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

Welcome and announcements
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Diagnosis Topics:

Contents

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**

	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

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:

	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.

	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.**

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.**

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

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Continuing Education Credits

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

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

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

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

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

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

Abnormal 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 RA[5]. Furthermore, individuals who have abnormal RA-related immunologic tests without IA are identified in growing numbers in clinical care. Notably, these individuals may have symptoms such as joint pain, stiffness or swelling, but no other objective evidence of IA[6-8]. There are current recommendations for medical follow-up and lifestyle changes (e.g., smoking cessation) that can be applied to these individuals. In addition, the predictive ability of RF and ACPA for future clinical RA has underpinned multiple clinical observational studies and prevention trials in RA[9-15], 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/arthralgia (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).

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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)

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 cells[1].

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 FDA[2]. 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:

- 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 used[6]. 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.

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

References

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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)

T45 Poisoning by, adverse effect of and underdosing of primarily systemic and hematological agents, not elsewhere classified

The appropriate 7th character is to be added to each code from category T45
A - initial encounter
D - subsequent encounter
S – sequela

T45.1 Poisoning by, adverse effect of and underdosing of antineoplastic and immunosuppressive drugs

Excludes1: poisoning by, adverse effect of and underdosing of tamoxifen (T38.6)

Add poisoning by, adverse effect of and underdosing of immune checkpoint inhibitors and immunostimulant drugs (T45.A)

New subcategory T45.A Poisoning by, adverse effect of and underdosing of immune checkpoint inhibitors and immunostimulant drugs

Add Excludes1: poisoning by, adverse effect of and underdosing of antineoplastic and immunosuppressive drug (T45.1)

New sub-subcategory T45.AX Poisoning by, adverse effect of and underdosing of immune checkpoint inhibitors and immunostimulant drugs

New code T45.AX1 Poisoning by immune checkpoint inhibitors and immunostimulant drugs, accidental (unintentional)

Add Poisoning by immune checkpoint inhibitors and immunosuppressive drugs NOS

New code T45.AX2 Poisoning by immune checkpoint inhibitors and immunostimulant drugs, intentional self-harm

New code T45.AX3 Poisoning by immune checkpoint inhibitors and immunostimulant drugs, assault

New code T45.AX4 Poisoning by immune checkpoint inhibitors and immunostimulant drugs, undetermined

New code T45.AX5 Adverse effect of immune checkpoint inhibitors and immunostimulant drugs

New code T45.AX6 Underdosing of immune checkpoint inhibitors and immunostimulant drugs

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)

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 re-recurrence and fecal incontinence.[7]

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

Takeda Pharmaceuticals America, Incorporated is proposing the following tabular modifications to enable better tracking of complex fistulas, facilitating greater understanding of anal fistula epidemiology, and improving treatment paradigms.

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TABULAR MODIFICATIONS

K50 Crohn's disease [regional enteritis]

Add — Use additional code to identify any associated fistulas, if applicable:
anal fistula (K60.3-)
anorectal fistula (K60.5-)
rectal fistula (K60.4-)

K51 Ulcerative colitis

Add — Use additional code to identify any associated fistulas, if applicable:
anal fistula (K60.3-)
anorectal fistula (K60.5-)
rectal fistula (K60.4-)

K60 Fissure and fistula of anal and rectal regions

Revise — **Excludes~~1~~2: abscess or cellulitis of anal and rectal regions (K61.-)**

Excludes2: anal sphincter tear (healed) (nontraumatic) (old) (K62.81)

K60.3 Anal fistula
Add Code first, if applicable:
Crohn's disease (K50.-)
ulcerative colitis (K51.-)
Add Excludes1: congenital fistula (Q43.6)

New code K60.30 Anal fistula, unspecified
Add Anal fistula NOS

New sub-subcategory K60.31 Anal fistula, simple
Add Low intersphincteric anal fistula
Add Superficial anal fistula

New code K60.311 Anal fistula, simple, initial
Add **Anal fistula, simple, new**

New code K60.312 Anal fistula, simple, persistent
Add Anal fistula, simple, chronic

New code K60.313 Anal fistula, simple, recurrent
Add Anal fistula simple, occurring following complete healing
New code K60.319 Anal fistula, simple, unspecified

New sub-subcategory K60.32 Anal fistula, complex
Add Extrasphincteric anal fistula
Add **High intersphincteric anal fistula**
Add Suprasphincteric anal fistula
Add Transsphincteric anal fistula

Add Code also, if applicable:
perianal abscess (K61.0)
rectovaginal fistula (N82.3)
stenosis of anus and rectum (K62.4)

New code K60.321 Anal fistula, complex, initial
Add **Anal fistula, complex, new**

New code K60.322 Anal fistula, complex, persistent
Add Anal fistula, complex, chronic

New code K60.323 Anal fistula, complex, recurrent
Add Anal fistula complex, occurring following complete healing

New code		K60.329	Anal fistula, complex, unspecified
	K60.4	Rectal fistula	
Add		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

New code		K60.423	Rectal fistula, complex, recurrent
Add			Rectal fistula complex occurring following complete healing
New code		K60.429	Rectal fistula, complex, unspecified
	K60.5	Anorectal fistula	
Add		Code first, if applicable: Crohn's disease (K50.-) ulcerative colitis (K51.-)	
Add		Excludes1: congenital fistula (Q43.6)	
New code	K60.50	Anorectal fistula, unspecified	
Add		Anorectal fistula NOS	
New sub-subcategory	K60.51	Anorectal fistula, simple	
Add		Low intersphincteric anorectal fistula	
Add		Superficial anorectal fistula	
New code		K60.511	Anorectal fistula, simple, initial
Add			**Anorectal fistula, simple, new**
New code		K60.512	Anorectal fistula, simple, persistent
Add			Anorectal fistula, simple, chronic
New code		K60.513	Anorectal fistula, simple, recurrent
Add			Anorectal fistula simple, occurring following complete healing
New code		K60.519	Anorectal fistula, simple, unspecified
New sub-subcategory	K60.52	Anorectal fistula, complex	
Add		Extrasphincteric anorectal fistula	
Add		**High intersphincteric anorectal fistula**	
Add		Suprasphincteric anorectal fistula	
Add		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		K60.521	Anorectal fistula, complex, initial
Add			**Anorectal fistula, complex, new**

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

INDEX MODIFICATIONS

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

Anosognosia

Anosognosia is 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 approaches[11] 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.

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

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

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

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

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

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

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

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TABULAR MODIFICATIONS

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

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

T78.08 Anaphylactic reaction due to eggs

New code T78.081 Anaphylactic reaction due to eggs

New code 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 T78.13 Other adverse food reaction due to eggs

New code 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

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).

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TABULAR MODIFICATIONS

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

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

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

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.

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.

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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~~

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

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 embolism[5] 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.

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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
	I26.09	Other pulmonary embolism with acute cor pulmonale
Add		Other thrombotic pulmonary embolism with acute cor pulmonale

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
	I26.99	Other pulmonary embolism without acute cor pulmonale
Add		Other thrombotic pulmonary embolism without acute cor pulmonale

Cholestatic Pruritus

This proposal was presented at the March 2023, ICD-10 Coordination and Maintenance Committee Meeting. Based on public comments, revisions have been made for consideration.

Cholestatic pruritus is the itching of the inside and outside of the skin as caused by liver disease. It is described by patients as intense itching on the outside and inside of the skin and persists even with the various drug treatments, making it almost untreatable.

Cholestatic pruritus has been defined as the impaired secretion of bile, due to liver diseases. This causes intense itching, loss of sleep, general discomfort, and in intense cases suicidal ideation to make the itching stop. This is a severe symptom of liver disease that can be drug-induced, which means one could be getting healthier but feeling worse due to symptoms from one's treatment.

Cholestatic pruritus is a very specific condition, and it would be beneficial to physicians, patients, and caregivers to have its own unique ICD-10-CM code.

The requested modification to the ICD-10-CM codes related to cholestatic pruritus is timely and relevant to align clinical documentation with the needs of clinical practice, patient and provider education, and epidemiology research. Medical and scientific research has been rapidly evolving and the public health implications are only now starting to be fully recognized.

The Global Liver Institute propose the following ICD-10-CM tabular modifications.

TABULAR MODIFICATIONS

	L29	Pruritus
		Excludes1: neurotic excoriation (L98.1)
		psychogenic pruritus (F45.8)
		L29.8 Other pruritus
New code		L29.81 Cholestatic pruritus
New code		L29.89 Other pruritus
Add		Code also, if applicable, type of liver disease
Add		Use additional code for adverse effect, if applicable, to identify drug (T36-T50 with fifth or sixth character 5)

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.

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WRISS data from Beth Hume, Injury Surveillance Program, Massachusetts Department of Public Health.

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

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 ⅓ 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.

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

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.

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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.-)

Option #2

	F88	Other disorders of psychological development		
Delete		~~Developmental agnosia~~		
Delete		~~Global developmental delay~~		
Delete		~~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	

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 ($p=4.09x10^{-5}$), 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,

facilitating their migration into tissues, thus reducing the number of circulating neutrophils an 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 membrance, 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.

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TABULAR MODIFICATIONS

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

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

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.

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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		E10.A1	Type 1 diabetes mellitus, presymptomatic, Stage 1
Add			Multiple confirmed islet autoantibodies with normoglycemia
New code		E10.A2	Type 1 diabetes mellitus, presymptomatic, Stage 2
Add			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.

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.

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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

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

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.

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.

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6. NCCN Clinical Practice Guidelines in Oncology: Breast Cancer, Version 4.2023.

TABULAR MODIFICATIONS

Malignant neoplasm of breast

C50 Malignant neoplasm of breast

Revise — Use additional code to identify estrogen, and other hormones and factors receptor status (Z17.~~ 0 ,~~ ~~Z17.1~~)

Revise — Z17 Estrogen, and other hormones and factors 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:
malignant neoplasm of breast (C50.-)
Add — 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
Add — Z17.21 Progesterone receptor positive status
PR+

New code
Add — Z17.22 Progesterone receptor negative status
PR-

New subcategory — Z17.3 Human epidermal growth factor 2 receptor

New code
Add — Z17.31 Human epidermal growth factor receptor 2 positive status
HER2+

New code
Add — Z17.32 Human epidermal growth factor receptor 2 negative status
HER2-

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

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.

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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

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]

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 and flank]
New code L02.227 Furuncle of flank

L02.23 Carbuncle of trunk
Revise L02.232 Carbuncle of back [any part, except buttock and flank]
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

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 Rl0.8A1 Right flank tenderness
New code Rl0.8A2 Left flank tenderness
New code R10.8A3 Suprapubic tenderness
New code R10.8A9 Flank tenderness, unspecified
Add Flank tenderness NOS

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

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

S30.85 Superficial foreign body of abdomen, lower back, pelvis, and external genitals

New code S30.85A Superficial foreign body of flank

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

New code S30.86A Insect bite (nonvenomous) of flank

S30.87 Other superficial bite of abdomen, lower back, pelvis, and external genitals

New code S30.87A Other superficial bite of flank

S30.9 Unspecified superficial injury of abdomen, lower back, pelvis, and external genitals

New code S30.9A Unspecified superficial injury of flank

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

New code	S31.157 Open bite of abdominal wall, left flank without penetration into peritoneal cavity
New code	S31.15A Open bite of abdominal wall, unspecified flank without penetration into peritoneal cavity
Add	Open bite of abdominal wall of flank NOS without penetration into peritoneal cavity
	S31.6 Open wound of abdominal wall with penetration into peritoneal cavity
	S31.60 Unspecified open wound of abdominal wall with penetration into peritoneal cavity
New code	S31.606 Unspecified open wound of abdominal wall, right flank with penetration into peritoneal cavity
New code	S31.607 Unspecified open wound of abdominal wall, left flank with penetration into peritoneal cavity
New code	S31.60A Unspecified open wound of abdominal wall, unspecified flank with penetration into peritoneal cavity
Add	Unspecified open wound of abdominal wall of flank NOS, with penetration into peritoneal cavity
	S31.61 Laceration without foreign body of abdominal wall with penetration into peritoneal cavity
New code	S31.616 Laceration without foreign body of abdominal wall, right flank with penetration into peritoneal cavity
New code	S31.617 Laceration without foreign body of abdominal wall, left flank with penetration into peritoneal cavity

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

New code S31.657 Open bite of abdominal wall, left flank with penetration into peritoneal cavity

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

Add Open bite of abdominal wall, flank NOS, with penetration into peritoneal cavity

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 repellant, 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.

TABULAR MODIFICATIONS

T75 Other and unspecified effects of other external causes

T75.8 Other specified effects of external causes

New sub-subcategory
T75.83 Effects of war theater
Add Use additional code to identify associated manifestations

New code
T75.830 Gulf war illness
Add Gulf war syndrome

New code
T75.838 Effects of other war theater

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

New Code
Z77.3 Contact with and (suspected) exposure to Gulf War theater
Add 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]

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.

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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-)

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 Association[3] and the American Association of Clinical Endocrinology[4] 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,

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.

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TABULAR MODIFICATIONS

E16 Other disorders of pancreatic internal secretion

E16.0 Drug-induced hypoglycemia without coma

Add — Use additional code for hypoglycemia level, if applicable (E16.A-)

E16.1 Other hypoglycemia

Add — Use additional code for hypoglycemia level, if applicable (E16.A-)

E16.2 Hypoglycemia, unspecified

Add — **Use additional code for hypoglycemia level, if applicable (E16.A-)**

New subcategory — E16.A Hypoglycemia level

New code — **E16.A1 Hypoglycemia level 1**
Add — **Decreased blood glucose level 1**

New code — **E16.A2 Hypoglycemia level 2**
Add — **Decreased blood glucose level 2**

New code — **E16.A3 Hypoglycemia level 3**
Add — **Decreased blood glucose level 3**

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-)

Injection Drug Use

Despite the devastating toll exacted by injection drug use (IDU) on morbidity and mortality[1-8], 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 specificity[9-12]. 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 SSPs[14]. 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 Association[15-19].

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 SSPs[20]. 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) infection[3-5]. 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.

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.

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TABULAR MODIFICATIONS

Z72 Problems related to lifestyle

Excludes2: Problems related to life-management difficulty (Z73 –
Problems related to socioeconomic and psychosocial circumstances (Z55 – Z65)

Z72.8 Other problems related to lifestyle

New sub-sub category	**Z72.83 Injection (non-prescribed) (illicit) drug use**
Add	**Code first: drug abuse (non-dependent) – see Alphabetic index: Abuse, drug** **drug dependence – see Alphabetic index: Dependence** **drug use (non-dependent) – see Alphabetic index: Use, drug** **poisoning – see Table of Drugs and Chemicals, by drug, poisoning**
Add	**Code also, if applicable, any associated condition, such as:** **acute bacterial endocarditis (I33.0)** **chronic hepatitis C virus infection (B18.8)** **human immunodeficiency virus infection (B20)**
Add	**Excludes2: Long term (current) drug therapy (Z79.-)**
New code	**Z72.830 Injection (non-prescribed) (illicit) drug use, active**
New code	**Z72.831 Injection (non-prescribed) (illicit) drug use, past**
New code	**Z72.839 Injection (non-prescribed) (illicit) drug use, unspecified**

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).

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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**

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

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

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

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

C81.4 Lymphocyte-rich Hodgkin lymphoma
Lymphocyte-rich classical Hodgkin lymphoma

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

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

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

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

C82 Follicular lymphoma
C82.0 Follicular lymphoma grade I

New code C82.0A Follicular lymphoma grade I, in remission
C82.1 Follicular lymphoma grade II

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

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

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

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

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

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

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

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

C83 Non-follicular lymphoma

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

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

C83.1 Mantle cell lymphoma
Centrocytic lymphoma

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

C83.3 Diffuse large B-cell lymphoma
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 C83.3A Diffuse large B-cell lymphoma, in remission
Add Anaplastic diffuse large B-cell lymphoma, in remission

Add		CD30-positive diffuse large B-cell lymphoma, in remission
Add		Centroblastic diffuse large B-cell lymphoma, in remission
Add		Diffuse large B-cell lymphoma, subtype not specified, in remission
Add		Immunoblastic diffuse large B-cell lymphoma, in remission
Add		Plasmablastic diffuse large B-cell lymphoma, in remission
Add		T-cell rich diffuse large B-cell lymphoma, in remission

C83.5 Lymphoblastic (diffuse) lymphoma
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

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

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

C85 Other specified and unspecified types of non-Hodgkin lymphoma
C85.1 Unspecified B-cell lymphoma

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

C85.2 Mediastinal (thymic) large B-cell lymphoma

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

C85.8 Other specified types of non-Hodgkin lymphoma

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

C85.9 Non-Hodgkin lymphoma, unspecified
Lymphoma NOS
Malignant lymphoma NOS
Non-Hodgkin lymphoma NOS

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

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

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

New code C86.01 Extranodal NK/T-cell lymphoma, nasal type, in remission

C86.1 Hepatosplenic T-cell lymphoma
Alpha-beta and gamma delta types

New code C86.10 Hepatosplenic T-cell lymphoma not having achieved remission
Add Hepatosplenic T-cell lymphoma NOS
Add Hepatosplenic T-cell lymphoma with failed remission

New code C86.11 Hepatosplenic T-cell lymphoma, in remission

C86.2 Enteropathy-type (intestinal) T-cell lymphoma
Enteropathy associated T-cell lymphoma

New code C86.20 Enteropathy-type (intestinal) T-cell lymphoma not having achieved remission
Add Enteropathy associated T-cell lymphoma 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

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

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

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

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.**8**1	Other malignant immunoproliferative diseases, in remission

C88.9 Malignant immunoproliferative disease, unspecified
Immunoproliferative disease NOS

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

Add Immunoproliferative disease NOS 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]

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]

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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

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 obstruction[2] 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 cartilage[5].

NVC may be further classified as either static or dynamic[6]. 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 life[7]. 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,

hypertrophied inferior turbinate, severely ptotic nasal tip, wide nasal columella, and/or a caudal septal deviation[8].

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

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TABULAR MODIFICATIONS

J34 Other and unspecified disorders of nose and nasal sinuses

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

New 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

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

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 ≥95th percentile as class 1 obesity, BMI ≥120% of the 95th percentile as class 2 obesity, and BMI ≥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.

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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 to less than 120% of the 95th percentile for age
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

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.

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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 colon~~ic~~ 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

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 effects[1]. 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 years[4].

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.

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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)

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.

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.

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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)

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.

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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)

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.

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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

TABULAR MODIFICATIONS PROPOSED ADDENDA

All approved modifications will be effective April 1, 2024

B04 Monkeypox

Add Mpox

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.-)

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.-)

Use additional code, if applicable, to identify:
acquired absence of pancreas (Z90.41-)

Delete — ~~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)~~

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.-)~~

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)~~ (J44.89)
Add other specified chronic obstructive pulmonary disease (J44.89)

J45 Asthma
Revise Excludes2: asthma with chronic obstructive pulmonary disease ~~(J44.9)~~ (J44.89)
Revise chronic asthmatic (obstructive) bronchitis ~~(J44.9)~~ (J44.89)
Revise chronic obstructive asthma ~~(J44.9)~~ (J44.89)
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-)

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 hyposecretion (K11.7)~~

Add Excludes2: salivary gland hyposecretion (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-)

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

TABULAR MODIFICATIONS PROPOSED ADDENDA

All approved modifications will be effective October 1, 2024

A18 Tuberculosis of other organs
A18.4 Tuberculosis of skin and subcutaneous tissue
Revise Lupus ~~excedens~~ exedens

A50 Congenital syphilis
A50.4 Late congenital neurosyphilis [juvenile neurosyphilis]
A50.45 Juvenile general paresis
Dementia paralytica juvenilis
Revise Juvenile ~~tabetoparetic~~ taboparetic neurosyphilis

A77 Spotted fever [tick-borne rickettsioses]
A77.4 Ehrlichiosis
Revise A77.41 Ehrlichiosis ~~chafeensis~~ chaffeensis [E. ~~chafeensis~~ chaffeensis]

E07 Other disorders of thyroid
E07.1 Dyshormogenetic goiter
Add Dyshormonogenetic goiter
Add Familial dyshormonogenetic goiter

E08 Diabetes mellitus due to underlying condition

Use additional code to identify control using:
Add injectable non-insulin antidiabetic drugs (Z79.85)

E09 Drug or chemical induced diabetes mellitus

Use additional code to identify control using:
Add injectable non-insulin antidiabetic drugs (Z79.85)

E11 Type 2 diabetes mellitus

Use additional code to identify control using:
Add injectable non-insulin antidiabetic drugs (Z79.85)

E13 Other specified diabetes mellitus
Use additional code to identify control using:
Add injectable non-insulin antidiabetic drugs (Z79.85)

F01 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
Delete ~~Code first the underlying physiological condition or sequelae of cerebrovascular disease.~~
Add Code first, if applicable, any causal condition

F02 Dementia in other diseases classified elsewhere

Code first the underlying physiological condition, such as:
Delete ~~dementia with Lewy bodies~~
Delete ~~dementia with Parkinsonism~~
Add neurocognitive disorder with Lewy bodies (G31.83)
Add other frontotemporal neurocognitive disorder (G31.90)

F03 Unspecified dementia
Delete ~~Excludes1: senility NOS (R41.81)~~
Excludes2: mild memory disturbance due to known physiological condition (F06.8)
Revise ~~senile~~ dementia with delirium or acute confusional state (F05)

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

F06 Other mental disorders due to known physiological condition
Revise Excludes1: unspecified dementia ~~(F03)~~ (F03.-)

F06.7 Mild neurocognitive disorder due to known physiological condition
Code first the underlying physiological condition, such as:
Alzheimer's disease (G30.-)
Revise other frontotemporal neurocognitive disorder (G31.09)
Revise Excludes1: dementia (F01.-, F02.-, ~~F03)~~ F03.-)

F20 Schizophrenia
Add Use additional code, if applicable, to identify:
other specified cognitive deficit (R41.84-)

F45 Somatoform disorders
F45.2 Hypochondriacal disorders
F45.22 Body dysmorphic disorder
Add Bigorexia
Add Muscle dysmorphia

H42 Glaucoma in diseases classified elsewhere
Code first underlying condition, such as:
Revise ~~Reiger's~~ Rieger anomaly (Q13.81)

G21 Secondary parkinsonism
Revise Excludes1: ~~dementia with Parkinsonism~~ neurocognitive disorder with Lewy bodies (G31.83)

G30 Alzheimer's disease
Revise Excludes1: senile dementia NOS ~~(F03)~~ (F03.-)

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
Revise Excludes1: dementia (F01.-, F02.-, ~~F03)~~ F03.-)

G93 Other disorders of brain
G93.4 Other and unspecified encephalopathy
Revise G93.42~~Megaloencephalic~~ Megalencephalic leukoencephalopathy with subcortical cysts

H44 Disorders of globe
H44.2 Degenerative myopia
H44.2A Degenerative myopia with choroidal neovascularization
Revise H44.2A3 Degenerative myopia with choroidal neovascularization, bilateral ~~eye~~

H44.2B Degenerative myopia with macular hole
Revise H44.2B3 Degenerative myopia with macular hole, bilateral ~~eye~~

H44.2C Degenerative myopia with retinal detachment
Revise H44.2C3 Degenerative myopia with retinal detachment, bilateral ~~eye~~

H44.2D Degenerative myopia with foveoschisis
Revise H44.2D3 Degenerative myopia with foveoschisis, bilateral ~~eye~~

H44.2E Degenerative myopia with other maculopathy
Revise H44.2E3 Degenerative myopia with other maculopathy, bilateral ~~eye~~

I21 Acute myocardial infarction
I21.A Other type of myocardial infarction
I21.A1 Myocardial infarction type 2
Revise Code first, if applicable, the underlying cause, such as:

I30 Acute pericarditis

Excludes1: Dressler's syndrome (I24.1)
rheumatic pericarditis (acute) (I01.0)
Revise viral pericarditis due to ~~Coxsakie~~ Coxsackie virus (B33.23)

J13 Pneumonia due to Streptococcus pneumoniae
Revise Code first, if applicable, associated influenza~~, if applicable~~ (J09.X1, J10.0-, J11.0-)
Revise Code also ~~associated abscess~~, if applicable, ~~(J85.1)~~ any associated condition such as:
Add abscess (J85.1)
Add aspiration pneumonia (J69.-)

J14 Pneumonia due to Hemophilus influenzae

Revise Code first, if applicable, associated influenza~~, if applicable~~ (J09.X1, J10.0-, J11.0-)
Revise Code also ~~associated abscess~~, if applicable, ~~(J85.1)~~ any associated condition such as:
Add abscess (J85.1)
Add aspiration pneumonia (J69.-)

J15 Bacterial pneumonia, not elsewhere classified
Revise Code first, if applicable, associated influenza~~, if applicable~~ (J09.X1, J10.0-, J11.0-)
Revise Code also ~~associated abscess~~, if applicable, ~~(J85.1)~~ any associated condition such as:
Add abscess (J85.1)
Add aspiration pneumonia (J69.-)

J16 Pneumonia due to other infectious organisms, not elsewhere classified
Revise Code first, if applicable, associated influenza~~, if applicable~~ (J09.X1, J10.0-, J11.0-)
Revise Code also ~~associated abscess~~, if applicable, ~~(J85.1)~~ any associated condition such as:
Add abscess (J85.1)
Add aspiration pneumonia (J69.-)

J17 Pneumonia in diseases classified elsewhere
Add Code also, if applicable, any associated condition such as:
Add abscess (J85.1)
Add aspiration pneumonia (J69.-)

J18 Pneumonia, unspecified organism
Revise Code first, if applicable, associated influenza~~, if applicable~~ (J09.X1, J10.0-, J11.0-)
Add Code also, if applicable, any associated condition such as:
Add aspiration pneumonia (J69.-)
Delete Excludes1: ~~abscess of lung with pneumonia (J85.1)~~
Delete ~~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 ~~lipid pneumonia (J69.1)~~
Delete ~~pneumonitis due to external agents (J67-J70)~~

Add Excludes2: abscess of lung with pneumonia (J85.1)
Add 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 lipid pneumonia (J69.1)
Add pneumonitis due to external agents (J67-J70)

J69 Pneumonitis due to solids and liquids
Add Codes also, if applicable, other types of pneumonias

J84 Other interstitial pulmonary diseases
Add Code also, if applicable, associated condition

K14 Diseases of tongue
Revise Excludes2:~~leukedema~~ leukoedema of tongue (K13.29)

K58 Irritable bowel syndrome

Revise K58.9 Irritable bowel syndrome, unspecified ~~without diarrhea~~
Irritable bowel syndrome NOS

M92 Other juvenile osteochondrosis

M92.8 Other specified juvenile osteochondrosis

Delete ~~Calcaneal apophysitis~~

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:

Add pyonephrosis (N13.6)

O24 Diabetes mellitus in pregnancy, childbirth, and the puerperium

Add Use additional code, if applicable (for):

Add injectable non-insulin antidiabetic drugs (Z79.85)

Add long-term (current) use of insulin (Z79.4)

O24.1 Pre-existing type 2 diabetes mellitus, in pregnancy, childbirth and the puerperium

Revise Use additional code ~~(for):~~ from category E11 to further identify any manifestations

Delete ~~long-term (current) use of insulin (Z79.4)~~

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

Add injectable non-insulin antidiabetic drugs (Z79.85)

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

Add injectable non-insulin antidiabetic drugs (Z79.85)
long-term (current) use of insulin (Z79.4)

O24.9 Unspecified diabetes mellitus in pregnancy, childbirth and the puerperium

Delete ~~Use additional code for long-term (current) use of insulin (Z79.4)~~

Use additional code (for):

Add injectable non-insulin antidiabetic drugs (Z79.85)
long-term (current) use of insulin (Z79.4)

P72 Other transitory neonatal endocrine disorders
Excludes1: congenital hypothyroidism with or without goiter (E03.0-E03.1)
Add dyshormonogenetic goiter (E07.1)

Q13 Congenital malformations of anterior segment of eye
Q13.8 Other congenital malformations of anterior segment of eye
Revise Q13.81 ~~Rieger's~~ Rieger anomaly
Add Axenfeld-Rieger syndrome
Add Code also any other associated congenital malformations such as cardiac defects

R41 Other symptoms and signs involving cognitive functions and awareness
R41.0 Disorientation, unspecified
Add Excludes1: delirium due to known physiological condition (F05)

R41.8 Other symptoms and signs involving cognitive functions and awareness
R41.84 Other specified cognitive deficit
Add Code first the underlying condition, if known, such as: schizophrenia (F20.-)

R54 Age-related physical debility
Revise Excludes1: senile psychosis ~~(F03)~~ (F03.-)

S06 Intracranial injury
S06.3 Focal traumatic brain injury
S06.34 Traumatic hemorrhage of right cerebrum
S06.341 Traumatic hemorrhage of right cerebrum with loss of consciousness of 30 minutes or less
Revise Traumatic hemorrhage of right cerebrum with brief loss of consciousness

T63 Toxic effect of contact with venomous animals and plants

T63.4 Toxic effect of venom of other arthropods
Add Use additional code, if applicable, for anaphylactic shock (T78.2)

T65 Toxic effect of other and unspecified substances
T65.3 Toxic effect of nitroderivatives and aminoderivatives of benzene and its homologues
Revise Toxic effect of ~~anilin~~ aniline [benzenamine]

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 ~~aide~~ aid (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 ~~omycophenolate~~ mycophenolate

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
Add — Code first, if applicable, adverse effect of underdosing (T36-T50)

Z93 Artificial opening status
Z93.2 Ileostomy status
Add — Ileal pouch status
Add — Kock pouch status

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) (cirsoid) (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

Revise Bacteriuria, bacteruria (asymptomatic) R82.71
Delete ~~- asymptomatic R82.71~~

Revise Bee sting (with allergic or anaphylactic shock) ~~-see Toxicity, venom, arthropod, 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

Revise Brion-Kayser disease -see Fever, ~~parathyroid~~ paratyphoid

Bursitis M71.9
Revise - ankle -see Enthesopathy~~, lower limb~~, ankle~~, specified type NEC~~ and tarsus

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 ~~Cotungo's~~ Cotugno disease -see Sciatica

Revise ~~Dacryocystoblenorrhea~~ Dacryocystoblennorrhea -see Inflammation, lacrimal, passages, chronic

Deficiency, deficient
Revise - ~~phosphomannomutuse~~ 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

- 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 - - - - ~~proprionate~~ propionate NEC E71.128
Revise - - - - ~~proprionic~~ 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 Ducreyi's chancre A57

Add Dysmorphia
Add - muscle F45.22

Ehrlichiosis A77.40
- due to
Revise - - E. ~~chafeensis~~ chaffeensis A77.41

	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	- ~~blenorrhagic~~ 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
Add	- - - 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
Add	- - Clostridium difficile
Revise	- - - not specified as recurrent A04.72
Revise	- - - recurrent A04.71
	- necrotizing K55.30
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

Fever (inanition) (of unknown origin) (persistent) (with chills) (with rigor) R50.9
- spotted A77.9
- - Ehrlichiosis A77.40
- - - due to
Revise - - - - E. ~~chafeensis~~ chaffeensis A77.41

Fitzhugh-Curtis syndrome
- due to
Revise - - Neisseria ~~gonorrhoea~~ gonorrhea (gonococcal peritonitis) A54.85

Fracture, traumatic (abduction) (adduction) (separation) -see also Fracture, pathological T14.8
- stress M84.30
Revise - - hip ~~M84.359~~ M84.35-

Hemoperitoneum K66.1
- traumatic S36.899
Revise - - with open wound -see Wound, open, abdominal, wall, by site if known, with penetration into peritoneal cavity

Hemorrhage, hemorrhagic (concealed) R58
- intracranial (nontraumatic) I62.9
- - intracerebral (nontraumatic) (in) I61.9
Revise - - - traumatic ~~(diffuse) -~~ see Injury, intracranial, ~~diffuse~~ intracerebral hemorrhage, traumatic
Delete ~~- - - - focal - see Injury, intracranial, focal~~

Hepatitis K75.9
Add - A – see Hepatitis, viral, type, A

- acute B17.9
Revise - - viral (see also, Hepatitis, viral) B17.9

- B B19.10
Revise - - with ~~hepatic coma B19.11~~
Add - - - delta (agent) – see Hepatitis, D
Add - - - hepatic coma B19.11

- 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 - - ~~myleitis~~ myelitis B00.82

History
- - malignant neoplasm (of) Z85.9
Revise - - - carcinoid -see History, personal (of), malignant neoplasm, by site, ~~carcinioid~~ 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 ~~Hypsarhythmia~~ Hypsarrhythmia -see Epilepsy, generalized, specified NEC

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) ~~(callinarum)~~ (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-~~Steid~~a Stieda disease or syndrome (calcification, knee joint) – see Bursitis, tibial collateral

Revise Larsen-Johansson disease or osteochondrosis -see Osteochondrosis, juvenile, patella

Leak, leakage
- device, implant or graft -see also Complications, by site and type, mechanical
- - genital NEC T83.498

Revise - - - penile prosthesis (cylinder) (implanted) (pump) ~~(resevoir)~~ (reservoir) T83.490

Revise Loa loa~~, loaiasis, loasis~~ (loiasis) B74.3

Long-term (current) (prophylactic) drug therapy (use of)

Revise - non-insulin antidiabetic drug, injectable ~~Z79.899~~ Z79.85

Revise - ~~o~~mycophenolate 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

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) ~~(resevoir)~~ (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) ~~(resevoir)~~ (reservoir) T83.490

Revise ~~Omenotocele~~ Omentocele -see Hernia, abdomen, specified site NEC

Osteonecrosis M87.9
- secondary NEC M87.30
- - due to

- - - hemoglobinopathy NEC D58.2 [M90.50]
Revise - - - - multiple sites D58.2 ~~[M90.58]~~ [M90.59]
Add - - - - 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) ~~(resevoir)~~ (reservoir) T83.490

Retinopathy (background) H35.00
- proliferative NEC H35.2-
Revise - - thaslassemia thalassemia H35.2

Revise Robinow-~~Silvermann~~ 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

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) ~~S23.420~~ S43.6-

Status (post) -see also Presence (of)
- hysterectomy (complete) (total) Z90.710
Revise - - partial (with remaining ~~cervial~~ 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 ~~gonorrhorea~~ gonorrhea (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]

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, intracranial, venous sinus

Revise - cavernous (venous) sinus -see Thrombosis, intracranial, venous sinus

Revise - cerebrovenous sinus -see also Thrombosis, intracranial, venous sinus

- intracranial (arterial) I66.9
- - venous sinus (any) G08
Add - - - nonpyogenic I67.6

Revise - lateral (venous) sinus -see Thrombosis, intracranial, venous sinus

Revise - longitudinal (venous) sinus -see Thrombosis, intracranial, venous sinus

Revise - sigmoid (venous) sinus -see Thrombosis, intracranial, venous sinus

Revise - sinus, intracranial (any) -see Thrombosis, intracranial, venous sinus

Revise Tuberosity, ~~enitre~~ 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 ~~C16.9~~ C49.A2

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-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 - - ~~hangun~~ handgun W32.1

Revise - handgun (parts) -see Explosion, firearm, ~~hangun~~ 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 - ~~firearn~~ firearm NEC W34.19

Revise Rape (attempted) (confirmed) T74.2-
Add - suspected T76.2-

Recoil

Revise - ~~firearn~~ 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~~ T43.581	~~T43.592~~ T43.582	~~T43.593~~ T43.583	~~T43.594~~ T43.584	~~T43.595~~ T43.585	~~T43.596~~ T43.586
Add - antiallergic	T45.0X1	T45.0X2	T45.0X3	T45.0X4	T45.0X5	T45.0X6

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						