaard M, Hagen M, Valor Mendez L, Tascilar K, Kroenke G, Schönau V, Simon D, Kleinert S, Baraliakos X, Braun J, Hueber A, Fleck M, Rubbert-Roth A, Behrens F, Feuchtenberger M, Zaenker M, Voll R, Glaser C, Filková M, Feist E, Burmester G, Karberg K, Strunk J, Canete J, Šenolt L, Naredo E, Schett G. Abatacept Significantly Reduces Subclinical Inflammation During Treatment (6 Months), This Persists After Discontinuation (12 Months), Resulting in a Delay in the Clinical Development of RA in Patients at Risk of RA (The ARIAA Study) [abstract 0530]. *Arthritis Rheumatol*. 2022.
15. Cope A, Jasencova M, Vasconcelos J, Filer A, Raza K, Qureshi S, D'agostino MA, Mcinnes I, Isaacs J, Pratt A, Fisher B, Buckley CD, Emery P, Ho P, Buch MH, Ciurtin C, Huizinga T, Van Schaardenburg D, Murphy C, Prevost T. ABATACEPT IN INDIVIDUALS AT RISK OF DEVELOPING RHEUMATOID ARTHRITIS: RESULTS FROM THE ARTHRITIS PREVENTION IN THE PRECLINICAL PHASE OF RA WITH ABATACEPT (APIPPRA) TRIAL (EULAR 2023 Abstract OP0130) DOI: 10.1136/annrhumdis-2023-eular.1751. 2023.

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

TABULAR MODIFICATIONS

M05 Rheumatoid arthritis with rheumatoid factor

New code	M05.A Abnormal antibody without a diagnosis of rheumatoid arthritis
Add	Abnormal anti-cyclic citrullinated peptide antibody and rheumatoid factor
Add	Excludes1: rheumatoid arthritis without rheumatoid factor (M06.0)

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Adverse Effects of Immune Checkpoint Inhibitor (ICI) Immunotherapy

Immunotherapy is a type of treatment which centers on boosting the body's own immune system. Although also used for other conditions, immunotherapy has become a significant modality for cancer treatment by unleashing the immune mechanisms that allow immune cells to locate, target and attack cancer cells¹.

Various types of antineoplastic immunotherapy are currently available. These include cancer vaccines, immune system modulators like interleukins and interferons, immune effector cell (IEC) therapy and, most commonly, immune checkpoint inhibitor (ICI) therapy.

Immune checkpoint inhibitor therapy has been in use since 2011, when the first immune checkpoint inhibitor drug was approved by the FDA². Particularly over the last five years, use has rapidly expanded with multiple additional immune checkpoint inhibitor drugs now approved to treat over 80 indications across 17 types of cancer. While initially directed toward unresectable, late stage, and metastatic cancers, immune checkpoint inhibitor therapy is now also being used in earlier stages of cancer and sometimes as a first-line treatment. The immune checkpoint inhibitor drug may be given as a single agent or combined with other therapies like chemotherapy or another immune checkpoint inhibitor.

Immune checkpoint inhibitor therapies work through surface proteins on T-cells, which are part of the body's immune system, to control immune system activation and suppression. These surface proteins are the "checkpoints." However, cancer cells may evade detection by the immune system when the checkpoints on cell surfaces are excessively elevated. The high levels of checkpoints has the effect of keeping the immune system inactive, which paradoxically signals the T-cells not to attack cancer cells. Immune checkpoint inhibitor drugs have the effect of obstructing these inactivation signals on either the T-cells or cancer cells. Inhibiting the inactivation signals allows the immune system to be activated which enables T-cells, along with other immune cells, to then properly recognize cancer cells and target them for attack.

Although both are types of cancer immunotherapy, immune effector cell (IEC) therapy and immune checkpoint inhibitor (ICI) therapy have significant differences. CAR-T cell therapy, the most prominent type of immune effector cell therapy, is customized to each patient and involves harvesting the patient's own T-cells, genetically modifying them in a laboratory, and then infusing them back into the patient. Conversely, immune checkpoint inhibitors are manufactured drugs, specifically monoclonal antibodies, and are used "off-the-shelf."

Immune checkpoint inhibitor therapy is also distinct from chemotherapy. Chemotherapy uses cytotoxic drugs to interfere with the uncontrolled cell division characteristic of cancer. Because their effect also impacts any rapidly dividing cells, particularly in the bone marrow, chemotherapy drugs are typically immunosuppressive. Immune checkpoint inhibitors are not immunosuppressive and actually have the opposite effect of activating the immune system.

As with all drugs, use of immune checkpoint inhibitors is subject to adverse effects, ranging from relatively mild to quite severe and affecting multiple body systems^{3,4,5}. By body system, observed adverse effects include but are not limited to:

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

- Cardiovascular: Pericarditis, myocarditis, vasculitis
- Skin: Maculopapular rash, bullous dermatitis, Stevens-Johnson like syndrome, toxic epidermal necrolysis
- Endocrine: Hypothyroidism, new-onset diabetes with ketoacidosis, adrenal insufficiency
- Gastrointestinal: Colitis, pancreatitis, hepatitis
- Systemic Immune: Cytokine release syndrome*
- Musculoskeletal: Inflammatory arthritis, polymyalgia rheumatica, giant cell arteritis
- Neurological: Aseptic meningitis, encephalitis, Guillain-Barre syndrome, myasthenia gravis, peripheral neuropathy, transverse myelitis
- Ocular: Vision changes, uveitis, scleritis
- Pulmonary: Pneumonitis
- Renal: Acute kidney injury

* Because cytokines play a role in T-cell activity, both immune effector cell therapy and immune checkpoint inhibitor therapy can lead to cytokine release syndrome.

These conditions have other etiologies for which there are standard treatment regimens. However, when arising as an adverse effect of immune checkpoint inhibitor therapy, treatment regimens are different. For example, for non-ICI-induced myocarditis, since the etiology is typically viral, the standard treatment is to abstain from significant exercise and follow-up with a cardiologist. In contrast, when caused by ICI therapy, the treatment regimen is high dose immunosuppression with close and frequent observation, assessing both the response to therapy and observing for complications related to profound immune suppression. There is also significant increased mortality risk for ICI-induced myocarditis.

Currently, best estimates for the incidence of serious adverse events are 14%-34%, depending on the type of immune checkpoint inhibitor drug used, and up to 55% when a combination of ICI drugs is used⁶. Because of immune checkpoint inhibitor therapy's unique mechanism of action, adverse effects of ICI drugs differ in their presentation and frequency from adverse effects of chemotherapy drugs. In particular, adverse effects of ICI may occur and persist long after ICI treatment has ended.

In recent years, immune checkpoint inhibitor therapy has become the standard of care for many types of cancer. Continued rapid uptake and expanding use has in turn increased the need for recognition and identification of ICI adverse effects in national databases to clearly track their occurrence, define their incidence, and inform clinical strategies for their management.

ICD-10-CM currently has unique codes for complications and adverse effects of other cancer treatments. For IEC, including CAR-T cell therapy, code T80.82, Complication of immune effector cell therapy, is available. For chemotherapy, codes T45.1X1–T45.1X6, Poisoning by, adverse effect of and underdosing of antineoplastic and immunosuppressive drugs, are available. Unique codes for adverse effects of immune checkpoint inhibitor therapy will complement the existing codes for these other modalities to create a more complete picture of adverse effects and complications of antineoplastic treatments.

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

This proposal is submitted by Project Data Sphere and Dr. Kerry Reynolds.

References

1. Hodi FS, O'Day SJ et al. Improved Survival with Ipilimumab in Patients with Metastatic Melanoma. *New England Journal of Medicine* 2010; 363:711-723. DOI: 10.1056/NEJMoa1003466
2. <https://www.fda.gov/drugs/resources-information-approved-drugs/oncology-cancer-hematologic-malignancies-approval-notifications>
3. National Comprehensive Cancer Network, NCCN Clinical Practice Guidelines in Oncology. Management of Immunotherapy-Related Toxicity. Version 1.2022, February 28, 2022.
4. Brahmer JR, Abu-Sbeih H, Ascierto PA, et al. Society for Immunotherapy of Cancer (SITC) clinical practice guideline on immune checkpoint inhibitor-related adverse events. *Journal for Immunotherapy of Cancer* 2021;9:e002435. doi:10.1136/jitc-2021-002435
5. Schneider BJ, Naidoo J, Santomaso BD et al. Management of immune-related adverse events in patients treated with immune checkpoint inhibitor therapy: ASCO guideline update. *Journal of Clinical Oncology* 39 (December 20, 2021) 4073-4126. doi:10.1200/JCO.21.01440
6. Arnaud-Coffin, P, Maillet D et al. A systematic review of adverse events in randomized trials assessing immune checkpoint inhibitors. *International Journal of Cancer*. 2019 Aug 1;145(3):639-648. doi: 10.1002/ijc.32132.

TABULAR MODIFICATIONS

D89 Other disorders involving the immune mechanism, not elsewhere classified

D89.8 Other specified disorders involving the immune mechanism, not elsewhere classified

D89.83 Cytokine release syndrome

Code first underlying cause, such as:
complications following infusion, transfusion and therapeutic injection (T80.89-)
complications of transplanted organs and tissue (T86.-)

Add

Use additional code for adverse effect, if applicable, to identify immune checkpoint inhibitors and immunostimulant drugs (T45.AX5)

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

T80 Complications following infusion, transfusion and therapeutic injection

T80.8 Other complications following infusion, transfusion and therapeutic injection

T80.82 Complication of immune effector cellular therapy
Complication of chimeric antigen receptor (CAR-T) cell therapy
Complication of IEC therapy

Add

Excludes2: adverse effect of immune checkpoint inhibitors and immunostimulant drugs (T45.AX5)
complication of bone marrow transplant (T86.0)
complication of stem cell transplant (T86.5)

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

Anal Fistula

This topic was presented at the March 2023 and at both 2022 Coordination and Maintenance meetings. 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. Changes are indicated in **BOLD**.

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 occurrence of a previously undiagnosed or untreated fistula tract is termed as an initial or new fistula. As such, the proposed initial/new fistula codes would apply when a new fistula tract is diagnosed and would continue to apply for the entire duration of treatment and recovery. If that fistula tract does not heal as expected or if the fistula tract heals and then recurs, one of the proposed persistent or recurrent codes would apply. If a new, distinct fistula (e.g., different location), occurs that fistula would also be diagnosed by an initial or new fistula code.

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 recurrence and fecal incontinence.⁷

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

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.

References:

1. Champagne, Bradley J. UpToDate, 2019, <http://www.uptodate.com/contents/operative-management-of-anorectal-fistulas>
2. American Gastroenterological Association Clinical Practice Committee. American Gastroenterological Association medical position statement: perianal Crohn's disease. *Gastroenterology*. 2003;125(5):1503-1507. doi:10.1016/j.gastro.2003.08.024
3. Vogel JD, Johnson EK, Morris AM, et al. Clinical Practice Guideline for the Management of Anorectal Abscess, Fistula-in-Ano, and Rectovaginal Fistula. *Dis Colon Rectum*. 2016;59(12):1117-1133. doi:10.1097/DCR.0000000000000733
4. Vogel, Jon D. UpToDate, 2020, <https://www.uptodate.com/contents/anorectal-fistula-clinical-manifestations-diagnosis-and-management-principles>
5. Panes J, Reinisch W, Rupniewska E, et al. Burden and outcomes for complex perianal fistulas in Crohn's disease: Systematic review. *World J Gastroenterol*. 2018;24(42):4821-4834. doi:10.3748/wjg.v24.i42.4821
6. Williams G, Williams A, Tozer P, et al. The treatment of anal fistula: second ACPGIBI Position Statement - 2018. *Colorectal Dis*. 2018;20 Suppl 3:5-31. doi:10.1111/codi.14054
7. Emile SH. Recurrent anal fistulas: When, why, and how to manage?. *World J Clin Cases*. 2020;8(9):1586-1591. doi:10.12998/wjcc.v8.i9.1586

TABULAR MODIFICATIONS

Add	K50	Crohn's disease [regional enteritis] Use additional code to identify any associated fistulas, if applicable: anal fistula (K60.3-) anorectal fistula (K60.5-) rectal fistula (K60.4-)
Add	K51	Ulcerative colitis Use additional code to identify any associated fistulas, if applicable: anal fistula (K60.3-) anorectal fistula (K60.5-) rectal fistula (K60.4-)
Revise	K60	Fissure and fistula of anal and rectal regions Excludes1: abscess or cellulitis of anal and rectal regions (K61.-) Excludes2: anal sphincter tear (healed) (nontraumatic) (old) (K62.81)

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Add	K60.3	Anal fistula Code first, if applicable: Crohn's disease (K50.-) ulcerative colitis (K51.-)
Add		Excludes1: congenital fistula (Q43.6)
New code Add	K60.30	Anal fistula, unspecified Anal fistula NOS
New sub-subcategory Add Add	K60.31	Anal fistula, simple Low intersphincteric anal fistula Superficial anal fistula
New code Add	K60.311	Anal fistula, simple, initial Anal fistula, simple, new
New code Add	K60.312	Anal fistula, simple, persistent Anal fistula, simple, chronic
New code Add	K60.313	Anal fistula, simple, recurrent Anal fistula simple, occurring following complete healing
New code	K60.319	Anal fistula, simple, unspecified
New sub-subcategory Add Add Add Add	K60.32	Anal fistula, complex Extrasphincteric anal fistula High intersphincteric anal fistula Suprasphincteric anal fistula 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 Add	K60.321	Anal fistula, complex, initial Anal fistula, complex, new
New code Add	K60.322	Anal fistula, complex, persistent Anal fistula, complex, chronic
New code Add	K60.323	Anal fistula, complex, recurrent Anal fistula complex, occurring following complete healing

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

New code		K60.329	Anal fistula, complex, unspecified
Add	K60.4	Rectal fistula	Code first, if applicable: Crohn's disease (K50.-) ulcerative colitis (K51.-)
Add		Excludes1:	congenital fistula (Q43.6)
New code	K60.40	Rectal fistula, unspecified	
Add		Rectal fistula NOS	
New sub-subcategory	K60.41	Rectal fistula, simple	
Add		Low intersphincteric rectal fistula	
Add		Superficial rectal fistula	
New code	K60.411	Rectal fistula, simple, initial	
Add		Rectal, fistula, simple, new	
New code	K60.412	Rectal fistula, simple, persistent	
Add		Rectal fistula, simple, chronic	
New code	K60.413	Rectal fistula, simple, recurrent	
Add		Rectal fistula simple, occurring following complete healing	
New code	K60.419	Rectal fistula, simple, unspecified	
New sub-subcategory	K60.42	Rectal fistula, complex	
Add		Extrasphincteric rectal fistula	
Add		High intersphincteric rectal fistula	
Add		Suprasphincteric rectal fistula	
Add		Transsphincteric rectal fistula	
Add		Code also, if applicable:	
		perianal abscess (K61.0)	
		rectovaginal fistula (N82.3)	
		stenosis of anus and rectum (K62.4)	
New code	K60.421	Rectal fistula, complex, initial	
Add		Rectal fistula, complex, new	
New code	K60.422	Rectal fistula, complex, persistent	
Add		Rectal fistula, complex, chronic	

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

New code Add	K60.423	Rectal fistula, complex, recurrent Rectal fistula complex occurring following complete healing
New code	K60.429	Rectal fistula, complex, unspecified
Add	K60.5	Anorectal fistula Code first, if applicable: Crohn's disease (K50.-) ulcerative colitis (K51.-) Excludes1: congenital fistula (Q43.6)
Add		
New code Add	K60.50	Anorectal fistula, unspecified Anorectal fistula NOS
New sub-subcategory Add Add	K60.51	Anorectal fistula, simple Low intersphincteric anorectal fistula Superficial anorectal fistula
New code Add	K60.511	Anorectal fistula, simple, initial Anorectal fistula, simple, new
New code Add	K60.512	Anorectal fistula, simple, persistent Anorectal fistula, simple, chronic
New code Add	K60.513	Anorectal fistula, simple, recurrent Anorectal fistula simple, occurring following complete healing
New code	K60.519	Anorectal fistula, simple, unspecified
New sub-subcategory Add Add Add Add	K60.52	Anorectal fistula, complex Extrasphincteric anorectal fistula High intersphincteric anorectal fistula Suprasphincteric anorectal fistula Transsphincteric anorectal fistula
Add		Code also, if applicable: perianal abscess (K61.0) rectovaginal fistula (N82.3) stenosis of anus and rectum (K62.4)
New code Add	K60.521	Anorectal fistula, complex, initial Anorectal fistula, complex, new

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

New code Add	K60.522	Anorectal fistula, complex, persistent Anorectal fistula, complex, chronic
New code Add	K60.523	Anorectal fistula, complex, recurrent Anorectal fistula complex, occurring following complete healing
New code	K60.529	Anorectal fistula, complex, unspecified

INDEX MODIFICATIONS

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

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Anosognosia

Anosognosia is a neuropsychiatric condition in which patients remain insistent that they do not have a severe illness.¹ Anosognosia is highly prevalent, affecting 50-98% of individuals with schizophrenia, approximately 40% with bipolar disorder, and about 80% with Alzheimer's disease.³ It also impacts 10-18% of those who develop one-sided paralysis after a stroke and has been found in patients with aphasia, Huntington's, Parkinson's, frontotemporal dementia, and severe traumatic brain injury.⁴ Recognized as a medical condition for over a century,⁵ anosognosia is distinct from psychological denial, arising from damage to the brain's self-image updating mechanism.⁶ Consequently, patients are neurologically incapable of perceiving loss of impaired neurological or neuropsychological function.⁷

Anosognosia substantially hinders treatment, compliance, therapist alliance development, and rehabilitation.⁸ Treatment effectiveness for mild to moderate dementia is impaired in those with anosognosia.⁹ Moreover, caregiver burden for Alzheimer's patients significantly increases with anosognosia.¹⁰ Treating patients with anosognosia often necessitates unique, varied approaches¹¹ and imposes additional responsibilities on medical professionals to overcome patient unawareness of their limitations.¹² Patients are often genuinely puzzled as to why family and caregivers want them to take medication and to participate in other treatments. As a result, patients often refuse them all. Untreated mental illness has harsh consequences, including suicide, rampage shootings, and abject disabilities in highly capable individuals. Such diagnosis will also help the courts in assessing whether involuntary commitment is appropriate.

This condition is currently an inclusion term at code R41.89, Other symptoms and signs involving cognitive functions and awareness. Considering the significant clinical challenges anosognosia imposes on medical professionals, caregivers, and patients, and its prevalence, a unique ICD-10-CM code is being requested.

This proposal has been reviewed and supported by the American Psychiatric Association.

References:

1 Acharya, A. B., & Sánchez-Manso, J. C. (2022, June 7). Anosognosia - StatPearls - NCBI Bookshelf. National Library of Medicine. Retrieved from <https://www.ncbi.nlm.nih.gov/books/NBK513361/>

2 2023 ICD-10-CM diagnosis code R41.89. (n.d.). Retrieved from <https://www.icd10data.com/ICD10CM/Codes/R00-R99/R40-R46/R41-R41.89>

3 Anosognosia: What it is, causes, symptoms & treatment. Cleveland Clinic. (n.d.). Retrieved from <https://my.clevelandclinic.org/health/diseases/22832-anosognosia>

4 Prigatano, G. P. (2009). Anosognosia: Clinical and ethical considerations. *Current Opinion in Neurology*, 22(6), 606–611. DOI: 10.1097/wco.0b013e328332a1e7

5 Weinstein, E. A. (1950). The Syndrome of Anosognosia. *Archives of Neurology and Psychiatry*, 64(6): 772-791, p. 772 n. 1. DOI: 10.1001/archneurpsyc.1950.02310300019002

6 Cleveland Clinic, *supra*; Prigatano, *supra*.

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

7 Prigatano, supra.

8 Id.

9 Koltai, C. C., Welsh-Bohmer, K. A., Schmechel, D. E. (2001). Influence of anosognosia on treatment outcome among dementia patients. *Neuropsychological Rehabilitation: An International Journal*, 11:3-4, 455-475, DOI: 10.1080/09602010042000097

10 Turró-Garriga, O., Garre-Olmo, J., Vilalta-Franch, J., Conde-Sala, J. L., de Gracia Blanco, M., & López-Pousa, S. (2012). Burden associated with the presence of Anosognosia in Alzheimer's disease. *International Journal of Geriatric Psychiatry*, 28(3), 291–297. DOI: 10.1002/gps.3824

11 Jenkinson, P. M., Preston, C., Ellis, S. J. (2011) Unawareness after stroke: A review and practical guide to understanding, assessing, and managing anosognosia for hemiplegia, *Journal of Clinical and Experimental Neuropsychology*, 33:10, 1079-1093, DOI: 10.1080/13803395.2011.596822

12 Id.; Moro, V., Scandola, M., Bulgarelli, C., Avesani, R., & Fotopoulou, A. (2014). Error-based training and emergent awareness in Anosognosia for Hemiplegia. *Neuropsychological Rehabilitation*, 25(4), 593–616. DOI: 10.1080/09602011.2014.951659

TABULAR MODIFICATIONS

R41	Other symptoms and signs involving cognitive functions and awareness
	R41.8 Other symptoms and signs involving cognitive functions and awareness
New Code	R41.85 Anosognosia
	R41.89 Other symptoms and signs involving cognitive functions and awareness
Delete	Anosognosia

Baked Egg Tolerance in Egg Allergy

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

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

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

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

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

There is also evidence that regular ingestion of baked egg in the diet may help children outgrow their egg allergy.^{11,13,16} In one study, subjects ingesting baked egg regularly were 14.6 times more likely than subjects in the comparison group ($P < .0001$) to develop regular egg tolerance, and they

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

developed tolerance earlier (median 50.0 vs 78.7 months; $P < .0001$). Baked egg ingestion was associated with immunologic changes, including decreasing skin prick testing to egg and egg-specific IgE levels, and increasing egg-specific IgG4 levels.¹¹ These immunologic changes parallel those seen in the natural resolution of egg allergy. The authors concluded that initiation of a baked egg diet accelerates the development of regular egg tolerance compared with strict avoidance. Therefore, differentiation of the different phenotypes of egg allergy can lead to different management, specifically earlier and sustained exposure to baked egg in tolerant patients as a form of possible treatment for egg allergy.

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

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

References

1. Rona RJ, Keil T, Summers C, et al. The prevalence of food allergy: a meta-analysis. *J Allergy Clin Immunol* 2007;120:638-46.
2. Savage JH, Matsui EC, Skripak JM, Wood RA. The natural history of egg allergy. *J Allergy Clin Immunol* 2007;120:1413-7.
3. Nowak-Wegrzyn A, Fiocchi A. Rare, medium, or well done? The effect of heating and food matrix on food protein allergenicity. *Curr Opin Allergy Clin Immunol* 2009;9:234-7.
4. Lomakina K, Mikova K. A Study of the Factors Affecting the Foaming Properties of Egg White – a Review. *Czech J Food Sci* 2006;24:110-8.
5. Osborne NJ, Koplin JJ, Martin PE, et al. Prevalence of challenge-proven IgE-mediated food allergy using population-based sampling and predetermined challenge criteria in infants. *J Allergy Clin Immunol* 2011;127:668-76 e1-2.
6. Lemon-Mule H, Sampson HA, Sicherer SH, Shreffler WG, Noone S, Nowak-Wegrzyn A. Immunologic changes in children with egg allergy ingesting extensively heated egg. *J Allergy Clin Immunol* 2008;122:977-83 e1.
7. Urisu A, Ando H, Morita Y, et al. Allergenic activity of heated and ovomucoid-depleted egg white. *J Allergy Clin Immunol* 1997;100:171-6.
8. Leonard SA, Caubet JC, Kim JS, Groetch M, Nowak-Wegrzyn A. Baked milk- and egg-containing diet in the management of milk and egg allergy. *The journal of allergy and clinical immunology In practice* 2015;3:13-23; quiz 4.
9. Vilar LK, Rolins Neto PR, Abdo MA, Cheik MFA, Afonso C, Segundo GRS. Baked egg tolerance: is it possible to predict? *J Pediatr (Rio J)* 2020;96:725-31.
10. Suprun M, Sicherer SH, Wood RA, et al. Mapping Sequential IgE-Binding Epitopes on Major and Minor Egg Allergens. *International archives of allergy and immunology* 2022;183:249-61.
11. Leonard SA, Sampson HA, Sicherer SH, et al. Dietary baked egg accelerates resolution of egg allergy in children. *J Allergy Clin Immunol* 2012;130:473-80 e1.
12. Leonard SA. Debates in allergy medicine: baked milk and egg ingestion accelerates resolution of milk and egg allergy. *The World Allergy Organization journal* 2016;9:1.
13. Peters RL, Dharmage SC, Gurrin LC, et al. The natural history and clinical predictors of egg allergy in the first 2 years of life: a prospective, population-based cohort study. *J Allergy Clin Immunol* 2014;133:485-91.
14. Jarvinen KM, Beyer K, Vila L, Bardina L, Mishoe M, Sampson HA. Specificity of IgE antibodies to sequential epitopes of hen's egg ovomucoid as a marker for persistence of egg allergy. *Allergy* 2007;62:758-65.
15. Bernhisel-Broadbent J, Dintzis HM, Dintzis RZ, Sampson HA. Allergenicity and antigenicity of chicken egg ovomucoid (Gal d III) compared with ovalbumin (Gal d I) in children with egg allergy and in mice. *J Allergy Clin Immunol* 1994;93:1047-59.
16. Sicherer SH, Wood RA, Vickery BP, et al. The natural history of egg allergy in an observational cohort. *J Allergy Clin Immunol* 2014;133:492-9

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

TABULAR MODIFICATIONS

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

T78.0 Anaphylactic reaction due to food
Anaphylactic reaction due to adverse food reaction
Anaphylactic shock or reaction due to nonpoisonous foods
Anaphylactoid reaction due to food

New code
New code

T78.08 Anaphylactic reaction due to eggs
T78.081 Anaphylactic reaction due to eggs
T78.082 Anaphylactic reaction due to eggs, but tolerant to
baked eggs

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

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

New code
New code

T78.13 Other adverse food reaction due to eggs
T78.14 Other adverse food reaction due to eggs, but tolerant to
baked eggs

Z91 Personal risk factors, not elsewhere classified

Z91.0 Allergy status, other than to drugs and biological substance

Z91.01 Food allergy status

New code

Z91.016 Allergy to eggs, but tolerant to baked egg

Baked Milk Tolerance in Milk Allergy

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

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

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

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

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

There is also evidence that regular ingestion of baked milk in the diet may help children outgrow their milk allergy. In one study, subjects ingesting baked milk regularly were 16 times more likely than subjects in the comparison group ($P < .0001$) to develop regular milk tolerance.¹⁰ Baked milk ingestion was associated with immunologic changes, including decreasing skin prick testing to CM, and increasing casein-specific IgG4 levels.^{5,10} These immunologic changes parallel those seen

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

in the natural resolution of milk allergy. The authors concluded that initiation of a baked milk diet accelerates the development of regular CM tolerance compared with strict avoidance. Therefore, differentiation of the different phenotypes of milk allergy can lead to different management, specifically earlier and sustained exposure to baked milk in tolerant patients as a form of possible treatment for milk allergy.

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

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

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

References

1. Rona RJ, Keil T, Summers C, et al. The prevalence of food allergy: a meta-analysis. *J Allergy Clin Immunol* 2007;120:638-46.
2. Skripak JM, Matsui EC, Mudd K, Wood RA. The natural history of IgE-mediated cow's milk allergy. *J Allergy Clin Immunol* 2007;120:1172-7.
3. Nowak-Wegrzyn A, Fiocchi A. Rare, medium, or well done? The effect of heating and food matrix on food protein allergenicity. *Curr Opin Allergy Clin Immunol* 2009;9:234-7.
4. Lomakina K, Mikova K. A Study of the Factors Affecting the Foaming Properties of Egg White – a Review. *Czech J Food Sci* 2006;24:110-8.
5. Nowak-Wegrzyn A, Bloom KA, Sicherer SH, et al. Tolerance to extensively heated milk in children with cow's milk allergy. *J Allergy Clin Immunol* 2008;122:342-7, 7 e1-2.
6. Leonard SA. Debates in allergy medicine: baked milk and egg ingestion accelerates resolution of milk and egg allergy. *The World Allergy Organization journal* 2016;9:1.
7. Bartnikas LM, Sheehan WJ, Hoffman EB, et al. Predicting food challenge outcomes for baked milk: role of specific IgE and skin prick testing. *Ann Allergy Asthma Immunol* 2012;109:309-13 e1.
8. Caubet JC, Nowak-Wegrzyn A, Moshier E, Godbold J, Wang J, Sampson HA. Utility of casein-specific IgE levels in predicting reactivity to baked milk. *J Allergy Clin Immunol* 2013;131:222-4 e1-4.
9. Leonard SA, Caubet JC, Kim JS, Groetch M, Nowak-Wegrzyn A. Baked milk- and egg-containing diet in the management of milk and egg allergy. *The journal of allergy and clinical immunology In practice* 2015;3:13-23; quiz 4.
10. Kim JS, Nowak-Wegrzyn A, Sicherer SH, Noone S, Moshier EL, Sampson HA. Dietary baked milk accelerates the resolution of cow's milk allergy in children. *J Allergy Clin Immunol* 2011;128:125-31 e2.
11. Wood RA, Sicherer SH, Vickery BP, et al. The natural history of milk allergy in an observational cohort. *J Allergy Clin Immunol* 2013;131:805-12.
12. Chatchatee P, Jarvinen KM, Bardina L, Beyer K, Sampson HA. Identification of IgE- and IgG-binding epitopes on alpha(s1)-casein: differences in patients with persistent and transient cow's milk allergy. *J Allergy Clin Immunol* 2001;107:379-83.
13. Caubet JC, Lin J, Ahrens B, et al. Natural tolerance development in cow's milk allergic children: IgE and IgG4 epitope binding. *Allergy* 2017;72:1677-85.

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

TABULAR MODIFICATIONS

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

T78.0 Anaphylactic reaction due to food
T78.07 Anaphylactic reaction due to milk and dairy products

New code T78.070 Anaphylactic reaction due to milk and dairy products

New code T78.071 Anaphylactic reaction due to milk and dairy products, but tolerant to baked milk

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

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

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

New code T78.12 Other adverse food reactions due to milk and dairy products, but tolerance to baked milk

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

Z91 Personal risk factors, not elsewhere classified

Z91.0 Allergy status, other than to drugs and biological substance

Z91.01 Food allergy status

Z91.011 Allergy to milk products

Excludes1: lactose intolerance (E73.-)

New code Z91.015 Allergy to milk products, but tolerant to baked milk

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Bicuspid Aortic Valve

Bicuspid aortic valve (BAV) is the most common congenital heart valve defect, occurring in approximately 1.5% of the population.¹ In BAV, the aortic valve has two, not the typical three, cusps (leaflets), thus markedly increasing the risk of aortic valve calcification and aortic aneurysm. The functional consequences of BAV are: 1) the aortic valve has impaired ability to prevent blood from leaking from the aorta back into the heart (aortic regurgitation); 2) the aortic valve becoming stiff and not opening as well, causing the heart to have to pump harder than usual to get blood past the valve (aortic stenosis); 3) the ascending aorta of individuals with BAV becoming enlarged (dilated or aneurysmal), leading to aortic dissection or an aortic aneurysm; and 4) increased risk of endocarditis (heart valve infection).²

The presence of a BAV is associated with an 80% lifetime risk of requiring aortic valve surgery and a 20% lifetime risk of requiring aortic surgery for aortic aneurysm.³ Individuals diagnosed with BAV should receive ongoing assessment and care to treat co-incident hypertension and heart failure and plan timing of surgery. The American Heart Association (AHA) and American College of Cardiology (ACC) Guidelines recommend longitudinal imaging of the aortic valve and ascending aorta (echocardiogram, CT scan, or MRI) in people with BAV.⁴ Further, the Guidelines recommend echocardiographic imaging of first-degree relatives of probands with BAV,^{4,5} which is rarely systematically performed.

The absence of a specific ICD-10-CM code for BAV impairs longitudinal follow-up. For illustration, of the 10,810 individuals with known BAV within the Mass General Brigham Healthcare System (Boston, MA) based on chart review, 55% do not have a recorded ICD-10-CM code for BAV (from category Q23 or I35).

The proposed Tabular Modifications below will:

- i. Increase accurate coding of bicuspid aortic valve disease
- ii. Separate congenital mitral valve disease from congenital aortic valve disease (at Q23)

The anticipated consequences of these changes will:

- i. Enable improved identification of individuals with BAV.
- ii. Improve longitudinal follow-up by facilitating implementation of EHR-based reminders to the patient and clinician for Guideline-driven follow-up.⁶
- iii. Assist in the identification of first-degree relatives of BAV probands
- iv. Enable EHR-based research of BAV and its morbidities.

A cleft mitral valve leaflet is a congenital heart valve abnormality that causes mitral regurgitation, but is currently coded to Q23.8, Other congenital malformations of aortic and mitral valves. BAV may also be coded to Q23.8, in particular in cases where congenital insufficiency of aortic valve is not present. In order to differentiate these two different valve abnormalities to facilitate accurate coding, it has been proposed also to create a separate code for congenital mitral valve cleft leaflet.

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

This proposal is made jointly, by the GenTAC Alliance, A Division of The Marfan Foundation (<https://www.gentacalliance.org/>), and the Bicuspid Aortic Foundation (<https://www.bicuspidfoundation.org/>), with input and support from the following clinical experts: Kim Eagle, University of Michigan, Ann Arbor, MI; Scott LeMaire, Baylor College of Medicine, Houston, TX; Siddharth Prakash, University of Texas, Houston, TX; Eric Isselbacher, Massachusetts General Hospital, Boston, MA; Simon Body, Boston University School of Medicine, Boston, MA.

References

1. Fedak PWM, Verma, S, David TE, et al. Clinical and pathophysiological implications of a bicuspid aortic valve. *Circulation*. 2002; 106:900-904. <https://doi.org/10.1161/01.CIR.0000027905.26586.E8>
2. Kiyota Y, Della Corte A, Montiero Vieira V, et al. Risk and outcomes of aortic valve endocarditis among patients with bicuspid and tricuspid aortic valves. *Open Heart*. 2017 May 16;4(1):e000545. <https://doi.org/10.1136/openhrt-2016-000545>.
3. Della Corte A, Body SC, Booher AM, et al. Surgical treatment of bicuspid aortic valve disease: knowledge gaps and research perspectives. *J Thorac Cardiovasc Surg*. 2014 Jun;147(6):1749-57, 1757.e1. <https://doi.org/10.1016/j.jtcvs.2014.01.021>.
4. Otto, CM, Nishimura, RA, Bonow, RO, et al. 2020 ACC/AHA guideline for the management of patients with valvular heart disease: A report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *Circulation*. 2021; 143:e72-e227. <https://doi.org/10.1161/CIR.0000000000000923>
5. Isselbacher EM, Preventza O, Hamilton Black J 3rd, et al. 2022 ACC/AHA Guideline for the Diagnosis and Management of Aortic Disease: A Report of the American Heart Association/American College of Cardiology Joint Committee on Clinical Practice Guidelines. *J Am Coll Cardiol*. 2022 Oct 28;S0735-1097(22)05769-2. <https://doi.org/10.1016/j.jacc.2022.08.004>.
6. Perri-Moore S, Kapsandoy S, Doyon K, et al. Automated alerts and reminders targeting patients: A review of the literature. *Patient Educ Couns*. 2016 Jun;99(6):953-9. <https://doi.org/10.1016/j.pec.2015.12.010>.

TABULAR MODIFICATIONS

	I35	Nonrheumatic aortic valve disorders
Add		Code also, if applicable, bicuspid aortic valve (Q23.81)
Delete		Excludes1: aortic valve disorder of unspecified cause but with diseases of mitral and/or tricuspid valve(s) (I08.-)
Delete		aortic valve disorder specified as congenital (Q23.0, Q23.1)
Delete		aortic valve disorder specified as rheumatic (I06.-)
Delete		hypertrophic subaortic stenosis (I42.1)
Add		Excludes2: aortic valve disorder of unspecified cause but with diseases of mitral and/or tricuspid valve(s) (I08.-)
Add		aortic valve disorder specified as congenital (Q23.0, Q23.1)
Add		aortic valve disorder specified as rheumatic (I06.-)
Add		hypertrophic subaortic stenosis (I42.1)
	Q23	Congenital malformations of aortic and mitral valves
	Q23.1	Congenital insufficiency of aortic valve
Delete		Bicuspid aortic valve

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Q23.8 Other congenital malformations of aortic and mitral valves

New code	Q23.81	Bicuspid aortic valve
Add		Bicuspid aortic valve at birth
Add		Congenital bicuspid aortic valve
Add		Unicuspid (congenital) aortic valve (at birth)
Add		Code also, if applicable, acquired aortic valve disorders, such as:
Add		aortic (valve) insufficiency (nonrheumatic) (I35.1)
Add		aortic (valve) stenosis (nonrheumatic) (I35.0)
Add		aortic (valve) stenosis with insufficiency (nonrheumatic) (I35.2)
Add		Excludes2: functional bicuspid aortic valve (with stenosis) (I35.0)
New code	Q23.82	Congenital mitral valve cleft leaflet
Add		Cleft mitral valve leaflet at birth
New code	Q23.88	Other congenital malformations of aortic and mitral valves

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Cement Pulmonary Embolism and Fat Pulmonary Embolism

The National Center for Health Statistics received a proposal requesting new ICD-10-CM codes for cement embolism of pulmonary artery and fat embolism of pulmonary artery with and without acute cor pulmonale from the Agency for Healthcare Research and Quality (AHRQ).

A fat embolism is when one or more droplet-like particles of fat enter the bloodstream and embolize through the systemic or pulmonary circulation.³ Fat emboli can form when an individual breaks a bone, particularly with more severe or complex fractures, although the reported incidence is generally below 10%,^{11,12} except in autopsy series.¹⁰ Long bone and pelvic fractures are the most frequent causes, but fat emboli can occur with other medical conditions or circumstances, such as burns, liposuction, lipo-injection, fat grafting, panniculitis, orthopedic surgeries, and cardiopulmonary resuscitation.¹³ Fat embolization occurs early, within 24-72 hours of lower extremity fractures and even earlier after surgical procedures requiring intramedullary reaming and insertion techniques.⁸

The main goal of treatment of a fat pulmonary embolism is to provide supportive care. There is no cure for fat pulmonary embolism, and treatments can include fluid resuscitation, oxygenation, and when indicated, noninvasive or invasive mechanical ventilation.¹³

Pulmonary cement embolism refers to embolization of polymethyl methacrylate (PMMA) into the lungs.⁴ The PMMA leaks into the vertebral vessels and then hardens in the pulmonary vasculature after traveling through the right side of the heart.⁵ A pulmonary cement embolism (PCE) is an iatrogenic form of pulmonary embolism⁵ caused by the use of PMMA during vertebroplasty procedures or to secure hip or knee prostheses during arthroplasty procedures.^{5,8} A study by Duran and colleagues of 128 percutaneous vertebroplasties found an incidence rate of pulmonary cement embolism of 6.8%.¹

The majority of PCEs are asymptomatic, but symptomatic PCEs often present with chest pain, tachycardia, signs of severe respiratory distress, and hypotension.⁹ This condition is known as bone cement implantation syndrome (BCIS), which is a poorly understood syndrome that may ultimately cause neurologic deficits and cardiac arrest.² The primary pathophysiologic mechanism for BCIS is debated, but probably involves increased pulmonary vascular resistance and right heart compromise or failure due to PCE. Treatments include embolectomy and supportive care and observation,⁹ depending on the patient's symptoms.⁷ Cement pulmonary embolism involves mechanical occlusion of pulmonary arteries and not a vascular clot.⁷ Computer tomography angiogram (CTA) allows visualization of cement within the pulmonary vasculature.⁹

Treatment of cement and fat pulmonary emboli is entirely different than treatment of an embolism due to blood clot. Specifically, there is no indication for systemic anticoagulation when the embolic material is cement or fat.

Creation of new ICD-10-CM codes for cement pulmonary embolism and fat pulmonary embolism with and without acute cor pulmonale will provide coding specificity for research, tracking, and trending.

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

References

- ¹ Duran C, Sirvanci M, Aydoğan M, Ozturk E, Ozturk C, Akman C. Pulmonary cement embolism: a complication of percutaneous vertebroplasty. *Acta Radiol.* 2007 Oct;48(8):854-9. doi: 10.1080/02841850701422153.
- ² Donaldson AJ, Thomson HE, Harper NJ, Kenny NW. Bone cement implantation syndrome. *Br J Anaesth.* 2009 Jan;102(1):12-22. doi: 10.1093/bja/aen328.
- ³ Maitre S. Causes, clinical manifestations, and treatment of fat embolism. *Virtual Mentor.* 2006 Sep 1;8(9):590-2. doi: 10.1001/virtualmentor.2006.8.9.cprl1-0609. PMID: 23234710.
- ⁴ Weerakkody Y, El-Feky M, El-Feky M, et al. Pulmonary cement embolism. Reference article, Radiopaedia.org (Accessed on 12 Dec 2022) <https://doi.org/10.53347/rID-22701>
- ⁵ Baston B, Sigler M, Cement emboli, 2016 December 26, <https://pulmonarychronicles.com/index.php/pulmonarychronicles/article/view/376>.
- ⁷ Satyanarayana S, Santosh H K, Sinha N, Padegal V, *Lung India.* 2015 Nov-Dec; 32(6): 602–605, doi: 10.4103/0970-2113.168119
- ⁸ Issack P S, Lauerman M H, Helfet D L, Sculco T P, Lane J M, 2009 February, *The American Journal of Orthopedics, Fat Embolism and Respiratory Distress Associated With Cemented Femoral Arthroplasty*, <https://cdn.mdedge.com/files/s3fs-public/Document/September-2017/038020072.pdf>
- ⁹ Waler A R, Sanchez K J, Parikh A A, Okorie O N, Hindawi, A Case of Pulmonary Cement Embolism Managed through Symptomatic Treatment. 2020 June 05, Volume 2020 | Article ID 2425973 | <https://doi.org/10.1155/2020/2425973>
- ¹⁰ Eriksson EA, Pellegrini DC, Vanderkolk WE, Minshall CT, Fakhry SM, Cohle SD. Incidence of pulmonary fat embolism at autopsy: an undiagnosed epidemic. *J Trauma.* 2011 Aug;71(2):312-5. doi: 10.1097/TA.0b013e3182208280.
- ¹¹ Müller C, Rahn BA, Pfister U, Meinig RP. The incidence, pathogenesis, diagnosis, and treatment of fat embolism. *Orthop Rev.* 1994 Feb;23(2):107-17. PMID: 8196970.
- ¹² Stein PD, Yaekoub AY, Matta F, Kleerekoper M. Fat embolism syndrome. *Am J Med Sci.* 2008 Dec;336(6):472-7. doi: 10.1097/MAJ.0b013e318172f5d2. PMID: 19092320.
- ¹³ Weinhouse GL. Fat embolism syndrome. *UpToDate.* 2023. <https://www.uptodate.com/contents/fat-embolism-syndrome>.

TABULAR MODIFICATIONS

	I26	Pulmonary embolism	
	I26.0	Pulmonary embolism with acute cor pulmonale	
New code	I26.03	Cement embolism of pulmonary artery with acute cor pulmonale	
New code	I26.04	Fat embolism of pulmonary artery with acute cor pulmonale	
Add	I26.09	Other pulmonary embolism with acute cor pulmonale Other thrombotic pulmonary embolism with acute cor pulmonale	

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

I26.9 Pulmonary embolism without acute cor pulmonale

Revise	I26.93	Single subsegmental <u>thrombotic</u> pulmonary embolism without acute cor pulmonale
Revise	I26.94	Multiple subsegmental <u>thrombotic</u> pulmonary emboli without acute cor pulmonale
New code	I26.95	Cement embolism of pulmonary artery without acute cor pulmonale
New code	I26.96	Fat embolism of pulmonary artery without acute cor pulmonale
Add	I26.99	Other pulmonary embolism without acute cor pulmonale Other thrombotic pulmonary embolism without acute cor pulmonale

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

Coding of Firearm Injuries Default

This is a representation of the proposal that was originally presented at the March 2023 ICD10 Coordination and Maintenance meeting.

The National Center for Health Statistics received a proposal to change the default in the Alphabetic Index for External Causes. The proposal that the default code listed in the Index for External Causes for “gunshot wound” be changed from “W34, accident by unspecified firearm,” to “X95.9, assault by unspecified firearm.”

The change is in the intent category to which firearm injuries would default to (assault) unless another intent category (accidental, intentional self-harm, legal intervention, terrorism or undetermined) is indicated by the medical documentation.

According to the National Electronic Injury Surveillance System (NEISS), 80% of nonfatal firearm injuries are assaults. For each of the other 22 broad mechanisms of injury (e.g., falls, motor vehicles, cutting/piercing instrument, poisoning, etc.) the vast majority (mean 91%, range 75-100%) are accidents. Default codes in the Alphabetic Index represent “that condition that is most commonly associated with the main term or is the unspecified code for the condition”.

Statewide and national hospital discharge data systems (like the Nationwide Emergency Department Sample [NEDS]) are vital for firearm injury surveillance but have one major flaw. According to a recent NORC report., NEDS reports that accidents are the leading type of firearm injury, while injury-focused data systems find assaults are the leading type.

The proposal is being requested by: Safe States and by Deborah Azrael, Catherine Barber (Harvard Injury Control Research Center, Harvard T. H. Chan School of Public Health); Matthew Miller (Northeastern University); Eric Goralnick, Erin MacPhaul, Ravali Yenduri, Li Zhou (Mass General Brigham Hospitals); Andrew Bowen, Steve Mooney, Ali Rowhani-Rahbar (Harborview Injury Prevention and Research Center, University of Washington)

Please note that coding generally requires direct documentation by the patient’s provider. However, the ICD-10-CM Official Guidelines for Coding and Reporting will be updated to show as an exception: *firearm injuries by intent, such as assault (X93-X95), or undetermined intent (Y22-Y24)*, to 1.B.14, Documentation by Clinicians Other than the Patient's Provider.

The proposal was reviewed by CDC/NCHS, Division of Analysis and Epidemiology (DAE), they are in support of option 2.

References

CDC National Center for Injury Prevention and Control and U.S. Consumer Product Safety Commission. National Electronic Injury Surveillance System ([NEISS](#)) All Injury Program, 2016. ICPSR [distributor], 2020-08-19.
Roman J & Cook P eds. [Improving Data Infrastructure to Reduce Firearms Violence](#). NORC at the University of Chicago, October 2021
Olufajo OA, Zeineddin A, Nonez H, et al. Trends in firearm injuries among children and teenagers in the United States. *J Surg Res*. 2020 Jan;245:529-536. doi: 10.1016/j.jss.2019.07.056. (Includes data on all age groups.)
WRISS data from Beth Hume, Injury Surveillance Program, Massachusetts Department of Public Health.

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Option #1 [Changing default to assault]

EXTERNAL CAUSE OF INJURY INDEX MODIFICATIONS

Discharge (accidental)

- Revise - firearm (~~accidental~~) ~~W34.00~~ X95.9
- Add -- accidental W34.00
- Revise -- handgun (pistol) (revolver) ~~W32.0~~ X93
- Add --- accidental W32.0
- Revise -- hunting rifle ~~W33.02~~ X94.1
- Add --- accidental W33.02
- Revise -- larger ~~W33.00~~ X94.9
- Add --- accidental W33.00
- Revise --- specified NEC ~~W33.09~~ X94.8
- Add ---- accidental W33.09
- Revise -- machine gun ~~W33.03~~ X94.2
- Add --- accidental W33.03
- Revise -- shotgun ~~W33.01~~ X94.0
- Add --- accidental W33.01

- Revise **Gunshot wound** (see also Discharge, firearm, by type) ~~W34.00~~ X95.9

Option #2 [Changing default to undetermined, shown below].

EXTERNAL CAUSE OF INJURY INDEX MODIFICATIONS

Discharge (accidental)

- Revise - firearm (~~accidental~~) ~~W34.00~~ Y24.9
- Add -- accidental W34.00
- Revise -- handgun (pistol) (revolver) ~~W32.0~~ Y22
- Add --- accidental W32.0
- Revise -- hunting rifle ~~W33.02~~ Y23.1
- Add --- accidental W33.02
- Revise -- larger ~~W33.00~~ Y23.9
- Add --- accidental W33.00
- Revise --- specified NEC ~~W33.09~~ Y23.8
- Add ---- accidental W33.09
- Revise -- machine gun ~~W33.03~~ Y23.3
- Add --- accidental W33.03
- Revise -- shotgun ~~W33.01~~ Y23.0
- Add --- accidental W33.01

- Revise **Gunshot wound** (see also Discharge, firearm, by type) ~~W34.00~~ Y24.9

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

DLG4-related synaptopathy

The SHINE Syndrome Foundation has requested creation of a specific ICD-10-CM code for DLG4-related synaptopathy, a form of non-specific syndromic intellectual disability. The letters of SHINE represent findings in this disorder: Sleep disruption, Hypotonia, Intellectual disability, Neurological disorders such as autism, and Epilepsy. While there is some variability, patients with DLG4-related synaptopathy often present with similar characteristics. Language development is variably impaired, with some children speaking with isolated words, associations of two or three words, or with short sentences, whereas others remain non-verbal. Some of the children show oral dyspraxia, which can result in some drooling or eating difficulties. The appearance and the growth of children with DLG4-related synaptopathy are not unusual. Children (and presumably adults) with DLG4-related synaptopathy continue along a normal growth curve. Several children, the majority of whose epilepsy is not well controlled, do regress; others can slowly continue to learn.

The gene *DLG4* encodes the protein PSD-95 (Postsynaptic Density Protein 95), which plays a major role in brain development and function through its implications in synaptic strength and plasticity. These mechanisms, along with PSD-95's role in organizing and interacting with other proteins, represent a gene with many capabilities that, when altered by variants, can be associated with DLG4-related synaptopathy. PSD-95 is found at the junctions between nerve cells, more specifically at the post synapses, where cell-to-cell communication occurs. PSD-95 helps regulate synapse adaptations and promotes proper brain wiring. The protein's function is particularly important during early brain development, affecting future cognitive ability.

Since initial description of DLG4-related synaptopathy in 2016 by Lelieveld et al., an increasing number of patients have been identified, suggesting that it may represent a more common cause of intellectual disability than first anticipated. At least 44 different *DLG4* variants have been found to cause it. In addition to mild-to-profound intellectual disability, this condition commonly features other neurological symptoms, including recurrent seizures and autism spectrum disorder. Most pathogenic *DLG4* variants are predicted to affect the production of functional PSD-95 protein from one allele, leading to haploinsufficiency. A reduction of PSD-95 can have multiple effects in nerve cells, leading to cognitive impairment and other neurological problems. Intellectual disability (ID) is a common diagnosis defined by the presence of significant limitations in both cognitive and adaptive behaviors with onset before the age of 18. Non-specific syndromic intellectual disability patients, including those associated with a *DLG4* variant, typically exhibit moderate to profound ID with varying degrees of epilepsy and/or autism spectrum disorders (ASD), and may also have attention deficits, hypotonia, impulsivity, and/or mood disorders.

Children with ID should be genetically screened for the potential involvement of genes. The presence of a generalized form of epilepsy (recognizable by physicians by the type of seizures and the EEG pattern) is consistent with the diagnosis and approximately 1/3 of DLG4 patients with epilepsy show ESES (Electrical Status Epilepticus during Slow-Wave Sleep). Brain imaging techniques such as MRI usually do not show any specific neural abnormalities, however some individuals with a diagnosis of DLG4-related synaptopathy present with delayed myelination, thinning of the corpus callosum, and/or white matter loss.

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

Given that the described clinical presentation is common and associated with numerous genetic diagnoses, mainly other synaptopathies, specific genetic testing for variants in *DLG4* results in a low yield. More recently, physicians are requesting more comprehensive genomic testing approaches, such as next-generation sequencing (NGS) techniques, exome sequencing (ES) and whole-genome sequencing (WGS), for the genetic diagnosis of children with global developmental delay (GDD)/ID. By interpreting the NGS data physicians are able to increase their likelihood of finding an underlying genetic etiology. Recommendations are necessary as there is an increasing number of children with this disorder. A team of *DLG4* clinicians and researchers recently published a chapter in GeneReviews highlighting these recommendations.

The symptoms caused by *DLG4* variants appear to be equally prevalent in males and females. The disorder is recognizable early during childhood. However, because affected children are generally healthy, this disorder may be as prevalent in the adult population as it is in children. Since this disorder is still quite rare, the data is limited and it is uncertain of prevalence within any one ethnic group.

DLG4-related synaptopathy may be considered together with developmental and epileptic encephalopathies (DEEs), certain of which were considered at the March 2023 C&M meeting. Following the approach and using genetic data from López-Rivera from 2020, as well as Lemke from 2020, the predicted U.S. incidence of new cases of DLG4-related synaptopathy among newborns would be in the range of 134 to 156 new cases annually, based on *de novo* mutation rates. However, this estimate could be either low or high for a number of reasons (see notes on DEEs from March 2023 C&M for more details). This would suggest that there could be as many as thousands of cases of existing undiagnosed DLG4-related synaptopathy in the U.S.

Like other forms of intellectual disability, there is no definitive cure for DLG4-related synaptopathy. Current practice is symptomatic treatment directed toward the specific symptoms that are apparent in each individual. Management may require the coordinated efforts of a team of specialists. Starting with early developmental intervention it is important to monitor any additional therapies that could be a positive resource dependent upon the specific abnormalities present. Following standard guidelines assessing for ADHD, mobility, strabismus, cerebral or cortical visual impairment (CVI), autism, anxiety, or sleep disturbances annually or as clinically indicated is potentially beneficial. Anti-seizure medications are usually effective in treating seizures for those patients that present with epilepsy; however, in a subset of patients, these medications do not work (refractory seizures) requiring non-pharmacological treatments such as epilepsy surgery or neurostimulation.

There is a concerted, worldwide effort to develop precision therapies for patients with genetic forms of non-specific syndromic intellectual disability. With respect to DLG4-related synaptopathy, evidence indicates that pathogenic *DLG4* variants disrupt the normal function of synapses. There are several current treatment options in development for *DLG4*, including AAV9 gene therapy, antisense oligonucleotide (ASO), and screening of pharmacological options which could alleviate some of the symptoms of individuals with *DLG4* variants.

The SHINE Syndrome Foundation has launched two online Natural History Study Registries. The Simons Searchlight registry compares patients with DLG4-related synaptopathy to other

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

neurodevelopmental disorders; it can be accessed through a convenient, secure, online platform for patients or their legally authorized representative to report cases of DLG4-related synaptopathy. The organization also launched a CoRDS registry (Coordination of Rare Diseases at Sanford registry) to get more personalized data specific to DLG4-related synaptopathy.

DLG4-related synaptopathy is a sporadic condition mostly caused by *de novo* variants of *DLG4*. Having a specific ICD-10-CM code for DLG4-related synaptopathy will allow physicians to better determine appropriate therapy for their patients and assess possible prognosis. This will also aid in the accuracy of monitoring and tracking efforts, enhancing clinicians' and researchers' efforts to identify prospective patients who will benefit from new treatments.

References

- Coordination of Rare Diseases at Sanford (CoRDS). Rare disease registry. <https://research.sanfordhealth.org/rare-disease-registry>
- Kniffin CL. Intellectual developmental disorder, autosomal dominant 62. OMIM. 2022. <https://www.omim.org/entry/618793>
- Lelieveld SH, Reijnders MRF, Pfundt R, et al. Meta-analysis of 2,104 trios provides support for 10 new genes for intellectual disability. Nat Neurosci. 2016 Sep;19(9):1194-6. Epub 2016 Aug 1. <https://doi.org/10.1038/nm.4352>
- Lemke JR. Predicting incidences of neurodevelopmental disorders. Brain. 2020 Apr 1;143(4):1046-1048. <https://doi.org/10.1093/brain/awaa079>
- López-Rivera JA, Pérez-Palma E, Symonds J, et al. A catalogue of new incidence estimates of monogenic neurodevelopmental disorders caused by de novo variants. Brain, Volume 143, Issue 4, April 2020, Pages 1099–1105. <https://doi.org/10.1093/brain/awaa051>
- SHINE Syndrome Foundation. <https://shinesyndrome.org/wp/>
- Simons Searchlight. <https://www.simonssearchlight.org/>
- Tümer Z, Dye TJ, Prada C, White-Brown AM, MacKenzie A, Levy AM. DLG4-Related Synaptopathy. GeneReviews®; Univ. of Wash, Seattle. NLM. NIH. Initial Posting: June 22, 2023. <https://www.ncbi.nlm.nih.gov/books/NBK592682/>

TABULAR MODIFICATIONS

Option #1

F78 Other intellectual disabilities

F78.A Other genetic related intellectual disabilities

New code F78.A2 DLG4-related synaptopathy

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

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Option #2

Delete	F88	Other disorders of psychological development
Delete		Developmental agnosia
Delete		Global developmental delay
		Other specified neurodevelopmental disorder
New subcategory	F88.0	Other genetic neurodevelopmental disorders
New code	F88.01	DLG4-related synaptopathy
Add		Code also any associated disorders, such as:
Add		attention-deficit hyperactivity disorders (F90.-)
Add		autism spectrum disorder (F84.0)
Add		epilepsy, by specific type (G40.-)
Add		intellectual disabilities (F70-F79)
Add		pervasive developmental disorders (F84.-)
New code	F88.09	Other genetic neurodevelopmental disorder
New code	F88.8	Other disorders of psychological development
Add		Developmental agnosia
Add		Global developmental delay
Add		Other specified neurodevelopmental disorder

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Duffy Phenotype

The American Society of Hematology (ASH) has requested creation of new ICD-10-CM codes to represent Duffy phenotype. In addition to Duffy antigens being one of the minor blood group antigens which can be associated with transfusion reactions, there is an association of the Duffy null phenotype with a lower range for the absolute neutrophil count (ANC).

The association between lower absolute neutrophil count (ANC) and African or Arab ancestry has been well established since the 1970s. Genome wide association sequencing in the 2000s established a genetic cause. Homozygosity for a single nucleotide polymorphism (SNP) rs2814778 in the promoter region of the *Duffy antigen receptor for chemokines DARC* gene (also known as the *atypical chemokine receptor ACKR1* gene) produces the Duffy null or Fy(a-b-) phenotype, which is the driver of the lower ANC observed in most of those of African and Arab ancestry.¹ The Fy+ allele at rs2814778 is found in 99.3% of Europeans but only 0.2% of Africans.¹ Further, the Fy(a-b-) phenotype is found in <1% of those with White or Asian ancestry but is very common in individuals from sub-Saharan Africa (80-100%) and the Arabian Peninsula (50-70%).²⁻⁶ Homozygosity of the SNP at rs2814788 is very strongly associated with lower neutrophil count ($p=4.09 \times 10^{-5}$), and association of lower ANC with race is abrogated when accounting for Duffy status.⁷ A prospective study in the U.S. among health Black individuals presenting for primary care found no difference in ANC among Black Duffy non-null individuals than the institutional reference range, but Black Duffy null individuals had a significantly lower median ANC (2820 cells/uL).⁸ Additionally, nearly a quarter of healthy Duffy null individuals had ANC below the institutional lower limit of normal.⁸ This normal, healthy variant of lower circulating neutrophils is now referred to as Duffy-null associated neutrophil count (DANC).⁸

Thus, the current ANC range in the U.S. is neither inclusive or nor accurate for a significant percentage of individuals with the Fy(a-b-) phenotype. An accurate ANC reference range is foundational to adequate care.

Inaccurate reference ranges lead to over-testing, unnecessary referrals, inappropriate medication discontinuation, and delays in chemotherapy administration.⁹⁻¹¹ Additionally, ANC is used to assess eligibility and toxicity grading in clinical trials: inaccurate ANC reference ranges impact enrollment and lead to reporting of false “adverse events.”^{12,13} In a study examining potential barriers to participation in prostate cancer trials, 47.2% of trials required an ANC of 1500 or higher for participation.¹⁴ A recent study recommended a new ANC reference range of 1210-5390 cells/uL for those with Duffy null phenotype, significantly different than the ANC reference range of 1900-7500 cells/uL for all other individuals.¹⁵ This hospital system currently publishes the Duffy null range as a comment below the institutional reference range as there is no simple demographic status or other way to enable an electronic medical record system to call up the Duffy null-specific reference range like there is, for instance, for sex-specific hemoglobin reference ranges.

There is much evidence that shows that Duffy null status is associated with lower circulating neutrophil counts without any known negative clinical consequences. Several studies have indicated that the Duffy-null genotype causes a change in the morphology of neutrophils,

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

facilitating their migration into tissues, thus reducing the number of circulating neutrophils and causing an apparent neutropenia.^{16,17} This mechanism is thought to be clinically benign because the production and functioning of neutrophils is not reduced and so their ability to fight infection remains unchanged.¹⁸ The importance of this is that it is an “apparent neutropenia” and not at all a true neutropenia.

There is a need for ways to represent the results of Duffy testing that will allow for accurate documentation, allay clinical concerns, and importantly, prevent unnecessary further testing. Some clinicians now may use ICD-10-CM code D70.9, Neutropenia, unspecified; however, this is not an accurate diagnosis code to report for someone with the Duffy null phenotype, as that code implies an abnormal state and assumes that all Duffy null patients will have an ANC below the traditional neutropenia threshold (<1500 cells/uL), which is only seen in about 10% of Duffy null patients.

Duffy is an antigen on the red blood cell membrane, easily tested and documented, and analogous to the major blood types or Rh status. ASH recommends creation of ICD-10-CM codes representing Duffy phenotype within the Z codes, and paralleling Rh status, with representation of the following phenotypes.

- Duffy null [Fy(a-b-)]
- Duffy a positive [Fy(a+b-)]
- Duffy b positive [Fy(a-b+)]
- Duffy a and b positive [Fy(a+b+)]

Specific codes will help document the Duffy status for individuals in a consistent and longitudinal manner. Specific codes will enable accurate documentation, appropriate clinical care and management, and augmented ability to conduct research. For prospective clinical trial participants, it will permit inclusion of diverse populations in research, currently excluded by inappropriate eligibility criteria. For patients, it will decrease duplicative testing and permit medication (e.g., chemotherapy) administration consistent with need. In the U.S. specifically, it will help redress an underappreciated cause of health disparities. Additionally, an ICD-10-CM code will be the bedrock for the development of electronic medical record advanced functions, enabling provision of an accurate ANC reference range that automatically populates based on Duffy status.

References

1. Reich D, Nalls MA, Kao WH, et al. Reduced neutrophil count in people of African descent is due to a regulatory variant in the Duffy antigen receptor for chemokines gene. *PLoS Genet.* 2009;5(1):e1000360. <https://doi.org/10.1371/journal.pgen.1000360>
2. Kibaya RS, Bautista CT, Sawe FK, et al. Reference ranges for the clinical laboratory derived from a rural population in Kericho, Kenya. *PLoS One.* 2008;3(10):e3327. <https://doi.org/10.1371/journal.pone.0003327>
3. Haddy TB, Rana SR, Castro O. Benign ethnic neutropenia: what is a normal absolute neutrophil count? *The Journal of laboratory and clinical medicine.* 1999;133(1):15-22. <https://doi.org/10.1053/jl.1999.v133.a94931>
4. Dhanak M. Racial differences in leukocyte counts. *Am J Psychiatry.* 1996;153(4):586-587. <https://doi.org/10.1176/ajp.153.4.586-a>
5. Reed WW, Diehl LF. Leukopenia, neutropenia, and reduced hemoglobin levels in healthy American blacks. *Arch Intern Med.* 1991;151(3):501-505. <https://doi.org/10.1001/archinte.1991.00400030063011>
6. Howes RE, Patil AP, Piel FB, et al. The global distribution of the Duffy blood group. *Nat Commun.* 2011;2:266. <https://doi.org/10.1038/ncomms1265>

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

7. Charles BA, Hsieh MM, Adeyemo AA, et al. Analyses of genome wide association data, cytokines, and gene expression in African-Americans with benign ethnic neutropenia. PLoS one. 2018;13(3):e0194400. <https://doi.org/10.1371/journal.pone.0194400>
8. Merz LE, Story CM, Osei MA, et al. Absolute Neutrophil Count by Duffy Status Among Healthy Black and African American Adults. Blood Advances. 2022;bloodadvances.2022007679. <https://doi.org/10.1182/bloodadvances.2022007679>
9. Dickson AL, Daniel LL, Jackson E, et al. Race, Genotype, and Azathioprine Discontinuation : A Cohort Study. Ann Intern Med. 2022;175(8):1092-1099. <https://doi.org/10.7326/m21-4675>
10. Oyogoa E, Mathews R, Olson S, DeLoughery T, Shatzel JJ, Martens KL. Clinical Outcomes of Patients Referred for Asymptomatic Neutropenia: A Focus on Racial Disparities in Hematology. European journal of haematology. 2023. <https://doi.org/10.1182/blood-2022-170414>
11. Nagalapuram V, McCall D, Palabindela P, et al. Outcomes of Isolated Neutropenia Referred to Pediatric Hematology-Oncology Clinic. Pediatrics. 2020;146(4). <https://doi.org/10.1542/peds.2019-3637>
12. Koen A, Jose L, Madhi SA, Fix A, Cryz S, Groome MJ. Neutrophil Counts in Healthy South African Infants: Implications for Enrollment and Adverse Event Grading in Clinical Trials in an African Setting. The journal of pediatrics: X. 2019;1:100005. <https://doi.org/10.1016/j.ympdx.2019.100005>
13. Kourtis AP, Bramson B, van der Horst C, et al. Low absolute neutrophil counts in African infants. Journal of the International Association of Physicians in AIDS Care (Chicago, Ill : 2002). 2005;4(3):73-76. <https://doi.org/10.1177/1545109705282591>
14. Vastola ME, Yang DD, Muralidhar V, et al. Laboratory Eligibility Criteria as Potential Barriers to Participation by Black Men in Prostate Cancer Clinical Trials. JAMA Oncol. 2018;4(3):413-414. <https://doi.org/10.1001/jamaoncol.2017.4658>
15. Merz LE, Osei M, Story CM, Freedman RY, Smeland-Wagman R, Kaufman RM, Achebe MO. Development of Duffy Null Specific Absolute Neutrophil Count Reference Ranges. Journal of the American Medical Association. 2023. <https://doi.org/10.1001/jama.2023.7467>
16. Duchene, J., Novitzky-Basso, I., Thiriot, A., Casanova-Acebes, M., Bianchini, M., Etheridge, S.L., Hub, E., Nitz, K., Artinger, K., Eller, K. et al. (2017) Atypical chemokine receptor 1 on nucleated erythroid cells regulates hematopoiesis. Nat. Immunol., 18, 753–761. <https://doi.org/10.1038/ni.3763>
17. Permanyer, M., Bosnjak, B. and Forster, R. (2018) Dual role for atypical chemokine receptor 1 in myeloid cell hematopoiesis and distribution. Cell. Mol. Immunol., 15, 399–401. <https://doi.org/10.1038/emi.2017.79>
18. Charles, B.A., Hsieh, M.M., Adeyemo, A.A., Shriner, D., Ramos, E., Chin, K., Srivastava, K., Zakai, N.A., Cushman, M., McClure, L.A. et al. (2018) Analyses of genome wide association data, cytokines, and gene expression in African-Americans with benign ethnic neutropenia. PLoS One, 13, e0194400. <https://doi.org/10.1371/journal.pone.0194400>

TABULAR MODIFICATIONS

	Z67	Blood type	
New subcategory		Z67.A	Duffy phenotype
New code Add		Z67.A1	Duffy null Duffy phenotype Fy(a-b-)

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

New code Add	Z67.A2	Duffy a positive Duffy phenotype Fy(a+b-)
New code Add	Z67.A3	Duffy b positive Duffy phenotype Fy(a-b+)
New code Add	Z67.A4	Duffy a and b positive Duffy phenotype Fy(a+b+)

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

Early-stage type 1 diabetes

Type 1 diabetes results from chronic autoimmune destruction of pancreatic beta cells that long predates the onset of symptoms. Clinical onset is preceded by a period of islet autoimmunity with one or more autoantibodies against islet autoantigens. Persistence of multiple islet autoantibodies predicts progression to symptomatic T1D with lifetime risk approaching 100%. [1-4] Those with asymptomatic dysglycemia are at very high risk of symptomatic T1D in the next 5 years. [5-7] These observations inform the designation of stages of type 1 diabetes, adopted by the American Diabetes Association (ADA), the Endocrine Society, the International Society for Pediatric and Adolescent Diabetes (ISPAD), and the JDRF (Juvenile Diabetes Research Foundation): [8-10].

Stage 1 T1D: Multiple islet autoantibodies, normal plasma glucose, presymptomatic.

Stage 2 T1D: Islet autoantibodies, abnormal glucose tolerance, presymptomatic.

Stage 3 T1D: Plasma glucose levels above ADA diagnostic thresholds. (currently captured at E10.9, Type 1 diabetes mellitus without complications). These individuals typically are symptomatic.

Presymptomatic T1D has been identified in 0.3% children screened at age 2 to 5 years with increased prevalence likely at older ages. [11] Recognition of early-stage (presymptomatic) T1D is beneficial to long-term health outcomes. First, identification and monitoring of children at early-stage T1D significantly lowers rates of diabetic ketoacidosis (DKA) at onset of stage 3 T1D. [12,13] Beyond acute morbidity and mortality, diagnosis prior to DKA has been associated with durable improvement in achieving glucose targets. [14] Second, individuals at stage 2 T1D may be eligible for treatment with recently FDA-approved teplizumab (Tzield®) to delay the need for insulin therapy; [15] other interventions are under active investigation. Third, early diagnosis allows for education and psychosocial support before onset of need for insulin.

ICD-10-CM currently has one code for “prediabetes” (R73.03) regardless of etiology. The immunopathology of T1D is distinct and there are also differences in typical progression of glycemic abnormalities, monitoring and intervention strategies, and eventual treatment. Confusion between T1D and T2D can lead to inappropriate management, particularly in the youngest children who are at highest risk of adverse outcomes. Code, R76.0, Raised antibody titer, may apply, but gives providers no indication that a patient is developing T1D.

An international group of endocrinologists and experts in diabetes are requesting new ICD-10-CM codes to capture early-stage, presymptomatic type 1 diabetes.

References:

1. Ziegler AG, Rewers M, Simell O, & et al. Seroconversion to multiple islet autoantibodies and risk of progression to diabetes in children. *JAMA* **309**, 2473–2479 (2013).
2. Orban, T. *et al.* Pancreatic Islet Autoantibodies as Predictors of Type 1 Diabetes in the Diabetes Prevention Trial–Type 1. *Diabetes Care* **32**, 2269–2274 (2009).
3. Steck, A. K. *et al.* Predictors of Progression From the Appearance of Islet Autoantibodies to Early Childhood Diabetes: The Environmental Determinants of Diabetes in the Young (TEDDY). *Diabetes Care* dc142426 (2015)
4. Frohnert, B. I. *et al.* Refining the Definition of Stage 1 Type 1 Diabetes: An Ontology-Driven Analysis of the Heterogeneity of Multiple Islet Autoimmunity. *Diabetes Care* Online Ahead of Print (2023)
5. Vehik, K. *et al.* Rising Hemoglobin A1c in the Nondiabetic Range Predicts Progression of Type 1 Diabetes As Well As Oral Glucose Tolerance Tests. *Diabetes Care* **45**, 2342–2349 (2022).

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

6. Krischer, J. P. The use of intermediate endpoints in the design of type 1 diabetes prevention trials 14. *Diabetologia* **56**, 1919–1924 (2013).
7. Helminen, O. *et al.* HbA1c Predicts Time to Diagnosis of Type 1 Diabetes in Children at Risk. *Diabetes* **64**, 1719–1727 (2015).
8. Insel, R. A. *et al.* Staging Presymptomatic Type 1 Diabetes: A Scientific Statement of JDRF, the Endocrine Society, and the American Diabetes Association. *Diabetes Care* **38**, 1964–1974 (2015).
9. ElSayed, N. A. *et al.* 2. Classification and Diagnosis of Diabetes: Standards of Care in Diabetes-2023. *Diabetes Care* **46**, S19–S40 (2023).
10. Besser, R. E. J. *et al.* ISPAD Clinical Practice Consensus Guidelines 2022: Stages of type 1 diabetes in children and adolescents. *Pediatr. Diabetes* **23**, 1175–1187 (2022).
11. Ziegler, A.-G. *et al.* Yield of a Public Health Screening of Children for Islet Autoantibodies in Bavaria, Germany. *JAMA* **323**, 339–351 (2020).
12. Jacobsen, L. M. *et al.* Heterogeneity of DKA Incidence and Age-Specific Clinical Characteristics in Children Diagnosed With Type 1 Diabetes in the TEDDY Study. *Diabetes Care* dc210422 (2022)
13. Elding Larsson, H. *et al.* Reduced prevalence of diabetic ketoacidosis at diagnosis of type 1 diabetes in young children participating in longitudinal follow-up. *Diabetes Care* **34**, 2347–52 (2011).
14. Duca, L. M., Wang, B., Rewers, M. & Rewers, A. Diabetic Ketoacidosis at Diagnosis of Type 1 Diabetes Predicts Poor Long-term Glycemic Control. *Diabetes Care* **40**, 1249–1255 (2017).
15. Herold, K. C. *et al.* Anti-CD3 monoclonal antibody in new-onset type 1 diabetes mellitus. *N Engl J Med* **346**, 1692–1698 (2002).

TABULAR MODIFICATIONS

	E10	Type 1 diabetes mellitus	
New subcategory	E10.A	Type 1 diabetes mellitus, presymptomatic	
New code	E10.A0	Type 1 diabetes mellitus, presymptomatic, unspecified	
New code Add	E10.A1	Type 1 diabetes mellitus, presymptomatic, Stage 1 Multiple confirmed islet autoantibodies with normoglycemia	
New code Add	E10.A2	Type 1 diabetes mellitus, presymptomatic, Stage 2 Confirmed islet autoimmunity with dysglycemia	

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

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

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

What is known is that bilateral salpingectomy substantially decreases ovarian cancer risk. Data from nested case-control and population-based retrospective cohort studies indicate that bilateral salpingectomy reduces the risk of ovarian cancer by at least 65 percent.^{7,8} In 2022, Canadian researchers published the first prospective evidence that opportunistic salpingectomy may substantially decrease the incidence of high-grade serous carcinoma in the general population. At the time of 9 years follow-up, no high-grade serous carcinoma was observed among the 25,889 women who underwent opportunistic salpingectomy during hysterectomy or in lieu of tubal ligation for surgical sterilization. This is significantly less than the expected rate as well as the rate seen in the 32,080 women who did not undergo bilateral salpingectomy.⁹ Studies that have compared the addition of opportunistic salpingectomy to a gynecological or pelvic procedure without salpingectomy have not found significant differences in ovarian endocrine function, surgical complications, operative time, or length of stay.^{10,11} In the US, over a million women undergo hysterectomy or surgical sterilization annually.^{12,13} By current projections, universal uptake of salpingectomy during hysterectomy and in lieu of tubal ligation could prevent nearly 2000 deaths from ovarian cancer per year.¹⁴ Expanding opportunistic salpingectomy to post-reproductive women undergoing non-gynecologic elective abdominal surgeries such as cholecystectomy, hernia repair, appendectomy, and gastrointestinal and urologic operations would at least double the impact of opportunistic salpingectomy on decreasing ovarian cancer incidence and mortality.

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

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

The Gynecologic Oncology Division of the Department of Gynecology and Obstetrics at Johns Hopkins University submitted the proposal.

This proposal was reviewed and is supported by the American College of Obstetricians and Gynecologists (ACOG).

References

- (1) American Cancer Society (2023, January 12). *Key Statistics for Ovarian Cancer*. Cancer.org. Retrieved June 7, 2023, from <https://www.cancer.org/cancer/types/ovarian-cancer/about/key-statistics.html>
- (2) Erickson BK, Conner MG, Landen CN. The role of the fallopian tube in the origin of ovarian cancer. *Am J Obstet Gynecol*. 2013 Nov;209(5):409-14.
- (3) George SH, Garcia R, Slomovitz BM. Ovarian Cancer: The fallopian tube as the site of origin and opportunities for prevention. *Front Oncol*. 2016 May 2;6:109.
- (4) ACOG Committee Opinion No. 774: Opportunistic salpingectomy as a strategy for epithelial ovarian cancer prevention. *Obstet Gynecol*. 2019 Apr;133(4):e279-e284.
- (5) Song H, et al. The contribution of deleterious germline mutations in BRCA1, BRCA2 and the mismatch repair genes to ovarian cancer in the population. *Hum. Mol. Genet*. 2014;23:4703–4709.
- (6) Jones MR, Kamara D, Karlan BY, Pharoah PDP, Gayther SA. Genetic epidemiology of ovarian cancer and prospects for polygenic risk prediction. *Gynecol. Oncol*. 2017;147:705–713.
- (7) Madsen, C., Baandrup, L., Dehlendorff, C., & Kjaer, S. K. (2015). Tubal ligation and salpingectomy and the risk of epithelial ovarian cancer and borderline ovarian tumors: A nationwide case-control study. *Acta Obstetrica et Gynecologica Scandinavica*, 94(1), 86-94.
- (8) Falconer, H., Yin, L., Gronberg, H., & Altman, D. (2015). Ovarian cancer risk after salpingectomy: A nationwide population-based study. *Journal of the National Cancer Institute*, 107(2).

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

- (9) Hanley GE, Pearce CL, Talhouk A, et al. Outcomes from opportunistic salpingectomy for ovarian cancer prevention. *JAMA Netw Open*. 2022;5(2):e2147343.
- (10) Kho, R. M., & Wechter, M. E. (2017). Operative outcomes of opportunistic bilateral salpingectomy at the time of benign hysterectomy in low-risk premenopausal women: A systematic review. *Journal of Minimally Invasive Gynecology*, 24(2), 218-229.
- (11) Song, T., Lee, S. H., Kim, W. Y., Heo, E. J., & Kim, T. J. (2016). Opportunistic salpingectomy does not affect ovarian reserve or surgical outcomes in patients undergoing laparoscopic myomectomy.
- (12) ACOG Practice Bulletin No 208: Benefits and risks of sterilization. *Obstet Gynecol*. 2019 Mar;133(3):e194-e207.
- (13) Simms KT, Yuill S, Killen J et al. Historical and projected hysterectomy rates in the USA: Implications for future observed cervical cancer rates and evaluating prevention interventions. *Gynecol Oncol*. 2020 Sep;158(3):710-718.
- (14) Naumann RW, Hughes BN, Brown J et al. The impact of opportunistic salpingectomy on ovarian cancer mortality and healthcare costs: a call for universal health coverage. *Am J Obstet Gynecol*. 2021 Oct;225(4):397.
- (15) Tomasch G, Lemmerer M, Oswald S et al. Prophylactic salpingectomy for prevention of ovarian cancer at the time of laparoscopic cholecystectomy. *Br J Surg*. 2020. Apr;107(5):519-524.
- (16) Alicja Zietek, Mogusiewicz M, Szumilo J et al. Opportunistic salpingectomy for prevention of sporadic ovarian cancer – a jump from basic science to clinical practice? *Ginekol Pol*. 2016;87(6):467-72.
- (17) Subramaniam A, Einerson BD, Blanchard CT et al. The cost-effectiveness of opportunistic salpingectomy versus standard tubal ligation at the time of cesarean delivery for ovarian cancer risk reduction. *Gynecol Oncol*. 2019 Jan;152(1): 127-32.
- (18) Rabin R. (2023, Feb. 1) To Prevent Cancer, More Women Should Consider Removing Fallopian Tubes, Experts Say. *The New York Times*.
<https://www.nytimes.com/2023/02/01/health/ovarian-cancer-fallopian-tubes.html>
- (19) Amenabar T, Goldstein A, Bever L. (2023, Feb. 2) Fallopian Tube Removal Advised for More Women to Prevent Ovarian Cancer. *The Washington Post*.
<https://www.washingtonpost.com/wellness/2023/02/02/fallopian-tube-removal-ovarian-cancer/>
- (20) Sakran JV, Long Roche K, Stone R. (2023, May 21) Having Their Fallopian Tubes Removed Will Spare a Large Number of Women from Ovarian Cancer. *Scientific American*.
<https://www.scientificamerican.com/article/having-their-fallopian-tubes-removed-will-spare-a-large-number-of-women-from-ovarian-cancer/>
- (21) Stone R, Sakran JV, Long Roche K. Salpingectomy in ovarian cancer prevention. *JAMA*. 2023 June 1. Doi: 10.1001/jama.2023.6979. Online ahead of print.

TABULAR MODIFICATIONS

- Z40 Encounter for prophylactic surgery
Excludes1: organ donations (Z52.-)
therapeutic organ removal-code to condition
- Z40.0 Encounter for prophylactic surgery for risk factors related to
malignant neoplasms
Admission for prophylactic organ removal
- Use additional code to identify risk factor

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Revise	Z40.03 Encounter for prophylactic removal of fallopian tube(s) <u>for persons with known genetic/familial risk factors</u>
New code	Z40.04 Encounter for prophylactic removal of fallopian tube(s) for persons with no known genetic/familial risk factors

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Estrogen and other hormones and factors receptor status

Clinical biomarkers are major factors in the workup and treatment of breast cancer. The three key biomarkers for breast cancer are:

- estrogen receptor (ER)
- progesterone receptor (PR)
- human epidermal growth factor receptor 2 (HER2)

These receptors can be positive or negative. Accordingly, triple negative breast cancer is a type of invasive breast cancer that is ER negative, PR negative, and HER2 negative.

Hormone receptors are proteins that bind to a specific hormone like estrogen or progesterone. These hormone receptors are found on the surface of breast cancer cells. If the breast cancer is hormone receptor positive, then it can be treated with hormone/endocrine therapy. The endocrine therapy may block the hormone receptor (eg, tamoxifen) or severely reduce the hormone level (eg, aromatase inhibitors) or the hormone receptor (eg, fulvestrant) blocking growth of the cancer cells. If the cancer is hormone receptor negative, then hormone/endocrine therapy is rarely effective.

Human epidermal growth factor receptor 2 (HER2) is not a hormone but rather a protein normally found on breast cells that is involved in normal cell growth. Interestingly, no specific molecule has yet been identified to which HER2 binds (ie, ligand). However, HER2 can become overexpressed in 15-20% of breast cancers and this overexpression is associated with aggressive biology. There are drugs that target the HER2 protein, eg, trastuzumab (Herceptin), that are highly active against breast cancers overexpressing HER2, ie, HER2 positive. These same agents are not generally effective if the cancer does not overexpress HER2, ie, HER2 negative.

It is standard of care to routinely examine breast tissue removed by biopsy or other surgical procedure for hormone receptor status and HER2 level of expression as part of the pathologic analysis. Hormone receptor status is typically determined by immunohistochemistry (IHC) tissue staining, with separate IHC stains for ER and PR. For HER2, status is typically determined by IHC and/or fluorescence in situ hybridization (FISH). Status can vary for each receptor.

To a significant extent, breast cancer management depends on the findings. All three of these biomarkers are used to determine prognosis and predict the course of breast cancer. All three are also used to select therapies most likely to be effective. The outcomes of HR positive breast cancer are improved with a wide variety of hormone/endocrine therapies, and the outcomes of HER2 positive breast cancers are improved with therapy targeting the HER2 receptor. Thus ER, PR, and HER2 status are key factors in medical decision-making relating to which treatment options to pursue, determining the overall prognosis, and predicting treatment response.

Receptor status is routinely noted on pathological reports. If either ER or PR are positive, this is generically referred to and documented as hormone receptor positive (HR+). Likewise, if both ER and PR are negative, this is referred to and documented as hormone receptor negative (HR-). HER2 status is usually documented separately, except in triple negative breast cancer.

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

Triple negative breast cancer (ER-negative, PR-negative, HER2-negative) accounts for about 10-20% of all breast cancers. It is more common in younger women under the age of 40, Black and Hispanic women, and women with the BRCA1 mutation. In particular, Black women have nearly a three times higher risk of triple negative breast cancer than white women.

Triple-negative breast cancer is a particularly aggressive type of breast cancer and likely to recur. Treatment options are also more limited, with chemotherapy being the mainstay. Unfortunately, prognosis is generally poor with a 5-year survival rate of about 25% in the US, compared to 95% for hormone receptor positive breast cancer. Black women have a worse prognosis than other groups with a significantly higher mortality rate.

The current American Joint Committee on Cancer (AJCC) Staging System and nationally accepted clinical practice treatment guidelines from the National Comprehensive Cancer Network (NCCN) both recognize the importance of the three biomarkers and incorporate them into the staging system and decision-making process for systemic therapies.

ICD-10-CM currently provides specific codes to capture estrogen receptor status. However, despite their impact on breast cancer management and prognosis, there are still no codes that identify progesterone receptor status or HER2 receptor status. More specifically, there is no code that identifies triple negative breast cancer despite its unique and dismal impact. The complete picture of receptor status is essential for breast cancer research, longitudinal review of data, and identifying the relationship clinical outcomes.

The proposal was submitted by National Minority Quality Forum.

References

1. Almansour NM (2022) Triple-Negative Breast Cancer: A Brief Review About Epidemiology, Risk Factors, Signaling Pathways, Treatment and Role of Artificial Intelligence. *Front. Mol. Biosci.* 9:836417. doi: 10.3389/fmolb.2022.836417
2. Aysola K, Desai A, Welch C et al. Triple Negative Breast Cancer – An Overview. *Hereditary Genet.*; 2013 (Suppl 2). doi:10.4172/2161-1041.S2-001
3. McCarthy AM, Friebel-Klingner T, Ehsan S, et al. Relationship of established risk factors with breast cancer subtypes. *Cancer Med.* 2021;10:6456–6467. <https://doi.org/10.1002/cam4.4158>
4. Giuliano AE, Connolly JL, et al. Breast Cancer-Major changes in the American Joint Committee on Cancer eighth edition cancer staging manual. *CA Cancer J Clin.* 2017 Jul 8;67(4):290-303. doi: 10.3322/caac.21393. Erratum in: *CA Cancer J Clin.* 2017 Jul 8;67(4):345. PMID: 28294295.
5. Burstein HJ, Curigliano G, et al; Panelists of the St Gallen Consensus Conference. Customizing local and systemic therapies for women with early breast cancer: the St. Gallen International Consensus Guidelines for treatment of early breast cancer 2021. *Ann Oncol.* 2021 Oct;32(10):1216-1235. doi: 10.1016/j.annonc.2021.06.023.
6. NCCN Clinical Practice Guidelines in Oncology: Breast Cancer, Version 4.2023.

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

TABULAR MODIFICATIONS

Malignant neoplasm of breast

	C50	Malignant neoplasm of breast
Revise		Use additional code to identify estrogen, <u>and other hormones and factors</u> receptor status (Z17. 0- , Z17. 1)
Revise	Z17	Estrogen, <u>and other hormones and factors</u> receptor status
Add		Note: Use one code, as available, for each receptor: Z17.0, Z17.1, Z17.2- and Z17.3-
Revise		Code first malignant neoplasm of breast (C50.-) , such as:
Add		<u>malignant neoplasm of breast (C50.-)</u> malignant neoplasm of ovary (C56.-)
	Z17.0	Estrogen receptor positive status [ER+]
	Z17.1	Estrogen receptor negative status [ER-]
New subcategory	Z17.2	Progesterone receptor status
New code	Z17.21	Progesterone receptor positive status
Add		PR+
New code	Z17.22	Progesterone receptor negative status
Add		PR-
New subcategory	Z17.3	Human epidermal growth factor 2 receptor
New code	Z17.31	Human epidermal growth factor receptor 2 positive status
Add		HER2+
New code	Z17.32	Human epidermal growth factor receptor 2 negative status
Add		HER2-

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

New subcategory Add	Z17.4 Combined receptor status Note: Assign a code from subcategory Z17.4- when only a combined receptor status is documented
New sub-subcategory Add	Z17.41 Hormone receptor positive HR+
New code Add	Z17.410 Hormone receptor positive with human epidermal growth factor receptor 2 positive status HR+ with HER2+
New code Add	Z17.411 Hormone receptor positive with human epidermal growth factor receptor 2 negative status HR+ with HER2-
New sub-subcategory Add	Z17.42 Hormone receptor negative HR-
New code Add	Z17.420 Hormone receptor negative with human epidermal growth factor receptor 2 positive status HR- with HER2+
New code Add Add Add	Z17.421 Hormone receptor negative with human epidermal growth factor receptor 2 negative status HR- with HER2- TNBC Triple negative breast cancer

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Family History of 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. A proposal to create a specific ICD-10-CM code for FAP was presented in Sept. 2022. There was also a separate proposal to create additional codes, including a code for family history of adenomatous and serrated polyps. These new codes will become effective Oct. 1, 2023.

FAP may often be inherited and necessitate for screening at a younger age for those with family history. Based on input following prior ICD10 Coordination and Maintenance meeting presentations, it is being proposed to create a separate new code for family history of familial adenomatous polyposis.

References

- Chung DC, Rodgers LH. Familial adenomatous polyposis: Screening and management of patients and families. UpToDate. 2022. <https://www.uptodate.com/contents/familial-adenomatous-polyposis-screening-and-management-of-patients-and-families>
- MedlinePlus. Familial adenomatous polyposis. National Library of Medicine, NIH. <https://medlineplus.gov/genetics/condition/familial-adenomatous-polyposis/>

TABULAR MODIFICATIONS

Z83 Family history of other specific disorders

Z83.7 Family history of diseases of the digestive system

New code Z83.72 Family history of familial adenomatous polyposis

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

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, September 2022 and March 2023 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 and flank]

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

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 flank (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

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

	R10.3 Pain localized to other parts of lower abdomen
Add	Excludes2: pain localized to flank R10.A-
Add	pelvic and perineal pain (R10.2-)
New subcategory	R10.A Pain localized to flank
Add	Lateral abdomen pain
Add	Lateral flank pain
Add	Latus region 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
	R10.81 Abdominal tenderness
	Abdominal tenderness NOS
Add	Excludes2: pain localized to other parts of lower abdomen (R10.3-)
Add	pain localized to upper abdomen (R10.1-)
	R10.82 Rebound abdominal tenderness
Add	Excludes2: pain localized to other parts of lower abdomen (R10.3-)
Add	pain localized to upper abdomen (R10.1-)
New subcategory	R10.8A Flank tenderness
New code	R10.8A1 Right flank tenderness
New code	R10.8A2 Left flank tenderness
New code	R10.8A3 Suprapubic tenderness
New code	R10.8A9 Flank tenderness, unspecified
Add	Flank tenderness NOS

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

New code	R10.85 Abdominal pain of multiple sites
Add	Excludes1: abdominal rigidity NOS (R19.3)
Add	generalized abdominal pain associated with acute abdomen (R10.0)
Add	generalized abdominal pain NOS (R10.84)
Add	localized abdominal pain (R10.1-R10.4-)
	R39.8 Other symptoms and signs involving the genitourinary system
New sub-category	R39.85 Costovertebral (angle) tenderness Add
	Excludes2: abdominal and pelvic pain (R10.-)
New code	R39.851 Costovertebral (angle) tenderness, right side
New code	R39.852 Costovertebral (angle) tenderness, left side
New code	R39.853 Costovertebral (angle) tenderness, bilateral
New code	R39.859 Costovertebral (angle) tenderness, unspecified side
S30	Superficial injury of abdomen, lower back, pelvis and external genitals
	S30.1 Contusion of abdominal wall and flank
Delete	Contusion of flank
Delete	Contusion of groin
New code	S30.11 Contusion of abdominal wall and flank
New code	S30.12 Contusion of groin
New code	S30.13 Contusion of latus region
	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

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

	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

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

	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

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

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
----------	---

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

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

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

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
New code	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
New code	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

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Gulf War Illness

This topic was presented at the March 2023 ICD10 Coordination and Maintenance meeting and based on comments received during the public comment period it is being represented for consideration. Gulf War Illness (GWI) is an exposure-induced chronic multisymptom illness affecting personnel who served in the 1990-1991 Persian Gulf conflict and tied to drug/environment exposures associated with that conflict. A solid body of evidence implicates such exposures, including dose-response and gene-environment interaction data. New, unique, and excessive exposures occurred to multiple agents, distributed differently across deployed personnel. This included exposure to organophosphate nerve agents (sarin, cyclosarin), prolonged exposure to multiple pesticides (including organophosphates and carbamates, among others), pyridostigmine bromide (PB) as a nerve agent pretreatment adjunct, anthrax and botulinum toxoid vaccines and multiple vaccines, permethrin-impregnated uniforms, high concentrate DEET insect repellent, depleted uranium, ciprofloxacin, and other agents. Especially strong evidence ties acetylcholinesterase inhibiting agents including organophosphates (as pesticides and nerve agents) and carbamates (such as PB and carbamate pesticides) to GWI.

Epidemiological studies affirm that deployment to the 1990-1991 conflict and associated exposures are tied to marked increases specifically in an empirically-defined debilitating multisymptom profile. Data show no difference in the fraction of individuals reporting symptoms that have persisted for at least six months in 1-2 defined symptom domains, but a marked difference in the fraction reporting concurrent symptoms across 3-6 distinct symptom domains in Gulf War-deployed personnel vs. Gulf War-era veterans (who were not deployed to the Gulf War Theater). Gulf War illness is distinguished by this identified complex of symptoms consistently characterized across 1990-91 Gulf War-deployed veteran populations.

This health condition requires deployment to the Gulf War Theater of Operations anytime between August 1, 1990, and July 31, 1991. It requires chronic symptoms for ≥ 6 months, arising during or after this deployment, in ≥ 3 of the 6 Kansas criteria questionnaire symptom domains of fatigue/sleep; neurological/cognitive/mood; pain; gastrointestinal; respiratory; and dermatologic. For a domain to qualify for this condition, symptoms in the domain must be either of at least moderate severity (not mild) and/or there must be multiple symptoms in that domain. Currently, there is no existing ICD-10-CM code which adequately captures this health condition.

The consortium of GWI clinicians, researchers, and patient advocates propose the following tabular modifications to aid clinical care of affected veterans, advance epidemiological tracking of this condition and improve health outcomes in affected veterans.

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

TABULAR MODIFICATIONS

T75 Other and unspecified effects of other external causes

T75.8 Other specified effects of external causes

New
sub-subcategory
Add

T75.83 Effects of war theater
Use additional code to identify associated manifestations

New code
Add

T75.830 Gulf war illness
Gulf war syndrome

New code

T75.838 Effects of other war theater

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

New Code
Add

Z77.3 Contact with and (suspected) exposure to Gulf War theater
Contact with and (suspected) exposure to Persian Gulf War theater

Hyperoxaluria

Hyperoxaluria is a condition which originates from a diverse group of disorders which all ultimately result in excess levels of oxalate, a toxic metabolite which cannot be broken down by the body and which forms insoluble calcium oxalate. In some types of hyperoxaluria, this may lead to recurrent, painful kidney stones and irreversible renal damage. In recent years, better understanding of these disorders has revealed significant differences in the causes, disease mechanisms, clinical presentation, diagnosis, severity, outcomes, and treatments among the various types and subtypes of hyperoxaluria. Primary Hyperoxaluria (PH) Types 1 (PH1), 2 (PH2), and 3 (PH3) are caused by distinct inherited genetic mutations and lead to different phenotypes of disease. Other forms of hyperoxaluria including enteric hyperoxaluria (EH) and dietary hyperoxaluria (DH) are caused by a variety of conditions which lead to accumulation of oxalate in the body. The subtypes of hyperoxaluria are significantly differentiated regarding clinical manifestation and the path to diagnosis and treatment.

Primary hyperoxaluria (PH) is a group of rare genetic disorders caused by mutations that cause the body to overproduce oxalate. PH1 is caused by a mutation in the AGXT gene and is the most common and severe form of PH, comprising 70-80 percent of known cases.^{1,2,3} PH1 impacts both children and adults, and can present as early as infancy.^{2,4} Infants with PH1 often present with failure to thrive, nephrocalcinosis, kidney failure, and kidney stones. In a survey of infants with PH1 in both developing and developed nations, the overall infant mortality rate was 52 percent.⁶ Older children and adults may present with frequent kidney stones due to build-up of oxalate often requiring emergency room visits, hospitalizations, and surgery. Children and adults can also present with chronic kidney disease (CKD) which leads to the need for dialysis and ultimately dual liver/kidney transplantation. In addition to frequent, painful kidney stones that often require surgery, progressive oxalate accumulation can lead to other physical consequences. People are often hospitalized for invasive stone-removal procedures (ureteroscopy, percutaneous nephrolithotomy), acute pain from ureteral stones or pyelonephritis, renal colic, urinary tract infection, or vomiting.¹ Patients may experience renal failure, including nephrocalcinosis. As renal failure progresses, oxalate accumulates and leads to systemic oxalosis (the spread of oxalate to organs and tissues outside of the kidneys), which can lead to joint damage, bone fractures, skin ulcers, vision loss, heart failure, and death.^{5,7,8}

Renal outcomes are meaningfully different among the different subtypes of PH. PH2, caused by a mutation in the GRHPR gene, and PH3, caused by a mutation in the HOGA1 gene, are less prevalent and usually less severe than PH1, but can both contribute to adverse outcomes for patients and are markedly different in terms of severity and age of onset. By age 30, renal survival decreases to 27%, 92%, and 95% for patients with PH1, PH2, and PH3, respectively. By age 60, renal survival decreases further to 12% and 66% for PH1 and PH2 patients, respectively, while PH3 renal survival generally doesn't worsen. PH1 is the most severe phenotype, with up to 70 percent of patients presenting with End Stage Kidney Disease.^{9,10,11} Though PH3 is generally the least severe and progresses the most slowly, it generally presents with the earliest symptoms, with a median age of 2.6 years (range from infancy to 31 years). PH1 and PH2 generally have a slightly later onset, with a median age of symptoms at 5.2 years for PH1 (range from infancy to 53 years) and 7.4 years for PH2 (range from infancy to 42 years).¹⁰

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

PH1 is the only form of PH caused by a liver-specific defect, and the only form of PH where oxalate production comes solely from the liver. The enzymes defective in PH2 and PH3 are also important outside of the liver, and thus excess oxalate production occurs in extra-hepatic sources in the body.^{10,12,13} As such, existing treatments such as liver transplantation or new liver-directed therapies targeting the defective AGXT gene which are, or will be, approved are appropriate only for treating PH1.¹⁴ As understanding of each type of PH evolves and the landscape develops to include distinct treatments for each type, a more granular set of codes will greatly facilitate patients' path to successful diagnosis and treatment.

Enteric hyperoxaluria (EH) occurs as a result of a variety of gastrointestinal disorders promoting fat malabsorption, leading to an excess absorption of dietary oxalate and increased urinary oxalate excretion.¹⁵ Dietary hyperoxaluria (DH) is caused by increased ingestion of oxalate and/or its precursors such as ascorbic acid.¹⁶ In some cases, the causes of hyperoxaluria may be unknown, or idiopathic. Patients with these non-genetic forms of hyperoxaluria can develop recurrent kidney stones or renal damage with progression to CKD or kidney failure depending on severity.¹⁵ Treatment for EH and DH is distinct from genetically-caused PH and depends on treatment of underlying gastrointestinal disorders, intensive dietary modifications, and/or the use of hydration and crystallization inhibitors.¹⁵ Currently, there is no effective way to capture these forms of hyperoxaluria in the coding set. If a specific type of hyperoxaluria is not indicated, coders may use R82.992 (Hyperoxaluria, a nonspecific code for abnormal findings in urine), or incorrectly use E72.53 (PH) for patients who actually have EH or DH.

Alnylam Pharmaceuticals, Incorporated is requesting the following new codes to further differentiate the various types of hyperoxaluria.

References:

1. Danese D, Murray R, Monpara A, et al. The importance of evaluating for potential underlying causes of kidney stones: a survey of physician experiences in diagnosing primary hyperoxaluria type 1. *Nephrol Dial Transplant*. 2019;34(Issue Supplement 1). doi:10.1093/ndt/gfz106.FP007
2. Hoppe B. An update on primary hyperoxaluria. *Nat Rev Nephrol*. 2012;8(8):467-475. doi:10.1038/nrneph.2012.113
3. Cochat P, Rumsby G. Primary hyperoxaluria [published correction appears in *N Engl J Med*. 2013 Nov 28;369(22):2168]. *N Engl J Med*. 2013;369(7):649-658. doi:10.1056/NEJMra1301564
4. Milliner DS, Harris PC, Sas DJ, et al. Primary Hyperoxaluria Type 1. Published 2002, updated 2022. Adam MP, Mirzaa GM, Pagon RA, et al., editors. *GeneReviews*®. Seattle (WA): University of Washington, Seattle; 1993-2023. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK1283/>
5. van der Hoeven SM, van Woerden CS, Groothoff JW. Primary hyperoxaluria type 1, a too often missed diagnosis and potentially treatable cause of end-stage renal disease in adults: results of the Dutch cohort. *Nephrol Dial Transplant*. 2012;27(10):3855-3862. doi:10.1093/ndt/gfs320
6. Cochat P, Koch Nogueira PC, Mahmoud MA, et al. Primary hyperoxaluria in infants: medical, ethical, and economic issues. *J Pediatr*. 1999;135(6):746-750. doi:10.1016/s0022-3476(99)70095-8
7. Lorenzo V, Torres A, Salido E. Primary hyperoxaluria. *Nefrologia*. 2014;34(3):398-412. doi:10.3265/Nefrologia.pre2014.Jan.12335
8. Milliner DS, McGregor TL, Thompson A, et al. End Points for Clinical Trials in Primary Hyperoxaluria. *Clin J Am Soc Nephrol*. 2020;15(7):1056-1065. doi:10.2215/CJN.13821119
9. Zhao F, Bergstralh EJ, Mehta RA, et al. Predictors of Incident ESRD among Patients with Primary Hyperoxaluria Presenting Prior to Kidney Failure. *Clin J Am Soc Nephrol*. 2016;11(1):119-126. doi:10.2215/CJN.02810315
10. Hopp K, Cogal AG, Bergstralh EJ, et al. Phenotype-Genotype Correlations and Estimated Carrier Frequencies of Primary Hyperoxaluria. *J Am Soc Nephrol*. 2015;26(10):2559-2570. doi:10.1681/ASN.2014070698
11. Mandrile G, van Woerden CS, Berchiolla P, et al. Data from a large European study indicate that the outcome of primary hyperoxaluria type 1 correlates with the AGXT mutation type. *Kidney Int*. 2014;86(6):1197-1204. doi:10.1038/ki.2014.222
12. Giafi CF, Rumsby G. Kinetic Analysis and Tissue Distribution of Human D-Glycerate

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

- Dehydrogenase/Glyoxylate Reductase and its Relevance to the Diagnosis of Primary Hyperoxaluria Type 2. *Ann Clin Biochem.* 1998;35(1):104-109. doi:10.1177/000456329803500114
13. Wood KD, Holmes RP, Erbe D, et al. Reduction in urinary oxalate excretion in mouse models of Primary Hyperoxaluria by RNA interference inhibition of liver lactate dehydrogenase activity. *Biochim Biophys Acta Mol Basis Dis.* 2019;1865(9):2203-2209. doi:10.1016/j.bbadis.2019.04.017
14. Groothoff JW, Metry E, Deesker L, et al. Clinical practice recommendations for primary hyperoxaluria: an expert consensus statement from ERKNet and OxalEurope. *Nat Rev Nephrol.* 2023;19:194-211. doi:10.1038/s41581-022-00661-1
15. Witting C, Langman CB, Assimios D, et al. Pathophysiology and Treatment of Enteric Hyperoxaluria. *Clin J Am Soc Nephrol.* 2021;16(3):487-495. doi:10.2215/CJN.08000520
16. Bhasin B, Ürekli HM, Atta MG. Primary and secondary hyperoxaluria: Understanding the enigma. *World J Nephrol.* 2015;4(2):235-244. doi:10.5527/wjn.v4.i2.235

TABULAR MODIFICATIONS

	E72	Other disorders of amino-acid metabolism	
	E72.5	Disorders of glycine metabolism	
	E72.53	Primary hyperoxaluria	
		Oxalosis	
		Oxaluria	
New code	E72.530	Primary hyperoxaluria, type 1	
New code	E72.538	Other specified primary hyperoxaluria	
Add		Primary hyperoxaluria, type 2	
Add		Primary hyperoxaluria, type 3	
New code	E72.539	Primary hyperoxaluria, unspecified	
	R82	Other and unspecified abnormal findings in urine	
	R82.9	Other and unspecified abnormal findings in urine	
	R82.99	Other abnormal findings in urine	
	R82.992	Hyperoxaluria	
Add		Dietary hyperoxaluria	
Add		Enteric hyperoxaluria	
Revise		Excludes1: primary hyperoxaluria (E72.53_)	

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Hypoglycemia level

This topic was presented at the March 2023 ICD-10 Coordination and Maintenance meeting and based on comments received during the public comment period is now being re-presented for consideration. Changes are indicated in **BOLD**.

Hypoglycemia is a condition in which the blood sugar (glucose) level is lower than what is considered to be the “normal range”; commonly referenced as below 70 mg/dL, with clinically present signs and symptoms.¹ Traditionally clinically diagnosed hypoglycemia was classified as mild, moderate, and severe. In 2017, a multi-stakeholder group, consisting of diabetes provider groups in the US, as well as patient advocacy groups, jointly published a consensus statement creating a “levels” classification for hypoglycemia.² The adopted hypoglycemia levels (1, 2 and 3) were published in peer-reviewed endocrine and diabetes journals. This standardized classification system has since been incorporated into the American Diabetes Association³ and the American Association of Clinical Endocrinology⁴ published standard of care.

Standardization of this language, along with accepted definitions has created a consistency in hypoglycemia reporting that is now recognized cross functionally in the diabetes care world by referencing Level 1, 2 or 3 hypoglycemic occurrences. This has created meaningful standardized outcome measures to demonstrate effectiveness and results in different care environments.

1. Level 1 hypoglycemia, defined as a glucose concentration < 70 mg/dL and should be used as an ‘alert value’ to help individuals avoid more severe hypoglycemia.
2. Level 2 hypoglycemia, defined as a glucose concentration < 54 mg/dL. This is the threshold at which neuroglycopenic symptoms begin to occur.
3. Level 3 hypoglycemia, defined as a severe event characterized by altered mental and/or physical functioning independently of the glycemic value, which requires third party assistance to treat (eg: the person experiencing this cannot treat their symptoms without assistance).

The level system was created by leading experts from the following clinical groups and non-profit organizations:²

- American Association of Clinical Endocrinologists
- Association of Diabetes Care and Education Specialists
- American Diabetes Association
- Endocrine Society
- Juvenile Diabetes Research Foundation (JDRF) International
- The Leona M. and Harry B. Helmsley Charitable Trust
- Pediatric Endocrine Society
- T1D Exchange
- International Society for Pediatric and Adolescent Diabetes

Hypoglycemia largely occurs in people living with diabetes (Type 1 and Type II). Hypoglycemia can be idiopathic (of uncertain origin), and it is also seen in non-diabetic patient populations,

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

including: pancreatic carcinoma, benign insulinoma, post bariatric surgery patients, chemotherapy patients, glycogen storage disease and congenial hyperinsulinism.

When the standardized hypoglycemia severity level is documented, it demonstrates the impact on patient management, diabetic research, and physician decisions to order continuous glucose monitoring systems (CGMs).

Dexcom is requesting new codes to better track the severity of these patients.

References:

1. <https://diabetes.org/healthy-living/medication-treatments/blood-glucose-testing-and-control/hypoglycemia>
2. <https://diabetesjournals.org/care/article/40/12/1622/36909/Standardizing-Clinically-Meaningful-Outcome>
3. American Diabetes Association Professional Practice Committee; 6. Glycemic Targets: *Standards of Medical Care in Diabetes—2022*. *Diabetes Care* 1 January 2022; 45 (Supplement_1): S83–S96. <https://doi.org/10.2337/dc22-S006>
4. Blonde, L., Umpierrez, G., Reddy, S.S., McGill, J.B., Berga, S.L., Bush, M., Chandrasekaran, S., DeFronzo, R.A., Einhorn, D., Galindo, R.J., Gardner, T.W., Garg, R., Garvey, W.T., Hirsch, I.B., Hurley, D.L., Izuora, K., Kosiborod, M., Olson, D., Patel, S.B., Pop-Busui, R., Sadhu, A.R., Samson, S.L., Stec, C., Tamborlane, W.V., Tuttle, K.R., Twining, C., Vella, A., Vellanki, P., & Weber, S.L. American Association of Clinical Endocrinology Clinical Practice Guideline: Developing a Diabetes Mellitus Comprehensive Care Plan—2022 Update. *Endocrine Practice*, Vol 28, Issue 10, 2022, Pages 923-1049, <https://doi.org/10.1016/j.eprac.2022.08.002>.

TABULAR MODIFICATIONS

	E16	Other disorders of pancreatic internal secretion
Add	E16.0	Drug-induced hypoglycemia without coma Use additional code for hypoglycemia level, if applicable (E16.A-)
Add	E16.1	Other hypoglycemia Use additional code for hypoglycemia level, if applicable (E16.A-)
Add	E16.2	Hypoglycemia, unspecified Use additional code for hypoglycemia level, if applicable (E16.A-)
New subcategory	E16.A	Hypoglycemia level
New code Add	E16.A1	Hypoglycemia level 1 Decreased blood glucose level 1
New code Add	E16.A2	Hypoglycemia level 2 Decreased blood glucose level 2
New code Add	E16.A3	Hypoglycemia level 3 Decreased blood glucose level 3

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

- E08 Diabetes mellitus due to underlying condition
- E08.6 Diabetes mellitus due to underlying condition with other specified complications
- E08.64 Diabetes mellitus due to underlying condition with hypoglycemia
Add Use additional code for hypoglycemia level, if applicable (E16.A-)
- E09 Drug or chemical induced diabetes mellitus
- E09.6 Drug or chemical induced diabetes mellitus with other specified complications
- E09.64 Drug or chemical induced diabetes mellitus with hypoglycemia
Add Use additional code for hypoglycemia level, if applicable (E16.A-)
- E10 Type 1 diabetes mellitus
- E10.6 Type 1 diabetes mellitus with other specified complications
- E10.64 Type 1 diabetes mellitus with hypoglycemia
Add Use additional code for hypoglycemia level, if applicable (E16.A-)
- E11 Type 2 diabetes mellitus
- E09.6 Type 2 diabetes mellitus with other specified complications
- E11.64 Type 2 diabetes mellitus with hypoglycemia
Add Use additional code for hypoglycemia level, if applicable (E16.A-)
- E13 Other specified diabetes mellitus
- E13.6 Other specified diabetes mellitus with other specified complications
- E13.64 Other specified diabetes mellitus with hypoglycemia
Add Use additional code for hypoglycemia level, if applicable (E16.A-)

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Injection Drug Use

Despite the devastating toll exacted by injection drug use (IDU) on morbidity and mortality¹⁻⁸, there is currently no existing diagnosis code to identify injection drug use (IDU). The impact of not having a specified code, does impede population-based surveillance, research, and tracking as well as proactive offering of prevention, testing, vaccination, treatment, and harm reduction services to these high-risk patients.

This proposal was presented at the March 2023 Coordination and Maintenance Meeting. Based on public comment, the proposal has been updated and resubmitted for reconsideration. **Changes are noted in bold.**

In revising the proposal, the requestor consulted with subject matter experts within the Veterans Health Administration (VHA), including VHA's coding experts within the Health Information Management office; the National SUD Program within VHA's Office of Mental Health and Suicide Prevention; and VHA's Specialty Care Program Office. As recommended, the requestor consulted with the Substance Abuse and Mental Health Services Administration (SAMHSA) for guidance on confidentiality issues, as well as the CDC's National Center for HIV, Viral Hepatitis, STD, and TB Prevention, as well as the CDC's Division of Viral Hepatitis.

In the absence of ICD-10-CM codes, surveillance efforts require the field to use various combinations of non-specific ICD-10-CM codes to identify IDU with the published literature rife with approaches showing widely varying degrees of sensitivity and specificity⁹⁻¹². The need for ICD-10-CM codes to capture IDU is critical given groundbreaking support from The White House for syringe services programs (SSPs) that "have a proven track record of reducing disease, increasing access to addiction treatment and improving public safety." ¹³.

Moreover, for the first time in its history, the 2022 National Drug Control Strategy includes a focus on evidence-based harm reduction, including SSPs¹⁴. SSPs are also endorsed by the Centers for Disease Control and Prevention (CDC), Surgeon General of the United States, World Health Organization (WHO), American Medical Association, and American Bar Association¹⁵⁻¹⁹.

Despite widespread need and support for SSPs, the experience of VHA early adopters of SSPs have identified a clear need for ICD-10-CM codes to enable case finding of people with IDU who would be appropriate for SSPs²⁰. Furthermore, not knowing how many persons have IDU also hampers public health from knowing how many SSPs are needed and where they are needed.

Lack of a code has been identified as a major barrier to implementation with patients often being identified too late, downstream, and usually as a result of a complication arising from IDU such as HIV and hepatitis C virus (HCV) infection³⁻⁵. This is concerning given that the number of acute HCV infections has more than doubled between 2013 to 2020 with IDU the number one risk behavior for these infections.²¹

A unique ICD-10-CM code would identify IDUs to facilitate surveillance, research, treatment, harm reduction and outcomes monitoring among this vulnerable population.

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

The Veterans Health Administration's (VHA) Office of Mental Health and Suicide Prevention (OMHSP) has reviewed and supports this proposal developed by Elizabeth Dinges, PharmD Karine Rozenberg, PharmD, and Elizabeth Oliva*, PhD, VHA National Opioid Overdose Education and Naloxone Distribution Coordinator, VHA OMHSP.

References

1. Handanagic S, Finlayson T, Burnett JC, et al. HIV Infection and HIV-Associated Behaviors Among Persons Who Inject Drugs - 23 Metropolitan Statistical Areas, United States, 2018. *MMWR Morb Mortal Wkly Rep.* 2021 Oct 22;70(42):1459-1465.
2. O'Donnell J, Tanz LJ, Gladden RM, et al. Trends in and Characteristics of Drug Overdose Deaths Involving Illicitly Manufactured Fentanyl - United States, 2019-2020. *MMWR Morb Mortal Wkly Rep.* 2021 Dec 17;70(50):1740-1746.
3. Zibbell JE, Iqbal K, Patel RC, et al. Increases in hepatitis C virus infection related to injection drug use among persons aged ≤30 years - Kentucky, Tennessee, Virginia, and West Virginia, 2006-2012. *MMWR Morb Mortal Wkly Rep.* 2015 May 8;64(17):453-8.
4. Harris AM, Iqbal K, Schillie S, et al. Increases in Acute Hepatitis B Virus Infections - Kentucky, Tennessee, and West Virginia, 2006-2013. *MMWR Morb Mortal Wkly Rep.* 2016 Jan 29;65(3):47-50.
5. Peters PJ, Pontones P, Hoover KW, et al. HIV Infection Linked to Injection Use of Oxycodone in Indiana, 2014-2015. *N Engl J Med.* 2016 Jul 21;375(3):229-39.
6. Dasgupta S, Tie Y, Lemons A, et al. Injection Practices and Sexual Behaviors Among Persons with Diagnosed HIV Infection Who Inject Drugs - United States, 2015-2017. *MMWR Morb Mortal Wkly Rep.* 2019 Aug 2;68(30):653-657.
7. Tookes H, Diaz C, Li H, et al. A Cost Analysis of Hospitalizations for Infections Related to Injection Drug Use at a County Safety-Net Hospital in Miami, Florida. *PLoS One.* 2015 Jun 15;10(6):e0129360.
8. Hall EW, Rosenberg ES, Jones CM, et al. Estimated number of injection-involved drug overdose deaths, United States, 2000 - 2018. *Drug Alcohol Depend.* 2022 May 1;234:109428.
9. Barnes E, Peacock J, Bachmann L. International Classification of Diseases (ICD) Codes Fail to Accurately Identify Injection Drug Use Associated Endocarditis Cases. *J Addict Med.* 2022 Jan-Feb 01;16(1):27-32.
10. Janjua NZ, Islam N, Kuo M, et al. Identifying injection drug use and estimating population size of people who inject drugs using healthcare administrative datasets. *Int J Drug Policy.* 2018 May;55:31-39.
11. Marks LR, Nolan NS, Jiang L, et al. Use of ICD-10 Codes for Identification of Injection Drug Use-Associated Infective Endocarditis Is Nonspecific and Obscures Critical Findings on Impact of Medications for Opioid Use Disorder. *Open Forum Infect Dis.* 2020 Sep 9;7(10):ofaa414.
12. McGrew KM, Carabin H, Garwe T, et al. Validity of ICD-based algorithms to estimate the prevalence of injection drug use among infective endocarditis hospitalizations in the absence of a reference standard. *Drug Alcohol Depend.* 2020 Apr 1;209:107906. doi: 10.1016/j.drugalcdep.2020.107906.
13. The White House. (2021, December 8). White House Releases Model Law to Help States Ensure Access to “Safe, Effective, and Cost-saving” Syringe Services Programs. Retrieved from <https://www.whitehouse.gov/ondcp/briefing-room/2021/12/08/white-house-releases-model-law-to-help-states-ensure-access-to-safe-effective-and-cost-saving-syringe-services-programs/>.
14. Office of National Drug Control Policy. National Drug Control Strategy. 2022. Washington, DC: Executive Office of the President.
15. Centers for Disease Control and Prevention. Syringe Services Programs. <https://www.cdc.gov/ssp/index.html>.
16. World Health Organization. Policy brief: Provision of sterile injecting equipment to reduce HIV transmission. 2004. World Health Organization No. WHO/HIV/2004.03.
17. Adams JM. Making the Case for Syringe Services Programs. *Public Health Reports.* 2020;135(1_suppl):10S-12S.
18. American Medical Association Policy H-95.958 (Syringe and Needle Exchange Programs), adopted 1995, retained as of 2016. See <https://policysearch.ama-assn.org/policyfinder/detail/syringe?uri=%2FAMADoc%2FHOD.xml-0-5337.xml>.
19. American Bar Association Resolution 106B, House of Delegates 1997 Annual Meeting, (Needle Exchange Programs), adopted August 1997, retained as of August 15, 2017. See https://www.americanbar.org/content/dam/aba/directories/policy/1997_am_106b.authcheckdam.pdf; <https://www.americanbar.org/content/dam/aba/directories/policy/annual-2017/2017-am-400b.pdf>, p.29.
20. Rife-Pennington T, Dinges E, Ho MQ. Implementing Syringe Services Programs Within the Veterans Health Administration: Facility Experiences and Next Steps. *Journal of the American Pharmacists Association.* 2022.
21. Centers for Disease Control and Prevention. Viral Hepatitis Surveillance Report – United States, 2020. <https://www.cdc.gov/hepatitis/statistics/2020surveillance/index.htm>. Published September 2022. Accessed December 1, 2022.

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Lymphoma in Remission

This proposal is being represented today and was previously presented at the September 2022 and March 2023 ICD-10 Coordination and Maintenance Committee Meetings. The modifications are in **bold**.

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 this proposal requesting new ICD-10-CM codes for lymphoma in remission from Alliance Dedicated Cancer Centers (ADCC).

References

¹ Mayo Clinic Staff. (n.d.). *Lymphoma*. Retrieved from Mayo Clinic:

[https://www.mayoclinic.org/diseasesconditions/lymphoma/symptoms-causes/syc-](https://www.mayoclinic.org/diseasesconditions/lymphoma/symptoms-causes/syc-20352638#:~:text=Lymphoma%20is%20a%20cancer%20of,other%20organs%20throughout%20the%20body.)

[20352638#:~:text=Lymphoma%20is%20a%20cancer%20of,other%20organs%20throughout%20the%20body.](https://www.mayoclinic.org/diseasesconditions/lymphoma/symptoms-causes/syc-20352638#:~:text=Lymphoma%20is%20a%20cancer%20of,other%20organs%20throughout%20the%20body.)

² *Treatment*. (n.d.). Retrieved from Leukemia & Lymphoma Society: [https://www.lls.org/lymphoma/non-hodgkin-](https://www.lls.org/lymphoma/non-hodgkin-lymphoma/treatment#:~:text=Complete%20remission%20means%20that%20all,show%20remaining%20sites%20of%20disease.)

[lymphoma/treatment#:~:text=Complete%20remission%20means%20that%20all,show%20remaining%20sites%20of%20disease.](https://www.lls.org/lymphoma/non-hodgkin-lymphoma/treatment#:~:text=Complete%20remission%20means%20that%20all,show%20remaining%20sites%20of%20disease.)

TABULAR MODIFICATIONS

	C81	Hodgkin lymphoma
	C81.0	Nodular lymphocyte predominant Hodgkin lymphoma
New code	C81.0A	Nodular lymphocyte predominant Hodgkin lymphoma, in remission
	C81.1	Nodular sclerosis Hodgkin lymphoma
		Nodular sclerosis classical Hodgkin lymphoma
New code	C81.1A	Nodular sclerosis Hodgkin lymphoma in remission
Add		Nodular sclerosis classical Hodgkin lymphoma in remission

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

	C81.2	Mixed cellularity Hodgkin lymphoma Mixed cellularity classical Hodgkin lymphoma
New code Add	C81.2A	Mixed cellularity Hodgkin lymphoma, in remission Mixed cellularity classical Hodgkin lymphoma, in remission
	C81.3	Lymphocyte depleted Hodgkin lymphoma Lymphocyte depleted classical Hodgkin lymphoma
New code Add	C81.3A	Lymphocyte depleted Hodgkin lymphoma, in remission Lymphocyte depleted classical Hodgkin lymphoma, in remission
	C81.4	Lymphocyte-rich Hodgkin lymphoma Lymphocyte-rich classical Hodgkin lymphoma
New code Add	C81.4A	Lymphocyte-rich Hodgkin lymphoma, in remission Lymphocyte-rich classical Hodgkin lymphoma, in remission
	C81.7	Other Hodgkin lymphoma Classical Hodgkin lymphoma NOS Other classical Hodgkin lymphoma
New code Add Add	C81.7A	Other Hodgkin lymphoma, in remission Classical Hodgkin lymphoma NOS, in remission 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

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

New code	C82.4 Follicular lymphoma grade IIIb C82.4A Follicular lymphoma grade IIIb, in remission
New code	C82.5 Diffuse follicle center lymphoma C82.5A Diffuse follicle center lymphoma, in remission
New code	C82.6 Cutaneous follicle center lymphoma C82.6A Cutaneous follicle center lymphoma, in remission
New code	C82.8 Other types of follicular lymphoma C82.8A Other types of follicular lymphoma, in remission
New code	C82.9 Follicular lymphoma, unspecified 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 Add Add Add Add	C83.0A Small cell B-cell lymphoma, in remission Lymphoplasmacytic lymphoma, in remission Nodal marginal zone lymphoma, in remission Non-leukemic variant of B-CLL, in remission Splenic marginal zone lymphoma, in remission
	C83.1 Mantle cell lymphoma Centrocytic lymphoma
New code Add	C83.1A Mantle cell lymphoma, in remission Centrocytic lymphoma, in remission
	C83.3 Diffuse large B-cell lymphoma Anaplastic diffuse large B-cell lymphoma CD30-positive diffuse large B-cell lymphoma Centroblastic diffuse large B-cell lymphoma Diffuse large B-cell lymphoma, subtype not specified Immunoblastic diffuse large B-cell lymphoma Plasmablastic diffuse large B-cell lymphoma T-cell rich diffuse large B-cell lymphoma
New code Add	C83.3A Diffuse large B-cell lymphoma, in remission Anaplastic diffuse large B-cell lymphoma, in remission

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

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
 B-precursor lymphoma
 Lymphoblastic B-cell lymphoma
 Lymphoblastic lymphoma NOS
 Lymphoblastic T-cell lymphoma
 T-precursor 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
 Atypical Burkitt lymphoma
 Burkitt-like 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
 Intravascular large B-cell lymphoma
 Lymphoid granulomatosis
 Primary effusion B-cell 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

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

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

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

New code Add	C84.9A	Mature T/NK-cell lymphomas, unspecified, in remission 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 Lymphoma NOS Malignant lymphoma NOS Non-Hodgkin lymphoma NOS
New code Add Add Add	C85.9A	Non-Hodgkin lymphoma, unspecified, in remission Lymphoma NOS, in remission Malignant lymphoma NOS, in remission 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 Add Add	C86.00	Extranodal NK/T-cell lymphoma, nasal type not having achieved remission Extranodal NK/T-cell lymphoma, nasal type NOS Extranodal NK/T-cell lymphoma, nasal type with failed remission
New code	C86.01	Extranodal NK/T-cell lymphoma, nasal type, in remission

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

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 not having achieved remission
Add Enteropathy associated T-cell lymphoma NOS
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

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Add		Blastic plasmacytoid dendritic cell neoplasm (BPDCN) not having achieved remission
Add		Blastic plasmacytoid dendritic cell neoplasm (BPDCN) NOS
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) not having achieved remission
Add		Angioimmunoblastic lymphadenopathy with dysproteinemia (AILD) NOS
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 not having achieved remission
Add		Lymphomatoid papulosis NOS
Add		Lymphomatoid papulosis with failed remission
Add		Primary cutaneous anaplastic large cell lymphoma not having achieved remission
Add		Primary cutaneous anaplastic large cell lymphoma NOS
Add		Primary cutaneous anaplastic large cell lymphoma with failed remission

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Add Primary cutaneous CD30-positive large T-cell lymphoma not having achieved remission

Add Primary cutaneous CD30-positive large T-cell lymphoma NOS

Add Primary cutaneous CD30-positive large T-cell lymphoma with failed remission

Add Primary cutaneous CD30-positive T-cell proliferations NOS

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 not having achieved remission

Add Lymphoplasmacytic lymphoma with IgM-production, NOS

Add Lymphoplasmacytic lymphoma with IgM-production with failed remission

Add Macroglobulinemia (idiopathic) (primary) not having achieved remission

Add Macroglobulinemia (idiopathic) (primary) NOS

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

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

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 not having achieved remission
Add		Franklin disease NOS
Add		Franklin disease with failed remission
Add		Gamma heavy chain disease not having achieved remission
Add		Gamma heavy chain disease NOS
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 not having achieved remission
Add		Mu heavy chain disease NOS
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 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 not having achieved remission
Add		Mediterranean lymphoma NOS
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

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

C88.4 Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue [MALT-lymphoma]
Lymphoma of bronchial-associated lymphoid tissue [BALT-lymphoma]
Lymphoma of skin-associated lymphoid tissue [SALT-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] not having achieved remission
Add		Lymphoma of bronchial-associated lymphoid tissue [BALT-lymphoma] NOS
Add		Lymphoma of bronchial-associated lymphoid tissue [BALT-lymphoma] with failed remission
Add		Lymphoma of skin-associated lymphoid tissue [SALT-lymphoma] not having achieved remission
Add		Lymphoma of skin-associated lymphoid tissue [SALT-lymphoma] NOS
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	C88.81	Other malignant immunoproliferative diseases, in remission

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

C88.9 Malignant immunoproliferative disease, unspecified
Immunoproliferative disease NOS

	C88.90	Malignant immunoproliferative disease, unspecified not having achieved remission
Add		Immunoproliferative disease NOS not having achieved remission
Add		Immunoproliferative disease NOS
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

Multifidus Muscle Dysfunction

Dysfunction of the multifidus muscles in the lumbar region is a recognized cause of chronic low back pain, which can be identified and can benefit from certain specific treatments. A request to create a specific code for this has been received from Brigham & Women's Hospital and Spine Center, Harvard Medical School, with support from the American Society of Pain & Neuroscience, American Society of Regional Pain & Anesthesia, International Neuromodulation Society, International Society for the Advancement of Spine Surgery, and North American Neuromodulation Society.

Motor control of the spine is typically described as the interplay between the structural (disco ligamentous), actuator (muscle) and control (neural) domains.¹ Altered motor control of the multifidus muscle is the primary mechanism of multifidus muscle dysfunction and involves the afferent somatosensory fibers of the peripheral nervous system, and the sensory and motor cortices of the central nervous system, as well as structural changes to the muscle.²

The lumbar multifidus muscles are important segmental stabilizers of the spine, and impaired function or weakness in these muscles is strongly associated with chronic low back pain (CLBP). The cascade of inhibitory and degenerative changes to the muscle and the consequences for CLBP have been recently summarized in several publications.²⁻⁴ In brief, dysfunction of the multifidus muscle presents as inhibition of muscle activation that results in delayed or absent activation during normal functional tasks. Structurally this ultimately may result in atrophy and replacement of the multifidus muscle cross-section with adipose and connective tissue, but there are multiple disease processes that must be more specifically captured by a classification of "multifidus muscle dysfunction."⁵ While there is a strong association between the presence of low back pain symptoms and multifidus muscle dysfunction together with muscle atrophy, there is also a cohort of patients who have back pain and multifidus dysfunction that do not have visible signs of atrophy.

When multifidus muscle dysfunction is associated with atrophy, that may be coded with M62.5A2, Muscle wasting and atrophy, not elsewhere classified, back, lumbosacral. Also, these together may collectively be mechanistically consistent with the diagnosis of 'spinal instability in the lumbar region' (coded with M53.2X6), and 'other low back pain' (M54.59). However, without the ability to identify multifidus muscle dysfunction, there is no way to specifically report the underlying cause of this phenotype of CLBP. It is important for diagnostic specificity, public health, and disease tracking to collect both atrophy and dysfunction independently.⁶

Pain and disability are the consequence of inadequate motor control resulting from multifidus muscle dysfunction. Conservative management such as motor control exercise for this condition can be beneficial when applied either early in the disease progression or in patients with mild to moderate symptoms.⁷ When multifidus muscle dysfunction is refractory to conservative care, there are treatment options that have been developed, including implantable, restorative neurostimulation.^{4,8}

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

Multifidus Muscle Dysfunction can be diagnosed clinically by the assessment of muscle function, and the observation of dysfunctional activation and recruitment.^{9,10} This aberrant activity diminishes the stability of the spine and predisposes the disco ligamentous structures of the spine to movements outside of physiological ranges leading to tissue overload.^{11,12} This in turn may activate tissue nociceptors resulting in pain and further muscle inhibition. The direct activation of nociceptors as a result of functional instability is fundamentally different from other commonly described phenotypes of low back pain such as neuropathic low back pain (LBP) from inflammation or injury to nerves, and nociplastic LBP arising from altered perception and processing of pain in the central nervous system.¹³ Multifidus dysfunction is primarily assessed through dedicated physical examination maneuvers such as the multifidus lift test and the prone instability test, techniques such as palpation, muscle strength testing, range of motion assessment, electromyography, and imaging studies such as MRI or ultrasound. The validity of these tests has been established and described in the literature.^{9,10}

It is proposed that a new code for dysfunction of the multifidus muscles in the lumbar region will be beneficial in conducting research, such as epidemiological studies, tracking for public health, and measuring quality, safety, and efficacy of care.¹⁴

References

1. Panjabi, M. M. The stabilizing system of the spine. Part I Function, Dysfunction, Adaptation and Enhancement. *J. Spinal Disord.* 5, 383–389 (1992). <https://doi.org/10.1097/00002517-199212000-00001>
2. Hodges, P. W. & Danneels, L. Changes in structure and function of the back muscles in low back pain: Different time points, observations, and mechanisms. *J. Orthop. Sports Phys. Ther.* 49, 464–476 (2019). <https://doi.org/10.2519/jospt.2019.8827>
3. Francio, V., Westerhaus, B. D., Carayannopoulos, A. G. & Sayed, D. Multifidus Dysfunction and Restorative Neurostimulation: A Scoping Review. *Pain Med.* (2023) <https://doi.org/10.1093/pm/pnad098>.
4. Lorio, M., Lewandrowski, K.-U., Coric, D., Phillips, F. & Shaffrey, C. I. International Society for the Advancement of Spine Surgery Statement: Restorative Neurostimulation for Chronic Mechanical Low Back Pain Resulting From Neuromuscular Instability. *Int J Spine Surg* (2023) <https://doi.org/10.14444/8525>.
5. Freeman, M. D., Woodham, M. A. & Woodham, A. W. The Role of the Lumbar Multifidus in Chronic Low Back Pain: A Review. *PM&R* 2, 142–146 (2010). <https://doi.org/10.1016/j.pmrj.2009.11.006>.
6. Kjaer, P., Bendix, T., Sorensen, J. S., Korsholm, L. & Leboeuf-Yde, C. Are MRI-defined fat infiltrations in the multifidus muscles associated with low back pain? *BMC Med.* 5, 1–10 (2007). <https://doi.org/10.1186/1741-7015-5-2>.
7. Macedo, L. G., Maher, C. G., Latimer, J. & McAuley, J. H. Motor control exercise for persistent, nonspecific low back pain: a systematic review. *Phys. Ther.* 89, 9–25 (2009). <https://doi.org/10.2522/ptj.20080103>.
8. Gilligan, C. et al. Three-Year Durability of Restorative Neurostimulation Effectiveness in Patients With Chronic Low Back Pain and Multifidus Muscle Dysfunction. *Neuromodulation* 26, 98–108 (2023). <https://doi.org/10.1016/j.neurom.2022.08.457>.
9. Hebert, J. J., Koppenhaver, S. L., Teyhen, D. S., Walker, B. F. & Fritz, J. M. The evaluation of lumbar multifidus muscle function via palpation: Reliability and validity of a new clinical test. *Spine J.* 15, 1196–1202 (2015). <https://doi.org/10.1016/j.spinee.2013.08.056>.
10. Denteneer, L., Stassijns, G., De Hertogh, W., Truijien, S. & Van Daele, U. Inter- and Intrarater Reliability of Clinical Tests Associated With Functional Lumbar Segmental Instability and Motor Control Impairment in Patients With Low Back Pain: A Systematic Review. *Arch. Phys. Med. Rehabil.* 98, 151–164.e6 (2017). <https://doi.org/10.1016/j.apmr.2016.07.020>.
11. Benoist, M. Natural history of the aging spine. *Eur. Spine J.* 12, s86–s89 (2003). <https://doi.org/10.1007/s00586-003-0593-0>.
12. Van Dieën, J. H., Selen, L. P. J. & Cholewicki, J. Trunk muscle activation in low-back pain patients, an analysis of the literature. *J. Electromyogr. Kinesiol.* 13, 333–351 (2003). [https://doi.org/10.1016/s1050-6411\(03\)00041-5](https://doi.org/10.1016/s1050-6411(03)00041-5).
13. Woolf, C. J. & Salter, M. W. Neuronal plasticity: Increasing the gain in pain. *Science* 288, 1765–1768 (2000). <https://doi.org/10.1126/science.288.5472.1765>.
14. Moore, K.L., Agur, A.M.R., Dalley, A.F. *Essential Clinical Anatomy* 5th Edition. (Wolters, Kluwer).

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

TABULAR MODIFICATIONS

M62 Other disorders of muscle

M62.8 Other specified disorders of muscle

New code M62.85 Dysfunction of the multifidus muscles, lumbar region

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Nasal Valve Collapse

More than 85 million adult Americans are estimated to suffer from some degree of nasal airway obstruction (NAO)¹. Of these NAO patients, 63.5% are estimated to have severe/extreme obstruction² as measured by a NOSE score >55 using the modified Cottle's maneuver. Among these patients, 73% presented with nasal valve collapse (NVC)³, which is defined as a weakness or narrowing of the nasal valve that is distinct from other forms of nasal airway obstruction.

The nasal valve is described as the narrowest portion of the human airway at the cross-sectional area of the nasal cavity with the greatest overall resistance to airflow. Anatomically, the nasal valve is composed of an internal and external nasal valve, and thus NVC can be broadly categorized as affecting either the internal or external valve (or both)⁴. The external nasal valve is located in the nasal vestibule, under the nasal ala, formed by the caudal septum, medial crura of the alar cartilages, alar rim, and nasal sill. The internal nasal valve is a structure formed by articulation of the anterior ridge of the upper lateral cartilage with the anterior septal edge. The internal nasal valve is located approximately 1.3 cm from the nares (nostril opening) and corresponds to the region under the upper lateral cartilages, bound medially by the dorsal septum, inferiorly by the head of the inferior turbinate, and laterally by the upper lateral cartilage⁵.

NVC may be further classified as either static or dynamic⁶. Static NVC is a narrowing of the of the nose at rest whereby the angle between the lateral cartilage and nasal septum is anatomically small and the area of the valve is reduced. Dynamic NVC is caused when the lateral nasal wall is pulled inward by increased pressure upon inhalation while static NVC results from a constantly narrowed airway with causes including inflamed tissue, scarring, and natural deformity.

Symptoms associated with NVC, either static or dynamic, may include difficulty in nasal inspiration, exercise intolerance, sleep disturbance and decreased quality of life⁷. Notably, the etiologic and pathologic characteristics of each patient's nasal airway problem are tightly correlated with determining the best course of action in treating the nasal airway. Historically, medical management (i.e., steroid spray) is an inadequate treatment unless accompanied by invasive surgical interventions. To date, surgical options to correct NVC have included bioabsorbable implants, grafts, flaps, and radiofrequency energy techniques.

Diagnosis of NVC typically includes an endoscopic visual assessment as well as a modified Cottle's maneuver assessment. The modified Cottle maneuver is performed by using a curette or cotton swab that is inserted into the nose and is gently pressed against the lateral nasal wall while the patient inhales to assess whether they experience relief. The dynamic nature of NVC as described above, and the fact that sometimes the external nasal findings are subtle may not lend themselves to photographic capture.

In 2010, the American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) published a consensus statement on the diagnosis and management of nasal valve compromise. Consensus was achieved that 1) NVC is a distinct clinical entity separate from other anatomic reasons for nasal airway obstruction and 2) NVC can be caused by collapse of the alar rim or lateral nasal wall, collapse of the cartilaginous portion of the nasal dorsum, a high septal deviation,

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

hypertrophied inferior turbinate, severely ptotic nasal tip, wide nasal columella, and/or a caudal septal deviation⁸.

Stryker, a medical technology company, is requesting the following new codes to align clinical documentation with clinical practice and to support epidemiology research.

References:

1. Value calculated based on 2020 US adult population estimate from US Census and World Bank data in conjunction with incidence numbers cited in Schuman TA, Senior BA. Treatment Paradigm for Nasal Airway Obstruction. *Otolaryngol Clin North Am.* 2018 Oct;51(5):873-882. doi: 10.1016/j.otc.2018.05.003. Epub 2018 Jun 23. PMID: 29941181.
2. Schuman TA, Senior BA. Treatment Paradigm for Nasal Airway Obstruction. *Otolaryngol Clin North Am.* 2018 Oct;51(5):873-882. doi: 10.1016/j.otc.2018.05.003. Epub 2018 Jun 23. PMID: 29941181.
3. Clark DW, DelSignore AG, Raithatha R, Senior B. Nasal airway obstruction: prevalence and anatomic contributors. *Ear Nose Throat J.* 2018; 97:173-176.
4. Chandra RK, Patadia MO, Raviv J. Diagnosis of nasal airway obstruction. *Otolaryngol Clin North Am.* 2009 Apr;42(2):207-25. vii. doi: 10.1016/j.otc.2009.01.004. PMID: 19328887.
5. Rhee JS, Weaver EM, Park SS, et al. Clinical Consensus Statement: Diagnosis and Management of Nasal Valve Compromise. *Otolaryngology-Head and Neck Surgery.* 2010, 143-48-59.
6. "Position Statement: Nasal Valve Repair." American Academy of Otolaryngology-Head and Neck Surgery (AAOHN), 23 Mar. 2023, www.entnet.org/resource/position-statement-nasal-valve-repair/.
7. Khanwalkar A, Welch K. Update on the Evaluation and Management of Nasal Valve Collapse. *Current Otorhinolaryngology Reports.* 2021.
8. Rhee JS, Weaver EM, Park SS, et al. Clinical Consensus Statement: Diagnosis and Management of Nasal Valve Compromise. *Otolaryngology-Head and Neck Surgery.* 2010, 143-48-59.

TABULAR MODIFICATIONS

J34 Other and unspecified disorders of nose and nasal sinuses

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

New
subcategory

J34.82 Nasal valve collapse

Add

Code first underlying cause, such as:
deviated nasal septum (J34.2)

New
sub-subcategory

J34.820 Internal nasal valve collapse

New code

J34.8200 Internal nasal valve collapse, unspecified

New code
Add

J34.8201 Internal nasal valve collapse, static
Narrowing of the septum, head of the
inferior turbinate and the upper lateral
cartilage

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

New code Add	J34.8202 Internal nasal valve collapse, dynamic Collapse or falling of the upper, middle sidewall of the nose on inspiration
New sub-subcategory	J34.821 External nasal valve collapse
New code	J34.8210 External nasal valve collapse, unspecified
New code Add	J34.8211 External nasal valve collapse, static Fixed narrowing of the caudal septum, lower lateral cartilage, alar rim and nasal sill
New code Add	J34.8212 External nasal valve collapse, dynamic Collapse or falling of the lower sidewall or nostril of the nose on inspiration
New code Add	J34.829 Nasal valve collapse, unspecified Nasal valve collapse, NOS

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

Pediatric Obesity Body Mass Index

Obesity is recognized as a highly prevalent chronic disease with complex inflammatory and endocrinological pathophysiology, with serious health and social consequences.¹⁻³ Previous proposal related to obesity classes was presented in Sept. 2022 and March 2023, and further clinical details are available from those proposals.

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. Obesity in children uses a classification system recognizing BMI \geq 95th percentile as class 1 obesity, BMI \geq 120% of the 95th percentile as class 2 obesity, and BMI \geq 140% of the 95th percentile as class 3 obesity.⁴

Obesity has been an ongoing problem in children and adolescents.^{5,6} It is recommended that adolescents with class 2 obesity together with a co-morbidity, or with class 3 obesity should be considered for bariatric surgery.⁷ In January 2023, the AAP released a Clinical Practice Guideline for the Evaluation and Treatment of Children and Adolescents With Obesity to inform pediatric healthcare providers about the standard of care for youth with overweight and obesity and related comorbidities.⁸

Proposed changes to the ICD-10-CM pediatric obesity codes have been based on input from both the American Academy of Pediatrics; and the Division of Nutrition, Physical Activity, and Obesity, of the National Center for Chronic Disease Prevention and Health Promotion, CDC; with further input from additional obesity experts. This proposal is a result of combined input, as well as comments from the previous presentations.

References

1. Mechanick JI, Garber AJ, Handelsman Y, Garvey WT. American Association of Clinical Endocrinologists' position statement on obesity and obesity medicine. *Endocr Pract* 2012; 18: 642-648.
2. American Medical Association. Recognition of obesity as a disease H-440.842. <https://policysearch.ama-assn.org/policyfinder/detail/obesity?uri=%2FAMADoc%2FHOD.xml-0-3858.xml>.
3. Centers for Disease Control and Prevention. Causes of Obesity. <https://www.cdc.gov/obesity/basics/causes.html>
4. Skinner AC, Ravanbakht SN, Skelton JA, et al. Prevalence of Obesity and Severe Obesity in US Children, 1999–2016. *Pediatrics* (2018) 141 (3): e20173459. <https://doi.org/10.1542/peds.2017-3459>
5. Skinner AC, Perrin EM, Moss LA, Skelton JA. Cardiometabolic Risks and Severity of Obesity in Children and Young Adults. *N Engl J Med*. 2015 Oct;373(14):1307-17. PMID: 26422721. <https://doi.org/10.1056/NEJMoa1502821>
6. Skinner AC, Skelton JA. Prevalence and trends in obesity and severe obesity among children in the United States, 1999-2012. *JAMA Pediatr*. 2014 Jun;168(6):561-6. PMID: 24710576. <https://doi.org/10.1001/jamapediatrics.2014.21>
7. Pratt JSA, Browne A, Browne NT, et al. ASMBS pediatric metabolic and bariatric surgery guidelines, 2018. *Surg Obes Relat Dis*. 2018 Jul;14(7):882-901. Epub 2018 Mar 23. PMID: 30077361 PMID: PMC6097871. <https://doi.org/10.1016/j.soard.2018.03.019>
8. Hampl SE, Hassink SG, Skinner AC, et al. Clinical Practice Guideline for the Evaluation and Treatment of Children and Adolescents With Obesity. *Pediatrics* 2023 Feb 1;151(2):e2022060640. <https://doi.org/10.1542/peds.2022-060640>

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

TABULAR MODIFICATIONS

Z68 Body mass index [BMI]

Z68.5 Body mass index [BMI] pediatric

Revise	Z68.54	Body mass index [BMI] pediatric, greater than or equal to 95th percentile for age <u>to less than 120% of the 95th percentile for age</u>
New code	Z68.55	Body mass index [BMI] pediatric, 120% of the 95th percentile for age to less than 140% of the 95th percentile for age
New code	Z68.56	Body mass index [BMI] pediatric, greater than or equal to 140% of the 95th percentile for age

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

Personal History of Colon Polyps

The National Center for Health Statistics received a proposal request during the public comment period of the September 2023 Coordination and Maintenance meeting to expand ICD-10-CM code Z86.010, Personal history of colonic polyps.

Polypectomies and surveillance are important to find added risk factors to prevent colorectal cancer and set up suitable surveillance.¹ Colorectal cancer often advances from precancerous polyps within the colon or rectum.²

Individuals with a history of removal of certain types of polyps during a procedure have higher cancer risk, and are recommended to have a repeat colonoscopy in 3 years, but some individuals may need repeat colonoscopy earlier or later than three years based on the type, size, and number of polyps previously removed.³

The colon polyps can be grouped as nonneoplastic and neoplastic. Nonneoplastic polyps include hyperplastic polyps, inflammatory polyps and hamartomatous polyps. Nonneoplastic polyps characteristically do not become cancerous.⁴

Neoplastic polyps include adenomas and serrated types. These polyps have the possibility to develop cancer with enough time to grow. These colon polyps often are adenomas. Serrated polyps could evolve to form cancer contingent on their size and site in the colon or rectum. Generally, larger neoplastic polyps have more risk or danger of developing into cancer.⁴

New ICD-10-CM codes will provide coding specificity for personal history of adenomatous polyps to identify individuals with risk factors.

This proposal is supported by American Gastroenterology Association.

References

- ¹ Hong, Sung Pil, et al. Risk Factors for Recurrent High-Risk Polyps after the Removal of High-Risk Polyps at Initial Colonoscopy. *Yansei Medical Journal*. [Online] January 15, 2015. <https://doi.org/10.3349/ymj.2015.56.6.1559>
- ² What Should I Know About Screening? Centers for Disease Control and Prevention. [Online] February 17, 2022. https://www.cdc.gov/cancer/colorectal/basic_info/screening/index.htm
- ³ American Cancer Society Guideline for Colorectal Cancer Screening. American Cancer Society. [Online]. <https://www.cancer.org/cancer/colon-rectal-cancer/detection-diagnosis-staging/acs-recommendations.html>
- ⁴ Staff, Mayo Clinic. Colon Polyps. Mayo Clinic. [Online]. <https://www.mayoclinic.org/diseases-conditions/colon-polyps/symptoms-causes/syc-20352875>

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

TABULAR MODIFICATIONS

Z86 Personal history of certain other diseases

Z86.0 Personal history of in-situ and benign neoplasms and neoplasms of uncertain behavior

Z86.01 Personal history of benign neoplasm

Revise	Z86.010	Personal history of colonie polyps
Add		Personal history of colorectal polyps
Add		Personal history of rectal polyps
New code	Z86.0100	Personal history of colon polyps, unspecified
Add		Personal history of colon polyps NOS
New code	Z86.0101	Personal history of adenomatous and serrated colon polyps
Add		Personal history of tubular adenoma polyps
Add		Personal history of sessile adenomatous colon polyp
Add		Personal history of sessile serrated colon polyp
Add		Personal history of tubulovillous adenoma polyps
Add		Personal history of villous adenoma polyps
Add		Personal history of traditional serrated adenoma polyps
New code	Z86.0102	Personal history of hyperplastic colon polyps
New code	Z86.0109	Other personal history of colon polyps

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Personal History of Immune Checkpoint Inhibitor (ICI) Immunotherapy

The first immune checkpoint inhibitor (ICI) drug was approved by the FDA in 2011 for treatment of melanoma. A dozen years later, there are multiple immune checkpoint inhibitor drugs approved to treat over 80 indications across 17 types of cancer. These include but are not limited to melanoma, non-small cell lung cancer, colorectal cancer, renal cell carcinoma, bladder, endometrial and cervical cancer, breast cancer, and esophageal carcinoma.

Immune checkpoint inhibitor therapy is now the standard of care for certain cancers and is increasingly being utilized as a first-line option in the metastatic stage. In addition, FDA approvals of ICI immunotherapy have expanded to earlier stages of disease in certain cases.

Immune checkpoint inhibitors (ICI) are a type of monoclonal antibody. Monoclonal antibodies are commonly used to treat many disorders, such as Crohn's disease, lupus, psoriasis, COVID-19, *C difficile*, hypercholesterolemia, and osteoporosis. Some monoclonal antibodies, such as immune checkpoint inhibitors, are used to treat cancer.

Like all drugs, immune checkpoint inhibitors have adverse effects¹. Conventionally, adverse effects arise while the patient is actively taking the drug and resolve after the drug is discontinued. However, this pattern differs significantly for some adverse effects of immune checkpoint inhibitors as they can have longstanding effects that turn into chronic conditions^{2,3}. Adverse effects can occur at any point during ICI therapy and can also arise after ICI therapy is completed, sometimes months or even years afterwards. For example, in one study, ocular adverse effects occurred one year after ICI therapy was discontinued and the maximum time for other adverse effects to appear was as late as three years⁴.

The ability to identify these patients in longitudinal data will enable better identification and monitoring of delayed adverse effects stemming from ICI therapy. This will allow for the identification of critical predictors associated with adverse effects, a more accurate assessment of outcomes, and a deeper understanding of treatment responses.

ICD-10-CM currently has unique codes for personal history of drug therapy and cellular therapy to similarly identify history of antineoplastic treatments. These include Z92.21, Personal history of antineoplastic chemotherapy and Z92.850, Personal history of Chimeric Antigen Receptor T-cell therapy. A unique code for personal history of immune checkpoint inhibitor therapy will likewise reflect the clinical importance of this history.

This proposal is submitted by Project Data Sphere and Kerry Reynolds, MD.

References

1. Brahmer JR, Abu-Sbeih H, Ascierto PA, et al. Society for Immunotherapy of Cancer (SITC) clinical practice guideline on immune checkpoint inhibitor-related adverse events. *Journal for Immunotherapy of Cancer* 2021;9:e002435. doi:10.1136/jitc-2021-002435
2. Couey MA, Bell RB, Patel AA et al. Delayed immune-related events (DIRE) after discontinuation of immunotherapy: diagnostic hazard of autoimmunity at a distance. *Journal for Immunotherapy of Cancer* (2019) 7:165 <https://doi.org/10.1186/s40425-019-0645-6>.

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

3. Tang S, Tang L et al, The Pattern of Time to Onset and Resolution of Immune-Related Adverse Events Caused by Immune Checkpoint Inhibitors in Cancer: A Pooled Analysis of 23 Clinical Trials and 8,436 Patients. *Cancer Research and Treatment*, *Cancer Research and Treatment*. 2021 Apr;53(2):339-354. <https://pubmed.ncbi.nlm.nih.gov/33171025/>
4. Patrinely JR, Johnson R et al. Chronic Immune-Related Adverse Events Following Adjuvant Anti-PD-1 Therapy for High-risk Resected Melanoma. *JAMA Oncol*. 2021 May; 7(5): 1–5. <https://pubmed.ncbi.nlm.nih.gov/33764387/>

TABULAR MODIFICATIONS

Z92	Personal history of other medical treatment	
	Z92.2 Personal history of drug therapy	
	Excludes2: long term (current) drug therapy (Z79.-)	
	Z92.22 Personal history of monoclonal drug therapy	
Add		Excludes2: personal history of immune checkpoint inhibitor therapy (Z92.26)
New code	Z92.26 Personal history of immune checkpoint inhibitor therapy	
	Personal history of ICI drug therapy	
	Z92.8 Personal history of other medical treatment	
	Z92.85 Personal history of cellular therapy	
Add		Excludes2: personal history of immune checkpoint inhibitor therapy (Z92.26)

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Post-exertional malaise/post-exertional symptom exacerbation

This proposal was originally presented at the March 2023 Coordination and Maintenance meeting and is being represented based on public comments.

The majority of Long COVID patients experience an exacerbation of some or all their symptoms and/or a further reduction in functioning following physical or cognitive exertion or emotional, positional, sensory, or other stressors that would have been tolerated before their illness.

This symptom is a defining characteristic of myalgic encephalomyelitis/chronic fatigue syndrome (ME/CFS) and is referred to as post-exertional malaise (PEM) or post-exertional symptom exacerbation (PESE). PEM is characterized by exacerbation of some or all a patient's symptoms or occurrence of new symptoms. Symptoms exacerbated can include physical fatigue, cognitive fatigue, exercise intolerance, problems thinking (e.g., slowed information processing speed, memory, concentration), unrefreshing sleep, muscle pain, joint pain, headaches, weakness/instability, light-headedness, flu-like symptoms, sore throat, nausea, orthostatic intolerance or other autonomic dysfunctions, sensory sensitivities, and other symptoms.

Characteristics include pathological loss of stamina and/or functional capacity that is not due to physical deconditioning. Onset can be immediate or delayed after the exertional stimulus by hours to days. There is a prolonged, unpredictable time to return to baseline that is not easily relieved by rest or sleep and may last days, weeks, months, or longer. Severity and duration of PEM/PESE is often out-of-proportion to the type, intensity, frequency, and/or duration of the exertion.

Even basic activities of daily living like toileting, bathing, dressing, communicating, and reading can trigger PEM/PESE. In some instances, the specific precipitant cannot be identified. The threshold for a precipitant to trigger PEM/PESE can vary between individuals as well as within the same individual, at different times during their illness.

This symptom has important implications for the diagnosis, treatment, disability assessment, morbidity tracking, and research using electronic health records for Long COVID and myalgic encephalomyelitis/chronic fatigue syndrome (ME/CFS) as that experience PEM. For instance:

- In its Long COVID guidance on fatigue and recommendations for exercise, the American Academy of Physical Medicine and Rehabilitation discusses the importance of identifying PEM and cautions against exercise programs that provoke PEM.
- Numerous Long COVID studies, including those in NIH's RECOVER Initiative and by the CDC, are now using electronic health records to identify important sequelae of an acute SARS CoV-2 infection. PEM/PESE is virtually invisible in these studies.

ICD-10 Coordination and Maintenance Committee Meeting September 12-13, 2023

Currently, the ICD-10-CM does not have a code for the symptom of PEM/PESE as described for Long COVID and (ME/CFS). As a result, this important symptom is not being identified or tracked in electronic health records. A code for this symptom would also facilitate education of physicians, improve communication between a patient, clinician and other healthcare providers. A new code would improve the accuracy of Long COVID and ME/CFS research and surveillance.

It's important to note that while PEM is included in a diagnosis of ME/CFS, not all patients with Long COVID and PEM/PESE have been given an ME/CFS diagnosis. This is because the patient has not yet met the 6-month diagnostic requirements or other ME/CFS criteria.

Patient-Led Research Collaborative, with support from #MEAction, Open Medicine Foundation, Solve ME/CFS Initiative, Massachusetts ME/CFS & FM Association and Pandora Org, are requesting a new ICD-10-CM code to identify patients with PEM/PESE without the diagnosis of ME/CFS.

References

1. Davis, H. E.; Assaf, G. S.; McCorkell, L.; et al. Characterizing Long COVID in an International Cohort: 7 Months of Symptoms and Their Impact. *EClinicalMedicine* 2021, 38, 101019. <https://doi.org/10.1016/j.eclinm.2021.101019>
2. Twomey, R.; DeMars, J.; Franklin, K.; et al. Chronic Fatigue and Postexertional Malaise in People Living With Long COVID: An Observational Study. *Physical Therapy* 2022, 102 (4), pzac005. <https://doi.org/10.1093/ptj/pzac005>
3. Kedor, C.; Freitag, H.; Meyer-Arndt, L.; et al. Chronic COVID-19 Syndrome and Chronic Fatigue Syndrome (ME/CFS) Following the First Pandemic Wave in Germany – a First Analysis of a Prospective Observational Study; preprint; *Infectious Diseases (except HIV/AIDS)*, 2021. <https://doi.org/10.1101/2021.02.06.21249256>
4. US Institute of Medicine. Beyond myalgic encephalomyelitis/ chronic fatigue syndrome: redefining an illness. Report guide for clinicians. The National Academies Press. February 2015. <https://www.nap.edu/resource/19012/MECFScliniciansguide.pdf>
5. Bateman, L.; Bested, A. C.; Bonilla, H. F.; et al. Myalgic Encephalomyelitis/Chronic Fatigue Syndrome: Essentials of Diagnosis and Management. *Mayo Clinic Proceedings* 2021, 0 (0). <https://doi.org/10.1016/j.mayocp.2021.07.004>
6. What Are ME/CFS Symptoms? Centers for Disease Control and Prevention. https://www.cdc.gov/me-cfs/pdfs/interagency/What-are-MECFS-Symptoms_508.pdf Accessed June 6, 2022
7. Stussman, B.; Williams, A.; Snow, J.; Gavin, A.; Scott, R.; Nath, A.; Walitt, B. Characterization of Post-Exertional Malaise in Patients with Myalgic Encephalomyelitis/Chronic Fatigue Syndrome. *Front. Neurol.* 2020, 11, 1025. <https://doi.org/10.3389/fneur.2020.01025>
8. Keller BA, Pryor JL, Giloteaux L. Inability of myalgic encephalomyelitis/chronic fatigue syndrome patients to reproduce VO2peak indicates functional impairment. *J Transl Med.* 2014;12(1):104.
9. Herrera, J. E.; Niehaus, W. N.; Whiteson, J.; et al. Multidisciplinary Collaborative Consensus Guidance Statement on the Assessment and Treatment of Fatigue in postacute Sequelae of SARS-CoV -2 Infection (PASC) Patients. *PM&R* 2021, 13 (9), 1027–1043. <https://doi.org/10.1002/pmri.12684>
10. World Physiotherapy response to COVID-19 Briefing paper 9. *World Physiotherapy.* June 2021. <https://world.physio/sites/default/files/2021-06/Briefing-Paper-9-Long-Covid-FINAL-2021.pdf>
11. Podell, R.; Dimmock, M. E.; Comerford, B. B. Documenting Disability in Myalgic Encephalomyelitis/Chronic Fatigue Syndrome(Me/Cfs). *WOR* 2020, 66 (2), 339–352. <https://doi.org/10.3233/WOR-203178>
12. Chu, L.; Fuentes, L. R.; Marshall, O. M.; Mirin, A. A. Environmental Accommodations for University Students Affected by Myalgic Encephalomyelitis/Chronic Fatigue Syndrome (ME/CFS). *WOR* 2020, 66 (2), 315–326. <https://doi.org/10.3233/WOR-203176>
13. Pfaff, E. R.; Girvin, A. T.; Bennett, T. D.; et al. Identifying Who Has Long COVID in the USA: A Machine Learning Approach Using N3C Data. *The Lancet Digital Health* 2022, S2589750022000486. [https://doi.org/10.1016/S2589-7500\(22\)00048-6](https://doi.org/10.1016/S2589-7500(22)00048-6)

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

TABULAR MODIFICATIONS

G93	Other disorders of brain
	G93.3 Postviral and related fatigue syndromes
	Use additional code, if applicable, for post COVID-19 condition, unspecified (U09.9)
	Excludes1: chronic fatigue NOS (R53.82) neurasthenia (F48.8)
	G93.31 Postviral fatigue syndrome
Add	Use additional code, if applicable, for post-exertional malaise (R68.85)
	G93.32 Myalgic encephalomyelitis/chronic fatigue syndrome
	Chronic fatigue syndrome ME/CFS Myalgic encephalomyelitis
Add	Excludes1: post-exertional malaise (R68.85)
	G93.39 Other post infection and related fatigue syndromes
Add	Use additional code, if applicable, for post-exertional malaise (R68.85)
	R68 Other general symptoms and signs
	R68.8 Other general symptoms and signs
New code	R68.85 Post-exertional malaise
Add	PEM
Add	PESE
Add	Post-exertional symptom exacerbation
Add	Code first underlying condition, if known, such as:
Add	other post infection and related fatigue syndromes (G93.39)
Add	postviral fatigue syndrome (G93.31)
Add	Use additional code, if applicable, for post COVID-19 condition, unspecified (U09.9)
Add	Excludes1: myalgic encephalomyelitis/chronic fatigue syndrome (G93.32)
	U09.9 Post COVID-19 condition, unspecified
	Post-acute sequela of COVID-19
	Code first the specific condition related to COVID-19 if known, such as:
Add	post-exertional malaise (R68.85)
Add	myalgic encephalomyelitis/chronic fatigue syndrome (G93.32)

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Serotonin Syndrome

Serotonin Syndrome is a rare but potentially life-threatening condition associated with excess serotonin in the central nervous system. Causes include various drug mechanisms such as therapeutic medication use, overdose, or drug interaction(s). Signs and symptoms can range from mild to life threatening, and include tachycardia, hypertension, diaphoresis, tachypnea, altered mental status, dry mucus membranes, flushed skin, shivering, headache, diarrhea, dilated pupils, nausea and vomiting, tremor, clonus, hyperreflexia, hallucinations, hyperthermia, muscle rigidity, agitated delirium. Life threatening manifestations include seizures, coma, disseminated intravascular coagulation, hypotension, ventricular tachycardia, and metabolic acidosis. Treatment ranges from observation and administration of benzodiazepines, hospital admission for cardiac monitoring and administration of Cyproheptadine, to the intensive care unit admission for cooling measures, sedation, skeletal muscle (SkM) paralysis, ventilation, along with treatment of Esmolol or nitroprusside.

There is currently no ICD-10-CM code to describe patients who present with Serotonin Syndrome, but there is an increasing incidence of this condition thought to correspond with the increasing use of serotonergic agents in medical care.

Assigning ICD-10-CM codes for the manifestations of Serotonin Syndrome (along with any adverse effect or poisoning code, as applicable) does captures a vague clinical picture, but fails to completely identify and describe the underlying cause of the manifestations. With the increasing incidence of Serotonin Syndrome, it is possible one of the reasons it is under-represented may be due to the lack of a specific ICD-10-CM code to identify the syndrome itself.

Creating a new code for Serotonin Syndrome would allow for improved data analysis, and a more accurate representation of the morbidity that Serotonin Syndrome carries with it.

The proposal was submitted by Bon Secours Mercy Health.

References

<https://www.everydayhealth.com/serotonin/guide/>

<https://www.thehealthy.com/healthcare/drugs-medicine/serotonin-syndrome-symptoms/>

<https://www.uptodate.com/contents/serotonin-syndrome-serotonin-toxicity>

<https://www.everydayhealth.com/depression-pictures/serotonin-syndrome-things-you-need-to-know.aspx>

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

TABULAR MODIFICATION

G90 Disorders of autonomic nervous system

G90.8 Other disorders of autonomic nervous system

New code G90.81 Serotonin syndrome

Add Serotonin toxicity

New Code G90.89 Other disorders of autonomic nervous system

Add Code first poisoning due to drug or toxin, if applicable
(T42-T43 with sixth character 1-4 or 6)

Add Use additional code, if applicable, to identify:

Add disseminated intravascular coagulation (D65)

Add hypertensive crisis (I16.-)

Add metabolic acidosis (E87.2.-)

Add shock, not elsewhere classified (R57.-)

Add toxic encephalopathy (G92.-)

Add ventricular tachycardia (I47.2.-)

Add Use additional code for adverse effect, if applicable, to identify
drug (T42-T43 with sixth character of 5)

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Usher Syndrome

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

- Type 1: Children have profound hearing loss or deafness at birth and may have severe balance problems that may lead to delayed motor milestones. Many obtain little or no benefit from hearing aids, but early use of cochlear implants may allow for development of speech. Decreased night vision by age 10, progressing to severe vision loss by midlife.
- Type 2: Moderate to severe hearing loss at birth. Normal balance. Decreased night vision by adolescence, progressing to severe vision loss by midlife.
- Type 3: Progressive hearing loss in childhood or early teens. Normal to near-normal balance in childhood. Chance of later problems. Vision loss varies in severity and age of onset; night vision problems often begin in teens and progress to severe vision loss by midlife.

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

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

References

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

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

TABULAR MODIFICATIONS

H91 Other and unspecified hearing loss

H91.8 Other specified hearing loss

New
sub-subcategory

H91.81 Usher syndrome

Add

Code also, if applicable, any associated retinal dystrophy
such as:

Add

other dystrophies primarily involving the sensory retina
(H35.53)

Add

pigmentary retinal dystrophy (H35.52)

New code

H91.810 Usher syndrome, type 1

New code

H91.811 Usher syndrome, type 2

New code

H91.812 Usher syndrome, type 3

New code

H91.818 Other Usher syndrome

New code

H91.819 Usher syndrome, unspecified

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

TABULAR MODIFICATIONS PROPOSED ADDENDA

All approved modifications will be effective April 1, 2024

Add	B04 Monkeypox <u>Mpox</u>	
	D68 Other coagulation defects D68.6 Other thrombophilia D68.61 Antiphospholipid syndrome	
Delete		Excludes1: anti-phospholipid antibody, finding without diagnosis (R76.0)
Delete		anti-phospholipid antibody with hemorrhagic disorder (D68.312)
Delete		lupus anticoagulant syndrome (D68.62)
Add		Excludes2: anti-phospholipid antibody, finding without diagnosis (R76.0)
Add		anti-phospholipid antibody with hemorrhagic disorder (D68.312)
Add		lupus anticoagulant syndrome (D68.62)
	D68.62	Lupus anticoagulant syndrome
Delete		Excludes1: anticardiolipin syndrome (D68.61)
Delete		antiphospholipid syndrome (D68.61)
Delete		lupus anticoagulant (LAC) finding without diagnosis (R76.0)
Delete		lupus anticoagulant (LAC) with hemorrhagic disorder (D68.312)
Add		Excludes2: anticardiolipin syndrome (D68.61)
Add		antiphospholipid syndrome (D68.61)
Add		lupus anticoagulant (LAC) finding without diagnosis (R76.0)
Add		lupus anticoagulant (LAC) with hemorrhagic disorder (D68.312)
	E27 Other disorders of adrenal gland E27.5 Adrenomedullary hyperfunction	
Add		Code also, if applicable, pheochromocytoma, by type (I15.-)

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

E89	Postprocedural endocrine and metabolic complications and disorders, not elsewhere classified
E89.1	Postprocedural hypoinsulinemia Postpancreatectomy hyperglycemia Postsurgical hypoinsulinemia
Add	Code first, if applicable, diabetes mellitus (postpancreatectomy) (postprocedural) (E13.-)
Delete	Use additional code, if applicable, to identify: acquired absence of pancreas (Z90.41-) diabetes mellitus (postpancreatectomy) (postprocedural) (E13.-)
I49	Other cardiac arrhythmias
Delete	Excludes1: neonatal dysrhythmia (P29.1-)
Delete	sinoatrial bradycardia (R00.1)
Delete	sinus bradycardia (R00.1)
Delete	vagal bradycardia (R00.1)
Add	Excludes2: bradycardia NOS (R00.1)
Add	neonatal dysrhythmia (P29.1-)
Add	sinoatrial bradycardia (R00.1)
Add	sinus bradycardia (R00.1)
Add	vagal bradycardia (R00.1)
J12	Viral pneumonia, not elsewhere classified
Delete	Excludes1: aspiration pneumonia due to anesthesia during labor and delivery (O74.0)
Delete	aspiration pneumonia due to anesthesia during pregnancy (O29)
Delete	aspiration pneumonia due to anesthesia during puerperium (O89.0)
Delete	aspiration pneumonia due to solids and liquids (J69.-)
Delete	aspiration pneumonia NOS (J69.0)
Delete	congenital pneumonia (P23.0)
Delete	congenital rubella pneumonitis (P35.0)
Delete	interstitial pneumonia NOS (J84.9)
Delete	lipid pneumonia (J69.1)
Delete	neonatal aspiration pneumonia (P24.-)

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Add	Excludes2: aspiration pneumonia due to anesthesia during labor and delivery (O74.0)
Add	aspiration pneumonia due to anesthesia during pregnancy (O29)
Add	aspiration pneumonia due to anesthesia during puerperium (O89.0)
Add	aspiration pneumonia due to solids and liquids (J69.-)
Add	aspiration pneumonia NOS (J69.0)
Add	congenital pneumonia (P23.0)
Add	congenital rubella pneumonitis (P35.0)
Add	interstitial pneumonia NOS (J84.9)
Add	lipid pneumonia (J69.1)
Add	neonatal aspiration pneumonia (P24.-)
	 J18.0 Bronchopneumonia, unspecified organism
	Excludes2: acute bronchiolitis (J21.-)
Revise	chronic bronchiolitis (J44.9) (<u>J44.89</u>)
Add	other specified chronic obstructive pulmonary disease (J44.89)
	 J45 Asthma
Revise	Excludes2: asthma with chronic obstructive pulmonary disease (J44.9) (<u>J44.89</u>)
Revise	chronic asthmatic (obstructive) bronchitis (J44.9) (<u>J44.89</u>)
Revise	chronic obstructive asthma (J44.9) (<u>J44.89</u>)
Add	other specified chronic obstructive pulmonary disease (J44.89)
	 K66 Other disorders of peritoneum
	K66.0 Peritoneal adhesions (postprocedural) (postinfection)
Delete	Excludes1: female pelvic adhesions [bands] (N73.6)
Add	Excludes2: female pelvic adhesions [bands] (N73.6)
Add	female pelvic postprocedural adhesions (N99.4)
	 O85 Puerperal sepsis
	Postpartum sepsis
	Puerperal peritonitis
	Puerperal pyemia
	Excludes1:
Delete	genital tract infection following delivery (O86.1-)
Delete	urinary tract infection following delivery (O86.2-)
	Excludes2:
Delete	sepsis during labor (O75.3)
Add	genital tract infection following delivery (O86.1-)

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Add		sepsis during labor (O75.3)
Add		urinary tract infection following delivery (O86.2-)
	R68	Other general symptoms and signs
		R68.2 Dry mouth, unspecified
		Excludes1: dry mouth due to dehydration (E86.0)
		dry mouth due to Sjögren syndrome (M35.0-)
Delete		salivary gland hyposalivation (K11.7)
Add		Excludes2: salivary gland hyposalivation (K11.7)
	Z59	Problems related to housing and economic circumstances
		Z59.1 Inadequate housing
Delete		Excludes1: problems related to the natural and physical environment (Z77.1-)
Add		Excludes2: problems related to the natural and physical environment (Z77.1-)

ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023

INDEX MODIFICATIONS PROPOSED ADDENDA
All approved modifications will be effective April 1, 2024

- Depression (acute) (mental) F32.A
Revise - central nervous system ~~R09.2~~ G98.8
- Anemia (essential) (general) (hemoglobin deficiency) (infantile) (primary) (profound) D64.9
Revise - myelodysplastic (see also Syndrome, myelodysplastic) D46.9
- Ascites (abdominal) R18.8
- due to
Revise - - hepatitis – see also Hepatitis
Revise - - - chronic active (see also Hepatitis, chronic active) ~~K71.51~~ R18.8
Add - - - with toxic liver disease K71.51
- Cachexia R64
Revise - cancerous (see also Cancer) ~~R64~~ E88.A
Revise - malignant (see also Cancer) ~~R64~~ E88.A
Revise - pituitary (see also Hypopituitarism) ~~E23.0~~ E88.A
Revise - pulmonary (see also specific underlying lung disease) ~~R64~~ E88.A
Revise - renal (see also specific underlying renal disease) ~~N28.9~~ E88.A
Revise - tuberculous NEC (-see also Tuberculosis) E88.A
- Calculus, calculi, calculous
- biliary -see also Calculus, gallbladder
Add - - with bile duct involvement - see also Calculus, bile duct
- kidney (impacted) (multiple) (pelvis) (recurrent) (staghorn) N20.0
- - with calculus, ureter N20.2
Add - - - with hydronephrosis N13.2
Add - - - with infection N13.6
- - congenital Q63.8
Add - - - with hydronephrosis N13.2
Add - - - with infection N13.6
- ureter (impacted) (recurrent) N20.1
Add - - with hydronephrosis N13.2
Add - - - with infection N13.6
- Monkeypox B04
Add - Mpox B04

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

TABULAR MODIFICATIONS PROPOSED ADDENDA

All approved modifications will be effective October 1, 2024

Revise	A18	Tuberculosis of other organs A18.4 Tuberculosis of skin and subcutaneous tissue Lupus exceedens <u>exedens</u>
Revise	A50	Congenital syphilis A50.4 Late congenital neurosyphilis [juvenile neurosyphilis] A50.45 Juvenile general paresis Dementia paralytica juvenilis Juvenile taboparetic <u>taboparetic</u> neurosyphilis
Revise	A77	Spotted fever [tick-borne rickettsioses] A77.4 Ehrlichiosis A77.41 Ehrlichiosis chafeensis <u>chaffeensis</u> [E. chafeensis <u>chaffeensis</u>]
Add Add	E07	Other disorders of thyroid E07.1 Dyshormogenetic goiter <u>Dyshormonogenetic</u> goiter Familial dyshormonogenetic goiter
Add	E08	Diabetes mellitus due to underlying condition Use additional code to identify control using: injectable non-insulin antidiabetic drugs (Z79.85)
Add	E09	Drug or chemical induced diabetes mellitus Use additional code to identify control using: injectable non-insulin antidiabetic drugs (Z79.85)
Add	E11	Type 2 diabetes mellitus Use additional code to identify control using: injectable non-insulin antidiabetic drugs (Z79.85)
Add	E13	Other specified diabetes mellitus Use additional code to identify control using: injectable non-insulin antidiabetic drugs (Z79.85)

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Delete	F01	<p>Vascular dementia Vascular dementia as a result of infarction of the brain due to vascular disease, including hypertensive cerebrovascular disease. Includes: arteriosclerotic dementia major neurocognitive disorder due to vascular disease multi-infarct dementia</p> <p>Code first the underlying physiological condition or sequelae of cerebrovascular disease.</p>
Add		Code first, if applicable, any causal condition
Delete	F02	<p>Dementia in other diseases classified elsewhere</p> <p>Code first the underlying physiological condition, such as:</p> <p>dementia with Lewy bodies</p>
Delete		dementia with Parkinsonism
Add		neurocognitive disorder with Lewy bodies (G31.83)
Add		other frontotemporal neurocognitive disorder (G31.90)
Delete	F03	<p>Unspecified dementia Excludes1: senility NOS (R41.81) Excludes2: mild memory disturbance due to known physiological condition (F06.8)</p>
Revise		senile dementia with delirium or acute confusional state (F05)
Delete	F05	<p>Delirium due to known physiological condition Excludes1: delirium NOS (R41.0)</p>
Revise	F06	<p>Other mental disorders due to known physiological condition Excludes1: unspecified dementia (F03) (F03.-)</p>
Revise	F06.7	<p>Mild neurocognitive disorder due to known physiological condition Code first the underlying physiological condition, such as: Alzheimer's disease (G30.-) <u>other</u> frontotemporal neurocognitive disorder (G31.09)</p>
Revise		Excludes1: dementia (F01.-, F02.-, F03) <u>F03.-</u>)
Add	F20	<p>Schizophrenia Use additional code, if applicable, to identify: other specified cognitive deficit (R41.84-)</p>
Add	F45	<p>Somatoform disorders F45.2 Hypochondriacal disorders F45.22 Body dysmorphic disorder Bigorexia Muscle dysmorphia</p>

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Revise	H42	Glaucoma in diseases classified elsewhere Code first underlying condition, such as: Reiger's <u>Rieger</u> anomaly (Q13.81)
Revise	G21	Secondary parkinsonism Excludes1: dementia with Parkinsonism <u>neurocognitive disorder with Lewy bodies</u> (G31.83)
Revise	G30	Alzheimer's disease Excludes1: senile dementia NOS (F03)-(F03.-)
Revise	G31	Other degenerative diseases of nervous system, not elsewhere classified G31.8 Other specified degenerative diseases of nervous system G31.84 Mild cognitive impairment of uncertain or unknown etiology Excludes1: dementia (F01.-, F02.-, F03)- <u>F03.-</u>)
Revise	G93	Other disorders of brain G93.4 Other and unspecified encephalopathy G93.42 Megalencephalic <u>Megalencephalic</u> leukoencephalopathy with subcortical cysts
Revise	H44	Disorders of globe H44.2 Degenerative myopia H44.2A Degenerative myopia with choroidal neovascularization H44.2A3 Degenerative myopia with choroidal neovascularization, bilateral eye
Revise	H44	H44.2B Degenerative myopia with macular hole H44.2B3 Degenerative myopia with macular hole, bilateral eye
Revise	H44	H44.2C Degenerative myopia with retinal detachment H44.2C3 Degenerative myopia with retinal detachment, bilateral eye
Revise	H44	H44.2D Degenerative myopia with foveoschisis H44.2D3 Degenerative myopia with foveoschisis, bilateral eye
Revise	H44	H44.2E Degenerative myopia with other maculopathy H44.2E3 Degenerative myopia with other maculopathy, bilateral eye

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Add Add Add	J17	Pneumonia in diseases classified elsewhere Code also, if applicable, any associated condition such as: abscess (J85.1) aspiration pneumonia (J69.-)
Revise Add Add Delete Delete Delete Delete Delete Delete Delete	J18	Pneumonia, unspecified organism Code first, <u>if applicable</u> , associated influenza, if applicable (J09.X1, J10.0-, J11.0-) Code also, if applicable, any associated condition such as: aspiration pneumonia (J69.-) Excludes1: abscess of lung with pneumonia (J85.1) aspiration pneumonia due to anesthesia during labor and delivery (O74.0) aspiration pneumonia due to anesthesia during pregnancy (O29) aspiration pneumonia due to anesthesia during puerperium (O89.0) aspiration pneumonia due to solids and liquids (J69.-) aspiration pneumonia NOS (J69.0) lipid pneumonia (J69.1) pneumonitis due to external agents (J67-J70)
Add Add Add Add Add Add Add		Excludes2: abscess of lung with pneumonia (J85.1) aspiration pneumonia due to anesthesia during labor and delivery (O74.0) aspiration pneumonia due to anesthesia during pregnancy (O29) aspiration pneumonia due to anesthesia during puerperium (O89.0) aspiration pneumonia due to solids and liquids (J69.-) aspiration pneumonia NOS (J69.0) lipid pneumonia (J69.1) pneumonitis due to external agents (J67-J70)
Add	J69	Pneumonitis due to solids and liquids Codes also, if applicable, other types of pneumonias
Add	J84	Other interstitial pulmonary diseases Code also, if applicable, associated condition
Revise	K14	Diseases of tongue Excludes2: leukedema <u>leukoedema</u> of tongue (K13.29)

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Revise	K58 Irritable bowel syndrome K58.9 Irritable bowel syndrome, <u>unspecified</u> without diarrhea Irritable bowel syndrome NOS
Delete	M92 Other juvenile osteochondrosis M92.8 Other specified juvenile osteochondrosis Caleaneal apophysitis
Add	N39 Other disorders of urinary system N39.0 Urinary tract infection, site not specified Use additional code (B95-B97), to identify infectious agent. Excludes1: urinary tract infection of specified site, such as: pyonephrosis (N13.6)
Add Add Add	O24 Diabetes mellitus in pregnancy, childbirth, and the puerperium Use additional code, if applicable (for): injectable non-insulin antidiabetic drugs (Z79.85) long-term (current) use of insulin (Z79.4)
Revise	O24.1 Pre-existing type 2 diabetes mellitus, in pregnancy, childbirth and the puerperium Use additional code (for) : from category E11 to further identify any manifestations
Delete	long-term (current) use of insulin (Z79.4)
Add	O24.3 Unspecified pre-existing diabetes mellitus in pregnancy, childbirth and the puerperium Use additional code (for): from category E11 to further identify any manifestation injectable non-insulin antidiabetic drugs (Z79.85)
Add	O24.8 Other pre-existing diabetes mellitus in pregnancy, childbirth, and the puerperium Use additional code (for): from categories E08, E09 and E13 to further identify any manifestation injectable non-insulin antidiabetic drugs (Z79.85) long-term (current) use of insulin (Z79.4)
Delete	O24.9 Unspecified diabetes mellitus in pregnancy, childbirth and the puerperium Use additional code for long-term (current) use of insulin (Z79.4)
Add	Use additional code (for): injectable non-insulin antidiabetic drugs (Z79.85) long-term (current) use of insulin (Z79.4)

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

Z45	Encounter for adjustment and management of implanted device Z45.3 Encounter for adjustment and management of implanted devices of the special senses Z45.32 Encounter for adjustment and management of implanted hearing device
Revise	Excludes1: Encounter for fitting and adjustment of hearing aid <u>aid</u> (Z46.1)
Z79	Long term (current) drug therapy Z79.6 Long term (current) use of immunomodulators and immunosuppressants Z79.62 Long term (current) use of immunosuppressant Z79.624 Long term (current) use of inhibitors of nucleotide synthesis
Revise	Long term (current) use of mycophenolate <u>mycophenolate</u>
Z91	Personal risk factors, not elsewhere classified Z91.1 Patient's noncompliance with medical treatment and regimen Z91.12 Patient's intentional underdosing of medication regimen
Delete	Excludes1: adverse effect of prescribed drug taken as directed—code to adverse effect poisoning (overdose)—code to poisoning
Z91.13	Patient's unintentional underdosing of medication regimen
Delete	Excludes1: adverse effect of prescribed drug taken as directed—code to adverse effect poisoning (overdose)—code to poisoning
Z91.14	Patient's other noncompliance with medication regimen Patient's underdosing of medication NOS Code first, if applicable, adverse effect of underdosing (T36-T50)
Add	
Z93	Artificial opening status Z93.2 Ileostomy status Ileal pouch status Kock pouch status
Add	
Add	

ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023

INDEX MODIFICATION PROPOSED ADDENDA
All approved modifications will be effective October 1, 2024

- Adenoma -see also Neoplasm, benign, by site
Revise - prostate -see Neoplasm, ~~benign~~, prostate, benign
- Allergy, allergic (reaction) (to) T78.40
Revise - bee sting (anaphylactic shock) -~~see Toxicity, venom, arthropod, bee T63.44-~~
- Anetoderma (maculosum) (of) L90.8
Revise - ~~Schweniger~~ Schweninger-Buzzi L90.1
- Aneurysm (anastomotic) (artery) (cirroid) (diffuse) (false) (fusiform) (multiple)
(saccular) I72.9
- brain I67.1
- - arteriovenous (congenital) (nonruptured) Q28.2
- - - acquired I67.1
Revise - - - - ruptured -see Aneurysm, arteriovenous, brain, ruptured ~~I60.8-~~
Revise - - - - ruptured -see Aneurysm, arteriovenous, brain, ruptured ~~I60.8-~~
- Apophysitis (bone) -see also Osteochondropathy
Revise - calcaneus ~~M92.8~~ M92.6
- Asplenia
Add - functional D73.0
- Bacteriuria, bacteruria (asymptomatic) R82.71
Delete -~~asymptomatic~~ ~~R82.71~~
- Bee sting (with allergic or anaphylactic shock) -~~see Toxicity, venom, arthropod,~~
Revise ~~bee T63.44-~~
- Breakdown
- device, graft or implant -see also Complications, by site and type, mechanical
T85.618
- - genital NEC T83.418
Revise - - - penile prosthesis (cylinder) (implanted) (pump) (~~resevoir~~) (reservoir) T83.410
- Brion-Kayser disease -see Fever, ~~parathyroid~~ paratyphoid
- Bursitis M71.9
Revise - ankle -see Enthesopathy, ~~lower limb~~, ankle, ~~specified type~~ NEC and tarsus

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

- Carcinoma (malignant) -see also Neoplasm, by site, malignant
- in
- Revise - - pleomorphic adenoma -see Neoplasm, salivary glands or duct, malignant
- Chancre (any genital site) (hard) (hunterian) (mixed) (primary) (seronegative)
(seropositive) (syphilitic) A51.0
- Add - ducreyi A57
- Revise Child – see also Problem, child
- Colitis (acute) (catarrhal) (chronic) (noninfective) (hemorrhagic) (see also
Enteritis) K52.9
- Add - Clostridioides difficile
- Add - - not specified as recurrent A04.72
- Add - - recurrent A04.71
- toxic NEC K52.1
- Revise - - due to Clostridium difficile
- Add - - - Clostridioides difficile
- Add - - - - not specified as recurrent A04.72
- Add - - - - recurrent A04.71
- Add - - - Clostridium difficile
- Revise - - - - not specified as recurrent A04.72
- Revise - - - - recurrent A04.71
- Complication(s) (from) (of)
- catheter (device) NEC -see also Complications, prosthetic device or implant
- - dialysis (vascular) T82.9
- Revise - - - intraperitoneal -see Complications, catheter, intraperitoneal dialysis
- joint prosthesis, internal T84.9
- - mechanical
- Add - - - osteolysis T84.059
- Add - - - - hip T84.05-
- Add - - - - knee T84.05-
- - - perforation - see Complications, joint prosthesis, mechanical, specified NEC
- Delete ~~_____ osteolysis T84.059~~
- Delete ~~_____ hip T84.05-~~
- Delete ~~_____ knee T84.05-~~
- Convulsions (idiopathic) (see also Seizure(s)) R56.9
- Revise - epileptiform, epileptoid - see ~~Seizure, epileptiform~~ Epilepsy
- Revise ~~Cotunngo's~~ Cotugno's disease -see Sciatica

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

- Revise ~~Dacryocystoblenorrhoea~~ Dacryocystoblenorrhoea -see Inflammation, lacrimal, passages, chronic
- Deficiency, deficient
- Revise - ~~phosphomannomutase~~ phosphomannomutase E74.818
- Degeneration, degenerative
- Revise - intervertebral disc ~~NOS~~
- Revise Delirium, delirious (acute or subacute) (not alcohol- or drug-induced) (~~with dementia~~) R41.0
- Add - with
- Add - - dementia (see also Dementia) F05
- Revise Dementia (degenerative (primary)) (~~old age~~) (persisting) (unspecified severity) (without behavioral disturbance, psychotic disturbance, mood disturbance, and anxiety) F03.90
- with
- Add - - acute confusional state F05
- Add - - old age (senile) F03
- Add - - Alzheimer's type – see Disease, Alzheimer's, late onset
- senile F03
- Delete ~~—with acute confusional state F05~~
- Diabetes, diabetic (mellitus) (sugar) E11.9
- due to drug or chemical E09.9
- - with
- Add - - - retina, hemorrhage E09.39
- - - retinopathy E09.319
- - - - with macular edema E09.311
- Revise - - - - _ resolved following treatment E09.37
- due to underlying condition E08.9
- - with
- - - retinopathy E08.319
- - - - with macular edema E08.311
- Revise - - - - _ resolved following treatment E08.37
- specified type NEC E13.9
- - with
- - - retinopathy E13.319
- - - - with macular edema E13.311
- Revise - - - - _ resolved following treatment E13.37

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

- type 1 E10.9

Add -- retina, hemorrhage E10.39

 - with
 - retinopathy E10.319
 - with macular edema E10.311

Revise ---- resolved following treatment E10.37

- type 2 E11.9

Add -- retina, hemorrhage E11.39

 - with
 - retinopathy E11.319
 - with macular edema E11.311

Revise ---- resolved following treatment E11.37

- Disease, diseased - see also Syndrome

Revise - Cotugno's - see Sciatica

Revise - myelodysplastic (see also Syndrome, ~~myelodysplasia~~ myelodysplastic) C94.6

- Disorder (of) -see also Disease

Revise - catatonic NOS F06.1

Revise -- organic NOS F06.1

 - metabolism NOS E88.9
 - amino-acid E72.9
 - aromatic E70.9

Revise ---- ~~propionate~~ propionate NEC E71.128

Revise ---- ~~propionic~~ propionic acidemia E71.121

- organic

Revise -- catatonic NOS F06.1

- Displacement, displaced

- device, implant or graft -see also Complications, by site and type, mechanical

T85.628 -- genital NEC T83.428

Revise --- penile prosthesis (cylinder) (implanted) (pump) (~~resevoir~~) (reservoir) T83.420

- Add Ducrey's chancre A57

- Add Dysmorphia

Add - muscle F45.22

- Ehrlichiosis A77.40

 - due to

Revise -- E. ~~chaffeensis~~ chaffeensis A77.41

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

- Embolism (multiple) (paradoxical) I74.9
 Revise - cavernous sinus (venous) -see Embolism, intracranial, venous sinus
- Endometritis (decidual) (nonspecific) (purulent) (senile) (atrophic) (suppurative) N71.9
 - with ectopic pregnancy O08.0
 - acute N71.0
 Revise - ~~hemorrhagic~~ blennorrhagic (gonococcal) (acute) (chronic) A54.24
- Encounter (with health service) (for) Z76.89
 Add - palliative care Z51.5
- Enteritis (acute) (diarrheal) (hemorrhagic) (noninfective) K52.9
 Add - Clostridioides difficile
 Add - - not specified as recurrent A04.72
 Add - - recurrent A04.71
- infectious NOS A09
 - - due to
 Add - - - Clostridioides difficile
 Add - - - - not specified as recurrent A04.72
 Add - - - - recurrent A04.71
- toxic NEC K52.1
 Revise - - due to ~~Clostridium difficile~~
 Add - - - Clostridioides difficile
 Add - - - - not specified as recurrent A04.72
 Add - - - - recurrent A04.71
- - - Clostridium difficile
 Revise - - - - not specified as recurrent A04.72
 Revise - - - - recurrent A04.71
- Enterocolitis (see also Enteritis) K52.9
 Revise - due to ~~Clostridium difficile~~
 Add - - Clostridioides difficile
 Add - - - not specified as recurrent A04.72
 Add - - - recurrent A04.71
- - Clostridium difficile
 Revise - - - not specified as recurrent A04.72
 Revise - - - recurrent A04.71
- necrotizing K55.30
 Revise - - due to ~~Clostridium difficile~~
 Add - - - Clostridioides difficile

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

- chronic K73.9
- - active NEC K73.2
- Add - - - toxic – see Disease, liver, toxic, with, hepatitis, chronic, active
- Add - - toxic - see Disease, liver, toxic, with, hepatitis, chronic
- Add - - viral - see Hepatitis, viral, chronic

- Add - D B16.1
- Add - - acute B16.1

- Add - - - with hepatic coma B16.0
- Add - - - in hepatitis B carrier B17.0
- Add - - chronic B18.0
- Add - delta (agent) – see Hepatitis, D

- viral, virus B19.9
- - chronic B18.9
- - - type
- Add - - - - D B18.0

- - type
- Add - - - D B16.1
- Add - - - - acute B16.1
- Add - - - - - with hepatic coma B16.0
- Add - - - - - in hepatitis B carrier B17.0
- Add - - - - chronic B18.0
- Add - - - delta (agent) – see Hepatitis, viral, type, D
- Herpes, herpesvirus, herpetic B00.9
- simplex B00.9
- Revise - - ~~myelitis~~ myelitis B00.82

- History
- - malignant neoplasm (of) Z85.9
- Revise - - - ~~carcinoid~~ -see History, personal (of), malignant neoplasm, by site, ~~carcinoid~~
carcinoid

- Hypertension, hypertensive (accelerated) (benign) (essential) (idiopathic)
(malignant) (systemic) I10

- due to
- Revise - - pheochromocytoma (see also Pheochromocytoma, by type) I15.2

- secondary NEC I15.9
- - due to
- Revise - - - pheochromocytoma (see also Pheochromocytoma, by type) I15.2

- Revise ~~Hyparrhythmia~~ Hypsarrhythmia -see Epilepsy, generalized, specified NEC

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

- Infection, infected, infective (opportunistic) B99.9
- Bacillus A49.9
- Revise - - ~~Ducrey's ducreyi~~ (any location) A57
- Revise - ~~Ducrey ducreyi~~ Haemophilus (any location) A57
- Add - Clostridioides
Add - - difficile
Add - - - as cause of disease classified elsewhere B96.89
Add - - - foodborne (disease)
Add - - - - not specified as recurrent A04.72
Add - - - - recurrent A04.71
Add - - - gas gangrene A48.0
Add - - - necrotizing enterocolitis
Add - - - - not specified as recurrent A04.72
Add - - - - recurrent A04.71
Add - - - sepsis A41.4
- due to or resulting from
- - device, implant or graft -see also Complications, by site and type, infection or inflammation T85.79
- Revise - - - penile (cylinder) (pump) (~~resevoir~~) (reservoir) T83.61
- Hemophilus
- Revise - - ~~ducrey ducreyi~~ (any location) A57
- Revise - Salmonella (aertrycke) (arizonae) (~~gallinarum~~) (cholerae-suis) (enteritidis) (gallinarum) (suipestifer) (typhimurium) A02.9
- Inflammation, inflamed, inflammatory (with exudation)
- due to device, implant or graft - see also Complications, by site and type, infection or inflammation
- Revise - - penile (cylinder) (pump) (~~resevoir~~) (reservoir) T83.61
- Intoxication
- Revise - hepatocerebral ~~intoxication~~ K76.82
- ~~Kerunoparalysis~~ Keraunoparalysis T75.09
- Revise Köhler-Pellegrini-Steida Stieda disease or syndrome (calcification, knee joint) – see Bursitis, tibial collateral
- Revise Larsen-Johansson disease or osteochondrosis -see Osteochondrosis, juvenile, patella

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

- Leak, leakage
- device, implant or graft -see also Complications, by site and type, mechanical
-- genital NEC T83.498
Revise --- penile prosthesis (cylinder) (implanted) (pump) (~~reservoir~~) (reservoir) T83.490
- Revise Loa loa, ~~loiasis, loasis~~ (loiasis) B74.3
- Revise Long-term (current) (prophylactic) drug therapy (use of)
- non-insulin antidiabetic drug, injectable ~~Z79.899~~ Z79.85
- Revise - omycophenolate Z79.624
- Malnutrition E46
- degree
-- severe (protein-energy) E43
Add --- with
Add ---- kwashiorkor E40
Add ---- marasmus E41
---- intermediate form E42
---- with
Delete ~~---- marasmus E41~~
- protein E46
-- calorie E46

---- severe E43
Add ---- with
Add ----- kwashiorkor E40
Add ----- marasmus E41
----- intermediate form E42
----- with
Delete ~~----- marasmus E41~~

-- energy E46
--- severe E43
Add ---- with
Add ----- kwashiorkor E40
Add ----- marasmus E41

----- intermediate form E42
----- with
Delete ~~----- marasmus E41~~
- Malnutrition E46
- severe (protein-energy) E43
-- with
Revise --- kwashiorkor (~~and marasmus~~) E42 E40

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

- Add - - - with marasmus E42
 Add - - intermediate form E42
 Add - - - with
 Add - - - kwashiorkor (and marasmus) E42
- Malposition
 - device, implant or graft -see also Complications, by site and type, mechanical
 T85.628
 - - genital NEC T83.428
 Revise - - - penile prosthesis (cylinder) (implanted) (pump) (~~reservoir~~) (reservoir) T83.420
- Mastopathy, mastopathia N64.9
 Revise - estrogenic, oestrogenica, oestrogenic N64.89
- Megacolon (acquired) (functional) (not Hirschsprung's disease) (in) K59.39
 - toxic NEC K59.31
 Revise - - due to ~~Clostridium difficile~~
 Add - - - Clostridioides difficile
 Add - - - - not specified as recurrent A04.72
 Add - - - - recurrent A04.71
 Add - - - Clostridium difficile
 Revise - - - - not specified as recurrent A04.72
 Revise - - - - recurrent A04.71
- Migraine (idiopathic) G43.909
 - not intractable G43.909
 Revise - - without status migrainosus ~~G43.919~~ G43.909
- Myelodysplastic syndrome -see also Syndrome, myelodysplastic D46.9
 - with
 Add - - pancytopenia, acquired see- Syndrome, myelodysplastic, pancytopenia
- Obstruction, obstructed, obstructive
 - device, implant or graft -see also Complications, by site and type, mechanical
 T85.698
 - - genital NEC T83.498
 Revise - - - penile prosthesis (cylinder) (implanted) (pump) (~~reservoir~~) (reservoir) T83.490
- Revise ~~Omentocoele~~ Omentocoele -see Hernia, abdomen, specified site NEC
- Osteonecrosis M87.9
 - secondary NEC M87.30
 - - due to

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

- Revise --- hemoglobinopathy NEC D58.2 [M90.50]
 Add ---- multiple sites D58.2 [~~M90.58~~] [M90.59]
 ---- specified NEC D58.2 [M90.58]
- Pancytopenia (acquired) D61.818
 - with
 Revise -- myelodysplastic syndrome -see Syndrome, myelodysplastic, pancytopenia (acquired)
- Paralysis, paralytic (complete) (incomplete) G83.9
 Revise - ~~subcapsularis~~ subscapularis G56.8-
 Poisoning (acute) -see also Table of Drugs and Chemicals
 - food NEC A05.9
 - - due to
 Revise --- salmonella (aertrycke) (~~callinarum~~) (choleraesuis) (enteritidis) (gallinarum) (paratyphi) (suipestifer) A02.9
- Pregnancy
 - complicated by (care of) (management affected by)
 - - anemia
 Add --- complicating childbirth O99.02
- Protrusion, protrusio
 - device, implant or graft -see also Complications, by site and type, mechanical
 T85.698
 - - genital NEC T83.498
 Revise --- penile prosthesis (cylinder) (implanted) (pump) (~~reservoir~~) (reservoir) T83.490
- Retinopathy (background) H35.00
 - proliferative NEC H35.2-
 Revise - - thaslassemia thalassemia H35.2
- Revise Robinow-~~Silverman~~-Silverman-Smith syndrome Q87.19
- Rupture, ruptured
 - globe (eye) (traumatic) -see Injury, eye, laceration
 Add - - nontraumatic (see also Disorder, globe) H44.89
- Schizophrenia, schizophrenic F20.9
 - catatonic (type) (excited) (withdrawn) F20.2
 Add - - catatonic NOS F06.1
- Revise Schweniger Schweninger-Buzzi anetoderma L90.1
- Secretion

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

- Revise - catecholamine, by pheochromocytoma (see also Pheochromocytoma, by type) E27.5
- hormone
- by
- Revise --- carcinoid tumor (see also Tumor, carcinoid) E34.0
- Revise --- pheochromocytoma (see also Pheochromocytoma, by type) E27.5
- Sprain (joint) (ligament)
- sternoclavicular joint S43.6-
- sternum S23.429
- Revise -- sternoclavicular (joint) (ligament) ~~S23.429~~ S43.6-
- Status (post) -see also Presence (of)
- hysterectomy (complete) (total) Z90.710
- Revise -- partial (with remaining ~~perineal~~ cervical stump) Z90.711
- Stenosis, stenotic (cicatricial) -see also Stricture
- artery NEC -see also Arteriosclerosis I77.1
- Revise -- celiac (compression) I77.4
- Add --- arteriosclerotic I70.8
- Add --- atherosclerosis I70.8
- Stricture -see also Stenosis
- artery I77.1
- Revise -- celiac (compression) I77.4
- Add --- arteriosclerotic I70.8
- Add --- atherosclerosis I70.8
- Surveillance (of) (for) -see also Observation
- Add - neoplasm - see Screening, neoplasm
- Syndrome -see also Disease
- Revise - cervicothoracic outlet (see also Syndrome, thoracic outlet) G54.0
- Fitzhugh-Curtis
- due to
- Chlamydia trachomatis A74.81
- Revise --- Neisseria ~~gonorrhoea~~ gonorrhoea (gonococcal peritonitis) A54.85
- Revise - Köhler-Pellegrini-~~Steida~~ Stieda -see Bursitis, tibial collateral
- Revise - ~~Lemiere~~ Lemierre I80.8
- myeloproliferative (chronic) D47.1
- Add -- pancytopenia (acquired), D47.1 [D68.818]

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

- Revise - outlet (thoracic) (see also Syndrome, thoracic outlet) G54.0
- thoracic outlet (compression) G54.0
- Add - - arterial I77.89
- Add - - neurogenic G54.0
- Add - - venous I87.1
- Threatened
- Revise - abuse (harm) -see Maltreatment, adult, threatened abuse; or Maltreatment, child, threatened abuse
- Thoracic -see also condition
- Revise - outlet syndrome (see also Syndrome, thoracic outlet) G54.0
- Thrombosis, thrombotic (bland) (multiple) (progressive) (silent) (vessel) I82.90
- brain (artery) (stem) -see also Occlusion, artery, cerebral
- Revise - - sinus -see Thrombosis, intracranial₂ venous sinus
- Revise - cavernous (venous) sinus -see Thrombosis, intracranial₂ venous sinus
- Revise - cerebrovenous sinus -see also Thrombosis, intracranial₂ venous sinus
- intracranial (arterial) I66.9
- - venous sinus (any) G08
- Add - - - nonpyogenic I67.6
- Revise - lateral (venous) sinus -see Thrombosis, intracranial₂ venous sinus
- Revise - longitudinal (venous) sinus -see Thrombosis, intracranial₂ venous sinus
- Revise - sigmoid (venous) sinus -see Thrombosis, intracranial₂ venous sinus
- Revise - sinus, intracranial (any) -see Thrombosis, intracranial₂ venous sinus
- Revise Tuberosity, ~~entire~~ entire maxillary M26.07
- Tumor -see also Neoplasm, unspecified behavior, by site
- carcinoid D3A.00
- Revise - - ~~mesentary~~ mesentery metastasis C7B.04
- Revise - salivary gland or duct type, mixed -see Neoplasm, salivary gland or duct, benign
- Revise - - malignant -see Neoplasm, salivary gland or duct, malignant
- stromal
- - gastric D48.1
- Revise - - - malignant ~~E16.9~~ C49.A2

**ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023**

- Revise - Warthin's -see Neoplasm, salivary gland or duct, benign
- Use (of)
- Revise - alcohol (see also Alcohol, alcoholic, by problem) F10.90
- - with
- Add - - - anxiety disorder F10.980
- Revise Valsuani's disease O99.03 -~~see Anemia, obstetric~~
- Vomiting R11.10
- Revise - fecal ~~mater~~ matter R11.13
- Revise Warthin's tumor -see Neoplasm, salivary gland or duct, benign

ICD-10 Coordination and Maintenance Committee Meeting
September 12-13, 2023

ICD-10-CM External Cause of Injuries Index
All approved modifications will be effective October 1, 2024

- Assault (homicidal) (by) (in) Y09
- bodily force Y04.8
Revise - - sexual (confirmed) T74.2- ~~—see subcategories T74.0, T76.0~~
Add - - - suspected T76.2-
Revise - rape (confirmed) T74.2-
Add - - suspected T76.2-
- Explosion (accidental) (of) (with secondary fire) W40.9
- aircraft (in transit) (powered) NEC V95.9
- - stated as
Revise - - - homicide (attempt) ~~Y03.8~~ Y08.81
- firearm (parts) NEC W34.19
Revise - - ~~handgun~~ handgun W32.1
- Revise - handgun (parts) -see Explosion, firearm, ~~handgun~~ handgun (parts)
- Exposure (to) X58
- fire, flames (accidental) X08.8
- - in, of, on, starting in
Revise - - - ~~street car~~ streetcar (in motion) V82.8
- Powder burn (by) (from)
Revise - ~~firearm~~ firearm NEC W34.19
- Revise Rape (attempted) (confirmed) T74.2-
Add - suspected T76.2-
- Recoil
Revise - ~~firearm~~ firearm NEC W34.19

ICD-10-CM TABLE of DRUGS and CHEMICALS

All approved modifications will be effective October 1, 2024

Substance	Poisoning Accidental (unintentional)	Poisoning Intentional self-harm	Poisoning Assault	Poisoning Undetermined	Adverse effect	Underdosing
Revise Hydroxyzine (anxiolytic)	T43.591 <u>T43.581</u>	T43.592 <u>T43.582</u>	T43.593 <u>T43.583</u>	T43.594 <u>T43.584</u>	T43.595 <u>T43.585</u>	T43.596 <u>T43.586</u>
Add - antiallergic	T45.0X1	T45.0X2	T45.0X3	T45.0X4	T45.0X5	T45.0X6

ICD-10 Coordination and Maintenance Committee Meeting

ICD-10-CM TABLE of NEOPLASMS
All approved modifications will be effective October 1, 2025

Neoplasm	Malignant Primary	Malignant Secondary	Benign	Ca in situ	Uncertain Behavior	Unspecified Behavior
Neoplasm, neoplastic						
Revise - odontogenic -see Neoplasm, jaw, bone						