CENTERS FOR DISEASE CONTROL AND PREVENTION

ICD-10 Coordination and Maintenance Committee Meeting
September 8-9, 2020
Diagnosis Agenda

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

Diagnosis Topics:

Contents

ICD-10 TIMELINE

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

September 8-9, 2020	The September 2020 ICD-10 Coordination and Maintenance Committee Meeting will be held fully virtual, with no in-person audience. Those who wish to attend must participate via Zoom Webinar or by dialing in.
September 2020	Recordings and slide presentations of the September 8-9, 2020 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, 2020	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/icd10cm.htm **Procedure addendum –** https://www.cms.gov/Medicare/Coding/ICD10/
October 9, 2020	**Deadline for receipt of public comments on proposed new codes discussed at the September 8-9, 2020 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on April 1, 2021.**
November 2020	Any new ICD-10 codes required to capture new technology that will be implemented on the following April 1 will be announced. Information on any new codes to be implemented April 1, 2021 will be posted on the following websites: https://www.cdc.gov/nchs/icd/icd10cm.htm https://www.cms.gov/Medicare/Coding/ICD10/

November 9, 2020	**Deadline for receipt of public comments on proposed new codes and revisions discussed at the September 8-9, 2020 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2021.**
December 4, 2020	**Deadline for requestors: Those members of the public requesting that topics be discussed at the March 9-10, 2021 ICD-10 Coordination and Maintenance Committee Meeting must have their requests submitted to CMS for procedures and to NCHS for diagnoses by this date.**
January 2021	Federal Register notice of March 9-10, 2021 ICD-10 Coordination and Maintenance Committee Meeting will be published.
February 2021	Tentative agenda for the Procedure portion of the March 9, 2021 ICD-10 Coordination and Maintenance Committee Meeting posted on CMS webpage as follows: https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html Tentative agenda for the Diagnosis portion of the March 10, 2021 ICD-10 Coordination and Maintenance Committee Meeting posted on NCHS homepage as follows: https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm
February 1, 2021	**On-line registration opens for the March 9-10, 2021 ICD-10 Coordination and Maintenance Committee Meeting at:** https://www.cms.gov/apps/events/default.asp. Please note that this meeting will be conducted virtually and registration is not required to attend. However, we are providing the ability to register on-line for those required to provide proof of attendance for continuing education purposes. The on-line registration will be available through March 1, 2021.
March 9-10, 2021	ICD-10 Coordination and Maintenance Committee Meeting.
March 2021	Recordings and slide presentations of the March 9-10, 2021 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, 2021	Any new ICD-10 codes to capture new diseases or technology will be implemented on April 1, 2021.
April 9, 2021	**Deadline for receipt of public comments on proposed new codes and revisions discussed at the March 9-10, 2021 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2021**.
April 2021	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 2022 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/index.html?redirect=/AcuteInpatientPPS/IPPS/list.asp
May/June 2021	Final addendum posted on web pages as follows: **Diagnosis addendum -** https://www.cdc.gov/nchs/icd/icd10cm.htm **Procedure addendum -** https://www.cms.gov/Medicare/Coding/ICD10/index.html
June 11, 2021	**Deadline for requestors: Those members of the public requesting that topics be discussed at the September 14-15, 2021 ICD-10 Coordination and Maintenance Committee Meeting, must have their requests submitted to CMS for procedures and NCHS for diagnoses.**
July 2021	Federal Register notice for the September 14-15, 2021 ICD-10 Coordination and Maintenance Committee Meeting will be published. This will include the tentative agenda.
August 1, 2021	Hospital Inpatient Prospective Payment System final rule 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, 2021. This rule can be accessed at: https://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/AcuteInpatientPPS/index.html
August 2021	Tentative agenda for the Procedure portion of the September 14, 2021 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 15, 2021 ICD-10 Coordination and Maintenance Committee Meeting will be posted on the NCHS webpage at - https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm
August 9, 2021	**On-line registration opens for the September 14-15, 2021 ICD-10 Coordination and Maintenance Committee Meeting at:** https://www.cms.gov/apps/events/default.asp. Please note that this meeting will be conducted virtually and registration is not required to attend. However, we are providing the ability to register on-line for those required to provide proof of attendance for continuing education purposes. The on-line registration will be available through September 9, 2021.
September 14-15, 2021	The September 2021 ICD-10 Coordination and Maintenance Committee Meeting will be held fully virtual, with no in-person audience. Those who wish to attend must participate via Zoom Webinar or by dialing in.
September 2021	Recordings and slide presentations of the September 14-15, 2021 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, 2021	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/icd10cm.htm **Procedure addendum –** https://www.cms.gov/Medicare/Coding/ICD10/
October 15, 2021	**Deadline for receipt of public comments on proposed new codes discussed at the September 14-15, 2021 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on April 1, 2022.**

November 2021	Any new ICD-10 codes required to capture new technology that will be implemented on the following April 1 will be announced. Information on any new codes to be implemented April 1, 2022 will be posted on the following websites: https://www.cdc.gov/nchs/icd/icd10cm.htm https://www.cms.gov/Medicare/Coding/ICD10/
November 15, 2021	**Deadline for receipt of public comments on proposed new codes and revisions discussed at the September 14-15, 2021 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2022.**

Contact Information

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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 Findings of Blood Amino-Acid Level

Currently, the ICD-10-CM classifies homocysteinemia and homocystinuria to the same code E72.11. The subcategory is E72.1, Disorders of sulfur-bearing amino-acid metabolism. The clinical knowledge of homocysteinemia has evolved and is clinically distinct from homocystinuria. NCHS received a proposal to create a unique code for abnormal findings of blood amino-acid level.

Homocystinuria is a term generally used to refer to an inborn error of metabolism, such as homocystinuria due to cystathionine beta-synthase deficiency and homocystinuria due to methylenetetrahydrofolate reductase deficiency. These disorders typically manifest with certain phenotypes and represent disease states.

Homocysteinemia (also referred to as "hyperhomocysteinemia") is generally used in the clinical world to refer to a state in which homocysteine has been measured in the blood and found to be above reference range. Thus, use of the term "homocysteinemia" in the clinical setting does not convey the same meaning as the term "homocystinuria."

While a finding of homocysteinemia typically occurs in the two disorders mentioned above (homocystinuria due to cystathionine beta-synthase deficiency and/or methylenetetrahydrofolate reductase deficiency), it is not limited to these two disorders. Homocysteinemia may also be a result of defects of vitamin B12 metabolism, associated with nutritional and other environmental factors.

Further clinical investigation is required to determine the underlying factors creating the abnormal blood levels of homocysteinemia. Over the last several decades, the abnormal finding of homocysteinemia has become strongly associated with higher than normal cardiovascular risk in patients without the neurologic, muscular, and other phenotypic findings of homocystinuria.

TABULAR MODIFICATIONS

	R79	Other abnormal findings of blood chemistry
	R79.8	Other specified abnormal findings of blood chemistry
	R79.81	Abnormal blood-gas level
	R79.82	Elevated C-reactive protein (CRP)
New code	R79.83	Abnormal findings of blood amino-acid level
Add		Homocysteinemia
Add		Excludes1: specific findings indicate disorder of amino-acid metabolism (E70-E72)
	R79.89	Other specified abnormal findings of blood chemistry

Acute Flaccid Myelitis

Acute flaccid myelitis (AFM) is a rare but serious neurologic condition. It affects the nervous system and is characterized by selective inflammation of the central gray matter of the spinal cord.

This condition causes the muscles and reflexes in the body to become weak. The sudden rapid onset of flaccid paralysis in a single limb or in multiple limbs occurs within hours to days. Sudden arm or leg weakness, loss of muscle tone, and loss of reflexes are the most common symptoms. Neurologic symptoms may be preceded by a febrile respiratory illness and sometimes pain in the extremities.

The most severe symptom of AFM is respiratory failure. This happens when the muscles involved with breathing become weak. Respiratory failure in AFM patients can result in the need for long-term mechanical ventilation.

Cranial nerve involvement may result in facial weakness, restricted eye movements, difficulty swallowing, difficulty with speech, and difficulty breathing. Most typically, there are no sensory or cognitive changes. Bowel and bladder dysfunction can occur. Severity of the disease can range from mild weakness in one limb to complete paralysis and inability to breathe. Some patients may be unable to swallow, support their own head, and may require feeding tubes and long-term ventilation. Prognosis is variable, often with some functional improvements, but most with the disease are left with persistent paralysis.

Most AFM cases (more than 90%) have been in young children with a median age of 5–9 years, depending on year. Since AFM affects predominantly children, AFM can lead to long-term disability over a lifetime. In severe cases a patient is rendered completely limp (flaccid) and paralyzed in most or all of the body. This requires the use of assistive devices for mobility (wheelchair, walker, braces, etc.). Likelihood of full recovery is minimal, even with intensive therapy, and most are left with residual disability. Seeking necessary rehabilitative resources without a specific diagnosis code that accurately depicts the profound impact and challenging prognosis creates additional hardship for families.

Sometimes AFM may be referred to as a “polio-like” condition in that it is a disease with features similar to poliomyelitis caused by poliovirus, but all the specimens from AFM patients tested negative for poliovirus by the CDC. The cases of AFM since 2014 are not caused by poliovirus.

The CDC has tracked Acute Flaccid Myelitis since 2014.

- As of July 31, 2020, there have been 16 confirmed cases in 2020. To date, there have been 38 reports of patients under investigation (PUIs) for AFM in 2020. Two patients with confirmed AFM died in the acute phase of their illness, one in 2017 and one in 2020. CDC has also learned of deaths in cases confirmed in previous years.

- There were 46 confirmed cases in 2019 out of 142 PUIs. CDC, state, and local health departments are still investigating some of the PUIs.

- There have been 633 confirmed cases since CDC began tracking AFM in August of 2014. CDC has been thoroughly investigating cases since that time. CDC has seen increases in AFM cases, mostly in young children, every two years.

Literature on AFM has been published since outbreaks of AFM in 2012 and 2014, though sporadic case reports date back earlier. Since this time the numbers tracked by CDC and WHO have grown with increasing biennial spikes in the summer to fall on even years, particularly in the United States.

The request for a unique and distinct code is to address disease specificity that does not already exist in current ICD-10-CM codes and sub-terms that are currently being used for this condition. A unique code for AFM will help to better support disease tracking, clinical care responses and associated mortality rates.

Current ICD-10-CM coding guidance refers to the index under the main term of Myelitis. Although the main term "Myelitis" has a non-essential modifier of acute, it does not include anything related to "flaccid". Therefore, the current default code suggested for AFM is G04.89 "Other myelitis". Utilization of other or more generic codes undermines disease specificity and decreases the opportunity to use ICD-10-CM codes for AFM surveillance.

A specific code for AFM, or an additional sub-term for "acute flaccid", combined with these symptoms would provide greater clinical context for documentation and case tracking. Research is continuing, and more is being learned about AFM every year thanks to CDC, researchers, and physicians who specialize in this disease. A specific ICD-10-CM code for AFM will also allow for standardized comparisons of cases throughout the United States and the world.

This proposal is jointly submitted by Dr. Kevin Messacar, MD, Associate Professor of Pediatrics Sections of Hospital Medicine and Pediatric Infectious Diseases, University of Colorado Children's Hospital Colorado and Ms. Robin Roberts, AFMAnow.org (Parent Advocacy Group).

This proposal has been reviewed and supported by the American Academy of Pediatrics, the American Neurology Association, and the AFM Team at the Centers for Disease Control, Division of Viral Diseases, National Center for Immunizations and Respiratory Diseases.

Resources:

Center for Disease Control: https://www.cdc.gov/acute-flaccid-myelitis/index.html

https://www.cdc.gov/ddid/bsc/afm/update-may-2019.html

https://wwwn.cdc.gov/nndss/conditions/acute-flaccid-myelitis/

Council for State and Territorial Epidemiology: https://cdn.ymaws.com/www.cste.org/resource/resmgr/2019ps/final/19-ID-05_AFM_final_7.31.19.pdf
Johns Hopkins Research Study: https://medicalxpress.com/news/2019-07-effort-acute-flaccid-myelitis.html

NIH Study: https://www.mdedge.com/pediatrics/article/205309/pediatrics/nih-launches-5-year-10-million-study-acute-flaccid-myelitis

Literature:
A Clinical Review of US Cases 2012–2015 https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5098271/

Association of Enterovirus D68 with Acute Flaccid Myelitis, Philadelphia, Pennsylvania, USA, 2009–2018 https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6711208/?report=classic

Clinical Differentiation https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6694036/?report=classic

AFM and The Evidence for Casualty https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6778404/

Increase in Acute Flaccid Myelitis https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6290805/

American Academy of Pediatrics Vigilance for acute flaccid myelitis https://www.aappublications.org/news/2019/07/09/afm070919

TABULAR MODIFICATIONS

A80 Acute poliomyelitis
Add Excludes1: acute flaccid myelitis (G04.82)

G04 Encephalitis, myelitis and encephalomyelitis

G04.8 Other encephalitis, myelitis and encephalomyelitis
Code also any associated seizure (G40.-, R56.9)

G04.81 Other encephalitis and encephalomyelitis
Noninfectious acute disseminated encephalomyelitis (noninfectious ADEM)

New Code G04.82 Acute flaccid myelitis
Add Code also, if known, other manifestations such as:
Add dysphagia (R13.10)
Add facial weakness (R29.810)
Add muscle weakness (R62.81)
Add Excludes1: transverse myelitis (G37.3)

G37 Other demyelinating diseases of central nervous system

G37.3 Acute transverse myelitis in demyelinating disease of central nervous system
Acute transverse myelitis NOS
Acute transverse myelopathy

Add Excludes1: acute flaccid myelitis (G04.82)
multiple sclerosis (G35
neuromyelitis optica [Devic] (G36.0)

Allergy to Mammalian Meats

Alpha-gal syndrome of late has been associated with allergic reactions to red meat and other products made from mammals. In the United States, the condition most often occurs from Lone Star or Deer Tick bites. The bite transmits a sugar molecule called alpha-gal into the person's body.[1] Alpha-gal is a carbohydrate found in all mammals but not in humans.[2] In some people, this triggers an immune system reaction that later produces mild to severe allergic reactions to red meat.[2] Examples of meats that have alpha-gal are beef, pork, lamb, venison, rabbit, goat, buffalo, bison, etc.[2] Alpha-gal is also present in the milk from these animal sources as well as their organ meats. Chicken, turkey, fish, and shellfish do not have alpha-gal and are safe for patients with Alpha-gal Syndrome (AGS) allergy to consume.[2]

The galactose-α-1,3-galactose (α-Gal) syndrome has many novel features that are relevant to diagnosis and management. In most cases, the diagnosis can be made on a history of delayed allergic reactions to mammalian meat and the blood test for IgE to the oligosaccharide α-Gal. In general, the diagnosis also dictates the primary treatment, that is, avoiding mammalian meat and dairy in some cases.[3] Blood levels of IgE to α-Gal often drop in patients who avoid recurrent tick bites, but the rate of decline is variable. Similarly, the delay before reactions is variable and the severity of the allergic reactions is not predicted by the delay or the titer of specific IgE.[3]

Some mammalian-derived products such as heart valves, gelatin-based plasma expanders, and pancreatic enzymes are relevant to only select patient groups. The minority of cases will prevent allergic reactions by avoiding a wide range of products that are prepared with mammalian-derived constituents, such as gelatin.[2]

Reasons and mechanisms for the delayed onset of food-related anaphylaxis and the preponderance of abdominal reactions are not clear, but could involve the kinetics of allergen digestion and processing or immunologic presentation via a different mechanism from usual immediate-type food allergy.[4]

Symptoms of allergic reactions to mammalian meats vary from person to person.[5]
Alpha-gal has a delayed reaction time while other foods (peanuts or shellfish) cause an allergic reaction within minutes of exposure. The alpha-gal syndrome reactions usually occur around three to six hours after exposure.[1]

Signs and symptoms of alpha-gal syndrome may include: hives, itching, eczema, swelling of the lips, face, tongue and throat, or other body parts, wheezing or shortness of breath, a runny nose, stomach pain, diarrhea, nausea or vomiting, sneezing, headaches, and a severe, potentially deadly allergic reaction that restricts breathing (anaphylaxis).[1]

Tickborne diseases increasingly threaten the health of people in the United States. The growing threat includes newly discovered disease-causing germs, an increasing number of reported tickborne illnesses, and expanding geographic ranges for ticks.[6] In 2018, 5,000 cases of Alpha-gal syndrome (AGS) had been diagnosed in the United States, and many more probably remain undiagnosed. In some tick-heavy regions, the prevalence of meat allergy is estimated to be at least 1 percent of the population.[7] The Lone Star tick is found predominantly in the southeastern United States, and most cases of alpha-gal syndrome occur in this region. The tick can also be found in the eastern and south-central United

States. The condition appears to be spreading farther north and west, however, as deer carry the Lone Star tick to new parts of the United States.[1]

Currently there is no ICD-10-CM code available to code and track food allergy to red meats. With the cumulative number of cases with the lifelong significant allergic reactions affecting people, a proposal was submitted for consideration for a new ICD-10-CM code for allergy to mammalian meats for coding and tracking the disease process. The following tabular modifications are being proposed:

References

[1] Mayo Clinic. Alpha-Gal Syndrome. mayoclinic.org. https://www.mayoclinic.org/diseases-conditions/alpha-gal-syndrome/symptoms-causes/syc-20428608. 11Aug. 2020

[2]Health and Human Services. Alpha-Gal Syndrome Subcommittee Report to the Tick-Borne Disease Working Group. HHS.gov. https://www.hhs.gov/ash/advisory-committees/tickbornedisease/reports/alpha-gal-subcomm-2020/index.html. 11 Aug. 2020

[3]The Journal of Allergy and Clinical Immunology, In Practice. Diagnosis and Management of Patients with the a-Gal Syndrome. jaci-inpractice.org. https://www.jaci-inpractice.org/article/S2213-2198(19)30795-0/abstract. 11 Aug. 2020

[4]Michael, Danijela, Tilo, Commins, Scott, Onyinye, Thomas, Eleonora, Marianne van, Wilson, Jeffrey. Galactose α-1,3-galactose phenotypes: Lessons from various patient populations. 26 Mar. 2019. https://pubmed.ncbi.nlm.nih.gov/30922956/ . 11 Aug. 2020

[5]American College of Allergy, Asthma, & Immunology. Meat Allergy. acaai.org. https://acaai.org/allergies/types/food-allergies/types-food-allergy/meat-allergy. 11 Aug. 2020

[6]Centers for Disrease Control and Prevention. Lyme and Other Tickborne Diseases Increasing. https://www.cdc.gov/media/dpk/diseases-and-conditions/lyme-disease/index.html. 13 Aug. 2020

[7]Velasqueez-Manoff. What the Myster of the Tick-Borne Meat Allergy Could Reveal. 24 July 2018. https://www.nytimes.com/2018/07/24/magazine/what-the-mystery-of-the-tick-borne-meat-allergy-could-reveal.html. 13 Aug. 2020

TABULAR MODICATIONS

Z91 Personal risk factors, not elsewhere classified

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

Z91.01Food allergy status

New code	Z91.014	Allergy to mammalian meats
Add		Allergy to red meats
Add		Allergy to beef
Add		Allergy to pork
Add		Allergy to lamb

Anaplastic Large Cell Lymphoma, ALK-negative of the breast

The American Society of Plastic Surgeons (ASPS) submitted this request seeking the creation of a unique code for Anaplastic large cell lymphoma, ALK-negative, of the breast, commonly called Breast Implant Associated Anaplastic Large Cell Lymphoma (BIA-ALCL). This was previously presented, as part of the addenda, at the March 2020 Coordination and Maintenance (C&M) meeting as an inclusion term at C84.7, Anaplastic large cell lymphoma, ALK-negative. ASPS feel that a unique code of the location is a better choice.

BIA-ALCL is not a disease of the lymph nodes nor the breast tissue but is instead a disease of the capsule surrounding a breast implant. It typically presents with swelling of the breast. Pathologically, BIA-ALCL is distinct from systemic ALCL and is identified as unique. It is considered an emerging disease by the World Health Organization (WHO), National Comprehensive Cancer Network (NCCN), and National Cancer Institute (NCI). It requires a multidisciplinary team approach for patient treatment. Statistical information captured via a unique ICD-10-CM code will help promote data collection, analysis and improve public health surveillance of BIA-ALCL in the United States.

The ASPS is committed to helping women get answers regarding BIA-ALCL and will be continuing work with advocacy groups to advance the science and improve patient care for BIA-ALCL.

TABULAR MODIFICATIONS

C84 Mature T/NK-cell lymphomas

C84.7 Anaplastic large cell lymphoma, ALK-negative

New code C84.7A Anaplastic large cell lymphoma, ALK-negative, of the breast

Add Breast implant associated anaplastic large cell lymphoma (BIA-ALCL)

Add Code also, if applicable:
Add breast implant status (Z98.82)
Add personal history of breast implant removal (Z98.86)

Cervicogenic Headache

Ccervicogenic headache is a type of headache resulting from referred pain perceived in the head from a source in the neck. A cervicogenic headache is a secondary headache, resulting from a disorder of the cervical spine and its component bone, disc and/or soft tissue elements, usually but not invariably accompanied by neck pain.

The term cervicogenic headache appears on occasion in the survey data of the National Ambulatory Medical Care Survey (NAMCS) conducted by the Division of Health Care Statistics (DHCS) of the National Center for Health Statistics (NCHS). There is no unique ICD-10-CM code for cervicogenic headache.

The Division of Health Care Statistics of the National Center for Health Statistics is requesting a new code to allow for reporting this distinct type of headache. The code also note is included in the proposal to provide instruction that the underlying cause of the headache should also be coded, if known.

TABULAR MODIFICATIONS

G44 Other headache syndromes

G44.8 Other specified headache syndromes

New code G44.86 Secondary headache, cervicogenic
Add Code also associated cervical spinal condition, if known

Complication of Immune Effector Cellular (IEC) Therapy

The Alliance of Dedicated Cancer Centers (ADCC) is requesting a single new code to capture the complication of Immune Effector Cellular (IEC) therapy. The request for a new code will enable the ability to report and track this important clinical information. The new code is requested to be added to Chapter 19 Injury, Poisoning and Certain Other Consequences of External Causes of ICD-10-CM in the T80-T88 section for complications of surgical and medical care, not elsewhere classified.

Currently there are no ICD-10-CM diagnosis codes for reporting a complication of IEC therapy at the level of specificity that is being requested. The creation of new codes will allow coding professionals to more accurately translate physician documentation and clinical terminology into codes that describe this condition.

The closest codes to report complications of IEC therapy is T86.898, Other complications of other transplanted tissue or T80.89, Other complications following infusion, transfusion and therapeutic injection. ICD-10-CM code T86.898 is for transplanted tissue and does not reflect that the complication of Cytokine Release Syndrome which is a result from an immune effector cellular therapy procedure. ICD-10-CM code T80.89, Other complications following infusion, transfusion and therapeutic injection also is not specific.

Without the creation of a new ICD-10-CM diagnosis code, hospitals are likely to code inconsistently. This will hamper the ability to make meaningful comparisons to help assess differences in patient care, resource consumption (i.e., use of the intensive care unit, overall length of stay, additional drugs, etc.), and outcomes for different types of IEC therapy cases.

ADCC clinicians and ADCC coding professionals support the need for new codes to adequately capture this important clinical information.

TABULAR MODIFICATIONS

T80 Complications following infusion, transfusion and therapeutic injection
Includes: complications following perfusion
Excludes2: bone marrow transplant rejection (T86.01)
febrile nonhemolytic transfusion reaction (R50.84)
fluid overload due to transfusion (E87.71)
posttransfusion purpura (D69.51)
transfusion associated circulatory overload (TACO) (E87.71)
transfusion (red blood cell) associated hemochromatosis (E83.111)
transfusion related acute lung injury (TRALI) (J95.84)

The appropriate 7th character is to be added to each code from category T80

A initial encounter
D subsequent encounter
S sequela

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

New Code T80.82 Complication of immune effector cellular therapy
Add Excludes2: complication of bone marrow transplant (T86.0)
Add complication of stem cell transplant (T86.5)

Add Use additional code to identify the specific complication, such as:
Add cytokine release syndrome (D89.83-)
Add immune effector cell-associated neurotoxicity syndrome (G92.0-)

Cough

The American Thoracic Society (ATS) and the American College of Chest Physicians (CHEST) Clinical Practice Committee jointly submit an updated proposal. **This proposal was presented at the March 2019 and March 2020 C&M Meeting. In response to public comments, this proposal is being resubmitted for consideration. The changes are in bold.**

Physiologically, cough arises following activation of a complex sensorimotor reflex arc. Coughing is part of the body's defense mechanism against inhaled irritants and respiratory infections, serving to clear the airways of foreign material and excess secretions (Chung and Pavord, 2008). In most cases, cough resolves after the inciting factor is eliminated. For some people, however, cough becomes persistent, impacting quality of life and prompting the patient to seek medical attention.

During clinical work-up, cough is initially classified by duration; different categories of cough duration have different diagnostic possibilities and thus different algorithms for evaluation and treatment. The classification of cough by duration was outlined by the world's first cough guideline developed by the CHEST Expert Cough Panel in 1998 and has persisted through the most recent 2018 update (Irwin et al, 1998, 2018).

Cough of less than 3 weeks duration in adults is defined as acute cough (Chung and Pavord, 2008). Though acute cough can be a sign of a life-threatening condition or an exacerbation of a pre-existing respiratory condition, most acute cough cases are associated with respiratory tract infections. The most common cause of acute cough is acute bronchitis, which is most often viral (Terasaki and Paauw, 2014). Cough associated with respiratory tract infections commonly resolves shortly after the infection itself and does not require targeted therapy. In fact, limited data exist that show any benefit of symptomatic relief for acute cough with traditional cough suppressants like dextromethorphan and codeine (Bolser, 2006). The efficacy of antitussive drugs has been challenged particularly in the case of cough associated with upper respiratory tract infection (URTI); specifically, CHEST advises against the use of antitussives in the case of URTI (Irwin et al, 2018).

Subacute cough is quite like acute cough as both may be related to URTI and typically resolve after the infection clears. Subacute cough also may be caused by post-infectious cough, pertussis, infection with Mycoplasma or Chlamydia, and – similarly to acute cough – exacerbations of other diseases such as asthma or COPD (Chung and Pavord, 2008). The defining difference between subacute and acute in adults is the duration of the cough, subacute being longer, lasting from three to eight weeks. In children, a cough is defined as chronic beginning at 8 weeks duration (Chang et al, 2017).

A significant minority of patients experience chronic cough that persists despite guideline-based treatment of underlying etiologies. This subset of chronic cough is defined as cough that persists after extensive medical investigation and is thus considered a diagnosis of exclusion (Gibson and Wang, 2016). While various terms have been used to describe this population, recent CHEST guidelines define Unexplained Chronic Cough (UCC) as cough that occurs under the following circumstances: 1) chronic

cough with no diagnosable cause, 2) explained but refractory chronic cough, and 3) unexplained and refractory chronic cough (Irwin et al, 2018).

Chronic cough can have wide-ranging effects on overall health and well-being. Some of the more severe symptoms include syncope, incontinence, vomiting, and sleep deprivation (Irwin, 2006). Literature indicates that the psychosocial impact of refractory chronic cough can also be profound – studies have demonstrated that 53% of patients with chronic cough exhibit depressive symptoms and are at risk for developing clinical depression (Dicpinigaitis et al, 2006; McGarvey et al, 2006). The prevalence of depressive symptoms among patients with refractory chronic cough is comparable to that seen in other chronic disorders, such as chronic obstructive pulmonary disease, chronic heart failure, and diabetes (Brignall et al, 2008

Since research indicates paroxysmal cough is normally seen during the second stage of pertussis (whooping cough), the submitters no longer recommend listing paroxysmal cough as an inclusion term under chronic cough, R05.3. Instead, they recommend consideration of paroxysmal cough as a potential inclusion term under both Whooping cough due to Bordetella pertussis, without pneumonia, A37.00, and Whooping cough due to Bordetella pertussis, with pneumonia, A37.01.

The following proposed tabular modifications will ensure that ICD-10-CM is better aligned with the current clinical guidelines for cough.

TABULAR MODIFICATIONS

A37 Whooping cough
A37.0 Whooping cough due to Bordetella pertussis
A37.00 Whooping cough due to Bordetella pertussis without pneumonia
Add Paroxysmal cough due to Bordetella pertussis without pneumonia
A37.01 Whooping cough due to Bordetella pertussis with pneumonia
Add Paroxysmal cough due to Bordetella pertussis with pneumonia
R05 Cough

Delete **Excludes1: ~~cough with hemorrhage (R04.2)~~**
Add **paroxysmal cough due to Bordetella pertussis (A37.0-)**
smoker's cough (J41.0)
Add **Excludes2: cough with hemorrhage (R04.2)**

New code R05.1 Acute cough

New code R05.2 Subacute cough
New code R05.3 Chronic cough
Add Persistent cough
Add Refractory cough
Add Unexplained cough

New code R05.4 Cough syncope
Code first: syncope and collapse (R55)

New code R05.8 Other specified cough

New code R05.9 Cough, unspecified

References

Bolser DC. Cough suppressant and pharmacologic protussive therapy: ACCP evidence-based clinical practice guidelines. CHEST. 2006; 120:238S-49S.

Brignall K, Jayaraman B, Birring SS. Quality of life and psychosocial aspects of cough. Lung. 2008; 186(Suppl1):S55-58.

Centers for Disease Control. ICD-10-CM Official Coding Guidelines – Supplement; Coding encounters related to E-cigarette, or vaping, product use. Available at https://www.cdc.gov/nchs/data/icd/Vapingcodingguidance2019_10_17_2019.pdf, last accessed 11/15/19.

Chang AB, Oppenheimer JJ, Weinberger MM et al. Use of management pathways or algorithms in children with chronic cough: CHEST Guideline and Expert Panel Report. Chest 2017; 151(4):884-890.

Chung KF, Pavord ID. Chronic Cough 1: Prevalence, pathogenesis, and causes of chronic cough. Lancet. 2008; 371:1364-74.

Dicpinigaitis PV, Tso R, Banauch G. Prevalence of depressive symptoms among patients with chronic cough. Chest 2006; 130:1839–1483.

Gibson P, Wang G, et al. Treatment of unexplained chronic cough, CHEST Guideline and Expert Panel Report. Chest 2016 Jan; 149(1): 27-44.

Irwin RS. Assessing cough severity and efficacy of therapy in clinical research: ACCP evidence-based clinical practice guidelines. Chest 2006; 129(1 Suppl):232S–237S

Irwin RS et al. Managing cough as a defense mechanism and as a symptom. A consensus panel report of the American College of Chest Physicians. Chest. 1998;114(2 suppl):133S-181S.

Irwin RS, French CL, et al. Classification of cough as a symptom in adults and management algorithms; CHEST Guideline and Expert Panel Report. Chest. 2018; 153(1):196-209.

McGarvey LPA, Carton C, Gamble LA, Heaney LG, Shepherd R, Ennis M, Macmahon J. Prevalence of psychomorbidity among patients with chronic cough. Cough; 2006; 2:4.

Terasaki G, Paauw DS. Evaluation and treatment of chronic cough. Med Clin N Am. 2014; 98:391-403.

COVID-19 Issues

COVID-19 caused by the virus SARS-CoV-2 is a significant public health issue, and multiple requests for related codes have been received. It is proposed to create specific codes related to COVID-19 screening, exposure, and personal history. **The implementation date is expected to be in January 2021. The comment deadline will be Oct. 9, 2020.**

For exposure to COVID-19, it is proposed to create a new code Z20.822, Contact with and (suspected) exposure to COVID-19. For screening for COVID-19, it is proposed to create a new code, Z11.52, Encounter for screening for COVID-19. In general, by its definition, screening would be for people who are asymptomatic (and a symptom code should generally be used otherwise). For those who have had COVID-19, it is proposed to create a new code Z86.16, Personal history of COVID-19.

References

CDC. Coronavirus (COVID-19). 2020. https://www.cdc.gov/coronavirus/2019-nCoV/index.html

TABULAR MODIFICATIONS

Z11 Encounter for screening for infectious and parasitic diseases

Z11.5 Encounter for screening for other viral diseases

New code Z11.52 Encounter for screening for COVID-19

Z20 Contact with and (suspected) exposure to communicable diseases

Z20.8 Contact with and (suspected) exposure to other communicable diseases

Z20.82 Contact with and (suspected) exposure to other viral communicable diseases

New code Z20.822 Contact with and (suspected) exposure to COVID-19

Add Contact with and (suspected) exposure to SARS-CoV-2

Z86 Personal history of certain other diseases

Z86.1 Personal history of infectious and parasitic diseases

New code Z86.16 Personal history of COVID-19

INDEX MODIFICATIONS

Infection …

Add - SARS-CoV-2 – see Infection, COVID-19

Depression NOS

The Division of Health Care Statistics (DHCS) of the National Center for Health Statistics (NCHS) is requesting a code for Depression NOS. ICD-9-CM had a unique code for unspecified depression (311). Though this is an unspecified term, it is frequently seen in medical records and needed to allow for data comparability across years. In 2014 and 2015, the ICD-9-CM code 311 was found on 662 records, 1.5% of the total records of the NCHS' National Ambulatory Medical Care Survey (NAMCS).

Currently in ICD-10-CM, the default for Depression NOS is code F32.9, Major depressive disorder, single episode, unspecified. However, this has been determined by subject matter experts to be clinically incorrect.

Having an unspecified term default to major depression will prevent the true incidence of depression NOS from being captured and will incorrectly increase the incidence of major depression in statistical data.

The American Psychiatric Association (APA) has reviewed and supports this proposal.

TABULAR MODIFICATIONS

Revise	F32	Depressive ~~Major depressive disorder, single~~ episode	
	F32.9	Major depressive disorder, single episode, unspecified	
Delete		~~Depression NOS~~	
Delete		~~Depressive disorder NOS~~	
		Major depression NOS	
New code	F32.A	Depression, unspecified	
Add		Depression NOS	
Add		Depressive disorder NOS	

INDEX MODIFICATION

Revise	Despondency ~~F32.9~~ F32.A

Endometriosis

The American College of Obstetricians and Gynecologists (ACOG) and American Association of Gynecologic Laparoscopists (AAGL) are requesting expansion of the N80 code category for endometriosis. Endometriosis is an often painful disorder in which tissue similar to the tissue that normally lines the inside of the uterus, the endometrium, grows outside the uterus. Endometriosis most commonly involves the ovaries, fallopian tubes and the tissue lining the pelvis. The primary symptom of endometriosis is pelvic pain, often associated with menstrual periods. Although many women experience cramping during their menstrual periods, those with endometriosis typically describe menstrual pain that's far worse than usual. Pain usually increases over time.

The description of superficial and deep:

- Superficial endometriosis: Ectopic growth of endometrial-like tissue that extends 5mm or less below the peritoneal surface. Lesions can vary in number (singular or in multiple locations).
- Deeply infiltrating endometriosis: Ectopic growth of endometrial-like tissue that extends greater than 5mm below the peritoneal surface. Lesions can vary in number (singular or in multiple locations). These lesions are commonly associated with deep fibrosis and adhesions.

Current ICD-10 codes for endometriosis do not provide details in terms of laterality, location, depth of invasion, volume of disease and specific organ(s) involved. The addition and use of these proposed codes to specifically describe the type and location of endometriosis will have direct implications on disease management and clinical outcomes.

ACOG and AAGL request the N80 code category to be expanded to provide additional specificity for appropriate diagnosis coding and to assist in measuring the incidence of these specific conditions. This will enable better tracking, measurement, and ultimately treatment for endometriosis.

TABULAR MODIFICATIONS

N80 Endometriosis

N80.0 Endometriosis of uterus

Delete	~~Adenomyosis~~
Add	Endometriosis of the cervix
	Excludes1: stromal endometriosis (D39.0)
New code	N80.00 Endometriosis of the uterus, unspecified
Add	Endometriosis of the cervix, unspecified

New code N80.01 Superficial endometriosis of the uterus
Add Superficial endometriosis of the cervix

New code N80.02 Adenomyosis of the uterus
Add Adenomyosis NOS

New code N80.03 Deep retrocervical endometriosis

N80.1 Endometriosis of ovary

New sub-subcategory N80.10 Superficial endometriosis of the surface of the ovary
New code N80.101 Superficial endometriosis of the surface of right ovary
New code N80.102 Superficial endometriosis of the surface of left ovary
New code N80.103 Superficial endometriosis of the surface of bilateral ovaries
New code N80.109 Superficial endometriosis of the surface of ovary, unspecified side

New sub-subcategory N80.11 Deep endometriosis of ovary
Add Deep ovarian endometriosis
Add Endometrioma

New code N80.111 Deep endometriosis of right ovary
New code N80.112 Deep endometriosis of left ovary
New code N80.113 Deep endometriosis of bilateral ovaries
New code N80.119 Deep endometriosis of unspecified ovary

N80.2 Endometriosis of fallopian tube

New sub-subcategory N80.20 Superficial endometriosis of fallopian tube
New code N80.201 Superficial endometriosis of right fallopian tube
New code N80.202 Superficial endometriosis of left fallopian tube
New code N80.203 Superficial endometriosis of bilateral fallopian tubes
New code N80.209 Superficial endometriosis of unspecified fallopian tube

New sub-subcategory	N80.21 Deep endometriosis involving muscular wall of fallopian tube
New code	N80.211 Deep endometriosis involving muscular wall of right fallopian tube
New code	N80.212 Deep endometriosis involving muscular wall of left fallopian tube
New code	N80.213 Deep endometriosis involving muscular wall of bilateral fallopian tubes
New code	N80.219 Deep endometriosis involving muscular wall of unspecified fallopian tube

N80.3 Endometriosis of pelvic peritoneum

New code	N80.30 Endometriosis of pelvic peritoneum, unspecified
Add	Endometriosis of unspecified part of the pelvic abdominal wall
New sub-subcategory	N80.31 Endometriosis of the anterior cul-de-sac
New code	N80.311 Superficial endometriosis of the anterior cul-de-sac
Add	Endometriosis of the anterior cul-de-sac, NOS
New code	N80.312 Deep endometriosis of the anterior cul-de-sac with obliteration of cul de sac
New code	N80.313 Deep endometriosis of the anterior cul-de-sac without obliteration of cul de sac
New code	N80.319 Endometriosis of the anterior cul-de-sac, unspecified
New sub-subcategory	N80.32 Endometriosis of the posterior cul-de-sac
New code	N80.321 Superficial endometriosis of the posterior cul-de-sac
Add	Endometriosis of the posterior cul-de-sac, NOS
New code	N80.322 Deep endometriosis of the posterior cul-de-sac with obliteration of cul de sac

New code	N80.323 Deep endometriosis of the posterior cul-de-sac without obliteration of cul de sac
New code	N80.329 Endometriosis of the posterior cul-de-sac, unspecified
New sub-subcategory	N80.33 Superficial endometriosis of the uterosacral ligament(s)
New code	N80.331 Superficial endometriosis of the right uterosacral ligament(s)
New code	N80.332 Superficial endometriosis of the left uterosacral ligament(s)
New code	N80.333 Superficial endometriosis of the bilateral uterosacral ligament(s)
New code	N80.339 Superficial endometriosis of the uterosacral ligament(s), unspecified
New sub-subcategory	N80.34 Deep endometriosis of the uterosacral ligament(s)
New code	N80.341 Deep endometriosis of the right uterosacral ligament(s)
New code	N80.342 Deep endometriosis of the left uterosacral ligament(s)
New code	N80.343 Deep endometriosis of bilateral uterosacral ligament(s)
New code	N80.349 Deep endometriosis of the uterosacral ligament(s), unspecified
New sub-subcategory	N80.35 Superficial endometriosis of the pelvic sidewall
New code	N80.351 Superficial endometriosis of the right pelvic sidewall
New code	N80.352 Superficial endometriosis of the left pelvic sidewall

New code	N80.353 Superficial endometriosis of bilateral pelvic sidewall
New code	N80.359 Superficial endometriosis of the pelvic sidewall, unspecified
New sub-subcategory	N80.36 Deep endometriosis of the pelvic sidewall
New code	N80.361 Deep endometriosis of the right pelvic sidewall
New code	N80.362 Deep endometriosis of the left pelvic sidewall
New code	N80.363 Deep endometriosis of the bilateral pelvic sidewall
New code	N80.369 Deep endometriosis of the pelvic sidewall, unspecified
New sub-subcategory	N80.3A Superficial endometriosis of the pelvic brim
New code	N80.3A1 Superficial endometriosis of the right pelvic brim
New code	N80.3A2 Superficial endometriosis of the left pelvic brim
New code	N80.3A3 Superficial endometriosis of bilateral pelvic brim
New code	N80.3A9 Superficial endometriosis of the pelvic brim, unspecified
New sub-subcategory	N80.3B Deep endometriosis of the pelvic brim
New code	N80.3B1 Deep endometriosis of the right pelvic brim
New code	N80.3B2 Deep endometriosis of the left pelvic brim
New code	N80.3B3 Deep endometriosis of bilateral pelvic brim
New code	N80.3B9 Deep endometriosis of the pelvic brim, unspecified
New code	N80.39 Endometriosis of the pelvic peritoneum, other specified sites

New subcategory N80.4 Endometriosis of rectovaginal septum and vagina
New code N80.41 Deep endometriosis of the rectovaginal septum
New code N80.42 Deep endometriosis of the pararectal fossa
New code N80.43 Deep endometriosis of the vagina
New code N80.49 Other endometriosis of rectovaginal septum and vagina

N80.5 Endometriosis of intestine
New code N80.50 Endometriosis of intestine, unspecified
New sub-subcategory N80.51Endometriosis of the rectum
New code N80.511 Superficial endometriosis of the rectum
Add Endometriosis of the rectum, NOS

New code N80.512 Deep endometriosis of the rectum
Add Deep endometriosis of the rectum, multifocal

New sub-subcategory N80.52 Endometriosis of the sigmoid colon
New code N80.521 Superficial endometriosis of the sigmoid colon
Add Endometriosis of the sigmoid colon, NOS

New code N80.522 Deep endometriosis of the sigmoid colon
Add Deep endometriosis of the sigmoid colon, multifocal

New sub-subcategory N80.53 Endometriosis of the cecum
New code N80.531 Superficial endometriosis of the cecum
Add Endometriosis of the cecum, NOS

New code N80.532 Deep endometriosis of the cecum
Add Deep endometriosis of the cecum, multifocal

New sub-subcategory N80.54 Endometriosis of the appendix
New code N80.541 Superficial endometriosis of the appendix
Add Endometriosis of the appendix, NOS
New code N80.542 Deep endometriosis of the appendix

New sub-subcategory N80.55 Endometriosis of the colon
New code N80.551 Superficial endometriosis of the colon
Add Endometriosis of the colon, NOS

New code N80.552 Deep endometriosis of the colon
Add Deep endometriosis of the colon, multifocal

New sub-subcategory N80.56 Endometriosis of the small intestine
New code N80.561 Superficial endometriosis of the small intestine
Add Endometriosis of the small intestine, NOS
New code N80.562 Deep endometriosis of the small intestine
Add Deep endometriosis of the small intestine, multifocal

N80.6 Endometriosis in cutaneous scar

New subcategory N80.A Endometriosis of the bladder and ureters
Add Deep visceral endometriosis

New code N80.A1 Superficial endometriosis of the surface of the bladder

New sub-subcategory N80.A2 Deep endometriosis of the bladder
New code N80.A21 Deep endometriosis of the bladder, partial thickness
New code N80.A22 Deep endometriosis of the bladder, full thickness

New sub-subcategory N80.A3 Extrinsic endometriosis of the ureter
Add Code also obstructive and reflux uropathy (N13.-)
New code N80.A31 Extrinsic endometriosis of the right ureter
New code N80.A32 Extrinsic endometriosis of the left ureter
New code N80.A33 Extrinsic endometriosis of the bilateral ureters

New code N80.A39 Extrinsic endometriosis of the unspecified ureter

New sub-subcategory N80.A4 Intrinsic endometriosis of the ureter
Add Code also obstructive and reflux uropathy (N13.-)
New code N80.A41 Intrinsic endometriosis of the right ureter
New code N80.A42 Intrinsic endometriosis of the left ureter
New code N80.A43 Intrinsic endometriosis of the bilateral ureters
New code N80.A49 Intrinsic endometriosis of the unspecified ureter

New subcategory	N80.B Endometriosis of the cardiothoracic space
Add	Endometriosis of thorax
Add	Code also, if applicable: catamenial pneumothorax (J93.83) catamenial hemothorax (J94.2)
New code	N80.B1 Endometriosis of the pleura
New code	N80.B2 Endometriosis of the lung
New sub-subcategory	N80.B3 Endometriosis of the diaphragm
New code	N80.B31 Superficial endometriosis of the diaphragm
New code	N80.B32 Deep endometriosis of the diaphragm
New code	N80.B39 Endometriosis of the diaphragm, unspecified
New code	N80.B4 Endometriosis of the pericardial space
New code	N80.B9 Other endometriosis of cardiothoracic space
Add	Endometriosis of the mediastinal space
New subcategory	N80.C Endometriosis of the nervous system
New code	N80.C0 Endometriosis peripheral nerve, unspecified
New code	N80.C1 Endometriosis of the peripheral nervous system
New code	N80.C2 Endometriosis of the sacral splanchnic nerves
New code	N80.C3 Endometriosis of the sacral nerve roots
New code	N80.C4 Endometriosis of the obturator nerve
New code	N80.C5 Endometriosis of the sciatic nerve
New code	N80.C6 Endometriosis of the pudendal nerve
New code	N80.C7 Endometriosis of the femoral nerve
New code	N80.C9 Endometriosis of other nerve

N80.8 Other endometriosis

Delete	~~Endometriosis of thorax~~
New code	N80.81 Endometriosis involving the skin or subcutaneous layer of the anterior abdominal wall
New code	N80.82 Endometriosis involving the fascia and or muscular layer of the anterior abdominal wall
New code	N80.83 Endometriosis of the umbilicus

New code	N80.84	Endometriosis of the inguinal canal
New code	N80.85	Endometriosis of other part of abdominal wall
Add		Endometriosis of abdominal peritoneum NOS
Add		Endometriosis of extra-pelvic abdominal peritoneum
New code	N80.89	Endometriosis of other site

N80.9 Endometriosis, unspecified

Esophageal Polyp

Hyperplastic polyps of the esophagus and esophagogastric junction region (EGJ) are characterized by hyperplastic epithelium (foveolar-type, squamous, or both) with variable amounts of inflamed stroma.

ICD-10-CM has specific codes to classify non-adenomatous or hyperplastic polyps. However, there are no unique codes for esophageal polyp or esophagogastric junction polyp. The two codes currently assigned to capture an esophagogastric junction polyp are K22.8, Other specified diseases of the esophagus and K31.7, Polyp of stomach and duodenum.

The submitter is requesting the following new codes to capture these polyps when not documented as non-adenomatous. A unique code for esophagogastric junction polyp is also been requested by the Editorial Advisory Board for Coding Clinic. The American Gastroenterological Association (AGA) has reviewed and supports this proposal.

TABULAR MODIFICATIONS

	K22 Other diseases of esophagus
New subcategory	K22.8 Other specified diseases of esophagus
Delete	~~Hemorrhage of esophagus NOS~~
New code	K22.81 Esophageal polyp
New code	K22.82 Esophagogastric junction polyp
New code	K22.89 Other specified disease of esophagus
Add	Hemorrhage of esophagus NOS

Fournier Disease of Vagina and Vulva

NCHS has received a proposal for a new code for Fournier disease or gangrene of the vagina and vulva. Fournier disease/gangrene is a severe infectious necrotizing condition. Currently this condition is coded to N76.89, Other specified inflammation of vagina and vulva. This code merely describes inflammation of the vagina and does not adequately reflect Fournier disease. Creation of a new code will more specifically classify this significant condition which is often a diabetic complication.

Fournier disease is a necrotizing fasciitis of the perineum, that occurs as a result of a breach in the integrity of the gastrointestinal or urethral mucosa. Fournier disease is a form of polymicrobial (type I) infection. Fournier gangrene typically begins abruptly with severe pain and may spread rapidly to the anterior abdominal wall and the gluteal muscles.

Early surgical debridement of necrotic tissues and antibiotics are fundamental in the treatment of FG. Despite advanced management mortality is still high and averages 20%–30%.6

The American College of Obstetricians and Gynecologists (ACOG) has reviewed and concurs with the request.

TABULAR MODIFICATIONS

N76 Other inflammation of vagina and vulva
Use additional code (B95-B97), to identify infectious agent
Excludes2: senile (atrophic) vaginitis (N95.2)
vulvar vestibulitis (N94.810)
N76.8 Other specified inflammation of vagina and vulva
N76.81 Mucositis (ulcerative) of vagina and vulva
Code also type of associated therapy, such as:
antineoplastic and immunosuppressive drugs (T45.1X-)
radiological procedure and radiotherapy (Y84.2)
Excludes2: gastrointestinal mucositis (ulcerative) (K92.81)
nasal mucositis (ulcerative) (J34.81)
oral mucositis (ulcerative) (K12.3-)

New code N76.82 Fournier disease of vagina and vulva
Add Fournier gangrene of vagina and vulva
Add Code also if applicable necrotizing fasciitis (M72.6)
Add Excludes 2: gangrene in diabetes mellitus (E08-E13 with .52)

Gastric Intestinal Metaplasia

This topic was previously presented at the September 2019 and March 2020 Coordination and Maintenance (C&M) meeting. Based on public comments received, the revised proposal is being presented for consideration.

Gastric cancer is the fourth most common cancer worldwide and the second leading cause of cancer deaths. It afflicts approximately 26,000 Americans yearly. The location of gastric intestinal metaplasia (IM) is a significant predictor for gastric cancer risk and as is one of the most important characteristics of the disease. Currently, there is no ICD-10-CM unique code for gastric IM. A similar precursor lesion for esophageal cancer, Barrett's esophagus (also known as esophageal intestinal metaplasia) has a unique code (K22.7-).

It is believed the risk for progression into gastric cancer is highest among patients with diffuse gastric IM (which involves both antrum and body). European guidelines use presence of diffuse gastric IM as a marker of higher risk. Gastric IM is categorized histopathologically into incomplete and complete types. Endoscopic gastric mapping to define extent of IM should be done for patients with incomplete IM to rule out dysplasia or adenocarcinoma. Once dysplasia is present, the location is less important as there is a higher risk regardless of location.

The American Gastroenterological Association (AGA) is requesting new codes to contribute to epidemiologic understanding and subsequent development of appropriate surveillance guidelines in the United States.

TABULAR MODIFICATIONS

K31 Other diseases of stomach and duodenum

New code K31.A Gastric intestinal metaplasia

New code K31.A0 Gastric intestinal metaplasia, unspecified
Add Gastric intestinal metaplasia indefinite for dysplasia
Add Gastric intestinal metaplasia NOS

New subcategory K31.A1 Gastric intestinal metaplasia without dysplasia

New code K31.A11 Gastric intestinal metaplasia without dysplasia, involving the antrum
New code K31.A12 Gastric intestinal metaplasia without dysplasia, involving the body (corpus)
New code K31.A13 Gastric intestinal metaplasia without dysplasia, involving the fundus
New code K31.A14 Gastric intestinal metaplasia without dysplasia, involving the cardia
New code K31.A15 Gastric intestinal metaplasia without dysplasia, involving-multiple sites

New code	K31.A19 Gastric intestinal metaplasia without dysplasia, unspecified site
New subcategory	K31. A2 Gastric intestinal metaplasia with dysplasia
New code	K31.A21 Gastric intestinal metaplasia with low grade dysplasia
New code	K31.A22 Gastric intestinal metaplasia with high grade dysplasia
New code	K31.A29 Gastric intestinal metaplasia with dysplasia, unspecified

Hematopoietic Stem Cell Transplant-Associated Thrombotic Microangiopathy

Hematopoietic Stem Cell Transplant-Associated Thrombotic Microangiopathy (HSCT-TMA), also known as Transplant-Associated Thrombotic Microangiopathy (TA-TMA), is a serious multisystem, life-threatening complication of hematopoietic stem cell transplantation [(1)]. HSCT-TMA is caused by endothelial damage as a result of the hematopoietic stem-cell transplant procedure, including conditioning regimens, exposure to immunosuppressants and associated infections as well as graft-versus-host disease (GVHD). This HSCT procedure takes place only in a limited number of select U.S. centers where patient survival rates, quality of care and complications are rigorously monitored.

Omeros Corporation is requesting creation of a specific ICD-10-CM code for Hematopoietic Stem Cell Transplant-Associated Thrombotic Microangiopathy (HSCT-TMA), otherwise known as Transplant-Associated Thrombotic Microangiopathy (TA-TMA). The Center for International Blood & Marrow Transplant Research (CIBMTR), the American Society for Transplantation and Cellular Therapy (ASTCT), academic transplant centers throughout the United States, and patient advocacy groups track epidemiology and clinical sequelae associated with a hematopoietic stem-cell transplant (HSCT). It will be easier for these organizations to fully characterize and determine the true incidence of HSCT-TMA with a specific ICD-10-CM code. HSCT-TMA is vastly underreported as evidenced by the extremely wide range (from 0.5% to 76%) of incidence of disease reported in the literature [(2, 3)]. As a complication of the HSCT procedure, HSCT-TMA differs clinically from other types of thrombotic microangiopathy (TMA), but these are all coded to the single ICD-10-CM code, M31.1, Thrombotic microangiopathy. Because HSCT-related complications and sequelae require detailed tracking by stakeholders, HSCT-TMA requires a unique code to differentiate it from other types of TMA. Due to the high severity of illness typical of patients who develop HSCT-TMA and the associated high mortality rate, appropriate classification of these patients is of paramount importance to align appropriate diagnostic and treatment strategies while avoiding inappropriate and potentially detrimental approaches. Creation of a specific ICD-10-CM code will improve the ability to track HSCT-TMA incidence, along with treatment and outcomes to improve patient care, and be useful for accurate capture of data for epidemiological research. This will enable clinicians to help explain differences in patient care delivery, resource consumption (e.g., overall length of stay, additional drugs required, etc.) and outcomes specifically related to HSCT-TMA.

Often HSCT-TMA occurs simultaneously with other diagnoses, such as graft vs. host disease (GVHD, D89.813), hepatic veno-occlusive disease/sinusoidal obstruction syndrome (VOD/SOS, K76.5), or atypical hemolytic uremic syndrome (aHUS, D59.3).

HSCT-TMA is caused by the HSCT procedure and has some unique comorbidities. HSCT-TMA is initiated when various factors associated with HSCT lead to endothelial cell damage [(1, 5, 12, 14)]. These factors include chemoradiotherapy (e.g., required conditioning regimens), cytokine release, immuno-suppressive therapies, bacterial endotoxins, engraftment syndrome and allogenic reactions with donor-derived immune cells (i.e., GVHD) [(13, 14)]. Injured endothelium releases procoagulant microparticles and activates the complement system's lectin pathway, resulting in microvascular platelet aggregation and thromboses (i.e., TMA) [(8, 12, 13, 14)]. These complement-mediated activities further amplify endothelial injury and dysfunction, causing or worsening clinical conditions [(12)]. The increasing endothelial damage leads to further microthrombi formation, mechanical damage to red blood cells and lumen-obstruction, leading to HSCT-TMA-related organ damage and organ failure [(12)].

Risk factors for developing HSCT-TMA include inherent/non-modifiable conditions such as female sex, African American race, and genetic variants, including variants of complement-related genes [3, 5, 12, 14]. There are also specific transplant and post-transplant risk factors including conditioning regimens, total body irradiation, unrelated donor or human leukocyte antigen (HLA) mismatched transplants, use of T-Cell inhibitors, GVHD and infection [3, 5, 12, 14].

Key warning signs for developing HSCT-TMA are reduction in platelet counts, elevated levels of lactate dehydrogenase (LDH), proteinuria, and hypertension that persist beyond what is typically observed with corticosteroid or calcineurin inhibitor use [11]. Other diagnostic criteria include the presence of schistocytes, decreased hemoglobin, negative Coombs test, and decreased haptoglobin levels [3, 11]. Diagnosis of HSCT-TMA calls for a high index of suspicion and requires routine prospective monitoring [2, 8, 11].

HSCT-TMA typically presents within the first 100 days after transplantation but has been diagnosed as late as 721 days post-transplant [4, 11, 12, 13, 15, 16]. The condition occurs in both autologous and allogeneic transplants but is more common in the allogeneic population [1, 8, 12]. In the United States, approximately 20,000 to almost 23,000 allogeneic transplants were performed annually in 2014-2017, and the procedural frequency is increasing each year [17]. HSCT-TMA generally presents with signs of microangiopathic hemolytic anemia, consumptive thrombocytopenia in the absence of coagulopathy, and microvascular thrombosis with end-organ damage that is associated with increased post-HSCT mortality and shorter survival time [1]. In a retrospective analysis, the risk of transplant-related mortality was approximately four times higher in patients with confirmed HSCT-TMA than in patients without HSCT-TMA [9]. The need for accurately tracking the incidence, disease presentation and treatment-associated outcomes of HSCT-TMA is clear given the varied reporting across and within patient populations.

A recent large retrospective review of adult patients who underwent allogeneic HSCT reported an incidence of 13% with definite HSCT-TMA and 26% with probable HSCT-TMA [9]. A prospective study of 100 pediatric patients found an incidence of 39% [4]. Of these, 92.3% of cases had a HSCT-TMA incidence within 100 days of transplant [4]. Patients with HSCT-TMA had a non-relapse mortality rate of 43.6% at 1-year post-HSCT [4]. In another study in adults, the median time for TMA onset following HSCT was 1.2 months for definite TMA and 0.7 months for probable TMA [5]. In an adult study, subjects with HSCT-TMA had a non-relapse mortality rate of 54% and 41% for definite and probable cases, respectively, over a median 48.2-month follow-up [5]. Overall survival was poor for patients with definite HSCT-TMA and their mean survival time after HSCT-TMA onset was 2.4 months [5]. Recent reports in both adult and pediatric allogeneic stem cell transplant populations have also found a HSCT-TMA incidence of up to 39 percent and at least one high-risk feature may be present in up to 80 percent of these patients [4-6]. Patients with severe disease may be defined by multiorgan impairment, uncontrolled hypertension, worsening renal function and a lack of response to therapeutic plasma exchange [4, 8]. In general, severe HSCT-TMA is fatal in 90% or more of cases [7]. Patients with nonlethal cases of HSCT-TMA have an increased risk of chronic organ injury and other conditions including CNS complications, hypertension, pulmonary hypertension, chronic kidney disease and gastrointestinal disease [1, 5, 10].

References

1. Jodele S, Laskin BL, Dandoy CE, Myers KC, El-Bietar J, Davies SM, Goebel J, Dixon BP. A new paradigm: diagnosis and management of HSCT-associated thrombotic microangiopathy as multisystem endothelial injury. *Blood Reviews*. 2015;29(3):191-204. https://doi.org/10.1016/j.blre.2014.11.001
2. Ho VT et al. Blood and marrow transplant clinical trials network toxicity committee consensus summary: thrombotic microangiopathy after hematopoietic stem cell transplantation. *Biol Blood Marrow Transplant.* 2005;11(8):571-575. https://doi.org/10.1016/j.bbmt.2005.06.001
3. Epperla N, Li A, et al. Incidence, risk factors for and outcomes of transplant-associated thrombotic microangiopathy. *Br J Haematol*. 2020 Mar 2. https://doi.org/10.1111/bjh.16457
4. Jodele S, Davies SM, Lane A, et al. Diagnostic and risk criteria for HSCT-associated thrombotic microangiopathy: a study in children and young adults. *Blood*. 2014;124(4):645-653. https://doi.org/10.1182/blood-2014-03-564997
5. Postalcioglu M et al. Impact of thrombotic microangiopathy on renal outcomes and survival after hematopoietic stem cell transplantation. *Biol Blood Marrow Transplant.* 2018;24(11):2344-2353. https://dx.doi.org/10.1016%2Fj.bbmt.2018.05.010
6. Willems E et al. Comparison of thrombotic microangiopathy after allogeneic hematopoietic cell transplantation with high-dose or nonmyeloablative conditioning. *Bone Marrow Transplant.* 2010;45(4):689-693. https://doi.org/10.1038/bmt.2009.230
7. Jodele S et al. Eculizumab therapy in children with severe hematopoietic stem cell transplantation associated thrombotic microangiopathy. *Biol Blood Marrow Transplant*. 2014;20(4):518-525. https://doi.org/10.1016/j.bbmt.2013.12.565
8. Jodele S et al. New approaches in the diagnosis, pathophysiology, and treatment of pediatric hematopoietic stem cell transplantation-associated thrombotic microangiopathy. *Transfus Apher Sci.* Published April 2016. 2016;54(2):181-190. https://doi.org/10.1016/j.transci.2016.04.007
9. Cho B-S et al. Validation of recently proposed consensus criteria for thrombotic microangiopathy after allogeneic hematopoietic stem cell transplantation. *Transplant.* 2010;90(8):918-926. https://doi.org/10.1097/tp.0b013e3181f24e8d
10. Bonardi M et al. brain imaging findings and neurologic complications after allogenic hematopoietic stem cell transplantation in children. *Radiographics.* 2018;38(4):1223-1238. https://doi.org/10.1148/rg.2018170139
11. Elsallabi O et al. Hematopoietic stem cell transplant-associated thrombotic microangiopathy. *Clin Appl Thromb Hemost.* 2016;22(1):12-20. https://doi.org/10.1177/1076029615598221
12. Khosla J et al. Hematopoietic stem cell transplant-associated thrombotic microangiopathy: current paradigm and novel therapies. *Bone Marrow Transplant.* 2018; 53(2):129-137. https://doi.org/10.1038/bmt.2017.207
13. Carreras E et al. The role of the endothelium in the short-term complications of hematopoietic SCT. *Bone Marrow Transplant*. 2011;46(12):1495-1502. https://doi.org/10.1038/bmt.2011.65
14. Dvorak CC et al. Transplant-associated thrombotic microangiopathy in pediatric hematopoietic cell transplant recipients: a practical approach to diagnosis and management. *Front Pediatr.* 2019; 7:133.
15. Sakellari I et al. Transplant-associated thrombotic microangiopathy: prevalence, prognostic factors and treatment outcomes in unrelated allogeneic transplant for hematologic diseases. *Blood.* 2016;128(22):982. https://doi.org/10.1182/blood.V128.22.982.982
16. Orlic L et al. Kidney complications due to hematopoietic stem cell transplantation-a disorder of an increasing incidence? *BANTAO Journal.* 2014;12(2):90-96. https://doi.org/10.2478/bj-2014-0018
17. Center for International Blood and Marrow Transplant Research – Transplant Activity Report Covering 2013-2017. Retrieved from https://bloodstemcell.hrsa.gov/sites/default/files/bloodstemcell/data/transplant-activity/transplants-year.pdf.

TABULAR MODIFICATIONS

M31 Other necrotizing vasculopathies

M31.1 Thrombotic microangiopathy

New code	M31.10	Thrombotic microangiopathy, unspecified
New code	M31.11	Hematopoietic stem cell transplantation-associated thrombotic microangiopathy [HSCT-TMA]
Add		Transplant-associated thrombotic microangiopathy [TA-TMA]
Add		Code first if applicable:
Add		Complications of bone marrow transplant (T86.0-)
Add		Complications of stem cell transplant (T86.5)
Add		Use additional code to identify specific organ dysfunction, such as:
Add		Acute kidney failure (N17.-)
Add		Acute respiratory distress syndrome (J80)
Add		Capillary leak syndrome (I78.8)
Add		Diffuse alveolar hemorrhage (R04.89)
Add		Encephalopathy (metabolic) (septic) (G93.41)
Add		Fluid overload, unspecified (E87.70)
Add		Graft versus host disease (D89.81-)
Add		Hemolytic uremic syndrome (D59.3)
Add		Hepatic failure (K72.0-)
Add		Hepatic veno-occlusive disease (K76.5)
Add		Idiopathic interstitial pneumonia (J84.111)
Add		Sinusoidal obstruction syndrome (K76.5)
New code	M31.19	Other thrombotic microangiopathy
Add		Thrombotic thrombocytopenic purpura

Hemolytic-Uremic Syndrome: Typical and Atypical

Hemolytic-uremic syndrome (HUS) is a rare but devastating disorder. Formerly considered largely a disease of childhood, it is now understood to affect children and adults of any age. HUS is a thrombotic microangiopathy, characterized by hemolytic anemia, thrombocytopenia, and acute kidney injury, although other organs may be affected. The cause of the acute kidney injury is the microthrombi formation in the kidney's arterioles and capillaries, leading to tissue ischemia.

There are two main categories of HUS: typical and atypical. Etiology, treatment, and outcomes differ markedly between typical HUS and atypical HUS.

Typical hemolytic-uremic syndrome (tHUS, STEC-HUS) is due to Shiga toxin-producing *E coli* (STEC). It most commonly occurs in children under 5 years old, but also occurs in adults infected with STEC. Most patients have a preceding illness that includes bloody diarrhea from the *E. coli* infection, with about 10-15% going on to develop tHUS. Acute kidney injury occurs in more than 70% of patients. The microthrombi may also cause cerebral ischemia, a major cause of mortality, as well as ischemic injury of other organs.

Treatment for tHUS consists of supportive care. Anemia is treated with blood transfusions as needed. Acute kidney injury is treated with maintenance of fluid and electrolyte balance, control of blood pressure, and dialysis when indicated. Additional treatment may include parenteral nutrition for gastroenteritis or pancreatitis as well as anticonvulsants if seizures develop. Antibiotics are not given to treat the *E. coli* infection since there is no evidence of benefit and some epidemiologic studies have suggested harm.

About 70% of patients with tHUS recover from the acute episode without major sequelae and require no further treatment or management. The prognosis for recovery of renal function following acute kidney injury is quite favorable, although some patients develop chronic kidney disease. A small percentage of patients develop end-stage kidney disease, but transplant is an effective treatment option since there is almost no risk of tHUS affecting the new kidney. Although some tHUS patients have residuals such as chronic kidney disease, tHUS itself is resolved and does not recur.

Atypical hemolytic-uremic syndrome (aHUS) occurs in both adults and children. It represents about 10% of HUS cases, although this varies significantly depending on the specific etiology. Unlike tHUS, there are multiple etiologies for aHUS. Probably the most common is derangements in the complement cascade, part of the immune system that works to mark pathogens for destruction, due to either gene mutations in complement regulatory proteins or production of autoantibodies directed against complement. Other causes of aHUS include non-STEC infections, adverse effects of drugs, non-complement genetic disorders, and coexisting states and conditions such as bone marrow transplant, malignancy, and systemic lupus erythematosus.

Treatment of aHUS depends on the underlying etiology. For aHUS due to complement derangements caused by gene mutations, treatment consists of medications that block or inhibit the complement system, specifically the monoclonal antibodies eculizumab (Soliris) and ravulizumab (Ultomiris). Patients with aHUS due to autoantibodies directed against the complement system may also be treated with immunosuppression and plasmapheresis. Because meningococcal infection is a known risk for

use of these medications, a meningococcal vaccine is administered at diagnosis and the patient is also placed on prophylactic antibiotics. For other causes of aHUS, treatment varies widely and may include treating the underlying condition or adjusting the medication triggering aHUS. In addition to the underlying disorder, acute kidney injury is treated with fluid and electrolyte balance, control of blood pressure, and dialysis when indicated.

Because underlying genetic abnormalities cannot be corrected or addressed, many patients with aHUS remain at significant risk after resolution of the acute episode. These patients are managed with long-term monitoring. Many patients also require long-term or life-long treatment with complement inhibitors to prevent recurrence of aHUS.

Differentiating typical HUS and atypical HUS will enable the prevalence, treatments, and outcomes for these two conditions to be individually tracked. This proposal to create specific codes for typical and atypical HUS was received from Dr. Laurence Greenbaum, of the Emory School of Medicine, in Atlanta, GA.

References

Schick, P., Nagalla, S. (2019). Hemolytic Anemia. Medscape. http://emedicine.medscape.com/article/201066-overview
National Library of Medicine (NLM) (2020). Hemolytic-uremic syndrome. MedlinePlus. National Institutes of Health. https://medlineplus.gov/ency/article/000510.htm

TABULAR MODIFICATIONS

D59 Acquired hemolytic anemia

D59.3 Hemolytic-uremic syndrome

Add — Code also, if applicable, any associated:
Add — acute kidney failure (N17.-)
Add — chronic kidney disease (N18.-)

Use additional code to identify associated:
E. coli infection (B96.2-)
Pneumococcal pneumonia (J13)
Shigella dysenteriae (A03.9)

New code — D59.31 Typical hemolytic-uremic syndrome
Add — Hemolytic-uremic syndrome NOS
Add — Shiga toxin *E. coli* related hemolytic uremic syndrome
Add — STEC-HUS

New code — D59.39 Other hemolytic-uremic syndrome
Add — Atypical hemolytic-uremic syndrome

D69 Purpura and other hemorrhagic conditions

Add Excludes1: hemolytic-uremic syndrome (D59.3-)

Acute kidney failure and chronic kidney disease (N17-N19)

Revise Excludes2: hemolytic-uremic syndrome (D59.3-)

Hereditary Alpha Tryptasemia

Hereditary alpha tryptasemia (HαT) is a recently described genetic trait that can be associated with mast cell activation (Lyons, 2016). It is characterized by an elevated basal serum tryptase, and genetic testing showing an increased copy number of the TPSAB1 gene encoding alpha tryptase (Lyons, 2018). Tryptase is a mast cell mediator, and its elevation is commonly associated with the diagnosis of mast cell associated diseases. Unlike patients with systemic mastocytosis, HαT patients have elevated serum tryptase in the absence of clonal mast cell expansion. HαT affects about 4-6% of the Caucasian population of the United States, and parts of Western Europe, and likely the entire diaspora of this racial group.

TPSAB1 gene replications and elevated basal tryptase are associated with multisystem complaints, including cutaneous flushing and pruritis, dysautonomia (dysfunctioning of the autonomic nervous system, potentially with swings in heart rate and blood pressure, and fainting), functional gastrointestinal symptoms, chronic pain, and connective tissue abnormalities including joint hypermobility in roughly 1/3 of patients diagnosed with HαT (Lyons, 2018). Those with HαT have an increasesd risk of severe allergic reactions to stinging insects, including anaphylactic reactions in some cases. CLIA-approved genetic testing is available in the U.S. through Gene by Gene, Inc. (Weiler, 2020).

The University of Mississippi in cooperation with the Mastocytosis Society and Gene by Gene have requested a new code for hereditary alpha tryptasemia, in order to track disease prevalence and patient outcomes. There is currently no specific ICD-10-CM code for hereditary alpha tryptasemia, although it would be appropriate now to use the code D89.49, Other mast cell activation disorder.

References

Lyons, J.J., et al. (2016). Elevated basal serum tryptase identifies a multisystem disorder associated with increased TPSAB1 copy number. Nat Genet, 48(12): 1564-1569. https://doi.org/10.1038/ng.3696

Lyons, J.J. (2018). Hereditary Alpha Tryptasemia: Genotyping and Associated Clinical Features. Immunol Allergy Clin North Am, 38(3): 483-495. https://doi.org/10.1016/j.iac.2018.04.003

Weiler, C.R. (2020). Hereditary Alpha Tryptasemia, Tryptase and Alpha Tryptasemia. The Mastocytosis Society. Retrieved from https://tmsforacure.org/overview/hereditary-alpha-tryptasemia-testing/

TABULAR MODIFICATIONS

D89 Other disorders involving the immune mechanism, not elsewhere classified

D89.4 Mast cell activation syndrome and related disorders

New code	D89.44	Hereditary alpha tryptasemia
Add		Use additional code, if applicable, for:

Add	allergy status, other than to drugs and biological substances (Z91.0-)
Add	personal history of anaphylaxis (Z87.892)

Immune Effector Cell Associated Neurotoxicity Syndrome (ICANS)

Chimeric Antigen Receptor T (CAR-T) Cell Therapy has been a welcome advancement in the treatment of relapsed or refractory leukemia and large b-cell lymphoma, however complications of the therapy have been observed. Two of the most prevalent complications are Cytokine Release Syndrome (CRS) and Immune effector Cell Associated Neurotoxicity Syndrome (ICANS). The Alliance of Dedicated Cancer Centers (ADCC) submits a request for new codes address this clinical condition. **This proposal was presented at the March 2020 C&M Meeting. In response to public comment, the proposal is being represented for consideration.**

ICANS is defined as "a disorder characterized by a pathologic process involving the central nervous system following any immune therapy that results in the activation or engagement of endogenous or infused T cells and/or other immune effector cells. "Signs and symptoms can be progressive and may include aphasia, altered level of consciousness, impairment of cognitive skills, motor weakness, seizures, and cerebral edema."[1]

Like CRS, some of the symptoms that occur as part of this syndrome are nonspecific, and it is of the opinion of the Alliance of Dedicated Centers (ADCC) and other clinical experts, that coding signs and symptoms of neurotoxicity will not be enough to understand which patients have this diagnosis and its severity. To enable research and comparisons, it is also important to have codes to describe the consensus grading scale grades of ICANS.

In January 2019, the ASTCT published a paper on the formal consensus grading in the official journal of the ASTCT, then named Biology of Blood and Marrow Transplantation.[2] Now that there is consensus on a grading scale, there is widespread agreement among clinicians and their institutions that unique ICD-10-CM diagnosis codes are essential to describe this frequent complication in patients in who receive immune effector cell therapy.

There are currently no ICD-10-CM diagnosis codes to report the ICANS complication of immune effector cell therapy, nor are there codes to report the severity of ICANS. The creation of new codes will allow coding professionals to accurately translate physician documentation and clinical terminology into the codes reported to describe the occurrence and severity of IEC therapy's most significant and common complications (i.e., the different grades of ICANS). This will allow hospitals and clinicians information needed to help explain differences in patient care delivery, resource consumption (i.e., use of the intensive care unit, overall length of stay, additional drugs, etc.), and outcomes for different types of IEC therapy cases.

1 Lee DW, Santomasso BD, Locke FL, et al., "ASTCT Consensus Grading for Cytokine Release Syndrome and Neurologic Toxicity Associated with Immune Effector Cells," Biol Blood Marrow Transplant. 2019 Apr;25(4):625-638. doi: 10.1016/j.bbmt.2018.12.758. Epub 2018 Dec 25. 2 Ibid.

TABULAR MODIFICATIONS

G92 Toxic encephalopathy

Delete ~~Toxic encephalitis~~
Delete ~~Toxic metabolic encephalopathy~~

Delete ~~Code first, if applicable, drug induced (T36-T50)~~
Delete ~~Code first (T51-T65) to identify toxic agent~~

Add Code first poisoning due to drug or toxin, if applicable, (T36-T65 with fifth or sixth character 1-4 or 6)

Add Use additional code for adverse effect, if applicable, to identify drug (T36-T50 with fifth or sixth character 5)

New subcategory G92.0 Immune effector cell-associated neurotoxicity syndrome

Add Code first underlying cause such as complications of immune effector cellular therapy (T80.82)

Add Code also associated signs and symptoms, such as seizures and cerebral edema

New code G92.00 Immune effector cell-associated neurotoxicity syndrome, grade unspecified
Add ICANS, grade unspecified

New code G92.01 Immune effector cell-associated neurotoxicity syndrome, grade 1
Add ICANS, grade 1

New code G92.02 Immune effector cell-associated neurotoxicity syndrome, grade 2
Add ICANS, grade 2

New code G92.03 Immune effector cell-associated neurotoxicity syndrome, grade 3
Add ICANS, grade 3

New code G92.04 Immune effector cell-associated neurotoxicity syndrome, grade 4
Add ICANS, grade 4

New code G92.05 Immune effector cell-associated neurotoxicity syndrome, grade 5
Add ICANS, grade 5

New code G92.8 Other toxic encephalopathy
Add Toxic encephalitis
Add Toxic metabolic encephalopathy

New code G92.9 Unspecified toxic encephalopathy

Immunization Counseling

Patients and caregivers seek counseling services without signs or symptoms, and unrelated to medical care, e.g. preventive care, for many reasons. While there are a number of ICD-10-CM codes for a variety of counseling services, currently there are no codes for counseling services related to immunizations. **This proposal was presented at the March 2020 Coordination and Maintenance Meeting. In response to public comments, this proposal is being resubmitted for consideration.**

A unique code is being requested to identify encounters where the parent/patient presents specifically for vaccine counseling. Typically, these parents are seeking an alternative vaccine, alternative vaccine schedule or spend time with the provider asking questions about vaccine safety. It is important to be able to show that counseling is being done particularly when a patient does not have an updated immunization record.

Vaccines, which have proven to be a safe and very effective preventive measure, are under constant fire through social media outlets with little to no scientific backing. Parents will read this information and decide not to vaccinate their children or want to discuss this with their child's provider. It has proven to be a public health issue when misinformation leads to many pediatric patients going unimmunized or underimmunized. This has recently been evident by the measles outbreaks in the US.

The American Academy of Pediatrics requests the addition of a specific code to identify this encounter when vaccinations are discussed at length with parents/patients.

TABULAR MODIFICATIONS

Z23 Encounter for immunization
Code first any routine childhood examination

Note: procedure codes are required to identify the types of immunizations given

Add **Code also, if applicable, encounter for immunization safety counseling (Z71.85)**

Z28 Immunization not carried out and underimmunization status
Includes: vaccination not carried out

Add **Code also, if applicable, encounter for immunization safety counseling (Z71.85)**

Z71 Persons encountering health services for other counseling and medical advice, not elsewhere classified
Excludes2: contraceptive or procreation counseling (Z30-Z31)
sex counseling (Z70.-)

Z71.8 Other specified counseling

New Code	Z71.85 Encounter for immunization **safety** counseling
Add	Encounter for vaccine product safety counseling
Add	Code also, if applicable, encounter for immunization (Z23)
Add	**Code also, if applicable, immunization not carried not (Z28.-)**
Add	**Excludes1: encounter for health counseling related to travel (Z71.84)**

Lipedema and Lipolymphedema

Lipedema, initially described at the Mayo clinic in 1940, is a loose, connective-tissue (fat) disease (lipomatosis) with a pathological deposition of fibrotic fatty tissue on the limbs of women sparing the trunk, hands and feet, resulting in a disproportionate body habitus. There is no specific ICD-10-CM code for lipedema. Deposition of lipedema fat increases with stage and body mass index (BMI) and likely involves sex hormones during times when weight is gained (puberty, pregnancy and menopause). Lipedema is inherited in 60% of women likely through genes affecting microvessels resulting in excess fluid bound to glycosaminoglycans in the interstitial space. Unique to lipedema is fat that is highly resistant to loss by diet, exercise, or bariatric surgery. Lipedema is often confused with secondary obesity or lymphedema. Women with lipedema and/or obesity can develop lymphedema called lipolymphedema, for which there is no ICD-10-CM code.

There is no cure for lipedema, but treatments aimed at reducing the lymphedema component of lipedema such as manual decongestive therapy, wrapping, exercise, compression garments and pumps, and some medical foods and medications are helpful. Expertly performed suction assisted lipectomy is the treatment of choice for suitable lipedema patients with an inadequate response to conservative and supportive measures. Lipedema is thought to affect 11% of the female population.

Lymphedema is a chronic and progressive swelling caused by a low output failure of the lymphatic system, resulting in the development of a high-protein edema in the tissues. Lymphedema is a lifelong condition for which no cure exists. Lymphedema can be either primary (hereditary) or secondary. Secondary lymphedema is the most common cause of the disease and affects approximately 1 in 1000 Americans. Complications of lymphedema include recurrent bouts of cellulitis and/or lymphangitis, bacterial and fungal infections, lymphangio-adenitis, deep venous thrombosis, poor wound healing, leg ulcers, severe functional impairment, disability, and necessary amputation. Patients with chronic lymphedema for 10 years have a 10% risk of developing lymphangiosarcoma. Praecox lymphedema is currently captured in ICD-10-CM as a secondary lymphedema; it is more accurately classified under code Q82.0: Hereditary lymphedema.

With support from the American Vein & Lymphatic Society (AVLS), the requestor is submitting the following modifications to identify and track lipedema and lipolymphedema patients.

References

1Allen, E. V., and Hines, E. A. J. (1940) Lipedema of the legs: A syndrome characterized by fat legs and orthostatic edema.. Proc Staff Meet Mayo Clin 15, 184-187

Wold, L. E., Hines, E. A., Jr., and Allen, E. V. (1951) Lipedema of the legs; a syndrome characterized by fat legs and edema. Ann Intern Med. 34, 1243-1250.

Cornely M. Lipoedema of arms and legs. Part 2: Conservative and surgical therapy of the lipoedema, Lipohyper- plasia dolorosa. Phlebologie 2011; 40:146-151.

Herbst K, Mirkovskaya L, Bharhagava A, Chava Y, Te CH. Lipedema Fat and Signs and Symptoms of Illness, Increase with Advancing Stage. Archives of Medicine. 2015;7(4:10):1-8.

Herbst KL. Subcutaneous Adipose Tissue Diseases: Dercum Disease, Lipedema, Familial Multiple Lipomatosis and Madelung Disease. In: Purnell J, Perreault L, eds. Endotext. Massachusetts: MDText.com; 2019.

Bast JH, Ahmed L, Engdahl R. Lipedema in patients after bariatric surgery. Surg Obes Relat Dis. 2016;12(5):1131-1132. doi: 1110.1016/j.soard.2016.1104.1013. Epub 2016 Apr 1114.

Pouwels S, Huisman S, Smelt HJM, Said M, Smulders JF. Lipoedema in patients after bariatric surgery: report of two cases and review of literature. Clin Obes. 2018;8(2):147-150. doi: 110.1111/cob.12239. Epub 12018 Jan 12225.

Pouwels S, Smelt HJ, Said M, Smulders JF, Hoogbergen MM. Mobility Problems and Weight Regain by Misdiagnosed Lipoedema After Bariatric Surgery: Illustrating the Medical and Legal Aspects. Cureus. 2019;11(8):e5388. doi: 5310.7759/cureus.5388.

Halk AB, Damstra RJ. First Dutch guidelines on lipedema using the international classification of functioning, disability and health. Phlebology. 2017;32(3):152-159

TABULAR MODIFICATIONS

E88 Other and unspecified metabolic disorders

New subcategory E88.2 Lipomatosis, not elsewhere classified

Delete ~~Lipomatosis NOS~~

Delete ~~Lipomatosis (Check) dolorosa [Dercum]~~

New code E88.21 Dercum disease
Add Lipomatosis dolorosa

New code E88.29 Lipomatosis NOS

E88.8 Other specified metabolic disorders

New subcategory E88.82 Lipedema

New code E88.821 Lipedema, Stage 1

New code E88.822 Lipedema, Stage 2

New code E88.823 Lipedema, Stage 3

New code E88.829 Lipedema NOS

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

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

I89 Other noninfective disorders of lymphatic vessels and lymph nodes

I89.0 Lymphedema, not elsewhere classified

Delete ~~Praecox lymphedema~~

New code I89.3 Lipolymphedema

Q82 Other congenital malformations of skin

Q82.0 Hereditary lymphedema

Add Praecox lymphedema

Lumbar and Lumbosacral Intervertebral Annular Fibrosis Defects

Intrinsic Therapeutics is proposing the creation of new ICD-10-CM diagnosis codes for describing large and small annular defects following discectomy within the lumbar and lumbosacral regions, consistent with policy guidance from the International Society for the Advancement of Spine Surgery (ISASS). Patient outcomes following lumbar and lumbosacral discectomy vary based on the presence and size of annular defects. Thus, it is important to measure and identify the size of the annular defect and determine if additional treatment may be needed to close the annular defect.

At present, research has shown clinical efficacy in closing and repairing large annular defects using an annular closure device. New codes to identify the size of the annular defect would help the clinical community, and support compliance with treatment guidelines (e.g., see ISASS Policy Guidelines) [(1)]. In addition, detailed coding describing both large and small annular defects will allow researchers and clinicians to track and further assess patient outcomes following lumbar/lumbosacral discectomy as well as outcomes associated with the treatment of large annular defects with bone-anchored annular closure devices.

Lumbar discectomy is the surgical standard for treating debilitating chronic pain caused by lumbar disc herniation (LDH) [(2)]. During discectomy, the affected vertebrae and disc are exposed, and the disc that has herniated is surgically removed to decompress the spinal cord and any impinged nerves. In some cases, the annulus fibrosus is ruptured during the herniation event. If the patient has a large annular defect, the remaining nucleus pulposus in the intervertebral space may reherniate through the annular defect.

Following lumbar discectomy, symptom recurrence related to reherniation is reported in 7 to 18% of patients [(3)]. Symptomatic lumbar disc reherniation is associated with poor clinical outcomes and often requires a technically demanding reoperation [(3)]. Patients with large postsurgical annular defects have a higher risk of symptom recurrence and reoperation compared to those with small defects – as much as 2.5 times greater.

Currently, there are no existing ICD-10-CM codes that are specific for small or large annular defects. New ICD-10-CM codes are needed to accurately describe both small (< 6 mm wide and < 4 mm high) and large (≥ 6 mm wide and ≥ 4 mm high) annular defects in patients with lumbar or lumbosacral disc herniation who have undergone discectomy surgery.

References

1. Lorio M, Kim C, Arachi A, Inzana J, Yue JJ. International Society for the Advancement of Spine Surgery Policy 2019—Surgical Treatment of Lumbar Disc Herniation with Radiculopathy. Int J Spine Surg. 2020 Feb; 14(1): 1–17. https://doi.org/10.14444/7001
2. Heindel P, Tuchman A, Hsieh PC, et al. Reoperation Rates After Single-level Lumbar Discectomy. *Spine.* 2017;42(8):E496-E501. https://doi.org/10.1097/BRS.0000000000001855
3. Miller L, McGirt MJ, Garfin SR, Bono CM. Association of Annular Defect Width After Lumbar Discectomy with Risk of Symptom Recurrence and Reoperation. *Spine*. 2018; Mar 1;43(5):E308-E315. https://doi.org/10.1097/BRS.0000000000002501

Ammerman J, Watters WC, Inzana JA, et al. Closing the Treatment Gap for Lumbar Disc Herniation Patients with Large Annular Defects: A Systematic Review of Techniques and Outcomes in this High- risk Population. *Cureus.* 2019;11:e4613. https://doi.org/10.7759/cureus.4613

Arts MP, Kursumovic A, Miller LE, et al. Comparison of treatments for lumbar disc herniation: Systematic review with network meta-analysis. *Medicine (Baltimore).* 2019;98:e14410. http://dx.doi.org/10.1097/MD.0000000000014410

Bouma GJ, Ardeshiri A, Miller LE, et al. Clinical performance of a bone-anchored annular closure device in older adults. Clin Interv Aging. 2019;14:1085-1094. https://doi.org/10.2147/CIA.S208098

Carragee EJ, Han MY, Suen PW, Kim D. Clinical outcomes after lumbar discectomy for sciatica: the effects of fragment type and anular competence. J Bone Joint Surg Am. 2003 Jan;85(1):102-8. PMID: 12533579

Cho PG, Shin DA, et al. Efficacy of a novel annular closure device after lumbar discectomy in Korean patients: a 24-month follow-up of a randomized controlled trial. J Korean Neurosurg Soc. 2019;62. https://doi.org/10.3340/jkns.2019.0071

Choy WJ, Phan K, Diwan AD, et al. Annular closure device for disc herniation: meta-analysis of clinical outcome and complications. *BMC Musculoskelet Disord.* 2018;19:290. https://doi.org/10.1186/s12891-018-2213-5

Kienzler JC, Klassen PD, Miller LE, et al. Three-year results from a randomized trial of lumbar discectomy with annulus fibrosus occlusion in patients at high risk for reherniation. Acta Neurochir (Wien). 2019;161:1389-1396. https://doi.org/10.1007/s00701-019-03948-8

Klassen PD, Lesage G, Miller LE, et al. Reoperation After Primary Lumbar Discectomy with or without Implantation of a Bone-Anchored Annular Closure Device: Surgical Strategies and Clinical Outcomes. World Neurosurg. 2019;130:e926-e932. https://doi.org/10.1016/j.wneu.2019.07.038

Miller LE, Allen RT, Duhon B, et al. Expert review with meta-analysis of randomized and nonrandomized controlled studies of Barricaid annular closure in patients at high risk for lumbar disc reherniation. Expert Rev Med Devices. 2020:1-9.

Nanda D, Arts MP, Miller LE, et al. Annular closure device lowers reoperation risk 4 years after lumbar discectomy. Med Devices (Auckl). 2019;12:327-335. https://doi.org/10.2147/MDER.S220151

Thome C, Klassen PD, Bouma GJ, et al. Annular closure in lumbar microdiscectomy for prevention of reherniation: a randomized clinical trial. Spine J. 2018;18:2278-2287. https://doi.org/10.1016/j.spinee.2018.05.003

TABULAR MODIFICATIONS

M51 Thoracic, thoracolumbar, and lumbosacral intervertebral disc disorders

New subcategory	M51.A	Other lumbar and lumbosacral annular disc defects
New code	M51.A1	Intervertebral annular fibrosis defect, small, lumbar region
Add		Intervertebral annular fibrosis defect, < 6 mm wide and < 4 mm high, lumbar region
Add		Code first lumbar disc herniation (M51.06, M51.16, M51.26)
New code	M51.A2	Intervertebral annular fibrosis defect, large, lumbar region
Add		Intervertebral annular fibrosis defect, ≥ 6 mm wide or ≥ 4 mm high, lumbar region
Add		Code first lumbar disc herniation (M51.06, M51.16, M51.26)
New code	M51.A3	Intervertebral annular fibrosis defect, small, lumbosacral region
Add		Intervertebral annular fibrosis defect, < 6 mm wide and < 4 mm high, lumbar region
Add		Code first lumbosacral disc herniation (M51.17, M51.27)

New code	M51.A4	Intervertebral annular fibrosis defect, large, lumbosacral region
Add		Intervertebral annular fibrosis defect, ≥ 6 mm wide or ≥ 4 mm high, lumbar region
Add		Code first lumbosacral disc herniation (M51.17, M51.27)

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	Defect
Add	- intervertebral annular fibrosis (see also Disease, intervertebral disc, by site) M51.9
Add	- - lumbar M51.86
Add	- - - large M51.A2
Add	- - - small M51.A1
Add	- - lumbosacral M51.87
Add	- - - large M51.A4
Add	- - - small M51.A3

Mild Cognitive Disorder Due to Known Physiological Conditions

Cognitive impairment related to aging occurs on a continuum ranging from the typical changes related to normal aging to cognitive deficits that exceed those expected given a person's age but yet not so severe as to be considered a dementia, and finally deficits of sufficient severity to warrant a dementia diagnosis.

Similarly, degenerative diseases of the nervous system typically evolve over time so that there may be a period of asymptomatic histopathological changes to a period of mild cognitive impairment (often protracted) on the way to the development of overt dementia. In recent years there has been great interest in identifying and potentially treating individuals during this pre-dementia period with the hope that clinical interventions might prevent the progression of the underlying illness. The American Psychiatry Association are requesting a new code subcategory and code expansion to capture this information.

Background: At the September 30, 2005 meeting of the ICD-9-CM Coordination and Maintenance Committee meeting, the American Academy of Neurology proposed the addition of a new code for mild cognitive impairment (MCI). In their proposal, they defined MCI as "a disease entity defined by an impairment in memory (or any other cognitive domain) that is beyond what is normal for age, with relatively intact function in the other domains." In explaining the need for this new code, they noted that using the standard set of criteria for MCI [(1)] patients progress to dementia at a rate of approximately 12% per year and when followed up at 6 years, approximately 80% of them will have converted to dementia, suggesting that this diagnosis identifies mildly cognitively impaired patients at high risk of developing dementia [(2)]. This rate was in marked distinction to incidence rates from a similar community progression rate of 1-2% per year and at the time this proposal was made, the underlying etiology of cases of MCI that progressed to dementia was presumed to be Alzheimer's disease [(3)].

Over the past fifteen years, presentations of mild cognitive impairment related to neurodegenerative diseases other than Alzheimer's disease as well as to other diseases in ICD-10-CM have garnered increased clinical and research interest, including MCI due to vascular disease [(4)], due to frontotemporal degeneration [(5)], due to HIV disease [(6),] due to Lewy body disease [(7)], due to traumatic brain injury [(8),] due to Parkinson's disease [(9)] and due to Huntington's disease [(10)]. However, there is currently no ICD-10-CM code for cases of mild cognitive disorder due to other medical conditions.

The American Psychiatric Association (APA) is proposing a new subcategory for "Mild cognitive disorder due to a known physiological condition" at code category F06, Other mental disorders due to known physiological condition.

This proposal is being modeled after F02.8, Dementia in diseases classified elsewhere, with a coding note instruction to "Code first the underlying physiological condition" in order to allow for the

specification of the underlying pathologic condition. The conditions listed under F02.8 is to be included as well, since the same conditions that can cause dementia can also cause mild cognitive disorder. It is also being proposed to use a modified version of the excludes1 note that is currently under G31.84, Mild cognitive impairment, so stated, since most of these are also applicable to proposed new code (F06.7-).

Finally, it is being proposed to include the provision of a 5th digit to indicate the presence (or absence) of a behavioral disturbance, a provision which is also modelled after F02.8. This new provision offers an important opportunity for the clinical documentation of progression of behavioral symptoms that have been increasingly recognized as a highly significant indicator of progression of the underlying disease along the continuum. (11,12).

APA is also recommending that G31.84, Mild cognitive impairment, so stated, be retained but that it apply only to cases of mild cognitive impairment which are presumed to be due to a medical etiology, but for which the etiology is currently uncertain or unknown. It is recommended to revise the code title of G31.84 from "Mild cognitive impairment, so stated" to "Mild cognitive impairment <u>of uncertain or unknown etiology</u>.".

References:

1. Petersen RC, Smith GE, Waring SC, Ivnik RJ, Tangalos EG, Kokmen E Mild cognitive impairment: clinical characterization and outcome. Arch Neurol. 1999 Mar;56(3):303-8.
2. Petersen RC, Morris JC. Clinical features. In: Petersen RC, ed. Mild Cognitive Impairment: Aging to Alzheimer's Disease. New York: Oxford University Press, Inc., 2003; 15–40
3. Petersen, RC. Mild cognitive impairment as a diagnostic entity. Journal of Internal medicine (2004) 256:183-194.
4. Al-Qazzaz NK, Ali SH, et al: Cognitive impairment and memory dysfunction after a stroke diagnosis: a post-stroke memory assessment. Neuropsychiatric Disease and Treatment 2014:10 1677–1691
5. de Mendonça A, Ribeiro F, et al., Frontotemporal Mild Cognitive Impairment J Alzheimers Dis. 2004 Feb;6(1):1-9.
6. Sheppard DP, Iudicello JE, et al. Elevated rates of mild cognitive impairment in HIV disease. J Neurovirol. 2015 Oct; 21(5): 576–584.
7. Goldman JG, Williams-Gray C, et. al, The spectrum of cognitive impairment in Lewy body diseases Mov Disord. 2014 Apr 15; 29(5): 608–621.
8. LoBoe C, Denney D, et al., Self-Reported Traumatic Brain Injury and Mild Cognitive Impairment: Increased Risk and Earlier Age of Diagnosis J Alzheimers Dis. 2016;51(3):727-36.
9. Litvan I, Goldman J, Troster A, Schmand B, Weintraub D, Petersen R, et al. Diagnostic criteria for mild cognitive impairment in Parkinson's disease: Movement Disorder Society Task Force Guidelines. Movement Disorders. 2012;27(3):349-56.
10. Duff K, Paulsen J, et al., Mild cognitive impairment in prediagnosed Huntington disease Neurology. 2010 Aug 10; 75(6): 500–507.
11. Geda YE, Roberts RO, Knopman DS, Petersen RC, Christianson TJ, Pankratz VS, Smith GE, Boeve BF, Ivnik RJ, Tangalos EG, Rocca WA. Prevalence of neuropsychiatric symptoms in mild cognitive impairment and normal cognitive aging: population-based study. Arch Gen Psychiatry. 2008 Oct;65(10):1193-8
12. Geda YE, Schneider LS, Gitlin LN, Miller DS, Smith GS, Bell J, Evans J, Lee M, Porsteinsson A, Lanctôt KL, Rosenberg PB, Sultzer DL, Francis PT, Brodaty H, Padala PP, Onyike CU, Ortiz LA, Ancoli-Israel S, Bliwise DL, Martin JL, Vitiello MV, Yaffe K, Zee PC, Herrmann N, Sweet RA, Ballard C, Khin NA, Alfaro C, Murray PS,

Schultz S, Lyketsos CG, Neuropsychiatric symptoms in Alzheimer's disease: past progress and anticipation of the future. Alzheimers Dement. 2013 Sep;9(5):602-8.

TABULAR MODIFCATIONS

	F06	Other mental disorders due to known physiological condition
New subcategory	F06.7	Mild cognitive disorder due to known physiological condition
Add		Mild cognitive impairment due to a known physiological condition
Add		Mild neurocognitive disorder due to a known physiological condition
Add		Code first the underlying physiological condition, such as:
Add		Alzheimer's (G30.-)
Add		cerebral lipidosis (E75.4)
Add		Creutzfeldt-Jakob disease (A81.0-)
Add		epilepsy and recurrent seizures (G40.-)
Add		frontotemporal dementia (G31.09)
Add		hepatolenticular degeneration (E83.0)
Add		human immunodeficiency virus [HIV] disease (B20)
Add		Huntington's disease (G10)
Add		hypercalcemia (E83.52)
Add		hypothyroidism, acquired (E00-E03.-)
Add		intoxications (T36-T65)
Add		Jakob-Creutzfeldt disease (A81.0-)
Add		Lewy body disease (G31.83)
Add		multiple sclerosis (G35)
Add		neurosyphilis (A52.17)
Add		niacin deficiency [pellagra] (E52)
Add		Parkinson's disease (G20)
Add		Pick's disease (G31.01)
Add		polyarteritis nodosa (M30.0)
Add		prion disease (A81.9)
Add		systemic lupus erythematosus (M32.-)
Add		traumatic brain injury (S06.-)
Add		trypanosomiasis (B56.-, B57.-)
Add		vitamin B deficiency (E53.8)
Add		Excludes1: age related cognitive decline (R41.81)
Add		altered mental status (R41.82)
Add		cerebral degeneration (G31.9)
Add		change in mental status (R41.82)
Add		cognitive deficits following (sequelae of) cerebral hemorrhage or infarction (I69.01-I69.11-, I69.21-I69.31-, I69.81-I69.91-)

Add — cognitive impairment due to intracranial or head injury (S06.-)
Add — dementia (F01.-, F02.-, F03)
Add — mild cognitive impairment due to unknown or unspecified etiology (G31.84)
Add — neurologic neglect syndrome (R41.4)
Add — personality change, nonpsychotic (F68.8)

New code — F06.70 Mild cognitive disorder due to known physiological condition without behavioral disturbance

New code — F06.71 Mild cognitive disorder due to known physiological condition with behavioral disturbance

G31 Other degenerative diseases of nervous system, not elsewhere classified
For codes G31.0-G31.83, G31.85-G31.9, use additional code to identify:
dementia with behavioral disturbance (F02.81)
dementia without behavioral disturbance (F02.80)

G31.8 Other specified degenerative diseases of nervous system

Revise — G31.84 Mild cognitive impairment of <u>uncertain or unknown etiology</u>, ~~so stated~~
Revise — Mild neurocognitive disorder of <u>uncertain or unknown etiology</u>

Add — Excludes1: mild neurocognitive disorder due to a known physiological condition (F06.7-)
Delete — ~~mild memory disturbance (F06.8)~~

Moisture Associated Skin Damage

The Wound Ostomy and Continence Nurses Society is proposing the creation of new codes for moisture associated skin damage. This proposal was originally presented at the March 2020 Coordination and Maintenance (C&M) meeting. Based on comments received during the public comment period, a revised proposal is being presented for consideration.

Among its multiple vital functions, the skin acts as a barrier to protect the body against mechanical trauma, noxious irritants, infectious pathogens and excessive fluids. Overexposure of the skin to moisture can compromise the integrity of the skin's epithelial barrier, disrupting the intricate molecular arrangement of intercellular lipids in the stratum corneum, the intercellular connections between epidermal cells (corneocytes) and the cutaneous microbiome. Once damaged, the skin is more permeable and susceptible to irritant penetration, leading to inflammation or dermatitis. The term moisture-associated skin damage (MASD) delineates a spectrum of injury characterized by the inflammation and erosion (or denudation) of the epidermis resulting from exposure to various sources of moisture and potential irritants (e.g. urine, stool, saliva or respiratory secretions and stoma or fistula secretions. With a shift in demographics toward an aging population worldwide, MASD is a common condition and its prevalence is likely to rise.

Moisture-associated skin damage is a complex, heterogenous condition, which includes irritant contact dermatitis (persistent erythema with or without erosion of superficial skin layers). Both urinary and fecal incontinence are known to cause irritant contact dermatitis on any portion of the skin. The term fecal, urinary or dual incontinence applies to adults and older children after successful toilet training. An individual with incontinence can have irritant contact dermatitis without wearing an absorbent product, such as a diaper. Irritant contact dermatitis has been shown to be an independent risk factor for full thickness pressure ulceration.

Similarly, skin around the stoma or fistula exposed to urinary, fecal or fistula secretions is at risk for irritant contact dermatitis impairing the efficacy of pouching systems and exposing the skin to other forms of damage. Saliva or respiratory secretions from the mouth, nose, a tracheostomy or a spit fistula may cause irritant contact dermatitis on nearby skin. Unique ICD-10-CM codes would improve data collection and facilitate research.

The following new codes are being requested to differentiate these conditions for data collection and research.

TABULAR MODIFICATIONS

	L24	Other and unspecified dermatitis
New subcategory	L24.A	Irritant contact dermatitis due to friction or contact with body fluids
Add		Excludes1: irritant contact dermatitis related to stoma or fistula (L24.B-)
Add		Excludes2: erythema intertrigo (L30.4)

New code	L24.A0	Irritant contact dermatitis due to friction or contact with body fluids, unspecified
New code	L24.A1	Irritant contact dermatitis due to saliva
New code	L24.A2	Irritant contact dermatitis due to fecal, urinary or dual incontinence
Add		Excludes1: diaper dermatitis (L22)
New code	L24.A9	Irritant contact dermatitis due friction or contact with other specified body fluids
Add		Wound fluids, exudate
New subcategory	L24.B	Irritant contact dermatitis related to stoma or fistula
Add		Use additional code to identify any artificial opening status (Z93.-), if applicable, for contact dermatitis related to stoma secretions
New code	L24.B0	Irritant contact dermatitis related to unspecified stoma or fistula
Add		Irritant contact dermatitis related to stoma NOS
Add		Irritant contact dermatitis related to fistula NOS
New code	L24.B1	Irritant contact dermatitis related to digestive stoma or fistula
Add		Irritant contact dermatitis related to gastrostomy
Add		Irritant contact dermatitis related to ileostomy
Add		Irritant contact dermatitis related to saliva or spit fistula
New code	L24.B2	Irritant contact dermatitis related to respiratory stoma or fistula
Add		Irritant contact dermatitis related to endotracheal tube
Add		Irritant contact dermatitis related to tracheostomy
New code	L24.B3	Irritant contact dermatitis related to fecal or urinary stoma or fistula
Add		Irritant contact dermatitis related to colostomy
Add		Irritant contact dermatitis related to ileostomy
Add		Irritant contact dermatitis related to enterocutaneous fistula

Multisystem Inflammatory Syndrome (MIS)

In April 2020, clinicians began recognizing a hyperinflammatory syndrome associated with past or present COVID-19 (SARS-CoV-2 infection). This syndrome has since been recognized in numerous locations around the world, including the United States.[1-6] In the United States, it is termed Multisystem Inflammatory Syndrome (MIS) and been predominantly described in children with less frequent case reports in adults. It has also been called Pediatric Inflammatory Multisystem Syndrome.[3]

On May 14, 2020, CDC released a Health Advisory to alert clinicians to MIS in children, provide background and a case definition, and recommended that healthcare providers report any patient who meets the case definition to local, state, and territorial health departments.[7] It is not clear why certain people develop MIS, although it appears that the significant systemic inflammation is linked to antecedent SARS-CoV-2 infection.

The current U.S. CDC case definition for MIS in children includes fever, laboratory markers of inflammation, severe illness requiring hospitalization with at least 2 organ systems involved and laboratory evidence of SARS-CoV-2 infection (by RT-PCR, serology, or antigen test) or a history of known exposure to a suspected or confirmed COVID-19 case within 4 weeks prior to symptom onset.

Given that this is a new condition, the true burden is unknown. While coding advice has been issued for this disorder, it is possible some cases may have been misdiagnosed or misclassified as other syndromes that share some clinical features (e.g., Kawasaki Disease, sepsis, or toxic shock syndrome). Surveillance for MIS is important for a number of reasons, including but not limited to defining the burden of severe SARS-CoV-2 infection, including this post-infectious inflammatory phase; assessing potential demand for therapeutics for MIS; and monitoring MIS as both an adverse event and related to potential vaccine effectiveness outcome in future once a SARS-CoV-2 vaccine has been developed.

Interim coding algorithms have been developed that advise on how MIS should be coded, but these are not sufficiently specific for MIS surveillance. CDC experts have proposed that the implementation date for this code be accelerated to the earliest date possible. A shorter timeline for implementation will ensure that surveillance for MIS can be ongoing in real time as the pandemic progresses. In addition, this will enable surveillance of MIS before and after vaccines are introduced to the market to be able to assess safety of vaccinations. **The implementation date is expected to be in January 2021. The comment deadline will be Oct. 9, 2020.**

References

1. Feldstein LR, Rose EB, Horwitz SM, et al. Multisystem Inflammatory Syndrome in U.S. Children and Adolescents. *N Engl J Med.* 2020; *383*:334-346. https://doi.org/10.1056/NEJMoa2021680
2. Dufort EM, Koumans EH, Chow EJ, et al. Multisystem Inflammatory Syndrome in Children in New York State. *N Engl J Med.* 2020; *383*:347-58. https://doi.org/10.1056/NEJMoa2021756.
3. Whittaker E, Bamford A, Kenny J, et al. Clinical Characteristics of 58 Children With a Pediatric Inflammatory Multisystem Syndrome Temporally Associated With SARS-CoV-2. *JAMA.* 2020; *324*(3):259-269. https://doi.org/10.1001/jama.2020.10369
4. Riphagen S, Gomez X, Gonzalez-Martinez C, Wilkinson N, Theocharis P. Hyperinflammatory shock in children during COVID-19 pandemic. *Lancet.* 2020; *395*(10237):1607-1608. https://doi.org/10.1016/S0140-6736(20)31100-4
5. Verdoni L, Mazza A, Gervasoni A, et al. An outbreak of severe Kawasaki-like disease at the Italian epicentre of the SARS-CoV-2 epidemic: an observational cohort study. *Lancet.* 2020; *395*(10239):1771-1778. https://doi.org/10.1016/S0140-6736(20)31103-X

6. Godfred-Cato S, Bryant B, Leung J, et al. COVID-19–Associated Multisystem Inflammatory Syndrome in Children — United States, March–July 2020. *MMWR Morb Mortal Wkly Rep*. 2020; *69*(32):1074-1080. https://www.cdc.gov/mmwr/volumes/69/wr/mm6932e2.htm?s_cid=mm6932e2_w.
7. CDC. Health Alert Network: Multisystem Inflammatory Syndrome in Children (MIS-C) Associated with Coronavirus Disease 2019 (COVID-19). 14 May 2020; https://emergency.cdc.gov/han/2020/han00432.asp.

TABULAR MODIFICATIONS

M35 Other systemic involvement of connective tissue

M35.8 Other specified systemic involvement of connective tissue

New code	M35.81	Multisystem inflammatory syndrome
Add		MIS-C
Add		Multisystem inflammatory syndrome in children
Add		Pediatric inflammatory multisystem syndrome
Add		PIMS
Add	Code first, if applicable, COVID-19 (U07.1)	
Add	Code also, if applicable, exposure to SARS-CoV-2 infection (Z20.822)	
Add	Use additional code(s) to identify any associated complications such as:	
Add	acute respiratory distress syndrome (J80)	
Add	acute hepatic failure (K72.0-)	
Add	acute kidney failure (N17)	
Add	acute myocarditis (I40)	
Add	cardiac arrhythmia (I49.9)	
Add	other viral pneumonia (J12.89)	
Add	severe sepsis (R65.2-)	
Add	viral pericarditis (B33.23)	
Add	viral cardiomyopathy (B33.24)	
New code	M35.89 Other specified systemic involvement of connective tissue	

Niemann-Pick Disease (NPD) Type A/B

The following proposal requests an update to the ICD-10-CM codes for Niemann-Pick category (E75.24) to add a specific diagnosis for Type A/B and to recognize another name, acid sphingomyelinase deficiency (ASMD), based on the biology of the enzyme deficit. This proposal was submitted by the Niemann-Pick Disease Foundation, Inc. (NNPDF) and Genzyme Corporation, a Sanofi company.

Acid sphingomyelinase deficiency (ASMD) is a rare, and potentially life-threatening lysosomal storage disorder.[1] Patients with ASMD have variable impairment in sphingomyelin metabolism due to pathogenic variants in *SMPD1*, the gene encoding acid sphingomyelinase (ASM), resulting in reduced ASM enzyme activity.[1] As ASM catalyzes the hydrolysis of sphingomyelin to ceramide and phosphocholine, reduced ASM activity results in the progressive lysosomal accumulation of sphingomyelin mostly within cells of the monocyte/macrophage lineage that reside in reticuloendothelial tissues, namely in the spleen, liver, lung, bone marrow, and lymph nodes. With severe disease, neurons may also be affected.[1,2]

ASMD is historically known as Niemann-Pick Disease (NPD), although the nomenclature is evolving. "ASMD" rather than "NPD" is predominating as the preferred term, as evidenced by a recent international consensus guideline for ASMD diagnosis[2] and the nomenclature used by patient support organizations including the National Organization for Rare Disorders (NORD).[3]

ASMD is an autosomal recessive single gene disease. It is known to generate a spectrum of phenotypes, which have been classified as Type A, Type B, and Type A/B. ASMD Type A is the early onset and acute neuropathic form of ASMD and results in failure to thrive, hepatosplenomegaly, rapidly progressive neurological degeneration, and death, usually before the age of 3 years.[1] ASMD Type B is usually diagnosed after the age of 2 years, after hepatosplenomegaly (the most common disease manifestation in all ASMD patients) is observed. It presents a slower progression with little or no neurological involvement. Other more variable features include liver dysfunction, pulmonary disease, retinal stigmata, and growth delays.

Patients can survive into adulthood. ASMD Type A/B includes patients with disease manifestations intermediate to Type A and Type B.[1] These patients may develop neurologic symptoms during childhood and can have dominant neurodegenerative and/or visceral manifestations aspects of ASMD with increased mortality.[2]

ASMD has an estimated birth prevalence of 0.4-0.6 per 100,000.[4] The ASMD nomenclature differentiates Niemann-Pick types A, B, and A/B from Niemann-Pick types C[5] and D, which are separate diseases.[6,7] With NPD Types C and D, although patients **do not** have a deficit of acid sphingomyelinase (ASM), they are unable to metabolize cholesterol and other lipids within the cell.[6,7]

Furthermore, the current lack of a specific Type A/B ICD-10-CM code results in clinicians needing to choose either Type A or B or an unspecified code, which does not accurately portray the varying disease burden of the patient and can result in untold emotional stress on patients and families who often remain misdiagnosed for years.

As the underlying defect and clinical spectrum of each type has been further understood for the group of Niemann-Pick Diseases, classification should be updated to reflect this evolution. Therefore, subcategorization of the specific Niemann-Pick diseases as ASMD Types A, B and Type A/B, via a diagnostic specific code, will allow further characterization of the clinical manifestations and the risks associated with each ASMD type. This knowledge can improve the diagnostic and treatment paradigms and improve treatment practices. Finally, these proposed changes maintain the link to Niemann-Pick disease, which is important from a historical perspective.

The American Academy of Pediatricians (AAP) and the American College of Medical Genetics and Genomics (ACMG) have reviewed and support the proposal.

1 McGovern MM, et al. Disease manifestations and burden of illness in patients with acid sphingomyelinase deficiency (ASMD). Orphanet Journal of Rare Diseases. 2017.
2 McGovern MM, et al. Consensus recommendation for a diagnostic guideline for acid sphingomyelinase deficiency. Genetics in medicine. Volume 19(9); September 2017.
3 https://rarediseases.org/rare-diseases/acid-sphingomyelinase-deficiency/. Accessed May 18, 2020. 4 Kingma SD, Bodamer OA, Wijburg FA. Best Pract Res Clin Endocrinol Metab. 2015;29(2):145–157.
5 https://www.mayoclinic.org/diseases-conditions/niemann-pick/symptoms-causes/syc-20355887. Accessed May 18, 2020.
6 https://nnpdf.org/overview/#NPC. Accessed May 18, 2020.
7 Greer WL, et al. The Nova Scotia (Type D) Form of Niemann-Pick Disease Is Caused by a G3097rT Transversion in NPC1. Am. J. Hum. Genet. 1998;63:52–54.

TABULAR MODIFICATIONS

E75 Disorders of sphingolipid metabolism and other lipid storage disorders

Excludes1: mucolipidosis, types I-III (E77.0-E77.1)

Refsum's disease (G60.1)

E75.2 Other sphingolipidosis

Excludes1: adrenoleukodystrophy [Addison-Schilder] (E71.528)

E75.21 Fabry (-Anderson) disease

E75.22 Gaucher disease

E75.23 Krabbe disease

E75.24 Niemann-Pick disease

E75.240 Niemann-Pick disease type A
Add Acid Sphingomyelinase Deficiency Type A (ASMD Type A)
Add Infantile neurovisceral

E75.241 Niemann-Pick disease type B
Add Acid Sphingomyelinase Deficiency Type B (ASMD Type B)
Add Chronic visceral

E75.242 Niemann-Pick disease type C

E75.243 Niemann-Pick disease type D

New code E75.244 Niemann-Pick disease type A/B
Add Acid Sphingomyelinase Deficiency Type A/B (ASMD Type A/B)
Add Chronic neurovisceral

E75.248 Other Niemann-Pick disease

E75.249 Niemann-Pick disease, unspecified

Nocturnal Polyuria

Nocturia is waking up at night to void. There are many potential causes, such as small bladder, BPH, making too much urine, overactive bladder, neurogenic bladder, poor sleeping habits. Treatments are varied, from addressing BPH or relaxing the bladder to behavioral changes to nerve modulation, sleeping aids, etc.

Polyuria is making too much urine but is not specific for time; one would assume day and night. Often this is due to drinking too many fluids or having diabetes.

Nocturnal polyuria is more specifically when the polyuria is only at night. There is not polyuria during the day. This may be due to fluid reabsorption or an oxytocin issue. The issue is that a person makes so much urine at night (> 1L to > 1.5L) that they need to wake up a number of times, but, in the absence of a neurogenic bladder, small bladder, BPH, overactive bladder or poor sleeping habits. There are specific medications to treat nocturnal polyuria, which are not used to treat other forms of nocturia or polyuria.

The specific symptom of nocturnal polyuria can be differentiated as being more specific and often not overlapping specifically with nocturia or polyuria.

The American Urological Association is requesting new codes to differentiate between nocturia which is caused by over active bladder and causes a patient to get up to go the bathroom frequently during the night versus nocturnal polyuria which causes the bladder to produce more urine than it can hold and causes a patient to get up during night to go to the bathroom.

TABULAR MODIFICATIONS

R35 Polyuria

New subcategory	R35.8	Other polyuria
Delete		~~Polyuria NOS~~
New code	R35.81	Nocturnal polyuria
New code	R35.89	Other polyuria
Add		Polyuria NOS

Non-Radiographic Axial Spondyloarthritis

Non-radiographic axial spondyloarthritis (nr-axSpA) is a potentially severe, chronic inflammatory arthritis that entails significant back pain and impairment in spinal mobility and physical function. It primarily affects the joints within the trunk (spine, chest, and pelvis). It causes unyielding non-mechanical back pain and progressive diminishing of spinal mobility and physical function. In contrast to mechanical back pain, the inflammatory back pain of non-radiographic axial spondyloarthritis is chronic, progressive, worse at night, and exacerbated after periods of immobility.

Non-radiographic axial spondyloarthritis is not the same as ankylosing spondylitis, although both are types of axial spondyloarthritis (axSpA). Clinically, spondyloarthritis (SpA) sits at the top of the established disease taxonomy. Then, axial spondyloarthritis (dorsal pathologies) stands next, above two distinct diseases. The two distinct types of axial spondyloarthritis patients are:

1. Non-Radiographic Axial Spondyloarthritis (nr-axSpA). These patients do not have definitive X-ray evidence identifying structural changes to their sacroiliac joints.
2. Ankylosing Spondylitis (AS). These patients have structural changes in the sacroiliac joints evidenced on an X-ray.

Some clinical differences between these are highlighted as follows. Non-Radiographic Axial Spondyloarthritis (Nr-AxSpA) has no radiographic (x-ray) evidence of sacroiliitis; elevated C-reactive protein level is less common; it is more common in women; HLA-B27 genetic variant is less common; and peripheral inflammation (arthritis, enthesitis) is more common. In contrast, Ankylosing Spondylitis (AS) has X-ray evidence of sacroiliitis required for diagnosis; elevated C-reactive protein is commonly seen; it is more common in men; HLA-B27 genetic variant is extremely common; and peripheral inflammation is less common

In axial spondyloarthritis (axSpA), inflammation usually starts in the sacroiliac joints [(1)]. Some axSpA patients develop structural changes in the sacroiliac joints over many years in the form of erosions, sclerosis, and/or ankyloses [(4)]. Patients with definitive structural changes in the sacroiliac joints visible on conventional X-rays and that fulfill the modified New York (mNY) criteria are classified as having AS [(2)]. The mNY criteria include both X-ray findings, along with at least one clinical criteria, such as low back pain and stiffness for more than 3 months that improves with exercise, but is not relieved by rest; or limitation of motion of the lumbar spine [(1)]. However, evidence of structural change in the sacroiliac joint takes years to develop, and the rate of development of structural change varies widely among patients [(3)]. Some patients develop only unilateral sacroiliitis, and others may never develop definitive sacroiliitis via x-ray despite significant disease burden and other signs and symptoms of the disease (e.g., spinal lesions, uveitis, enthesitis, and peripheral arthritis) [(4)]. Only40% of patients with nr-axSpA may convert to AS over 10 years [(5)]. While some nr-axSpA patients appear to develop AS over time, a significant portion of nr-axSpA patients (~60%) may never convert to AS despite suffering equally significant symptoms [(6)].

Although conversion from nr-axSpA to AS may require many years if it ever occurs, patients with nr-axSpA experience a high disease burden and a level of disease activity comparable to AS [(5)]. Namely, patients with nr-axSpA have demonstrated a similar level of clinical burden of disease and quality of life impairment when compared to AS patients across multiple outcomes, including: [(1)] level of inflammation (CRP), [(2)] physical function (BASFI), [(3)] disease activity (BASDAI), [(4)] patient global

assessment (GADA), and [5] back pain. Radiographic (x-ray) evidence of structural damage to the sacroiliac joint does not correlate with the severity of disease [5].

Classification of AS and nr-axSpA in ICD-11 is not specific, but both are grouped together within a subcategory, FA92.0, Axial spondyloarthritis, within the broader category of Inflammatory spondyloarthritis (FA92). The map back to ICD-10 from ICD-11 for axial spondyloarthritis goes to the category M45, Ankylosing spondylitis.

A request to create specific codes for non-radiographic-axial spondyloarthritis has been received from UCB, Inc., a multinational biopharmaceutical company. This proposal is based on this request, although changes have been made from the original request, for consistency with the structure of the classification. Addition of new codes for non-radiographic-axial spondyloarthritis, will enable better identification and tracking of this distinct set of patients, which is anticipated will advance the clinical understanding of non-radiographic-axial spondyloarthritis, and subsequently improve the diagnostic and treatment paradigms. The request states that this accurate identification is critical to facilitate research that will further elucidate the marked needs and characteristics of non-radiographic axial spondyloarthritis, and that more importantly, this coding request aligns with the clinical consensus reached by clinical experts in the field of rheumatology.

References

1. Sieper J, Rudwaleit M, Baraliakos X, et al The Assessment of SpondyloArthritis international Society (ASAS) handbook: a guide to assess spondyloarthritis *A Ann Rheum Dis.* 2009;68:ii1-ii44. https://doi.org/10.1136/ard.2008.104018
2. van der Heijde D, Baraliakos X, Hermann KA, et al. Limited radiographic progression and sustained reductions in MRI inflammation in patients with axial spondyloarthritis: 4-year imaging outcomes from the RAPID-axSpA phase III randomised trial. *Ann Rheum Dis* 2018;77:699-705. https://doi.org/10.1136/annrheumdis-2017-212377
3. Rudwaleit M, Sieper J. Referral strategies for early diagnosis of axial spondyloarthritis. *Nat Rev Rheumatol.* 2012 Apr 10;8(5):262-8. https://doi.org/10.1038/nrrheum.2012.39
4. Sieper J, Poddubnyy D. New evidence on the management of spondyloarthritis. *Nat Rev Rheumatol.* 2016 May;12(5):282-95. https://doi.org/10.1038/nrrheum.2016.42
5. Protopopov M, et al. Radiographic progression in non-radiographic axial spondyloarthritis. *Expert Rev Clin Immunol.* 2018;14(6):525-533. https://doi.org/10.1080/1744666x.2018.1477591
6. Sieper J, van der Heijde D. Review: Non-radiographic axial spondyloarthritis: new definition of an old disease? *Arthritis Rheum.* 2013 Mar;65(3):543-51. https://doi.org/10.1002/art.37803

TABULAR MODIFICATIONS

M45 Ankylosing spondylitis

New subcategory	M45.A	Non-radiographic axial spondyloarthritis	
New code	M45.A0	Non-radiographic axial spondyloarthritis of unspecified sites in spine	
New code	M45.A1	Non-radiographic axial spondyloarthritis of occipito-atlanto-axial region	
New code	M45.A2	Non-radiographic axial spondyloarthritis of cervical region	
New code	M45.A3	Non-radiographic axial spondyloarthritis of cervicothoracic region	
New code	M45.A4	Non-radiographic axial spondyloarthritis of thoracic region	
New code	M45.A5	Non-radiographic axial spondyloarthritis of thoracolumbar region	
New code	M45.A6	Non-radiographic axial spondyloarthritis of lumbar region	
New code	M45.A7	Non-radiographic axial spondyloarthritis of lumbosacral region	
New code	M45.A8	Non-radiographic axial spondyloarthritis of sacral and sacrococcygeal region	
New code	M45.AB	Non-radiographic axial spondyloarthritis of multiple sites in spine	

Pneumonia due to Coronavirus Disease 2019 (COVID-19)

The Federal Drug Administration (FDA) is requesting to create a new code for pneumonia due to coronavirus disease 2019 (COVID-19) caused by the 2019 novel coronavirus (SARS-CoV-2) that distinguishes this novel disease from pneumonia due to SARS-associated coronavirus or other viral-associated pneumonia. Currently, there is no specific code for pneumonia due COVID-19.

On March 11, 2020, WHO has classified the COVID-19 as a global pandemic. An outbreak of coronavirus disease 2019 (COVID-19) caused by the 2019 novel coronavirus (SARS-CoV-2) started in China with a cluster of pneumonia cases reported in December 2019 and is currently ongoing around the world. [1-6] COVID-19 cases have been confirmed worldwide, including United States, and the number of reported cases continues to rise. [6-8]

COVID-19 is generally a respiratory disease and symptoms can include cough, fever, myalgia, fatigue, shortness of breath, and dyspnea. [3, 10-13] Although many infected persons may not develop symptoms immediately due to a long incubation period of up to two weeks, COVID-19 is considered serious and frequently results in pneumonia and may lead to death.[2, 14-16]

Around the world number of COVID-19 cases and deaths continue to rise and have long surpassed cases with SARS, severe acute respiratory syndrome. [7, 12, 16, 18, 19] As of May 6, 2020, the coronavirus infections have been reported in over 120 countries with more than 3.5 million reported cases and over 250,000 deaths [19]. As of May 6, 2020, U.S. had over 1 million confirmed or presumptive cases and over 70,000 deaths from 50 states, District of Columbia, Puerto Rico, Guam, Northern Marianas, and US Virgin Islands, as reported by CDC [7]. It is expected that cumulatively number of COVID-19 cases will continue to rise and may become seasonal due to efficient human-to-human transmission [20] and likely persistence on inanimate surfaces [21].

In summary, COVID-19 and related complications (e.g., pneumonia, mortality) will likely continue to pose public safety concern in the United States and around the world, and therefore will need continued surveillance of the disease and related serious complications (e.g., pneumonia).

The objective of the request is to improve coding specificity for pneumonia due to coronavirus disease 2019 (COVID-19) caused by the 2019 novel coronavirus (SARS-CoV-2) as it can lead to very serious health consequences for patients including pneumonia and mortality. As of now, there is no specific ICD-10-CM diagnosis code for COVID-19 related pneumonia. The new pneumonia specific code will allow physicians to diagnose Pneumonia due to COVID-19 accurately as well as enable FDA and other public health organizations to monitor the Pneumonia due to COVID-19 in the United States nationally and locally, by state and county of residence, using real-world evidence (e.g., large databases).

Although interim guideline advises the use of two diagnosis codes, one for COVID-19 and one for ‘other viral pneumonia’, the investigation using CMS data suggest that using two codes may substantially under record pneumonia-related COVID-19, with <50% of recorded COVID-19 cases having had ‘other viral pneumonia’ recorded. Also, CDC COVID-19 mortality data shows that less than 50% of COVID-related deaths have pneumonia recorded, suggesting a potentially substantial under recording of pneumonia due to COVID-19. [22] As such, the introduction of the specific code for Pneumonia due to COVID-19 will allow a more accurate surveillance of COVID-19 related

pneumonia, including geographic distribution and severity of pneumonia-related cases, and will help to develop prevention strategies and assure public health safety in the U.S. The new code will also allow to distinguish the pneumonia due to COVID-19 from the previously approved SARS-associated pneumonia code and from other viral pneumonia code(s) and thus will improve accuracy of diagnosis and coding.

Since many COVID patients may be asymptomatic, the new pneumonia specific coding will also be helpful in identification of the COVID-19 risk to blood supply and if needed, to develop donor testing strategies to prevent potential transfusion-transmission and assure the safety of blood supply as studies have shown potential risk of COVID-19 to blood supply. [23-26]

In summary, the introduction of a disease-specific code will improve coding accuracy, increase provider and consequently population awareness of this serious consequence of COVID-19. In addition a new code will provide the ability to monitor the occurrence of the Pneumonia due to COVID-19 both locally and nationally. Ultimately, this will help in development and evaluation of appropriate prevention and treatment strategies.

The implementation date is expected to be in January 2021. The comment deadline will be Oct. 9, 2020.

References

1. World Health Organization. Novel coronavirus(2019-nCoV). Situation report 15. Geneva, Switzerland: World Health Organization; 2020. https://www.who.int/docs/default-source/coronaviruse/situation-reports/20200204-sitrep-15-ncov.pdf?sfvrsn=88fe8ad6_2pdf icon

2. Patel A, Jernigan DB. Initial Public Health Response and Interim Clinical Guidance for the 2019 Novel Coronavirus Outbreak — United States, December 31, 2019–February 4, 2020. MMWR Morb Mortal Wkly Rep 2020;69:140–146. DOI: http://dx.doi.org/10.15585/mmwr.mm6905e1

3. Bajema KL, Oster AM, McGovern OL, et al. Persons Evaluated for 2019 Novel Coronavirus — United States, January 2020. MMWR Morb Mortal Wkly Rep. ePub: February 7, 2020. http://dx.doi.org/10.15585/mmwr.mm6906e1

4. Wang C, Horby P, Hayden F, Gao G. A novel coronavirus outbreak of global health concern pdf icon[4 pages]. The Lancet. Published online January 24, 2020. https://doi.org/10.1016/S0140-6736(20)30185-9external icon

5. Paules CI, Marston HD, Fauci AS. Coronavirus Infections—More Than Just the Common Cold. JAMA. 2020;323(8):707–708. http://doi.org/10.1001/jama.2020.0757

6. Jernigan DB. Update: Public Health Response to the Coronavirus Disease 2019 Outbreak — United States, February 24, 2020. MMWR Morb Mortal Wkly Rep 2020;69:216–219. DOI: http://dx.doi.org/10.15585/mmwr.mm6908e1external icon

7. U.S. CDC: About Coronavirus Disease 2019 (COVID-19). [Accessed March 4, 2020 updated March 4 2020]. https://www.cdc.gov/coronavirus/2019-ncov/cases-in-us.html

8. del Rio C, Malani PN. 2019 Novel Coronavirus—Important Information for Clinicians. JAMA. Published online February 05, 2020. Available at: https://jamanetwork.com/journals/jama/fullarticle/2760782?utm_campaign=articlePDF%26utm_medium%3darticlePDFlink%26utm_source%3darticlePDF%26utm_content%3djama.2020.1490

9. Zhou P, Yang XL,Wang XG, et al. Discovery of a novel coronavirus associated with the recent pneumonia outbreak in humans and its potential bat origin. Preprint. https://www.biorxiv.org/content/10.1101/2020.01.22.914952v2.full.pdf Posted January 23, 2020. Accessed February 3, 2020

10. Chan JF, Yuan S, Kok K, et al. A familial cluster of pneumonia associated with the 2019 novel coronavirus indicating person-to-person transmission: a study of a family cluster pdf icon[1.4 MB, 10 pages]. The Lancet. Published online January 24, 2020. https://doi.org/10.1016/S0140-6736(20)30154-9external icon

11. Chen N, Zhou M, Dong X, et al. Epidemiological and clinical characteristics of 99 cases of 2019 novel coronavirus pneumonia in Wuhan, China: a descriptive study pdf icon[7 pages]. The Lancet. Published online January 29, 2020. https://doi.org/10.1016/S0140-6736(20)30211-7external icon

12. Chang D, Lin M, Wei L, et al. Epidemiologic and Clinical Characteristics of Novel Coronavirus Infections Involving 13 Patients Outside Wuhan, Chinaexternal icon. JAMA. Published online February 7, 2020. Available at: https://jamanetwork.com/journals/jama/fullarticle/2761043https://jamanetwork.com/journals/jama/fullarticle/2761043
13. US Centers for Disease Control and Prevention. 2019 Novel coronavirus, Wuhan, China: symptoms. https://www.cdc.gov/coronavirus/2019-ncov/about/symptoms.html. Published January 26, 2020. Accessed January 27, 2020.

14. Li Q, Guan X,Wu P, et al. Early transmission dynamics in Wuhan, China, of novel coronavirus-infected pneumonia. N Engl J Med.Published online January 29, 2020. http://dx.doi.org/10.1056/NEJMoa2001316

15. Wang D, Hu B, Hu C, et al. Clinical Characteristics of 138 Hospitalized Patients with 2019 Novel Coronavirus-Infected Pneumonia in Wuhan, Chinaexternal icon. JAMA. Published online February 7, 2020. Available at: https://jamanetwork.com/journals/jama/fullarticle/2761044

16. Wu Z, McGoogan JM. Characteristics of and Important Lessons From the Coronavirus Disease 2019 (COVID-19) Outbreak in China: Summary of a Report of 72 314 Cases From the Chinese Center for Disease Control and Prevention. JAMA. Published online February 24, 2020. http://doi.org/10.1001/jama.2020.2648

17. US Centers for Disease Control and Prevention. People Who Are at Higher Risk for Severe Illness. Available at: https://www.cdc.gov/coronavirus/2019-ncov/need-extra-precautions/people-at-higher-risk.html

18. Noah C Peeri, Nistha Shrestha, Siddikur Rahman. The SARS, MERS and novel coronavirus (COVID-19) epidemics, the newest and biggest global health threats: what lessons have we learned? International Journal of Epidemiology, 2020, 1–10. http://doi.org/10.1093/ije/dyaa033

19. Dong E, Du H, Gardner L. An interactive web-based dashboard to track COVID-19 in real time. Lancet Infect Dis; published online Feb 19. https://doi.org/10.1016/S1473-3099(20)30120-1. https://gisanddata.maps.arcgis.com/apps/opsdashboard/index.html?utm_source=Nature+Briefing&utm_campaign=ee09f96136-briefing-dy-20200130&utm_medium=email&utm_term=0_c9dfd39373-ee09f96136-44431161#/bda7594740fd40299423467b48e9ecf6

20. US Centers for Disease Control and Prevention. How Coronavirus Spreads. https://www.cdc.gov/coronavirus/2019-ncov/prevent-getting-sick/how-covid-spreads.html?CDC_AA_refVal=https%3A%2F%2Fwww.cdc.gov%2Fcoronavirus%2F2019-ncov%2Fabout%2Findex.html

21. Kampf G, Todt D, Pfaender S, Steinmann E. Persistence of coronaviruses on inanimate surfaces and its inactivation with biocidal agentsexternal icon. J Hosp Infect. February 6, 2020. pii: S0195-6701(20)30046-3. Available at https://www.ncbi.nlm.nih.gov/pubmed/32035997

22. CDC: National Center for Health Statistics. National Center for Vital Statistics. Provisional Death Counts for Coronavirus Disease (COVID-19). Available at: https://www.cdc.gov/nchs/nvss/vsrr/COVID19/index.htm

23. L. Chang, Y. Yan and L.Wang, Coronavirus Disease 2019: Coronaviruses and Blood Safety, Transfusion Medicine Reviews. Available at: https://doi.org/10.1016/j.tmrv.2020.02.003

24. R.Y. Dodd and S.L. Stramer, COVID-19 and Blood Safety:Helpwith a Dilemma, Transfusion Medicine Reviews. Available at: https://doi.org/10.1016/j.tmrv.2020.02.004

25. Chang L, Zhao L, Gong H, Wang Lunan, Wang L. Severe acute respiratory syndrome coronavirus 2 RNA detected in blood donations. Emerg Infect Dis. 2020 Jul [date cited]. https://doi.org/10.3201/eid2607.200839 https://wwwnc.cdc.gov/eid/article/26/7/20-0839_article

26. Kwon, S.-Y., Kim, E.-J., Jung, Y.S., Jang, J.S. and Cho, N.-S. (2020), Post-donation COVID-19 identification in blood donors. Vox Sang. doi:10.1111/vox.12925 https://onlinelibrary.wiley.com/doi/full/10.1111/vox.12925

TABULAR MODIFICATIONS

J12 Viral pneumonia, not elsewhere classified

J12.8 Other viral pneumonia

J12.81 Pneumonia due to SARS-associated coronavirus

New code	J12.82 Pneumonia due to coronavirus disease 2019
Add	Pneumonia due to COVID-19
Add	Pneumonia due to 2019 novel coronavirus (SARS-CoV-2)

J12.89 Other viral pneumonia

Postprocedural Anastomotic Leak of Digestive System Organ or Structure Following a Procedure

A proposal for a new code for anastomotic leak of a digestive system organ following a procedure has been requested. Currently there is not a unique code for intestinal anastomotic leaks that are not dehiscence. There are codes for complications of devices, implants, and grafts, and codes for dehiscence, but none of these are accurate for leakage from an intact intestinal anastomosis.

Anastomotic leak is defined as a leak of intra-luminal contents secondary to a surgical connection between two walls of the intestine. Anastomotic leaks are the most important complication to recognize following gastrointestinal surgery. Early diagnosis and treatment of an anastomotic leak is key. It can be a life-threatening condition if not identified in a timely fashion.

Creation of new code K91.88 will specifically describe anastomotic leak and allow K91.89 to be used for other digestive complications that can be further described with an additional code.

The American Gastrointestinal Association (AGA) has reviewed the proposal and is in support of the creation of a unique code.

TABULAR MODIFICATIONS

K91 Intraoperative and postprocedural complications and disorders of digestive system, not elsewhere classified

K91.8 Other intraoperative and postprocedural complications and disorders of digestive system

New Code K91.88 Postprocedural anastomotic leak of digestive system organ or structure following a procedure

Add Excludes1: disruption or dehiscence of closure of internal organ or other internal tissue (T81.32-)

mechanical complication of gastrointestinal device, implant, or graft (T85.5-)

Pyruvate Kinase (PK) Deficiency

Red blood cell (RBC) pyruvate kinase (PK) deficiency is a rare congenital hemolytic anemia caused by mutations in the PKLR gene that encodes PK (Grace 2018a). Pyruvate kinase deficiency is inherited via an autosomal recessive pattern and has a wide geographic distribution (Grace 2019). Pyruvate kinase catalyzes the final step in the glycolysis pathway, ultimately leading to the production of adenosine triphosphate (ATP), which is responsible for the maintenance of RBC energy levels, multiple cellular functions, and cellular integrity (Grace 2018a, Grace 2019). In patients with PK deficiency, a reduction in production of ATP leads to shortened reticulocyte and RBC life span resulting in chronic hemolysis and anemia (Grace 2018a). Although rare, PK deficiency is the most common red cell glycolytic enzyme defect causing hereditary non-spherocytic hemolytic anemia (van Wijk 2005).

The diagnosis of PK deficiency is currently hindered by the clinical heterogeneity of disease manifestations and a lack of awareness among providers (Al-Samkari 2020, Bianchi 2019). It is therefore likely underdiagnosed or underreported and, accordingly, its exact prevalence is unknown. Estimates have reported the prevalence of heterozygous carriers to range from 1% to 3% in the Caucasian population, with PK deficiency itself having an estimated prevalence of about 1 in 20,000, for diagnosed and undiagnosed cases together in the Western population (based on gene frequency analysis, with the caveats that this could be somewhat increased by inbreeding, but likely decreased by lower survival in those with PK deficiency). Diagnosed cases are estimated to have somewhat lower prevalence, but as noted above, there is likely under diagnosis (Beutler 2000, Carey 2000, Secrest 2020).

The clinical course of PK deficiency ranges from fully compensated or mildly symptomatic anemia to significant anemia requiring regular transfusions. Diagnosis of PK deficiency is confirmed with detection of reduced PK enzymatic activity and/or compound heterozygous and homozygous PKLR gene mutations and may occur at any point from infancy through young adulthood (Grace 2019, Bianchi 2019). Symptoms in newborns may present as in utero complications such as intrauterine growth retardation, hydrops fetalis, or prematurity, or after birth as neonatal jaundice, hyperbilirubinemia, and significant hemolysis (Grace 2019). In adults, the most common symptoms include complications of hemolysis such as splenomegaly, jaundice, and gallstones, but may also include aplastic crisis, bone deformities, extramedullary erythropoiesis, delayed puberty, hyperpigmentation, leg ulcers, and pulmonary hypertension, among others (Grace 2019).

PK deficiency manifestations can be unpredictable throughout life and the potential for serious complications requires frequent monitoring of patients by a hematologist, regardless of the degree of anemia (Grace 2019). Iron overload is one such serious complication that is commonly seen in PK deficiency associated with, and independent of, transfusions, and requires close follow-up from a young age (van Beers, Grace 2018a).

Patients with PK deficiency are at a high risk for a range of complications and experience a significant burden of disease and negative impact on quality of life (Boscoe 2019, Grace 2018b). In an age- and gender-matched comparison, patients with PK deficiency had higher lifetime rates of hypertension (4.6% vs 0.3%), osteoporosis (15.6% vs 0%), and liver cirrhosis (5.6% vs 0.4%) (Boscoe 2019). Other examples of complications include reticulocytosis, cognitive difficulties, liver cirrhosis, blood-borne viral infections, osteopenia, endocrinopathies, including thyroid dysfunction, sex hormone

abnormalities, and diabetes, and extramedullary hematopoiesis (Grace 2019, Grace 2015, Grace 2018a).

There are currently no guidelines for the treatment of patients with PK deficiency. Treatment approaches are supportive, including RBC transfusions and/or splenectomy (Grace 2018a, Grace 2019). These treatment approaches are not curative and are associated with serious complications such as iron overload, blood-borne infections or sepsis, and thrombotic events.

Other supportive medications that can be provided for PK deficiency patients include folic acid to support erythropoiesis, iron chelation therapies to reduce hemolysis- or transfusion-related iron overload, and prophylactic antibiotics and aspirin, particularly in patients with a history of splenectomy who are at higher risk of infections from encapsulated organisms and thrombotic events (Grace 2018a, Grace 2019).

Due to the lack of central reporting and rarity of disease, long term follow-up and reporting of patients with PK deficiency has been limited. Therefore, the possible effects of lifelong complications from PK deficiency and the burden of these patients on the healthcare system are not yet fully understood.

There is currently no unique ICD-10-CM code for PK deficiency, although there is the inclusion term Pyruvate kinase [PK] deficiency anemia, at the code D55.2, Anemia due to disorders of glycolytic enzymes. Agios Pharmaceuticals has requested creation of a specific code for pyruvate kinase deficiency, and believes that creation of a unique code will aid in improved accuracy of reporting PK deficiency among other hemolytic anemias, along with improved treatment and follow-up of patients, appropriate claims adjudication, as well as support future research for patients with PK deficiency. The American Society of Hematology (ASH) represents more than 18,000 clinicians and scientists worldwide committed to the study and treatment of blood and blood-related diseases. ASH has reviewed and supports this application for a new code for PK deficiency to best determine the incidence and prevalence of the disease, and to lead to improved treatment for this patient population.

References

Al-Samkari H, van Beers EJ, Kuo KHM, et al. The variable manifestations of disease in pyruvate kinase deficiency and their management. Haematologica. 2020. https://doi.org/10.3324/haematol.2019.240846

Beutler E, Gelbart T. Estimating the prevalence of pyruvate kinase deficiency from the gene frequency in the general white population. Blood. 2000; 95(11):3585-3588.

Bianchi P, Fermo E, Glader B, et al. Addressing the diagnostic gaps in pyruvate kinase deficiency: consensus recommendations on the diagnosis of pyruvate kinase deficiency. Am J Hematol. 2019; 94(1):149-161. https://doi.org/10.1002/ajh.25325

Boscoe AN, Yan Y, Hedgeman E, et al. Comorbidities and complications in adults with pyruvate kinase deficiency. Blood. 2019; 134(Suppl_1):2175. https://doi.org/10.1182/blood-2019-123069

Carey PJ, Chandler J, Hendrick A, et al. Prevalence of pyruvate kinase deficiency in northern European population in the north of England. Norther Region Haematologists Group. Blood. 2000; 96(12):4005-4006.

Grace RF, Bianchi P, van Beers EJ, et al. Clinical spectrum of pyruvate kinase deficiency: data from the Pyruvate Kinase Deficiency Natural History Study. Blood. 2018a; 131(20):2183-2192. https://doi.org/10.1182/blood-2017-10-810796

Grace RF, Cohen J, Egan S, et al. The burden of disease in pyruvate kinase deficiency: patients' perception of the impact on health-related quality of life. Eur J Haematol. 2018b; 101(6):758-765. https://doi.org/10.1111/ejh.13128

Grace RF, Layton DM, Barcellini W. How we manage patients with pyruvate kinase deficiency. Br J Hematol. 2019; https://doi.org/10.1111/bjh.15758

Grace RF, Zanella A, Neufel EJ. Erythrocyte pyruvate kinase deficiency: 2015 status report. Am J Hematol. 2015; 90:825-830. https://doi.org/10.1002/ajh.24088

Secrest MH, Storm M, Carrington C, et al. Prevalence of pyruvate kinase deficiency: a systematic literature review. Eur J Haematol. 2020. https://doi.org/10.1111/ejh.13424

van Beers EJ, van Straaten S, Morton DH, et al. Prevalence and management of iron overload in pyruvate kinase deficiency: report from the Pyruvate Kinase Deficiency Natural History Study. Haematologica. 2019;104:e51. https://doi.org/10.3324%2Fhaematol.2018.196295

van Wijk R, van Solinge WW. The energy-less red blood cell is lost: erythrocyte enzyme abnormalities. Blood. 2005;106:4034-4042. https://doi.org/10.1182/blood-2005-04-1622

TABULAR MODIFICATIONS

D55 Anemia due to enzyme disorders

D55.2 Anemia due to disorders of glycolytic enzymes

Delete ~~Hemolytic nonspherocytic (hereditary) anemia, type II~~
Delete ~~Hexokinase deficiency anemia~~
Delete ~~Pyruvate kinase [PK] deficiency anemia~~
Delete ~~Triose-phosphate isomerase deficiency anemia~~

Excludes1: disorders of glycolysis not associated with anemia (E74.81-)

New code D55.21 Anemia due to pyruvate kinase deficiency
Add PK deficiency anemia
Add Pyruvate kinase deficiency anemia

New code D55.29 Anemia due to other disorders of glycolytic enzymes
Add Hemolytic nonspherocytic (hereditary) anemia, type II
Add Hexokinase deficiency anemia
Add Triose-phosphate isomerase deficiency anemia

Rapid Destructive Osteoarthritis

Rapid destructive osteoarthritis (RDOA) is a rare arthropathy of unknown etiology which can progress to abnormal destruction of subchondral bone and joint collapse. Patients with RDOA typically present with pain and significant radiographic joint damage. Sequential radiographs demonstrate a rapid narrowing of the joint space with eventually subchondral bone loss and joint collapse. The rate of RDOA is increasing in the United States as the population ages. The incidence of RDOA is not well understood, as no large longitudinal studies have been conducted.

RDOA is characterized by rapid joint destruction. Other terms to describe the disease are Rapidly Progressive Osteoarthritis Type 2 (RPOA Type 2), destructive arthropathy, rapid destructive coxarthrosis, and Postel's osteoarthritis. RDOA can mimic septic arthritis and inflammatory arthritis. The condition is typically described in the hip but has also been reported in the knee and shoulder. It usually occurs in elderly women or in patients who have sustained trauma. Rapidly destructive osteoarthritis is a rare form of degenerative hip disease distinct from osteonecrosis. It is also distinct from Rapidly Progressive Osteoarthritis Type 1 (RPOA Type 1), a significant loss of joint space width ≥ 2mm within approximately 1 year, without gross structural failure.

Recent studies with monoclonal antibody nerve growth factor inhibitors (NGF inhibitors), a new class of biologics in development for the management of pain, have shown that NGF inhibitors increase the risk factor for developing RDOA. The pathophysiology is currently unknown.

Pfizer, Incorporated, and Eli Lilly and Company, pharmaceutical based companies, are requesting new codes to clarify the differences between osteonecrosis and RDOA.

TABULAR MODIFICATIONS

M16 Osteoarthritis of hip

New code	M16.A	Bilateral rapid destructive osteoarthritis of hip
Add		Bilateral rapid progressive osteoarthritis Type 2 of hip
Add		Exclude1: osteoarthritis of hip, unspecified (M16.9)
Add		osteoarthritis resulting from hip dysplasia (M16.2, M16.3.-)
Add		osteonecrosis (M87.-)
Add		post-traumatic osteoarthritis of hip (M16.4, M16.5-)
Add		primary osteoarthritis of hip (M16.0, M16.1-)
Add		secondary osteoarthritis of hip (M16.6, M16.7)

New subcategory	M16.B	Unilateral rapid destructive osteoarthritis of hip
Add		Unilateral rapid progressive osteoarthritis Type 2 of hip
Add		Exclude1: osteoarthritis of hip, unspecified (M16.9)
Add		osteoarthritis resulting from hip dysplasia (M16.2, M16.3.-)

Add osteonecrosis (M87.-)
Add post-traumatic osteoarthritis of hip (M16.4, M16.5-)
Add primary osteoarthritis of hip (M16.0, M16.1-)
Add secondary osteoarthritis of hip (M16.6, M16.7)

New code M16.B0 Unilateral rapid destructive osteoarthritis, unspecified hip
New code M16.B1 Unilateral rapid destructive osteoarthritis, right hip
New code M16.B2 Unilateral rapid destructive osteoarthritis, left hip

M17 Osteoarthritis of knee

New code M17.A Bilateral rapid destructive osteoarthritis of knee

Add Bilateral rapid progressive osteoarthritis Type 2 of knee
Add Exclude1: osteoarthritis of knee, unspecified (M17.9)
Add osteonecrosis (M87.-)
Add post-traumatic osteoarthritis of knee (M17.2, M17.3-)
Add primary osteoarthritis of knee (M17.0, M17.1-)
Add secondary osteoarthritis of knee (M17.4, M17.5)

New subcategory M17.B Unilateral rapid destructive osteoarthritis of knee

Add Unilateral rapid progressive osteoarthritis Type 2 of knee
Add Exclude1: osteoarthritis of knee, unspecified (M17.9)
Add osteonecrosis (M87.-)
Add post-traumatic osteoarthritis of knee (M17.2, M17.3-)
Add primary osteoarthritis of knee (M17.0, M17.1-)
Add secondary osteoarthritis of knee (M17.4, M17.5)

New code M17.B0 Unilateral rapid destructive osteoarthritis, unspecified knee
New code M17.B1 Unilateral rapid destructive osteoarthritis, right knee
New code M17.B2 Unilateral rapid destructive osteoarthritis, left knee

M19 Other and unspecified osteoarthritis

New code M19.A Rapid destructive osteoarthritis of other joints
Add Rapid progressive osteoarthritis Type 2 of other joints
Add Exclude1: osteonecrosis (M87.-)
Add post-traumatic osteoarthritis of other joints (M19.1-)
Add primary osteoarthritis of other joints (M19.0-)
Add secondary osteoarthritis of other joints (M19.2-)

New subcategory M19.A1 Rapid destructive osteoarthritis, shoulder

New code M19.A11 Rapid destructive osteoarthritis, right shoulder
New code M19.A12 Rapid destructive osteoarthritis, left shoulder
New code M19.A19 Rapid destructive osteoarthritis, unspecified shoulder

New subcategory M19.A7 Rapid destructive osteoarthritis, ankle and foot

New code M19.A71 Rapid destructive osteoarthritis, right ankle and foot
New code M19.A72 Rapid destructive osteoarthritis, left ankle and foot
New code M19.A79 Rapid destructive osteoarthritis, unspecified ankle and foot

M19.9 Osteoarthritis, unspecified site

New code M19.94 Rapid destructive osteoarthritis, unspecified site
Add Rapid destructive osteoarthritis NOS

M87 Osteonecrosis

Add Excludes1: rapid destructive osteoarthritis of hip (M16.-)
rapid destructive osteoarthritis of knee (M17.-)
rapid destructive osteoarthritis of other and unspecified site (M19.-)

Refractory Angina Pectoris

Chronic angina pectoris, refractory to medical and interventional therapies, is a common and disabling medical condition, and a major public health problem that affects millions of patients worldwide [1]. The clinical burden of refractory angina (RA) is growing due to an aging population and improved survival from coronary artery disease (CAD). Estimates suggest that in the US up to 1.8 million patients suffer from RA [2].

Refractory angina is conventionally defined as a chronic condition characterized by angina in the setting of coronary artery disease (CAD), which cannot be controlled by a combination of optimal medical therapy, angioplasty or bypass surgery, and where reversible myocardial ischemia has been clinically established to be the cause of the symptoms [3].

An increasing number of patients, particularly those with advanced, chronic coronary artery disease [4] have severe symptoms of angina despite optimal medical therapy. However, refractory angina is common not only in patients who are not good candidates for revascularization, but also in patients following successful revascularization. Persistence or recurrence of angina after PCI or CABG surgery is well recognized and may affect 20–40% of patients during short and medium-term [5,6,7,8,9,10,11].

When further revascularization options are limited, these patients are frequently described as being no option for treatment, and as having refractory angina. The care of these patients is challenging, and the guidance available from national practice guidelines is limited.

The American College of Cardiology/American Heart Association Task Force on Practice Guidelines, and the American Association for Thoracic Surgery, Preventive Cardiovascular Nurses Association, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons 2012 Guideline for Diagnosis and Management of Patients with Stable Ischemic Heart Disease and the 2014 Focused Update includes treatment of patients with refractory angina.

The European Society of Cardiology (ESC) released guidelines in 2019 for treatment of patients with refractory angina.

There are currently no diagnosis codes available to describe refractory angina pectoris. There are diagnosis codes available to describe unstable angina (I20.0), angina pectoris with documented spasm (I20.1), other forms of angina pectoris (I20.8) and angina pectoris, unspecified (I20.9).

A specialty medical device company submitted this proposal requesting the creation of a specific code for angina pectoris, refractory to distinguish refractory angina pectoris from other angina diagnoses, thus improving data collection and management of the disease.

This proposal has the support of the American College of Cardiology.

References

1 Benck L, Henry TD. CD34+ Cell Therapy for No-Option Refractory Disabling Angina: Time for FDA Approval? Cardiovascular revascularization medicine: including molecular interventions 2019;20:177-178.

2 Mannheimer C, Camici P, Chester MR et al. The problem of chronic refractory angina; report from the ESC Joint Study Group on the Treatment of Refractory Angina. Eur Heart J 2002;23:355-70.

3 Ibid

4. Henry TD, Satran D, Jolicoeur EM.Treatment of refractory angina in patients not suitable for revascularization. Nat Rev Cardiol 2014;11:78-95.

5 Abdallah MS, Wang K, Magnuson EA et al. Quality of Life After Surgery or DES in Patients With 3-Vessel or Left Main Disease. J Am Coll Cardiol 2017;69:2039-2050.

6 Al-Lamee R, Howard JP, Shun-Shin MJ et al. Fractional Flow Reserve and Instantaneous Wave-Free Ratio as Predictors of the Placebo-Controlled Response to Percutaneous Coronary Intervention in Stable Single-Vessel Coronary Artery Disease. Circulation 2018;138:1780-1792

7 Crea F, Bairey Merz CN, Beltrame JF et al. Mechanisms and diagnostic evaluation of persistent or recurrent angina following percutaneous coronary revascularization. Eur Heart J2019;40:2455-2462

8 Serruys PW. Assessing percutaneous intervention: re-appraising the significance of residual angina. EuroIntervention: Journal of EuroPCR in collaboration with the Working Group on Interventional Cardiology of the European Society of Cardiology 2015;10:1253.

9 Venkitachalam L, Kip KE, Mulukutla SR et al. Temporal trends in patient-reported angina at 1 year after percutaneous coronary revascularization in the stent era: a report from the National Heart, Lung, and Blood Institute-sponsored 1997-2006 dynamic registry. Circ Cardiovasc Qual Outcomes 2009;2:607-15.

10 Weintraub WS, Spertus JA, Kolm P et al. Effect of PCI on quality of life in patients with stable coronary disease. N Engl J Med 2008;359:677-87.

11 Niccoli G, Montone RA, Lanza GA, Crea F. Angina after percutaneous coronary intervention: The need for precision medicine. International Journal of Cardiology 2017;248:14-19.

TABULAR MODIFICATIONS

I20 Angina pectoris

I20.0 Unstable angina
I20.1 Angina pectoris with documented spasm
New code I20.2 Refractory angina pectoris

I25 Chronic ischemic heart disease

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

I25.111 Atherosclerotic heart disease of native coronary artery with unstable angina pectoris

Excludes1: angina pectoris with documented spasm without atherosclerotic heart disease (I20.0)

New code

I25.112 Atherosclerosis heart disease of native coronary artery with refractory angina pectoris

I25.7 Atherosclerosis of coronary artery bypass graft(s) and coronary artery of transplanted heart with angina pectoris

I25.70 Atherosclerosis of coronary artery bypass graft(s), unspecified, with angina pectoris

I25.701 Atherosclerosis of coronary artery bypass graft(s), unspecified, with angina pectoris with documented spasm

Excludes1: angina pectoris with documented spasm without atherosclerosis of coronary artery bypass graft (I20.1)

New code

I25.702 Atherosclerosis of coronary artery bypass graft(s), unspecified, with refractory angina pectoris

Short Bowel Syndrome and Intestinal Failure

Short bowel syndrome (SBS) is a condition that occurs when your body is unable to absorb enough nutrients from the foods you eat because you do not have enough small intestine. Short bowel syndrome is caused by the physical absence or loss of massive portions of intestine (typically to < 200 cm of residual intestine). Many, but not all individuals with SBS may also develop intestinal failure (IF), which is the inability to absorb enough nutrients and/or fluid necessary to maintain nutritional autonomy. Conversely, not all patients with IF suffer from SBS, but may have a myriad of different malabsorptive disorders.

Treatment of patients with SBS and IF is complex, including the management of intravenous nutrition and fluids, and the prevention and treatment of nutrient deficiencies and dehydration. Additional complications of the disease can affect the liver, kidney, brain, and bones. It is likely that lack of knowledge and understanding of both SBS and IF has led to misclassification of these diseases under various identifiers, thereby commonly reported prevalence numbers may be either over- or under-representations of actual disease prevalence.

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

Currently, this proposal has been recommended for approval by the Medical and Scientific Advisory Committee (MASC) of the World Health Organization (WHO) and is awaiting review by the ICD11 Classifications and Statistics Advisory Committee to ensure suitability.

Alan Buchman, MD, with technical support from Takeda Pharmaceuticals, is proposing the following tabular modifications for better delineation in these conditions and tracking these patients.

TABULAR MODIFICATIONS

	K90	Intestinal malabsorption
	K90.8	Other intestinal malabsorption
New code		K90.82 Short bowel syndrome
New code		K90.83 Intestinal failure

Short Stature Due to Endocrine Disorder

Changes in normal growth patterns may be a sign of a pathologic condition. As such, physicians monitor linear as well as skeletal growth from birth on through adolescence. Experiences and exposures encountered in the intrauterine environment may also influence growth from birth to two-to-three years of age, and occasionally throughout childhood and adolescence. Postnatally both growth hormone (GH) and insulin-like growth factor-I (IGF-I) drive statural growth. In puberty sex steroid hormones facilitate the pubertal growth spurt.[1]

With low concentrations of GH and IGF-I hormones, short stature develops. Short stature is defined by a height/length that is two standard deviation scores (SDS) away from the mean height/length of the age group, therefore affecting ~2.5% of children.[2] Of those, approximately 5%, or 1:1,000 children have short stature due to endocrine disorders.[3]

Between birth and puberty, a normal growth rate depends on an adequate secretion and action of growth hormone, which is released from the pituitary gland in response to several factors: hypothalamic GH releasing hormone (GHRH), ghrelin, and somatostatin.[4] Growth hormone binds to GH receptors (GHR), mainly on cells in the liver, although most tissues contain GHRs.[5] The interaction between GH and the GHR induces formation and release of IGF-I. Both the circulating IGF-I, secreted from the liver into the circulation, and locally produced IGF-I, then exert the growth-promoting effects at the level of skeletal muscle, cartilage, bone, and other tissues.[4]

When these coordinated growth events are altered, short stature may occur. Short stature has a variety of causes and the first step in the diagnostic evaluation of growth impairment leading to short stature due to an endocrine disorder will be to rule out other causes of growth failure, including genetic syndromes such as Turner syndrome, and several other secondary causes like malnutrition and inflammatory disorders.[6]

The most common hormonal disorder of the GH/IGF-I axis is GH deficiency (GHD), which is characterized by short stature due to a lack of growth hormone production/action.[7] Its prevalence is estimated to be between 1:4,000 to 1:10,000.[4] It is most often due to low-to-negligible growth hormone secretion from the pituitary gland, as is seen in hypopituitarism, but also exists in an isolated form.

To diagnose growth hormone deficiency, growth hormone provocation testing is used in combination with additional testing of IGF-I production, as well as measuring the binding protein(s) for IGF-I, as the concentrations of these peptides are highly dependent on GH secretion.[8] Some studies suggest imaging the hypothalamic-pituitary region via MRI may be more helpful in diagnosing growth hormone deficiency than laboratory assays.[9] GHD is treated with recombinant human GH (rhGH), also known as somatotropin.[7]

Growth hormone deficiency must also be ruled out in order to diagnose constitutional short stature, which, along with familial short stature, is a form of normal variant short stature often classified as idiopathic short stature (ISS).[8,10,11] Constitutional short stature or constitutional growth delay describes

patients with an unknown cause of short stature. This diagnosis depends on ruling out other causes of short stature, and is further characterized by specific auxological characteristics.[11] Approximately 70% of children with a short stature diagnosis have some type of idiopathic short stature, including constitutional short stature, but also with other unknown etiologies.[3] In some situations of ISS (not constitutional short stature or benign familial short stature), use of supplemental rhGH can increase the growth potential despite normal endogenous GH production.[8]

For those children who do not have GHD despite having IGF-I deficiency, primary IGF-I deficiency (PIGFD) may be the underlying etiology.[8] Severe PIGFD (SPIGFD) is defined by height and circulating IGF-I concentrations below -3 SDS.[12,13] A subset of patients with SPIGFD have mutations in the GH receptor gene and have Laron-type short stature.[14] The prevalence rate of SPIGFD in children suspected of having a growth abnormality is approximately 1%.[15] In some situations, patients with GHD who develop GH inactivating antibodies are considered GH insensitive, also have IGFD, and could also benefit from treatment with rhIGF.[16,17]

Currently E23.0 Hypopituitarism, would be used for those with short stature specifically caused by altered (decreased) pituitary hormone secretion, including GH. E34.3 Short stature due to endocrine disorder covers all other short stature diagnoses. Updates to guidelines for treatment of short stature from the Drug and Therapeutics Committee of the Pediatric Endocrine Society specifically call out SPIGFD as a separate diagnosis from GHD and ISS, because of the availability of a specific treatment and the opportunity to make a specific diagnosis.[8] Providers currently map SPIGFD to any of the following codes (E23.0 Hypopituitarism, E34.3 Short stature due to endocrine disorder, and R62.52 Short stature (child)), which has negative implications on tracking and disease management efforts.

As it stands, E34.3 broadly describes short stature due to all other endocrine disorders, which, again, may be detrimental for disease tracking purposes. Constitutional short stature is also included in the inclusion notes under E34.3. The table below demonstrates the differences between constitutional short stature and other short stature types due to endocrine disorder.
The cause, diagnostic approach, and treatment needs, and modalities differ significantly between constitutional short stature and SPGIFD.

Based on the above information, an expansion of code E34.3 would establish more precise disease-specific coding used to better identify and track patients. More specifically, separating out constitutional short stature from other types of short stature due to endocrine disorder, such as the narrowly defined short stature condition of SPIGFD, would also more closely align with the current Pediatric Endocrine Society recommendations for diagnosis and management of growth disorders. Modifying the existing ICD-10-CM code will help ensure more precise coding and alignment with current data from clinical practice, research databases and registries, and peer reviewed literature.

This proposal is submitted by Ipsen Biopharmaceuticals and supported by the Pediatric Endocrine Society.

References:

[1].Barstow C, Rerucha C. Evaluation of Short and Tall Stature in Children. *Am Fam Physician.* 2015;92(1):43-50.

[2].Stanley T. Diagnosis of growth hormone deficiency in childhood. *Curr Opin Endocrinol Diabetes Obes.* 2012;19(1):47-52.

[3]. Lindsay R, Feldkamp M, Harris D, Robertson J, Rallison M. Utah Growth Study: growth standards and the prevalence of growth hormone deficiency. *J Pediatr.* 1994;125(1):29- 35.

[4].Ranke MB, Wit JM. Growth hormone - past, present and future. *Nat Rev Endocrinol.* 2018;14(5):285-300.

[5].Dehkhoda F, Lee CMM, Medina J, Brooks AJ. The Growth Hormone Receptor: Mechanism of Receptor Activation, Cell Signaling, and Physiological Aspects. *Front Endocrinol (Lausanne).* 2018;9:35.

[6].Rhee N, Oh KY, Yang EM, Kim CJ. Growth hormone responses to provocative tests in children with short stature. *Chonnam Med J.* 2015;51(1):33-38.

[7].NORD. Growth Hormone Deficiency. 2020.

[8].Grimberg A, DiVall SA, Polychronakos C, et al. Guidelines for Growth Hormone and Insulin-Like Growth Factor-I Treatment in Children and Adolescents: Growth Hormone Deficiency, Idiopathic Short Stature, and Primary Insulin-Like Growth Factor-I Deficiency. *Horm Res Paediatr.* 2016;86(6):361-397.

[9].Maghnie M, Lindberg A, Koltowska-Haggstrom M, Ranke MB. Magnetic resonance imaging of CNS in 15,043 children with GH deficiency in KIGS (Pfizer International Growth Database). *Eur J Endocrinol.* 2013;168(2):211-217.

[10].Ranke MB. Towards a consensus on the definition of idiopathic short stature. *Horm Res.* 1996;45 Suppl 2:64-66.

[11].Cohen P, Rogol AD, Deal CL, et al. Consensus statement on the diagnosis and treatment of children with idiopathic short stature: a summary of the Growth Hormone Research Society, the Lawson Wilkins Pediatric Endocrine Society, and the European Society for Paediatric Endocrinology Workshop. *J Clin Endocrinol Metab.* 2008;93(11):4210-4217.

[12]. Cohen J, Blethen S, Kuntze J, Smith SL, Lomax KG, Mathew PM. Managing the child with severe primary insulin-like growth factor-1 deficiency (IGFD): IGFD diagnosis and management. *Drugs R D.* 2014;14(1):25-29.[13].INCRELEX® (mecasermin) FDA Approved Label. 2019.

[13]Laron Z: Laron syndrome (primary growth hormone resistance or insensitivity): the personal experience 1958-2003. J Clin Endocrinol Metab 2004;89:1031-1044.

[14].Teissier R, Flechtner I, Colmenares A, et al. Characterization and prevalence of severe primary IGF1 deficiency in a large cohort of French children with short stature. *Eur J of Endocrinology.* 2014; 170 (6): 847-54.

[15].Ahangari G, Ostadali MR, Rabani A, Rashidian J, Sanati MH, Zarindast MR. Growth hormone antibodies formation in patients treated with recombinant human growth hormone. *Int J Immunopathol Pharmacol.* 2004;17(1):33-38.

[16].David A, Hwa V, Metherell LA, et al. Evidence for a continuum of genetic, phenotypic, and biochemical abnormalities in children with growth hormone insensitivity. *Endocr Rev.* 2011;32(4):472-497.

TABULAR MODIFICATIONS

E23 Hypofunction and other disorders of the pituitary gland

Includes: the listed conditions whether the disorder is in the pituitary or the hypothalamus

Excludes1: postprocedural hypopituitarism (E89.3)

Add short stature due to endocrine disorder (E34.3-)

E34 Other endocrine disorders

Excludes1: pseudohypoparathyroidism (E20.1)

New subcategory E34.3 Short stature due to endocrine disorder

Delete ~~Constitutional short stature~~

Delete ~~Laron-type short stature~~

New Code E34.30 Short stature due to endocrine disorder unspecified

New code E34.31 Idiopathic short stature

New code E34.32 Constitutional short stature

Add Constitutional delay of growth and puberty/maturation

New code E34.33 Primary insulin-like growth factor-1 (IGF-1) deficiency

Add Acid-labile subunit gene (*IGFALS*) defect

Add Growth hormone gene 1 (*GH*1) defect with growth hormone neutralizing antibodies

Add Growth hormone insensitivity syndrome (GHIS)

Add Insulin-like growth factor 1 gene (*IGF1*) defect

Add Laron type short statue

Add Severe primary insulin-like growth factor-1 deficiency (SPIGFD)

Add		Signal transducer and activator of transcription 5B gene (*STAT5b*) defect
New code	E34.34	Insulin-like growth factor-1 (IGF-1) resistance
Add		Genetic syndrome with resistance to insulin-like growth factor-1
Add		Insulin-like growth factor-1 receptor (*IGF-1R*) defect
Add		Post-insulin-like growth factor-1 receptor signaling defect
New code	E34.39	Other short stature due to endocrine disorder

Thrombocytosis and Essential Thrombocythemia

This topic was previously presented in March 2020 and has been modified based on comments received. It proposes to create separate codes for other thrombocytosis (including secondary thrombocytosis and reactive thrombocytosis), and unspecified thrombocytosis, at a new subcategory D75.83 (instead of a single new code as previously proposed). As before, it also separates these conditions from primary or essential thrombocytosis, or primary or essential thrombocythemia, which are a neoplastic condition, involving cancer of the blood or bone marrow (the hematopoietic system).

Entries changed from the prior presentation are shown in bold.

TABULAR MODIFICATIONS

D47 Other neoplasms of uncertain behavior of lymphoid, hematopoietic and related tissue

D47.3 Essential (hemorrhagic) thrombocythemia
Essential thrombocytosis
Idiopathic hemorrhagic thrombocythemia

Add Excludes1: **reactive thrombocytosis (D75.838)**
Add secondary thrombocytosis (**D75.838**)
Add **thrombocythemia NOS (D75.839)**
Add thrombocytosis NOS (**D75.839**)

D75 Other and unspecified diseases of blood and blood-forming organs

D75.8 Other specified diseases of blood and blood-forming organs

New subcategory Add **D75.83 Thrombocytosis**
Excludes1: Essential thrombocytemia (D47.3)

New code Add **D75.838 Other thrombocytosis**
Reactive thrombocytosis
Secondary thrombocytosis

New code Add **D75.839 Thrombocytosis, unspecified**
Thrombocythemia NOS
Thrombocytosis NOS

Vertebrogenic Pain

Relievant Medsystems is proposing the creation of new codes for describing low back pain that are identified by Modic type endplate changes as seen on MRI. Low back pain (M54.5) is a diagnosis code that is broad and non-specific. Until recently, an objective biomarker that could improve diagnostic accuracy of low back pain did not exist.

Scientific research involving Modic changes that began decades ago [(1)] and continues today [(2, 3)] supports the correlation of Modic type endplate changes with a specific low back pain subtype (vertebrogenic pain). Our request for a new diagnosis code relates to the knowledge gained from this research.

These changes will enable better identification and tracking of this specific condition, and thus allow differentiating it from non-specific causes of back pain. This will be of clinical value, particularly related to enabling specific identification of cases for use of new therapy for this type of back pain.

References
1. Modic, MT, Steinberg PM, Rosse JS, et al. Degenerative disk disease: assessment of changes in vertebral body marrow with MRI imaging. Radiology 1988; 166 (1Pt1): 193-9. https://doi.org/10.1148/radiology.166.1.3336678
2. Antonacci MD, Mody DR, Heggeness NH. Innervation of the human vertebral body: a histologic study. J Spinal Disord 1998; 11(6):526-31. PMID: 9884299.
3. Dudli S, Fields AJ, Samartzis D, et al. Pathobiology of Modic changes. Eur Spine J 2016; 25(11):3723-34. https://doi.org/10.1007/s00586-016-4459-7 https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5477843/

TABULAR MODIFICATIONS

M54 Dorsalgia

M54.5 Low back pain

Delete	~~Loin pain~~	
Delete	~~Lumbago NOS~~	
New code	M54.50	Low back pain, unspecified
Add		Loin pain
Add		Lumbago NOS
New code	M54.51	Vertebrogenic low back pain
New code	M54.59	Other low back pain

INDEX MODIFICATIONS

	Low
Revise	- back syndrome M54.59
Revise	Lumbago, lumbalgia M54.50
	Pain(s) (see also Painful) R52
Revise	- loin M54.50
Revise	- low back M54.50
Add	- - vertebrogenic M54.51
Revise	- lumbar region M54.50
Add	- - vertebrogenic M54.51
	- spine M54.9
Revise	- - low back M54.50
Add	- - vertebrogenic M54.51
	Syndrome - see also Disease
	- low
Revise	- - back M54.59

Vulvovaginal Candidiasis Recurrent

Vulvovaginal candidiasis (VVC), also commonly known as vaginal yeast infection, is inflammation of the vulva and vagina due to Candida, typically *C albicans*. Familiar signs and symptoms include pruritus, vaginal soreness, dyspareunia, external dysuria, vulvar edema and erythema, and abnormal vaginal discharge.

An estimated 75% of women[1] will have at least one episode of vulvovaginal candidiasis in their lifetime. However, most episodes of VVC are uncomplicated with mild to moderate symptoms that are quickly and successfully addressed via over-the-counter topical antifungal creams and/or a short course of oral fluconazole, an antifungal drug. Among uncomplicated cases of VVC, most are diagnosed on the basis of symptoms alone, and many are self-diagnosed and self-treated.
A smaller but significant subgroup of women develop a more complicated form of vulvovaginal candidiasis.[2] Complicated vulvovaginal candidiasis refers to severe disease, infection in an immune-compromised woman, or infection with a non-*C albicans* species. Most prominently, it refers to recurrent vulvovaginal candidiasis (RVVC), defined as 3-4 or more episodes of symptomatic infection within one year. [2,3,4,5] Prevalence of RVVC has been variously estimated in literature reviews and surveys at 5-9% of women.[3,4,6,7]

The population of women with recurrent vulvovaginal candidiasis is clinically distinct in multiple respects. While several risk factors such as antibiotic use, diabetes, or pregnancy are known, the vast majority of women with RVVC develop the infection without having any risk factor. This implies that a genetic component likely plays an important role in susceptibility to RVVC.[8]
An episode of uncomplicated VVC is often considered a nuisance that is easily resolved. However, women with RVVC typically endure multiple relapses and require months of treatment with a significant impact on their lives.[5] Although new drugs and regimens are being developed, current treatment for RVVC typically consists of topicals or oral fluconazole for 10 to 14 days, followed by a maintenance regimen of oral fluconazole once a week for at least 6 months. This controls symptoms in the great majority of patients, but cessation is followed by another episode of VVC in approximately 50% of women within three to four months, and likely a higher percentage over time. [2,3,5]

RVVC is debilitating for patients, both physically and in terms of their mental health. The physical symptoms interfere with normal elements of life, from urinating to sexual activity. Women with RVVC reported missing an average of about 6 hours of work for each recurrent episode. As surveyed, health-related quality of life is significantly worse for women with RVVC than in the general population.[6] Over two-thirds of women with RVVC report depression and anxiety during recurrent episodes and over half report anxiety between episodes.[6] A sense of feeling “dirty” and suspecting sexually transmitted diseases from their partners is commonly reported.[7] Overall, women with RVVC ranked it similar to asthma and COPD and even higher than migraine for its negative impact on their quality of life.[6]

As often noted in the literature, the availability of over-the-counter treatment create difficulties in accurately determining the frequency of recurrent vulvovaginal candidiasis.[3,4] It is not currently

possible to clearly differentiate recurrent vulvovaginal candidiasis from uncomplicated vulvovaginal candidiasis[7] or to track cases of this clinically significant population in the data.[5]

The Mycoses Study Group Education and Research Consortium (MSGERC), is requesting new codes to uniquely identify recurrent vulvovaginal candidiasis. The American College of Obstetricians and Gynecologists (ACOG) has reviewed and support the proposed changes.

1. https://www.cdc.gov/std/tg2015/candidiasis.htm
2. Pappas PG, Kauffman CA, Andes DR et al. Clinical practice guidelines for the management of candidiasis: 2016 update by the Infectious Diseases Society of America, Clin Infect Dis 2016
3. Sobel, JD. Recurrent vulvovaginal candidiasis. Am. J. Obstet. Gynecol. 2016; 214,15–21.
4. Blostein F, Levin-Sparenberg E, Wagner J, Foxman B. Recurrent vulvovaginal candidiasis. Ann Epidemiol. 2017;27(9):575–82.
5. Crouss T, Sobel JD, Smith K, Nyirjesy P. Long term outcomes of women with recurrent vulvovaginal candidiasis after a course of maintenance antifungal therapy. J Low Genit Tract Dis. 2018;22:382–386.
6. Aballéa S, Guelfucci F, Wagner J, et al. Subjective health status and health-related quality of life among women with recurrent vulvovaginal candidosis (RVVC) in Europe and the USA. Health Qual Life Outcomes 2013; 11: 169.
7. Denning DW, Kneale M, Sobel JD, Rautemaa-Richardson R. Global burden of recurrent vulvovaginal candidiasis: a systematic review. Lancet Infect Dis. 2018;18(11):e339–e47.
8. Rosati D, Bruno M, Jaeger M, ten Oever J, Netea M. Recurrent Vulvovaginal Candidiasis: An Immunological Perspective. Microorganisms 2020, 8(2), 144

TABULAR MODIFICATIONS

	B37	Candidiasis
New subcategory	B37.3	Candidiasis of vulva and vagina
		Candidal vulvovaginitis
		Monilial vulvovaginitis
		Vaginal thrush
New code	B37.30	Candidiasis of vulva and vagina, not specified as recurrent
Add		Candidiasis of vulva and vagina NOS
New code	B37.31	Candidiasis of vulva and vagina, chronic
Add		Candidiasis of vulva and vagina, recurrent
New code	B37.39	Other candidiasis of vulva and vagina
Add		Acute candidiasis of vulva and vagina

ICD-10-CM TABULAR OF DISEASES - PROPOSED ADDENDA

All proposed effective October 1, 2021

B74 Filariasis

Excludes2: onchocerciasis (B73)

Revise tropical (pulmonary) eosinophilia NOS ~~(J82) (~~J82.89)

B97.4 Respiratory syncytial virus as the cause of diseases classified elsewhere

Delete ~~Excludes2: acute bronchiolitis due to respiratory syncytial virus (RSV) (J21.0)~~

Delete ~~acute bronchitis due to respiratory syncytial virus (RSV) (J20.5)~~

Delete ~~respiratory syncytial virus (RSV) pneumonia (J12.1)~~

Add Excludes1: acute bronchiolitis due to respiratory syncytial virus (RSV) (J21.0)

Add acute bronchitis due to respiratory syncytial virus (RSV) (J20.5)

Add respiratory syncytial virus (RSV) pneumonia (J12.1)

C25 Malignant neoplasm of pancreas

Revise Code also, if applicable exocrine pancreatic insufficiency (K86.81)

D47 Other neoplasms of uncertain behavior of lymphoid, hematopoietic and related tissue

D47.Z Other specified neoplasms of uncertain behavior of lymphoid, hematopoietic and related tissue

D47.Z2 Castleman disease

Revise Code also, if applicable human herpesvirus 8 infection (B10.89)

D57 Sickle-cell disorders

D57.4 Sickle-cell thalassemia

D57.41 Sickle-cell thalassemia with crisis

D57.419 Sickle-cell thalassemia, unspecified, with crisis

Sickle-cell thalassemia with (painful) crisis NOS

Revise Sickle-cell thalassemia with vasoocclusive pain NOS

D57.43 Sickle-cell thalassemia beta zero with crisis

D57.439 Sickle-cell thalassemia beta zero with crisis, unspecified

HbS-beta zero with other specified complication

Sickle-cell beta zero with crisis unspecified

Sickle-cell thalassemia beta zero with (painful) crisis NOS

Revise Sickle-cell thalassemia beta zero with vasoocclusive pain NOS

D57.45 Sickle-cell thalassemia beta plus with crisis

D57.459 Sickle-cell thalassemia beta plus with crisis, unspecified

HbS-beta plus with crisis with unspecified complication

Sickle-cell beta plus with crisis with unspecified complication

Sickle-cell thalassemia beta plus with (painful) crisis NOS

Revise Sickle-cell thalassemia beta plus with vasoocclusive pain NOS

D57.8 Other sickle-cell disorders

D57.81Other sickle-cell disorders with crisis

D57.813 Other sickle-cell disorders with cerebral

vascular involvement

Revise Code also, if applicable~~:~~ ~~-~~cerebral infarction (I63.-)

D72 Other disorders of white blood cells

D72.1 Eosinophilia

Revise Excludes2: Löffler's syndrome ~~(~~J82~~)~~ ~~(~~J82.89)

Revise pulmonary eosinophilia ~~(~~J82~~)~~ (J82.-)

E21 Hyperparathyroidism and other disorders of parathyroid gland

Delete Excludes1: ~~familial hypocalciuric hypercalcemia (E83.52)~~

Add Excludes2: familial hypocalciuric hypercalcemia (E83.52)

F10 Alcohol related disorders

F10.1 Alcohol abuse

Revise F10.13 Alcohol abuse~~,~~ ~~-~~with withdrawal

F84 Pervasive developmental disorders

Delete ~~Use additional code to identify any associated medical condition and intellectual disabilities~~

Add Code also to identify any associated medical condition and intellectual disabilities

G02 Meningitis in other infectious and parasitic diseases classified elsewhere

Revise Excludes1: infectious mononucleosis complicated by meningitis (B27.- with ~~fourth~~ fifth character 2)

G63 Polyneuropathy in diseases classified elsewhere

Excludes1: infectious mononucleosis (B27.0-B27.9

Revise with fifth character 1)

G71 Primary disorders of muscles

G71.2 Congenital myopathies

Revise G71.20 Congenital myopathy, ~~unspecifed~~ unspecified

H35 Other retinal disorders

H35.0 Background retinopathy and retinal vascular changes

Revise Code also any associated hypertension (I10~~.~~)

Ischemic heart diseases (I20-I25)

Delete ~~Use additional code to identify presence of hypertension (I10-I16)~~

Add Code also to identify presence of hypertension (I10-I16)

J05 Acute obstructive laryngitis [croup] and epiglottitis

Revise Code also influenza, if present such as:

influenza due to identified novel influenza A virus with other respiratory manifestations (J09.X2)

influenza due to other identified influenza virus with other respiratory manifestations (J10.1)

influenza due to unidentified influenza virus with other respiratory manifestations (J11.1)

J09 Influenza due to certain identified influenza viruses

J09.X Influenza due to identified novel influenza A virus

J09.X2 Influenza due to identified novel influenza A virus with other

Add Influenza due to other identified influenza virus with other respiratory manifestations

J10 Influenza due to other identified influenza virus

J10.1 Influenza due to other identified influenza virus with other

Add — Influenza due to unidentified influenza virus with other respiratory manifestations

J44 Other chronic obstructive pulmonary disease

Revise — Includes: chronic bronchitis with ~~airways~~ airway obstruction

J45 Asthma

Revise — Excludes1: eosinophilic asthma ~~(J82)~~ (J82.83)

J47 Bronchiectasis

J47.0 Bronchiectasis with acute lower respiratory infection

Bronchiectasis with acute bronchitis

Delete — ~~Use additional code to identify the infection~~

Add — Code also to identify infection, if applicable

K37 Unspecified appendicitis

Revise — Excludes1: - unspecified appendicitis with peritonitis (K35.2-, K35.3-)

K59 Other functional intestinal disorders

K59.0 Constipation

K59.03 Drug induced constipation

Revise — Use additional code for adverse effect, if applicable, to identify drug (T36-T50 with fifth or sixth character 5)

K72 Hepatic failure, not elsewhere classified

Revise K72.1Chronic hepatic failure (end stage liver disease)

K77 Liver disorders in diseases classified elsewhere

Revise Excludes1: infectious mononucleosis with liver disease

(B27.0-B27.9 ~~with .9~~)

L89 Pressure ulcer

L89.0 Pressure ulcer of elbow

L89.01 Pressure ulcer of right elbow

L89.019 Pressure ulcer of right elbow, unspecified stage

Healing pressure ulcer of right of elbow NOS

Delete ~~Healing pressure ulcer of right elbow, unspecified stage healing pressure of right elbow~~

L89.02 Pressure ulcer of left elbow

L89.029 Pressure ulcer of right elbow, unspecified stage

Healing pressure ulcer of right of elbow NOS

Delete ~~Healing pressure ulcer of right elbow, unspecified stage healing pressure of right elbow~~

L99 Other disorders of skin and subcutaneous tissue in diseases classified elsewhere

Code first underlying disease, such as: amyloidosis (E85.-)

Revise Excludes1: skin disorders in diabetes (E08-E13 ~~with .62~~)

M34 Systemic sclerosis [scleroderma]

M34.8 Other forms of systemic sclerosis

M34.81 Systemic sclerosis with lung involvement

Add Code also if applicable:

Add other interstitial pulmonary diseases (J84.89)

Add secondary pulmonary arterial hypertension (I27.21)

M40 Kyphosis and lordosis

M40.1 Other secondary kyphosis

Add Code first underlying disease

M41 Scoliosis

M41.5 Other secondary scoliosis

Add Code first underlying disease

O10 Pre-existing hypertension complicating pregnancy, childbirth and the puerperium

O10.0 Pre-existing essential hypertension complicating pregnancy, childbirth and the puerperium

Revise O10.01 Pre-existing essential hypertension complicating pregnancy~~,~~

O22 Venous complications and hemorrhoids in pregnancy

O22.3 Deep phlebothrombosis in pregnancy

Deep vein thrombosis, antepartum

Revise Use additional code to identify the deep vein thrombosis

(I82.4-, I82.5-, I82.62~~-~~, I82.72-)

O87 Venous complications and hemorrhoids in the puerperium

O87.1 Deep phlebothrombosis in the puerperium

Revise Use additional code to identify the deep vein thrombosis

(I82.4-, I82.5-, I82.62~~-~~, I82.72-)

P08 Disorders of newborn related to long gestation and high birth weight

P08.1 Other heavy for gestational age newborn

Revise Excludes1: syndrome of infant of mother with gestational diabetes (P70.0)~~.~~

Q21 Congenital malformations of cardiac septa

Q21.8 Other congenital malformations of cardiac septa

Eisenmenger's defect

Pentalogy of Fallot

Revise Code also, if applicable:

Eisenmenger's complex (I27.83)

Eisenmenger's syndrome (I27.83)

Q61 Cystic kidney disease

Q61.5 Medullary cystic kidney

Revise ~~Nephronopthisis~~ Nephronophthisis

S23 Dislocation and sprain of joints and ligaments of thorax

S23.1 Subluxation and dislocation of thoracic vertebra

Revise Code also any associated:

open wound of thorax (S21.-)

spinal cord injury (S24.0-, S24.1-)

S82 Fracture of lower leg, including ankle

Revise Excludes2: periprosthetic fracture around internal ~~of~~ prosthetic ~~implant of~~ knee joint ~~(M97.0-)~~ (M97.1)

T63 Toxic effect of contact with venomous animals and plants

T63.6 Toxic effect of contact with other venomous marine animals

Revise T63.61 Toxic effect of contact with ~~Portugese~~ Portuguese Man-o-war

Toxic effect of contact with bluebottle

Revise T63.611 Toxic effect of contact with ~~Portugese~~ Portuguese Man-o-war, accidental (unintentional)

Revise T63.612 Toxic effect of contact with ~~Portugese~~ Portuguese Man-o-war, intentional self-harm

Revise T63.613 Toxic effect of contact with ~~Portugese~~ Portuguese Man-o-war, assault

Revise T63.614 Toxic effect of contact with ~~Portugese~~ Portuguese Man-o-war, undetermined

Z09 Encounter for follow-up examination after completed treatment

for conditions other than malignant neoplasm

Medical surveillance following completed treatment

Revise Use additional code to identify any applicable history of

disease code (Z86.-~~,~~ Z87.-)

Z13 Encounter for screening for other diseases and disorders

Z13.2 Encounter for screening for nutritional, metabolic and other endocrine disorders

Z13.29 Encounter for screening for other suspected endocrine disorder

Delete ~~Excludes1: encounter for screening for diabetes mellitus (Z13.1)~~

Add Excludes2: encounter for screening for diabetes mellitus (Z13.1)

Z79 Long term (current) drug therapy

Z79.4 Long term (current) use of insulin

Delete ~~Excludes1: long term (current) use of oral antidiabetic drugs (Z79.84)~~

Delete ~~long term (current) use of oral hypoglycemic drugs~~

~~(Z79.84)~~

Add Excludes2: long term (current) use of oral antidiabetic drugs (Z79.84)

Add long term (current) use of oral hypoglycemic drugs (Z79.84)

Z92 Personal history of medical treatment

Z92.2 Personal history of drug therapy

Revise Z92.25 Personal history of immunosuppression therapy

Z99 Dependence on enabling machines and devices, not elsewhere classified

Delete ~~Excludes1: cardiac pacemaker status (Z95.0)~~

ICD-10-CM INDEX OF DISEASES - PROPOSED ADDENDA

All proposed effective October 1, 2021

Anemia (essential) (general) (hemoglobin deficiency) (infantile) (primary) (profound) D64.9
- postoperative (postprocedural)
Revise - - specified NEC ~~D64.9~~ J64.89

Arthritis, arthritic (acute) (chronic) (nonpyogenic) (subacute) M19.90
Add - facet joint (see also Spondylosis) M47.819

Arthropathy -*see also* Arthritis M12.9
Add - facet joint (see also Spondylosis) M47.819

Aspiration
Delete ~~food or foreign body (with asphyxiation) -*see* Asphyxia, food~~
Add - food or foreign body-*see* Foreign body, by site

Atrophy, atrophic (of)
Revise - nutritional ~~E41~~ E43
Add - - with marasmus E41

Burn (electricity) (flame) (hot gas, liquid or hot object) (radiation) (steam) (thermal) T30.0
Delete - ~~corrosion (external) (internal) -*see* Corrosion, by site~~
Add - corrosion (external) (internal) – see by site, second degree
Delete ~~- partial thickness - code as Burn, unspecified degree, by site~~
Add - partial thickness - code as Burn, by site, second degree

Bursitis M71.9
Revise - radiohumeral ~~M77.8~~ M70.3-

Cachexia R64
- pituitary E23.0
Add - pulmonary R64

Calcification
- adrenal (capsule) (gland) E27.49
Revise - - tuberculous ~~E35 [B90.8]~~ A18.7

Calicectasis N28.89
Add - due to obstruction – see Hydronephrosis

Revise Chylothorax (nonfilarial) ~~I89.8~~ J94.0

Complication(s) (from) (of)
- catheter (device) NEC -see also Complications, prosthetic device or implant
- - dialysis (vascular) T82.9
Revise - - - intraperitoneal ~~-see Complications, catheter, intraperitoneal~~
- see Complication, catheter, intraperitoneal dialysis
- joint prosthesis, internal T84.9
- - mechanical
Delete ~~- - - perforation -*see* Complications, joint prosthesis, mechanical, specified NEC~~
Add - - - periprosthetic
Delete ~~- - - - osteolysis T84.059~~
Add - - - - osteolysis by site T84.05
Revise - - intraperitoneal (dialysis) catheter ~~-see Complications, catheter, intraperitoneal~~
- see Complication, catheter, intraperitoneal dialysis
- - mechanical NEC T85.698
- - - dialysis catheter (vascular) -see also Complication, catheter, dialysis, mechanical
Revise - - - - peritoneal ~~-see Complication, catheter, intraperitoneal, mechanical~~
- see Complication, catheter, intraperitoneal dialysis
Delete ~~- periprosthetic -see Complications, joint prosthesis, mechanical,~~
Add - periprosthetic, osteolysis, by site
- prosthetic device or implant T85.9
Revise - - intraperitoneal (dialysis) catheter ~~-see Complications, catheter, intraperitoneal~~
- see Complication, catheter, intraperitoneal dialysis

Cyst (colloid) (mucous) (simple) (retention)
- nervous system NEC G96.89
Delete ~~- perineural G96.191~~
-neuroenteric (congenital) Q06.8
- pericoronal K09.0
Add - perineural G96.191
- periodontal K04.8
- prosthetic device or implant T85.9

Deformity Q89.9
- meninges or membrane (congenital) Q07.9
Revise - - spinal cord (congenital) ~~G96.198~~ Q06.-

Dermatitis (eczematous) L30.9
- contusiformis L52
Add - desquamative L30.8

- dysmenorrheica N94.6

Delete ~~- desquamative L30.8~~

Revise - eyelid – (see Also Dermatosis, eyelid) H01.9

Add - - specified NEC H01.8

Dependence (on) (syndrome) F19.20
- alcohol (ethyl) (methyl) (without remission) F10.20

Add - - in remission F10.21

Dermatosis L98.9
- eyelid (noninfectious) H01.9

Revise ~~- - dermatitis -see Dermatitis, eyelid~~
- eyelid ~~– see also Dermatosis, eyelid~~ H01.9

Diabetes, diabetic (mellitus) (sugar) E11.9
- with

Add - - coma due to

Add - - - hypoglycemia E11.641

Add - - - hyperosmolarity E11.01

Add - - - ketoacidosis E11.11

Dilatation
- calyx N28.89

Add - due to obstruction – see Hydronephrosis
- kidney N28.89

Add - due to obstruction – see Hydronephrosis

Disease, diseased -*see also* Syndrome
- joint -see also Disorder, joint

Add - - facet joint (see also Spondylosis) M47.819
- - end stage K72.90

Add - - - with coma K72.91

Add - - - acute K72.00

Add - - - - with coma K72.01

Add - - - chronic K72.10

Add - - - - with coma K72.11

Revise - myelodysplastic, not classified ~~C94.6~~ D46.-
- wasting NEC R64

Revise - - due to malnutrition ~~E41~~ E43

Add - - - with marasmus E41

Disorder (of) -*see also* Disease
Revise - ~~gluecose~~ glucose transport E74.819

Delete ~~Duplay's bursitis or periarthritis -*see* Tendinitis, calcific, shoulder~~
Add Duplay's bursitis or periarthritis M75.0

Effusion
Revise - amniotic fluid -see Pregnancy, complicated by, ~~prematue~~ premature rupture of membranes

Revise Emaciation (due to malnutrition) (see also Cachexia) E43
Add - with marasmus E41

Enthesopathy (peripheral)
Revise - ankle and tarsus ~~M77.9~~ M77.8
Revise - elbow region (see also Bursitis, elbow) M77.8
Revise - foot NEC ~~M77.9~~ M77.5-
Revise - lower limb (excluding foot) ~~M76.9~~ M76.8-
Revise - shoulder (region) –(see also Lesion, shoulder) M75.9
Add - - adhesive M75.0
Add - - specified type NEC M75.8

Examination
Delete ~~- forced sexual exploitation Z04.81~~
- forced labor exploitation Z04.82
Add - forced sexual exploitation Z04.81

- medicolegal reason NEC Z04.89
- - following
Delete ~~- - - forced sexual exploitation Z04.81~~
- - - forced labor exploitation Z04.82
Add - - - forced sexual exploitation Z04.81

Add Exploitation
Add - labor
Add - - confirmed
Add - - - adult forced T74.61
Add - - - child forced T74.62
Add - - suspected
Add - - - adult forced T76.61
Add - - - child forced T76.62
Add - sexual
Add - - confirmed

Add - - - adult forced T74.51
Add - - - child T74.52
Add - - suspected
Add - - - adult forced T76.51
Add - - - child T76.52

Erosion
Revise - implanted mesh -see Complications, prosthetic ~~devise~~ device or implant, mesh

Failure, failed
- heart (acute) (senile) (sudden) I50.9
- - with
- - - acute pulmonary edema -*see* Failure, ventricular, left
Revise - - - decompensation ~~– *see also* Failure, heart, by type as diastolic or systolic, acute and chronic~~ I50.9
Add - - - - with
Add - - - - - normal ejection fraction I50.33
Add - - - - - preserved ejection fraction I50.33
Add - - - - - reduced ejection fraction I50.23
Add - - - - - - with diastolic dysfunction I50.43
Add - - - - combined systolic and diastolic I50.43
Add - - - - diastolic I50.33
Add - - - - right I50.813
Add - - - - systolic I50.23

Gastropathy K31.9
Revise -congestive portal (see also, hypertension, portal) K31.89
Revise -portal hypertensive (see also, hypertension, portal) K31.89

Hepatitis K75.9
- fulminant NEC (viral) -see Hepatitis, viral
Add - ischemia, ischemic- K72.00
- reactive, nonspecific K75.2
Add - shock K72.00

Hypertrophy, hypertrophic
Add - facet joint (see also Spondylosis) M47.819

Inhalation
Revise - steam (see also Burn, respiratory tract) T59.-

Injury -see also specified injury type T14.90
- nerve ...

Revise - - lumbar spinal ~~- see Injury, nerve, spinal, lumbar~~
Add - - - peripheral S34.6
Add - - - root S34.21
Add - - - sympathetic S34.5
Revise - - sacral spinal ~~- see Injury, nerve, spinal, sacral~~
Add - - - peripheral S34.6
Add - - - root S34.22
Add - - - sympathetic S34.5

Long-term (current) (prophylactic) drug therapy (use of)
Add - non-insulin antidiabetic drug, injectable Z79.899

Migraine (idiopathic) G43.909
- menstrual G43.829
Revise - - not intractable ~~4G43.829~~ G43.829

Mycobacterium, mycobacterial (infection) A31.9
Revise - tuberculosis (human, bovine) – ~~seeTuberculosis~~ see Tuberculosis

Obstruction, obstructed, obstructive
Revise - kidney (calices) (see also Hydronephrosis) N28.89
Revise - renal (see also Hydronephrosis) N28.89

Osteoarthritis
Revise - generalized (multiple joints) M15.9

Delete ~~- primary M19.91~~
Add - primary generalized M15.0

Osteoarthropathy (hypertrophic) M19.90
Delete ~~- multiple site - *see* Osteoarthritis, primary, multiple joint~~
Add - multiple sites - see Osteoarthritis, generalized (multiple joints), primary M15.0

Osteolysis M89.50
Delete - ~~joint prosthesis (periprosthetic) - see Complications, joint prosthesis, mechanical,~~
- - ~~periprosthetic, osteolysis, by site~~

Osteomyelitis
- acute M86.10
- - hematogenous M86.00
Revise - - - ilium ~~M86.059~~ M86.08
Revise - - - ischium ~~M86.059~~ M86.08

Pancreatitis (annular) (apoplectic) (calcareous) (edematous) (hemorrhagic) (malignant)
- recurrent
Revise - - acute ~~K85.50~~ K85.9-

Polyneuropathy
- in (due to)
Revise - - transthyretin-related (ATTR) familial amyloid E85.1

Presence (of)
Revise - ICD (cardioverter-~~defribillator~~ defibrillator) Z95.810

Schizophrenia, schizophrenic F20.9
Add - chronic F20.89

Revise Sciatica (infective) M54.3

Spondylosis M47.9
- specified NEC M47.899
Add - - facet joint (see also Spondylosis) M47.819

Stenosis, stenotic (cicatricial) -*see also* Stricture
Revise - heart valve ~~(congenital) Q24.8~~ (see Stenosis, valve)
Delete ~~- - aortic Q23.0~~
Delete ~~- - mitral Q23.2~~
Delete ~~- - pulmonary Q22.1~~
Delete ~~- - tricuspid Q22.4~~

Syndrome -*see also* Disease
Add - facet joint (see also Spondylosis) M47.819
- hypereosinophilic (HES)
- - lymphocytic variant (LHES) D72.111
Add - - myeloid D72.118

Test, tests, testing (for)
Revise - blood-alcohol ~~Z04.89~~ Z02.83
Revise - blood-drug ~~Z04.89~~ Z02.83

Thrombosis, thrombotic (bland) (multiple) (progressive) (silent) (vessel) I82.90
- vein (acute) I82.90
- - deep (DVT) I82.40-
Revise - - - upper leg I82.4Y.-
Revise - - - - chronic I82.5~~y~~Y--

Varix
Revise - leg (asymptomatic) ~~I83.90~~ ~~– I83.9-~~
- - with
Revise - - - ulcer ~~I83.009~~ ~~I83.-~~
Revise - - - - with inflammation ~~I83.209~~ ~~I83.2-~~

Virus, viral -see also condition
- as cause of disease classified elsewhere B97.89
Revise - - respiratory syncytial virus (RSV) ~~-see Virus, infection, respiratory syncytial (RSV)~~ See Virus, respiratory syncytial (RSV)

Wasting
- disease R64
Revise - - due to malnutrition ~~E41-~~ E43
Add - - - with marasmus E41
Revise - extreme (due to malnutrition) ~~E41~~ E43
Add - - with marasmus E41

Wernicke-Korsakoff's syndrome or psychosis (alcoholic) F10.96
Add -Wernicke-Korsakoff (nonalcoholic) F04

Xerosis
Revise - cutis (dry skin) L85.3