CENTERS FOR DISEASE CONTROL AND PREVENTION

ICD-10 Coordination and Maintenance Committee Meeting
Diagnosis Agenda
March 5-6, 2019
Part 2

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

Diagnosis Topics:

Contents

ICD-10 TIMELINE

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

March 5-6, 2019	ICD-10 Coordination and Maintenance Committee Meeting.
	Those who wish to attend the ICD-10 Coordination and Maintenance Committee meeting **must have registered for the meeting online by February 22, 2019.** You must bring an official form of picture identification (such as a driver's license) in order to be admitted to the building.
	In compliance to The Real ID Act, enacted in 2005, (http://www.dhs.gov/real-id-enforcement-brief) the following states/territories: Maine, Minnesota, Missouri, Montana and Washington State **will not** gain access into any Federal Agencies using the **above states** driver's license or ID. This means CMS visitors from these states/territories will need to provide alternative proof of identification (**such as a passport**) to gain entrance into Baltimore-based and Bethesda CMS buildings, as well as the Humphrey Building in Washington.
March 2019	Webcast of the March 5-6, 2019 ICD-10 Coordination and Maintenance Committee meeting will be posted on the CMS webpage as follows: https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html
April 1, 2019	There were no requests for ICD-10 codes to capture new diagnoses or new technology for implementation on April 1, 2019. Therefore, there will be no new ICD-10 diagnosis or procedure codes implemented on April 1, 2019.
April 5, 2019	**Deadline for receipt of public comments on proposed new ICD-10-PCS codes and revisions discussed at the March 5-6, 2019 ICD-10**

	Coordination and Maintenance Committee meetings for implementation on October 1, 2019.
April 2019	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 finalized FY 2020 ICD-10-CM diagnosis and ICD-10-PCS procedure codes 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: http://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/AcuteInpatientPPS/index.html?redirect=/AcuteInpatientPPS/IPPS/list.asp
May 10, 2019	**Deadline for receipt of public comments on proposed new diagnoses codes and revisions discussed at the March 5-6, 2019 ICD-10 Coordination and Maintenance Committee meetings for implementation on October 1, 2020.**
June 2019	Final addendum posted on web pages as follows: Diagnosis addendum - http://www.cdc.gov/nchs/icd/icd10cm.htm Procedure addendum - http://cms.hhs.gov/Medicare/Coding/ICD10/index.html
June 14, 2019	**Deadline for requestors: Those members of the public requesting that topics be discussed at the September 10-11, 2019 ICD-10 Coordination and Maintenance Committee meeting must have their requests submitted to CMS for procedures and NCHS for diagnoses.**
August 1, 2019	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, 2019. This rule can be accessed at:

	http://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/AcuteInpatientPPS/index.html?redirect=/AcuteInpatientPPS/IPPS/list.asp
August 2019	Tentative agenda for the Procedure part of the September 10-11, 2019 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 part of the September 10-11, 2019 ICD-10 Coordination and Maintenance Committee meeting will be posted on the NCHS webpage at - http://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm
	Federal Register notice for the September 10-11, 2019 ICD-10 Coordination and Maintenance Committee meeting will be published. This will include the tentative agenda.
August 2, 2019	**On-line registration opens for the September 10-11, 2019 ICD-10 Coordination and Maintenance Committee meeting at: https://www.cms.gov/apps/events/default.asp**
September 2, 2019	Because of increased security requirements, those wishing to attend the September 10-11, 2019 ICD-10 Coordination and Maintenance Committee meeting must register for the meeting online at: **https://www.cms.gov/apps/events/default.asp** **Attendees must register online by September 2, 2019; failure to do so may result in lack of access to the meeting.**
September 10-11, 2019	ICD-10 Coordination and Maintenance Committee Meeting.

	Those who wish to attend the ICD-10 Coordination and Maintenance Committee meeting **must have registered for the meeting online by September 2, 2019.** You must bring an official form of picture identification (such as a driver's license) in order to be admitted to the building.
September 2019	Webcast of the September 10-11, 2019 ICD-10 Coordination and Maintenance Committee meeting will be posted on the CMS webpage as follows: https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html
October 1, 2019	New and revised ICD-10-CM and ICD-10-PCS codes go into effect along with DRG changes. Final addendum available on web pages as follows: Diagnosis addendum – http://www.cdc.gov/nchs/icd/icd10cm.htm Procedure addendum – http://www.cms.gov/Medicare/Coding/ICD10/
October 11, 2019	**Deadline for receipt of public comments on proposed new ICD-10-CM codes discussed at the September 10-11, 2019 ICD-10 Coordination and Maintenance Committee meetings for implementation on April 1, 2020.**
November 2019	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, 2020 will be posted on the following websites: http://www.cdc.gov/nchs/icd/icd10cm.htm http://www.cms.gov/Medicare/Coding/ICD10/

Webcast and Dial-In Information

- Day 1: March 5, 2019: The meeting will begin at 9:00 AM ET and will end promptly at 1:00 PM ET. There will not be a lunch break for this session. The meeting will be webcast via CMS at http://www.cms.gov/live/.
- Day 2: March 6, 2019: The meeting will begin at 9:00 AM ET and will end at 5:00 PM ET. Lunch will be held from 11:30 AM ET to 1:00 PM ET. The meeting will be webcast via CMS at http://www.cms.gov/live/.
- Toll-free dial-in access is available for listen-only participants who cannot join the webcast:
  Day 1-March 5, 2019: Phone: 1-877-267-1577; Meeting 990 668 147.
  Day 2-March 6, 2019: Phone: 1-877-267-1577; Meeting 990 668 147.
  We encourage you to join early, as the number of phone lines is limited.

Contact Information
Mailing address:

National Center for Health Statistics
ICD-9-CM Coordination and Maintenance Committee
3311 Toledo Road
Hyattsville, Maryland 20782
Fax: (301) 458-4022

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

Donna Pickett	(301) 458-4434
David Berglund	(301) 458-4095
Cheryl Bullock	(301) 458-4297
Shannon McConnell-Lamptey	(301) 458-4612
Traci Ramirez	(301) 458-4454

Continuing Education Credits

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

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

If you plan to 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 a 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.

Babesiosis

Human babesiosis is a tick-borne zoonosis caused by intra-erythrocytic protozoa of the genus *Babesia*. Babesiosis can also be transmitted by transfusion of blood and blood components collected from an infected donor. Although the majority of U.S. babesiosis cases are caused by *B. microti,* which is prevalent in the Northeast and upper Midwest, other *Babesia* species such as *B. duncani, B. divergens,* and other species have been implicated in transmission in multiple U.S. states. This proposal is based on a request for new specific codes for multiple species of *Babesia,* received from Office of Biostatistics and Epidemiology, Center for Biologics Evaluation and Research (CBER), of the Food and Drug Adminstration (FDA).

As the majority of Babesia infections are presumed to be asymptomatic and may never be diagnosed, especially in younger persons, babesiosis may persist undetected. Transfusion of blood and blood components collected from asymptomatic donors may result in transfusion-transmitted babesiosis (TTB), leading to potentially fatal clinical illness in blood transfusion recipients. It has been also determined by FDA that babesiosis is a transfusion-transmitted infection as the disease agent was identified to be fatal or life-threatening and is transmissible by blood or blood components.

Over the past decade, there have been a growing number of reported cases in the United States, including TTB cases. Babesiosis infections have also been spreading to non-endemic states. In response, efforts are being made to mitigate the risk of human babesiosis infections, including the development of donor screening tests and testing strategies. On March 6, 2018, FDA licensed two independent assays for screening donors for *B. microti:* the Imugen *Babesia microti* Arrayed Fluorescent Immunoassay (AFIA) for the detection of *B. microti*-specific antibodies and the Imugen *Babesia microti* Nucleic Acid Test (NAT) for the detection of DNA of *B. microti*. On January 24, 2019, FDA approved a nucleic acid amplification test for donor screening, to detect specific *Babesia* species: *B. microti*, *B. duncani*, *B. divergens*, and *B. venatorum*

Creation of new codes for species of *Babesia* would improve *Babesia* infection coding granularity, to allow physicians to code the *Babesia spp.* accurately and to help FDA monitor safety of the blood supply for different *Babesia spp.* The addition of species-specific *Babesia* coding granularity will also allow FDA to more easily provide appropriate donor testing recommendations depending on geographic distribution and spread of different *Babesia spp.* in various regions of the country. Specifically, the improved coding granularity will enable the FDA to monitor spread of different *Babesia* species in the U.S. by states and counties of residence over time as well as from disease-endemic areas to non-endemic areas, thus allowing FDA to recommend appropriate donor testing strategies to prevent transfusion transmission of different *Babesia* species as tests for babesiosis are currently species-specific (i.e. no cross-reactivity between species). The improved *Babesia* coding granularity will also stimulate development of new *Babesia* species-specific donor tests and tests that can detect multiple Babesia species, which will further improve detection of blood donors positive for *Babesia spp.* and prevent transfusion-transmission. In summary, the added *Babesia* coding granularity will allow FDA to assess distribution of different *Babesia spp.* in the United States using large databases as required by FDAAA of 2007, make suitable recommendations for appropriate donor testing, stimulate development of species-specific testing, and prevent transfusion-transmitted *Babesia* infections and donor loss, thus helping to assure a safe and adequate blood

supply. Finally, the improved coding granularity will enhance *Babesia* coding accuracy and level of detail allowing physicians to make species-specific diagnosis.

References

Conrad, P.A., et al., Description of Babesia duncani n.sp. (Apicomplexa: Babesiidae) from humans and its differentiation from other piroplasms. Int J Parasitol, 2006. 36(7):779-789. https://doi.org/10.1016/j.ijpara.2006.03.008

FDA. Recommendation for reducing the risk of transfusion-transmitted babesiosis: draft guidance for industry [Internet]. Silver Spring (MD): US Food and Drug Administration;2018 [cited 2018 Aug 24]. Available from: https://www.fda.gov/downloads/BiologicsBloodVaccines/GuidanceComplianceRegulatoryInformation/Guidances/Blood/UCM614734.pdf

Gubernot DM, Lucey CT, Lee KC, Conley GB, Holness LG, Wise RP. Babesia infection through blood transfusions: reports received by the U.S. Food and Drug Administration, 1997–2007. Clin Inf Dis 2009;48:25–30. https://doi.org/10.1086/595010

Gubernot DM, Nakhasi HL, Mied PA, Asher DM, Epstein JS, Kumar S. Transfusion-transmitted babesiosis in the United States: summary of a workshop. Transfusion 2009 Dec;49(12):2759-71. https://doi.org/10.1111/j.1537-2995.2009.02429.x

Herwaldt, B.L., et al., Transfusion-associated babesiosis in the United States: a description of cases. Ann Intern Med, 2011. 155(8):509-519. https://doi.org/10.7326/0003-4819-155-8-201110180-00362

Krause, P.J., et al., Increasing health burden of human babesiosis in endemic sites. Am J Trop Med Hyg, 2003. 68(4):431-436. Retrieved from http://www.ajtmh.org/cgi/pmidlookup?view=long&pmid=12875292

Menis M, et. al. Babesiosis Occurrence among the Elderly in the United States, as Recorded in Large Medicare Databases during 2006-2013. PLoS One. 2015 Oct 15;10(10):e0140332. https://doi.org/10.1371/journal.pone.0140332

Menis M, Anderson SA, Izurieta HS, Kumar S, Burwen DR, Gibbs J, et al. Babesiosis among elderly Medicare beneficiaries, United States, 2006–2008. Emerg Infect Dis. 2012 Jan;18(1):128-31. https://doi.org/10.3201/eid1801.110305

Persing, D.H., et al., Infection with a babesia-like organism in northern California. N Engl J Med, 1995. 332(5):298-303. https://doi.org/10.1056/NEJM199502023320504

Vannier EG, Diuk-Wasser MA, Ben Mamoun C, Krause PJ. Babesiosis. Infect Dis Clin North Am. 2015 Jun;29(2):357-70. https://doi.org/10.1016/j.idc.2015.02.008

Vannier, E. and P.J. Krause, Human babesiosis. N Engl J Med, 2012. 366(25): 2397-2407. https://doi.org/10.1056/NEJMra1202018

TABULAR MODIFICATIONS

B60 Other protozoal diseases, not elsewhere classified

B60.0 Babesiosis
Piroplasmosis

New code	B60.00	Babesiosis, unspecified
Add		Babesiosis due to unspecified Babesia species
Add		Piroplasmosis, unspecified
New code	B60.01	Babesiosis due to Babesia microti
Add		Infection due to B. microti
New code	B60.02	Babesiosis due to Babesia duncani
Add		Infection due to B. duncani and B. duncani-type species
New code	B60.03	Babesiosis due to Babesia divergens
Add		Infection due to B. divergens and B. divergens-like strains
Add		Babesiosis due to Babesia MO-1
New code	B60.09	Other babesiosis and piroplasmosis
Add		Babesiosis due to Babesia venatorum
Add		Babesiosis due to Babesia KO-1
Add		Infection due to other Babesia species
Add		Infection due to other protozoa of the order Piroplasmida

C3 Glomerulopathy

The Renal Physicians Association (RPA) has requested new ICD-10-CM codes for C3 glomerulopathy (C3G), a newly classified, uncommon kidney disorder characterized by the deposition of complement component 3 (C3) within the glomeruli. C3G is comprised of two distinct clinical subtypes: C3 glomerulonephritis (C3GN) and dense deposit disease (DDD). While DDD has specific ICD-10-CM codes, there are no specific codes for C3G or the subtype C3GN.

Glomerulonephritis (GN) involves inflammation of kidney glomeruli, which are responsible for filtering the blood. This enables the kidneys to maintain electrolyte balance, remove waste, and control fluid homeostasis. The etiology of GN results from different sources of inflammation, with autoimmune disorders identified as a leading cause. GN is classified into pathogenic types, which have been defined by the classification forms seen in renal biopsy. One of these types is C3 glomerulopathy.

Membranoproliferative glomerulonephritis (MPGN) has a histopathological pattern of renal injury characterized by the thickening of capillary walls and mesangial enlargement secondary to increased cellularity and matrix deposition. The traditional classification of MPGN was based on the pattern and location of deposits visible via electron microscopy rather than the specific etiology of those deposits. MPGN type 1 has mesangial and subendothelial electron dense deposits, MPGN type 2 has electron dense material in the glomerular basement membrane, and MPGN type 3 has subepithelial deposits with basement membrane spikes. However, with that basis, the current codes for glomerulonephritis (GN) do not include a specific diagnosis code for C3GN.

Specific coding for the C3G two subtypes DDD and C3GN is critical for identifying appropriate disease etiology, patient segmentation, and therapeutic selection. More importantly, this coding aligns with clinical consensus reached by a group of experts in renal pathology, nephrology, complement biology, and complement therapeutics.

References

Bomback AS, Appel GB. Pathogenesis of the C3 glomerulopathies and reclassification of MPGN. *Nat Rev Nephrol.* 2012 Nov;8(11):634-642. https://doi.org/10.1038/nrneph.2012.213

Pickering, MC., et al. C3 glomerulopathy: consensus report. *Kidney International*, 84(6):1079-1089, 2013. https://dx.doi.org/10.1038%2Fki.2013.377

Sethi S, et al. Mayo Clinic/Renal Pathology Society Consensus Report on Pathologic Classification, Diagnosis, and Reporting of GN. *J Am Soc Nephrol*. 27:1278-1284, 2016. https://doi.org/10.1681/ASN.2015060612

TABULAR MODIFICATIONS

N00 Acute nephritic syndrome

N00.5 Acute nephritic syndrome with diffuse mesangiocapillary glomerulonephritis
Add — Excludes1: Acute nephritic syndrome with C3 glomerulopathy (N00.A)
Add — Acute nephritic syndrome with C3 glomerulonephritis (N00.A)

N00.6 Acute nephritic syndrome with dense deposit disease
Add — Acute nephritic syndrome with C3 glomerulopathy with dense deposit disease
Revise — Acute nephritic syndrome with membranoproliferative glomerulonephritis, type_2

New code — N00.A Acute nephritic syndrome with C3 glomerulonephritis
Add — Acute nephritic syndrome with C3 glomerulopathy, NOS
Add — Excludes1: Acute nephritic syndrome (with C3 glomerulopathy) with dense deposit disease (N00.6)

N01 Rapidly progressive nephritic syndrome

N01.5 Rapidly progressive nephritic syndrome with diffuse mesangiocapillary glomerulonephritis
Add — Excludes1: Rapidly progressive nephritic syndrome with C3 glomerulopathy (N01.A)
Add — Rapidly progressive nephritic syndrome with C3 glomerulonephritis (N01.A)

N01.6 Rapidly progressive nephritic syndrome with dense deposit disease
Add — Rapidly progressive nephritic syndrome with C3 glomerulopathy with dense deposit disease

New code — N01.A Rapidly progressive nephritic syndrome with C3 glomerulonephritis
Add — Rapidly progressive nephritic syndrome with C3 glomerulopathy, NOS
Add — Excludes1: Rapidly progressive nephritic syndrome (with C3 glomerulopathy) with dense deposit disease (N01.6)

N02 Recurrent and persistent hematuria

N02.5 Recurrent and persistent hematuria with diffuse mesangiocapillary glomerulonephritis

Add Excludes1: Recurrent and persistent hematuria with C3 glomerulopathy (N02.A)

Add Recurrent and persistent hematuria with C3 glomerulonephritis (N02.A)

N02.6 Recurrent and persistent hematuria with dense deposit disease

Add Recurrent and persistent hematuria with C3 glomerulopathy with dense deposit disease

New code N02.A Recurrent and persistent hematuria with C3 glomerulonephritis

Add Recurrent and persistent hematuria with C3 glomerulopathy

Add Excludes1: Recurrent and persistent hematuria (with C3 glomerulopathy) with dense deposit disease (N02.6)

N03 Chronic nephritic syndrome

N03.5 Chronic nephritic syndrome with diffuse mesangiocapillary glomerulonephritis

Add Excludes1: Chronic nephritic syndrome with C3 glomerulopathy (N03.A)

Add Chronic nephritic syndrome with C3 glomerulonephritis (N03.A)

N03.6 Chronic nephritic syndrome with dense deposit disease

Add Chronic nephritic syndrome with C3 glomerulopathy with dense deposit disease

New code N03.A Chronic nephritic syndrome with C3 glomerulonephritis

Add Chronic nephritic syndrome with C3 glomerulopathy

Add Excludes1: Chronic nephritic syndrome (with C3 glomerulopathy) with dense deposit disease (N03.6)

N04 Nephrotic syndrome

N04.5 Nephrotic syndrome with diffuse mesangiocapillary glomerulonephritis

Add Excludes1: Nephrotic syndrome with C3 glomerulopathy (N04.A)

Add Nephrotic syndrome with C3 glomerulonephritis (N04.A)

N04.6 Nephrotic syndrome with dense deposit disease

Add Nephrotic syndrome with C3 glomerulopathy with dense deposit disease

New code N04.A Nephrotic syndrome with C3 glomerulonephritis
Add Nephrotic syndrome with C3 glomerulopathy
Add Excludes1: Nephrotic syndrome (with C3 glomerulopathy) with dense deposit disease (N04.6)

N05 Unspecified nephritic syndrome

N05.5 Unspecified nephritic syndrome with diffuse mesangiocapillary glomerulonephritis
Add Excludes1: Unspecified nephritic syndrome with C3 glomerulopathy (N05.A)
Add Unspecified nephritic syndrome with C3 glomerulonephritis (N05.A)

N05.6 Unspecified nephritic syndrome with dense deposit disease
Add Unspecified nephritic syndrome with C3 glomerulopathy with dense deposit disease

New code N05.A Unspecified nephritic syndrome with C3 glomerulonephritis
Add Unspecified nephritic syndrome with C3 glomerulopathy
Add Excludes1: Unspecified nephritic syndrome (with C3 glomerulopathy) with dense deposit disease (N05.6)

N06 Isolated proteinuria with specified morphological lesion

N06.5 Isolated proteinuria with diffuse mesangiocapillary glomerulonephritis
Add Excludes1: Isolated proteinuria with C3 glomerulopathy (N06.A)
Add Isolated proteinuria with C3 glomerulonephritis (N06.A)

N06.6 Isolated proteinuria with dense deposit disease
Add Isolated proteinuria with C3 glomerulopathy with dense deposit disease

New code N06.A Isolated proteinuria with C3 glomerulonephritis
Add Isolated proteinuria with C3 glomerulopathy
Add Excludes1: Isolated proteinuria (with C3 glomerulopathy) with dense deposit disease (N06.6)

N07 Hereditary nephropathy, not elsewhere classified

N07.5 Hereditary nephropathy, not elsewhere classified with diffuse mesangiocapillary glomerulonephritis

Add — Excludes1: Hereditary nephropathy, not elsewhere classified with C3 glomerulopathy (N07.A)

Add — Hereditary nephropathy, not elsewhere classified with C3 glomerulonephritis (N07.A)

N07.6 Hereditary nephropathy, not elsewhere classified with dense deposit disease

Add — Hereditary nephropathy, not elsewhere classified with C3 glomerulopathy with dense deposit disease

New code — N07.A Hereditary nephropathy, not elsewhere classified with C3 glomerulonephritis

Add — Hereditary nephropathy, not elsewhere classified with C3 glomerulopathy

Add — Excludes1: Hereditary nephropathy, not elsewhere classified (with C3 glomerulopathy) with dense deposit disease (N07.6)

Eosinophilic Gastrointestinal Diseases

Specific codes for certain eosinophilic gastrointestinal diseases have been requested by the American Partnership for Eosinophilic Disorders (APFED), a 501(c)3 non-profit patient advocacy group, and the International Eosinophil Society, Inc. (IES), an organization of scientists and clinicians interested in the eosinophil and its roles in health and disease.

In 2008, new ICD-9-CM codes were created for certain eosinophilic gastrointestinal diseases, including codes 558.41, Eosinophilic gastroenteritis; 558.42, Eosinophilic colitis; 535.70, Eosinophilic gastritis, without mention of hemorrhage; and 535.71, Eosinophilic gastritis, with hemorrhage. However, in the transition to ICD-10-CM, the coding for eosinophilic gastritis and eosinophilic gastroenteritis were combined as one code, K52.81, Eosinophilic gastritis or gastroenteritis. In eosinophilic gastritis, elevated levels of eosinophils cause injury and inflammation to the stomach. In eosinophilic gastroenteritis, elevated levels of eosinophils cause injury and inflammation to both the stomach and small intestine, and potentially other parts of the gastrointestinal system. These disorders both are often associated with an increase in the number of blood eosinophils. It has been requested that K52.81 be expanded, to create two separate ICD-10-CM codes for these conditions.

Eosinophilic colitis typically presents with painful lower GI symptoms, and elevated levels of colonic or rectal eosinophils defined by histopathology on biopsy obtained during colonoscopy. The code K52.82 is titled Eosinophilic colitis, but it currently has inclusions for certain other very different conditions: allergic proctocolitis, food-induced eosinophilic proctocolitis, food protein-induced proctocolitis, and milk protein-induced proctocolitis.

It is important to differentiate these conditions. Thus, food protein induced enterocolitis syndrome (FPIES), allergic proctocolitis, and milk protein-induced proctocolitis should be excluded from eosinophilic colitis. FPIES rarely requires colonoscopy and typically is not associated with eosinophils. The conditions allergic proctocolitis, and milk protein-induced proctocolitis are similarly not specifically related to eosinophilic colitis, which is diagnosed specifically by levels of eosinophils found in the colon. Allergic proctocolitis and milk-protein colitis generally do not require colonoscopy, often occur in newborns, and are usually temporary (self-resolving diseases). It is proposed allergic proctocolitis and milk-protein proctocolitis has specific codes created at subcategory K52.2, Allergic and dietetic gastroenteritis and colitis, and that excludes notes be created for these under K52.82.

TABULAR MODIFICATIONS

K52 Other and unspecified noninfective gastroenteritis and colitis

K52.2 Allergic and dietetic gastroenteritis and colitis

Delete — Excludes2: ~~food protein-induced proctocolitis (K52.82)~~

New code — K52.23 Milk protein-induced proctocolitis

New code — K52.24 Allergic proctocolitis
Food-induced eosinophilic proctocolitis
Food protein-induced proctocolitis

K52.29 Other allergic and dietetic gastroenteritis and colitis
Food hypersensitivity gastroenteritis or colitis
Immediate gastrointestinal hypersensitivity

K52.8 Other specified noninfective gastroenteritis and colitis

K52.81 Eosinophilic gastritis or gastroenteritis

Delete — ~~Eosinophilic enteritis~~

New code — K52.810 Eosinophilic gastroenteritis

Add — Eosinophilic enteritis

New code — K52.811 Eosinophilic gastritis

K52.82 Eosinophilic colitis

Delete — ~~Allergic proctocolitis~~

Delete — ~~Food-induced eosinophilic proctocolitis~~

Delete — ~~Food protein-induced proctocolitis~~

Delete — ~~Milk protein-induced proctocolitis~~

Add — Excludes2: Allergic proctocolitis (K52.24)

Add — Food-induced eosinophilic proctocolitis (K52.24)

Add — Food protein-induced proctocolitis (K52.24)

Add — Food protein-induced enterocolitis syndrome (FPIES) (K52.21)

Add — Milk protein-induced proctocolitis (K52.23)

Food Insecurity

The Blue Cross Blue Shield of Vermont and the Yale School of Nursing are requesting new ICD-10-CM codes related to food insecurity. They note that there is broad national consensus that assessing the social determinants of health (SDH) is a key facet of improving both individual and population health outcomes. BCBS of Vermont notes that this is represented first in the policy aims of government programs such as Healthy People 2020 (Secretary's Advisory Committee on National Health Promotion and Disease Prevention Objectives for 2020, 2010) and the Center for Medicaid and Medicare Services Accountable Health Communities Project (Alley, Asomugha, Conway, & Sanghavi, 2016; Center for Medicare and Medicaid Innovation, 2017).

Within the social determinants, food insecurity is an established concept of concern. The United States Department of Agriculture publishes a yearly report on the state of food insecurity (Coleman-Jensen, Rabbitt, Gregory, & Singh, 2017). Professional organizations such as the American Academy of Family Physicians (AAFP), the American Academy of Pediatrics (AAP) and the Academy of Nutrition and Dietetics have policy statements on the need for social determinant and food insecurity screening and assessment (American Academy of Family Physicians, 2012; American Academy of Pediatrics, 2015; Holben & Marshall, 2017). To aid clinicians in practice, both the AAFP and the AAP have toolkits with specific steps for SDH and food insecurity screening and assessment (American Academy of Family Physicians, 2018; American Academy of Pediatrics & Food Research and Action Center, 2017)

Although ICD-10-CM has a code for food and water insufficiency (Z59.4, Lack of adequate food and drinking water), the concepts are joined, which makes tracking of each individual issue impossible. In addition, Z59.4 does not specifically represent the social and economic conditions that are inherent in the United States Department of Agriculture 2017 conceptual and operational definition of food insecurity. Lastly, there is no code to represent when patients with prescribed dietary needs are unable to follow a dietary regimen because of cost.

The submitter states that as we work toward more sensitive and specific screening and assessment for the social determinants of health in clinical practice, specific language for the mature concept of food insecurity is a necessary step.

The Blue Cross Blue Shield of Vermont and Yale School of Nursing request to aid patient and population care and study, is to divide Z59.4 capture food and water concepts separately, add a specific code at subcategory Z59.4 for the concern of food insecurity, and add a unique code at Z91.1 and Z71.8 to capture lack of compliance to dietary regimen due to financial hardship, a sequela of food insecurity.

This proposal is supported by a number of organizations and individuals including: the Academy of Nutrition and Dietetics, Colorado Prevention Alliance, Connecticut State Medical Society Blue Cross Blue Shield New York, and the Oregon Primary Care Association.

REFERENCES:

Alley, D. E., Asomugha, C. N., Conway, P. H., & Sanghavi, D. M. (2016). Accountable health communities - addressing social needs through medicare and medicaid. *New England Journal of Medicine, 374*, 8-11. doi:10.1056/NEJMp1512532

American Academy of Family Physicians. (2012). Social determinants of health policy. Retrieved from http://www.aafp.org/about/policies/all/social-determinants.html
American Academy of Family Physicians. (2018). The everyone project: Advancing health equity in every community. Retrieved from https://www.aafp.org/patient-care/social-determinants-of-health/cdhe/everyone-project/tools.html
American Academy of Pediatrics. (2015). Promoting food security for all children. *Pediatrics, 136*, e1431. doi:10.1542/peds.2015-3301
American Academy of Pediatrics, & Food Research and Action Center. (2017). *Addressing food insecurity: A toolkit for pediatricians.* Retrieved from http://frac.org/aaptoolkit
Center for Medicare and Medicaid Innovation. (2017). *The accountable health communities health-related social needs screening tool*. Retrieved from https://innovation.cms.gov/Files/worksheets/ahcm-screeningtool.pdf
Coleman-Jensen, A., Rabbitt, M., Gregory, C., & Singh, A. (2017). *Household food security in the united states in 2016 (err-237).* Retrieved from https://www.ers.usda.gov/webdocs/publications/84973/err-237.pdf
Holben, D. H., & Marshall, M. B. (2017). Position of the academy of nutrition and dietetics: Food insecurity in the united states. *Journal of the Academy of Nutrition and Dietetics, 117*, 1991-2002. doi:10.1016/j.jand.2017.09.027
Secretary's Advisory Committee on National Health Promotion and Disease Prevention Objectives for 2020. (2010). *Healthy people 2020: An opportunity to address societal determinants of health in the united states.* Retrieved from http://www.healthypeople.gov/2010/hp2020/advisory/SsocietalDeterminantsHealth.htm.

TABULAR MODIFICATION

Z59 Problems related to housing and economic circumstances

New subcategory

Z59.4 Lack of adequate food and safe drinking water
Inadequate drinking water supply
Excludes1:effects of hunger (T73.0)
inappropriate diet or eating habits (Z72.4)
malnutrition (E40-E46)

New code

Z59.41 Lack of adequate food
Inadequate food
Lack of food

New code

Z59.42 Food insecurity

New code

Z59.43 Lack of safe drinking water
Inadequate supply of drinking water

Z71 Persons encountering health services for other counseling and medical advice, not elsewhere classified

Z71.8 Other specified counseling

New code

Z71.85 Counseling for socioeconomic factors

Z91 Personal risk factors, not elsewhere classified

Z91.1 Patient's noncompliance with medical treatment and regimen

Z91.11 Patient's noncompliance with dietary regimen

New code	Z91.110 Patient's noncompliance with dietary regimen due to financial hardship

Glut1 Deficiency

Glucose Transporter Protein Type 1 Deficiency Syndrome (Glut1 Deficiency, Glut1 DS, G1D, or De Vivo Syndrome) is a treatable genetic disorder of brain metabolism where glucose does not reach and fuel the brain properly. A wide range of neurological symptoms may result, including intellectual disability, developmental delay, seizures, motor dysfunction, speech and language impairments, microcephaly, abnormal eye-head movements, hemiplegia, migraines, and other issues. Symptoms may be constant, transient, or episodic. A ketogenic diet is the standard of care treatment as it provides ketones as an alternate source of brain energy.

G1D is caused by a mutation in the SLC2A1 gene (solute carrier family 2, facilitated glucose transporter member 1), which encodes the glucose transporter protein type 1, or Glut1. The Glut1 protein is responsible for transporting glucose across the blood brain barrier, across cell membranes, and into cells. Mutations in this gene impair glucose transport, which affects brain metabolism, development, and function. Approximately 90% of G1D patients have an identifiable mutation in the SLC2A1 gene. In the absence of a detectable gene mutation, a diagnosis may be made based on clinical symptoms and low glucose levels in the cerebrospinal fluid. It is estimated that 90% of patients have a de novo mutation, although SLC2A1 mutations may also be inherited through an autosomal dominant pattern or, in very rare cases, through autosomal recessive inheritance.

There is a phenotypic spectrum found in G1D, with a continuum of findings. The classic phenotype may be referred to as GLUT1 deficiency syndrome 1, infantile onset, and is characterized by infantile-onset seizures, developmental delay, acquired microcephaly, and complex movement disorders, with potential variability. A less severe phenotype may be referred to as GLUT1 deficiency syndrome 2, childhood onset; or alternatively as paroxysmal exercise-induced dyskinesia with or without epilepsy or hemolytic anemia (previously known as dystonia 18). Another phenotype is paroxysmal choreoathetosis with spasticity (previously known as dystonia 9). Different types of seizures may occur, including generalized tonic/clonic, myoclonic, atonic, or atypical absence seizures, as well as movement disorders including ataxia, choreoathetosis, dystonia, myoclonus, and paralysis.

Previous estimates put the prevalence of G1D at 1 in 90,000. However, recent research (awaiting publishing) through a large scale genetic testing program suggests that the prevalence may be as high as 1 in 24,000. Thus, up to 14,000 patients in the United States may be affected by G1D, with most cases undiagnosed.

There is currently no specific ICD-10-CM code for G1D, but it should be coded to E74.8, Other specified disorders of carbohydrate metabolism. Related neurological and developmental issues should also be coded.

The Glut1 Deficiency Foundation, a nonprofit patient advocacy organization, has requested creation of a specific ICD-10-CM code for G1D. A specific and unique code will be beneficial in more accurately capturing and tracking prevalence, enhancing research efforts, and aiding in proper diagnosis, treatment, and services for patients. It is proposed to create a new sub-subcategory at E74.81, for Disorders of glucose transport, not elsewhere classified, with a specific code E74.810, Glucose transporter protein type 1 deficiency.

References:

Kniffen, CL. GLUT1 deficiency syndrome 1; GLUT1DS1. 2015. *Online Mendelian Inheritance in Man*. Retrieved from: https://www.omim.org/entry/606777

Kniffen, CL. GLUT1 deficiency syndrome 2; GLUT1DS2. 2015. *Online Mendelian Inheritance in Man*. Retrieved from: https://www.omim.org/entry/612126

Kniffen, CL. Dystonia 9; DYT9. GLUT1 deficiency syndrome 2; GLUT1DS2. 2012. *Online Mendelian Inheritance in Man*. Retrieved from: https://www.omim.org/entry/601042

NLM. Glut1 Deficiency Syndrome. *Genetics Home Reference* [Internet]. 2014. Bethesda, MD: National Library of Medicine (US). Retrieved from: https://ghr.nlm.nih.gov/condition/glut1-deficiency-syndrome

Wang D, Pascual JM, De Vivo D. Glucose Transporter Type 1 Deficiency Syndrome. 2002 Jul 30 [Updated 2018 Mar 1]. In: Adam MP, Ardinger HH, Pagon RA, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2018. Retrieved from: https://www.ncbi.nlm.nih.gov/books/NBK1430/

TABULAR MODIFICATIONS

E74 Other disorders of carbohydrate metabolism

E74.8 Other specified disorders of carbohydrate metabolism

Delete ~~Essential pentosuria~~
Delete ~~Renal glycosuria~~

New Sub-subcategory **E74.81 Disorders of glucose transport, not elsewhere classified**

New Code **E74.810 Glucose transporter protein type 1 deficiency**
Add De Vivo Syndrome
Add GLUT1 deficiency syndrome 1, infantile onset
Add GLUT1 deficiency syndrome 2, childhood onset
Add Glucose transport defect, blood-brain barrier
Add Glut1 deficiency

New Code **E74.818 Other disorders of glucose transport**
Add (Familial) renal glycosuria

New Code **E74.819 Disorders of glucose transport, unspecified**

New Code **E74.89 Other specified disorders of carbohydrate metabolism**
Add essential pentosuria

Hepatic Fibrosis

Nonalcoholic steatohepatitis (NASH) is a disease that causes fat to build up in the liver, leading to inflammation and fibrosis. Hepatic fibrosis can become advanced, and can ultimately lead to liver cirrhosis and failure. NASH is part of the broader spectrum of nonalcoholic fatty liver disease (NAFLD), distinguished from other NAFLD by the presence of hepatocyte injury.

Hepatic fibrosis associated with NASH has a classification accepted by the Food & Drug Administration as an endpoint for clinical trials, in the NASH Clinical Research Network (CRN) staging system. The NASH CRN classification evaluates fibrosis stage on a scale from F0 to F4 as follows: F0 – no fibrosis; F1 – perisinusoidal or periportal; F2 – perisinusoidal and portal/periportal; F3 – bridging fibrosis or pre-cirrhosis; and F4 – cirrhosis.

In the U.S., it has been estimated that 16 million adults have NASH. Of these, about 20% (3.3 million people) have advanced fibrosis (fibrosis stages F3 or F4). Due to low levels of awareness, and lack of simple diagnostic testing, along with the often asymptomatic nature of NASH, diagnosis of advanced fibrosis due to NASH is often delayed. Patients are most often diagnosed when they reach End Stage Liver Disease (ESLD), increasing the likelihood that they will incur higher healthcare costs than if diagnosis occurred earlier in the disease process. A significant increase in the prevalence of advanced fibrosis due to NASH is expected in the U.S., with an expected rise by 2030 in the number of people with F3 fibrosis due to NASH to 4 million people, and in the number of people with cirrhosis (F4 fibrosis) due to NASH expected to rise from to 3.5 million people.

Advanced fibrosis due to NASH is associated with increased risk of liver-related complications, including liver-related mortality, as well overall mortality. Patients with advanced fibrosis due to NASH also have a reduced quality of life. They have increased risk of liver cancer, and increased risk of hospitalization.

Assessment of hepatic fibrosis has traditionally depended on liver biopsy, and that is the gold standard. However, there are now non-invasive tests that can be used for assessment of the stages of hepatic fibrosis. These will be helpful in detecting and differentiating early and advanced fibrosis.

A proposal has been received from the Global Liver Institute, a 501(c)3 nonprofit advocacy organization, to create specific codes for hepatic fibrosis that is early, and hepatic fibrosis that is advanced, along with a note for these to be coded with NASH. These revisions will enable liver fibrosis to be classified more accurately and more consistently with current clinical perspectives and documentation, and will enable enhanced tracking and research into progression of disease and the impact of treatment.

References

Estes C, Razavi H, Loomba R, et. al. Modeling the epidemic of nonalcoholic fatty liver disease demonstrates an exponential increase in burden of disease. Hepatology. 2018 Jan;67(1):123-133. doi: 10.1002/hep.29466.

Kleiner DE1, Brunt EM, Van Natta M, et al.; Nonalcoholic Steatohepatitis Clinical Research Network. Design and validation of a histological scoring system for nonalcoholic fatty liver disease. Hepatology. 2005 Jun;41(6):1313-21. doi: 10.1002/hep.20701.

Rinella ME. Nonalcoholic fatty liver disease: a systematic review. JAMA. 2015 Jun 9;313(22):2263-73. doi: 10.1001/jama.2015.5370.

TABULAR MODIFICATIONS

K74 Fibrosis and cirrhosis of liver

K74.0 Hepatic fibrosis

Add — Code first underlying liver disease, such as:
Nonalcoholic steatohepatitis (NASH) (K75.81)

New Code — K74.00 Hepatic fibrosis, unspecified

New Code — K74.01 Hepatic fibrosis, early fibrosis
Hepatic fibrosis, stage F1 or stage F2

New Code — K74.02 Hepatic fibrosis, advanced fibrosis
Hepatic fibrosis, stage F3
Excludes1: cirrhosis of liver (K74.6-)
hepatic fibrosis, stage F4 (K74.6-)

K75 Other inflammatory liver disease

K76.8 Other specified inflammatory liver diseases

K75.81 Nonalcoholic steatohepatitis (NASH)

Add — Code also, if applicable, hepatic fibrosis (K74.0-)

Hypereosinophilic Syndromes

Hypereosinophilic syndromes (HES) are a clinically and pathogenically heterogeneous group of disorders characterized by elevated blood eosinophil count and eosinophil-mediated end-organ damage. A request for new codes for certain hypereosinophilic syndromes has been received, from the American Partnership for Eosinophilic Disorders (APFED), a 501(c)3 non-profit patient advocacy group, and the International Eosinophil Society, Inc. (IES), an organization of scientists and clinicians interested in the eosinophil and its roles in health and disease.

The clinical manifestation of HES varies according to the target organ systems affected, most commonly cutaneous, pulmonary, gastrointestinal, cardiac, and nervous system tissues. The diagnosis of HES is made based on an elevated eosinophil count ($\geq 1.50 \times 10^9$/L) on at least 2 occasions, associated with evidence of eosinophil-induced organ damage or tissue infiltration after other secondary causes of hypereosinophilia such as allergic, parasitic, and non-hematological malignant disorders have been excluded. The disease burden of HES is due to both acute disease activity as well as chronic end organ damage and fibrosis.

Considerable progress has been made in the pathogenesis and treatment of different HES subtypes, with development of kinase inhibitors, biologics, and other therapies, with various therapies specific for particular disorders. Specific ICD codes for these specific disorders, types of HES, will facilitate the management of HES patients by providing a more detailed context for understanding disease natural history and optimal therapy.

Myeloid Hypereosinophilic Syndrome (MHES)

MHES is defined as HES with features more typical of myeloproliferative disorders, including increased serum vitamin B_{12} and tryptase levels, chromosomal abnormalities, anemia and/or thrombocytopenia, hepatomegaly, splenomegaly, and circulating leukocyte precursors. Approximately half of MHES is caused by a specific fusion gene (FIP1L1-PDGFRA) An additional population of approximately 10-20% of MHES is associated with other fusion genes or mutations (involving PDGFRA, PDGFRB, FGFR1, or JAK2). Additionally, a minority of MHES patients have no characterized mutation.

Compared to other HES subtypes, MHES has a higher frequency of cardiac involvement and evolution into a more aggressive myeloproliferative disorder, both of which lead to shortened survival. MHES is typically resistant to corticosteroid therapy, and response to other treatments can depend on the subtype (e.g., FIP1L1-PDGFRA positive MHES is highly responsive to low dose imatinib).

Lymphocytic Variant Hypereosinophilic Syndrome (LHES)

LHES is defined by the presence of clonal or aberrant T lymphocytes that produce Th2 cytokines, such as interleukin-5, that drive eosinophilia. Skin is the most commonly involved organ system, but other organs are frequently affected. Compared to Idiopathic Hypereosinophilic Syndrome (IHES; described below), LHES often requires higher doses of corticosteroid and off-label use of biologic or

other immunosuppressive therapy to achieve an acceptable clinical response. LHES may progress into lymphoma in 5-25% of patients.

Idiopathic Hypereosinophilic Syndrome (IHES)

Prior to the discovery of the specific HES subtypes noted above, all HES was considered idiopathic, IHES. It still accounts for approximately 70% of HES and is not associated with the laboratory findings that define MHES and LHES. As its name implies, the etiology of IHES is unknown. Corticosteroids are the first line treatment for IHES.

Episodic Angioedema with Eosinophilia (EAE), also known as Gleich's Syndrome

EAE (Gleich's Syndrome) was first described in 1984 as a cyclic disorder characterized by recurrent episodes of fever, angioedema, weight gain and peripheral eosinophilia with a frequency of every 4-6 weeks. It is a rare eosinophilic disorder with multilineage cell cycling with unknown prevalence. EAE has traditionally been considered a variant of HES; patients presenting with this disorder have a unique and unusual clinical phenotype distinct from other hypereosinophilic syndromes. Patients have an aberrant T cell phenotype, increase in polyclonal immunoglobulin M and frequently have a clonal T cell population in the blood. Some patients with this disorder have developed organ-system involvement and lymphoma.

EAE is rare, although it is felt that many patients may go undiagnosed or are treated as HES prior to receiving a definitive diagnosis. Based on the number of cases seen at a major referral center for HES, EAE likely makes up <1% of patients with HES. Disease onset may occur in childhood though presentation is more common in adulthood and is associated with delays in diagnosis due to the unusual nature of the presentation. Untreated patients will often have diuresis and resolution of symptoms in between cycles; however, symptoms may be severe enough to warrant daily glucocorticoid treatment.

References

Bochner BS. Novel therapies for eosinophilic disorders. Immunol Allergy Clin North Am 2015; 35:577-98. https://dx.doi.org/10.1016%2Fj.iac.2015.05.007

Crane MM, et al. Incidence of myeloproliferative hypereosinophilic syndrome in the United States and an estimate of all hypereosinophilic syndrome incidence. J Allergy Clin Immunol 2010; 126:179-81. https://dx.doi.org/10.1016%2Fj.jaci.2010.03.035

Gleich, G.J., Schroeter, A.L., Marcoux, J.P., Sachs, M.I., O'Connell, E.J. and Kohler, P.F. 1984. Episodic angioedema associated with eosinophilia. The New England Journal of Medicine 310(25), pp. 1621–1626. https://doi.org/10.1056/NEJM198406213102501

Gotlib J. World Health Organization-defined eosinophilic disorders: 2017 update on diagnosis, risk stratification, and management. Am J Hematol. 2017 Nov;92(11):1243-1259. https://doi.org/10.1002/ajh.24880

References, cont.

Khoury, P., Herold, J., Alpaugh, A., Dinerman, E., Holland-Thomas, N., Stoddard, J., Gurprasad, S., Maric, I., Simakova, O., Schwartz, L.B., Fong, J., Lee, C.-C.R., Xi, L., Wang, Z., Raffeld, M. and Klion, A.D. 2015. Episodic angioedema with eosinophilia (Gleich syndrome) is a multilineage cell cycling disorder. Haematologica 100(3), pp. 300–307. https://doi.org/10.3324/haematol.2013.091264

Klion AD. How I treat hypereosinophilic syndromes. Blood 2015; 126:1069-77. https://doi.org/10.1182/blood-2014-11-551614

Valent P, Gleich GJ, Reiter A, Roufosse F, Weller PF, Hellmann A, et al. Pathogenesis and classification of eosinophil disorders: a review of recent developments in the field. Expert Rev. Hematol. 2012; 5:157-76. https://doi.org/10.1586/ehm.11.81

Valent P, Klion AD, Horny HP, Roufosse F, Gotlib J, Weller PF, et al. Contemporary consensus proposal on criteria and classification of eosinophilic disorders and related syndromes. J Allergy Clin Immunol 2012; 130:607-12 e9. https://dx.doi.org/10.1016%2Fj.jaci.2012.02.019

TABULAR MODIFICATIONS

D72 Other disorders of white blood cells

D72.1 Eosinophilia

Add — Excludes1: Hypereosinophilic syndromes (D72.A-)

New subcategory — D72.A Hypereosinophilic syndromes

New code — D72.A0 Idiopathic hypereosinophilic syndrome
Add — Hypereosinophilic syndrome, NOS

New code — D72.A1 Myeloid hypereosinophilic syndrome [MHES]

New code — D72.A2 Lymphocytic Variant Hypereosinophilic Syndrome [LHES]

New code — D72.A9 Other hypereosinophilic syndrome
Add — Episodic Angioedema with Eosinophilia [EAE]
Add — Gleich Syndrome

Intracranial Hypotension

A proposal to create new codes for intracranial hypotension was previously presented at the March 2018 ICD-10 Coordination and Maintenance Committee meeting. This proposal has been modified based on comments from that prior presentation. Intracranial hypotension results from a loss of cerebrospinal fluid volume, and it is an under-recognized and under-diagnosed central nervous system disorder. Intracranial hypotension is most often associated with a cerebrospinal fluid leak at the level of the spine and is not causally associated with cerebrospinal fluid leaks arising from the skull base.

The causes of intracranial hypotension include:

- Spontaneous cerebrospinal fluid leaks at the level of the spine (most under-recognized category)
- Iatrogenic holes or defects in the spinal dura from:
  - Intentional diagnostic or therapeutic spinal dural punctures
  - Inadvertent spinal dural puncture during epidural injection procedures
  - Inadvertent or intentional spinal durotomies during spinal or other surgeries
- Over-drainage of cerebrospinal fluid shunting devices
- Traumatic spinal dural tears or defects resulting in spinal cerebrospinal fluid leaks

While headache is the most common symptom of intracranial hypotension, the presence of headache is not universal, nor is it the only symptom. A range of symptoms, signs and complications may occur as a result of effects on the brain and other intracranial structures, cranial nerve roots, spinal cord and spinal nerve roots. Using the term “intracranial hypotension” is more precise and inclusive. When serious complications occur, such as subdural hematomas, stroke, cerebral venous thrombosis, fronto-temporal dementia, or syringomyelia, these should also be coded. It is also of note that cranial MRI findings, or spinal imaging findings may or may not be evident.

While the headache and other symptoms associated with intracranial hypotension often have a positional component, not all patients with a positional (orthostatic) component to their symptoms have intracranial hypotension. An additional code for “headache with orthostatic component, not elsewhere classified” is being proposed for cases in which the diagnosis remains unclear.

The International Classification of Headache Disorders, 3rd edition, recognizes spontaneous intracranial hypotension as a secondary headache disorder, as well as classifying iatrogenic and traumatic cases.

Currently, intracranial hypotension is captured in ICD-10-CM using a number of codes that are not specific, such as those related to CSF leaks, and headaches. Iatrogenic causes of spinal CSF leaks are currently included at category G97, Intraoperative and postprocedural complications and disorders of nervous system, not elsewhere classified; some are also classified related to obstetrics, including subcategories O29.4, Spinal and epidural anesthesia induced headache during pregnancy; and O29.5, Other complications of spinal and epidural anesthesia during pregnancy; and codes O89.4, Spinal and epidural anesthesia-induced headache during the puerperium; O89.5, Other complications of spinal and epidural anesthesia during the puerperium; O74.5, Spinal and epidural anesthesia-induced

headache during labor and delivery; and O74.6, Other complications of spinal and epidural anesthesia during labor and delivery.

These proposed changes have the potential to improve diagnostic accuracy, be supportive of improved understanding of requirements for diagnostic testing and treatments, and to better track prevalence of intracranial hypotension, and outcomes. They will also enable better differentiation of cranial and spinal cerebrospinal fluid leaks, which have different underlying causes, symptoms, complications, diagnostic testing and treatments.

Specific codes and expansion in ICD-10-CM has been requested by a team of key opinion leaders, including Timothy Amhrein, MD, Peter G. Kranz, MD and Linda-Gray Leithe, MD, Neuroradiology / Spine Intervention at Duke University Medical Center; Ian Carroll, MD, MS, Anesthesiology / Pain Medicine at Stanford; Connie Deline, MD, General Medicine, Spinal CSF Leak Foundation; Charles Louy, PhD, MD, MBA, Anesthesiology / Pain Medicine at Cedars-Sinai; Marcel Maya, MD, Neuroradiology at Cedars-Sinai; Wouter Schievink, MD, Neurosurgery at Cedars-Sinai; Stephen Silberstein, MD, Headache Neurology at Jefferson Headache Center; and Deborah Friedman, MD, MPH, Neurology and Ophthalmology at UT Southwestern Medical Center.

References:

Deline CR, Schievink WI. Spontaneous Intracranial Hypotension. Rare Disease Database. NORD. 2017. https://rarediseases.org/rare-diseases/spontaneous-intracranial-hypotension/

Headache Classification Committee of the International Headache Society (IHS). The International Classification of Headache Disorders, 3rd edition. Cephalalgia. 2018:38(1):1-211. https://www.ichd-3.org/

Kranz PG, Gray L, Amrhein TJ. Spontaneous intracranial hypotension: 10 myths and misperceptions. Headache. 2018 Jul;58(7):948–959. https://onlinelibrary.wiley.com/doi/abs/10.1111/head.13328

Kranz PG, Malinzak MD, Amrhein TJ, Gray L. Update on the diagnosis and treatment of spontaneous intracranial hypotension. Curr Pain Headache Rep. 2017 Aug;21(8):37. https://link.springer.com/article/10.1007%2Fs11916-017-0639-3

Mokri B. Spontaneous Intracranial Hypotension. Continuum (Minneap, Minn). 2015 Aug;21(4 Headache):1086–1108. https://insights.ovid.com/crossref?an=00132979-201508000-00015

Schievink WI. Spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension. JAMA. 2006;295(19):2286-2296. https://doi.org/doi:10.1001/jama.295.19.2286

Schievink WI, Dodick DW, Mokri B, Silberstein S, Bousser M-G, Goadsby PJ. Diagnostic criteria for headache due to spontaneous intracranial hypotension: a perspective. Headache. 2011 Oct;51(9):1442–1444. https://doi.org/10.1111/j.1526-4610.2011.01911.x

TABULAR MODIFICATIONS

G96 Other disorders of central nervous system

G96.0 Cerebrospinal fluid leak

Add Code also spontaneous intracranial hypotension, if applicable (G96.81)

Excludes1: cerebrospinal fluid leak from spinal puncture (G97.0)

New code G96.00 Cerebrospinal fluid leak, unspecified

New code G96.01 Cranial cerebrospinal fluid leak, spontaneous
Cerebrospinal fluid leak from skull base
CSF otorrhea
CSF rhinorrhea
otorrhea due to CSF leak
rhinorrhea due to CSF leak

New code G96.02 Spinal cerebrospinal fluid leak, spontaneous
Cerebrospinal fluid leak from spine

New code G96.09 Other cerebrospinal fluid leak

G96.1 Disorders of meninges, not elsewhere classified

G96.11 Dural tear

Add Code also spontaneous intracranial hypotension, if applicable (G96.81)

G96.8 Other specified disorders of central nervous system

New sub-subcategory G96.81 Intracranial hypotension

Add Code also any associated diagnoses, such as:
brachial amyotrophy (G54.5)
cerebrospinal fluid leak from spine (G96.02)
cranial nerve disorders in diseases classified elsewhere (G53)
nerve root and compressions in diseases classified elsewhere (G55)
nonpyogenic thrombosis of intracranial venous system (I67.6)
nontraumatic subdural hemorrhage (I62.0-)
nontraumatic intracerebral hemorrhage (I61.-)
other and unspecified cord compression (G95.2-)
other secondary parkinsonism (G21.8)
reversible cerebrovascular vasoconstriction syndrome (I67.841)
spinal cord herniation (G95.89)
stroke (I63.-)
syringomyelia (G95.0)

New code G96.810 Intracranial hypotension, unspecified

New code G96.811 Intracranial hypotension, spontaneous

New code G96.819 Other intracranial hypotension

New code G96.89 Other specified disorders of central nervous system

G97 Intraoperative and post-procedural complications and disorders of nervous system, not elsewhere classified

G97.0 Cerebrospinal fluid leak from spinal puncture
Add Code also any associated diagnoses or complications, such as:
Add Intracranial hypotension following a procedure (G97.83)

G97.1 Other reaction to spinal and lumbar puncture
Add Other reaction to spinal dural puncture
Add Code also if applicable any associated headache with orthostatic component (R51.0)

G97.2 Intracranial hypotension following ventricular shunting
Add Code also any associated diagnoses or complications

G97.4 Accidental puncture and laceration of a nervous system organ or structure during a procedure

G97.41 Accidental puncture or laceration of dura during a procedure
Incidental (inadvertent) durotomy
Add Code also any associated diagnoses or complications

G97.8 Other intraoperative and postprocedural complications and disorders of nervous system
Use additional code to further specify disorder

New code G97.83 Intracranial hypotension following lumbar cerebrospinal fluid shunting
Add Code also any associated diagnoses or complications

New code G97.84 Intracranial hypotension following other procedure
Add Code also, if applicable:
Add accidental puncture or laceration of dura during a procedure (G97.41)
Add cerebrospinal fluid leak from spinal puncture (G97.0)

G44.8 Other specified headache syndromes
Add Excludes2: headache with orthostatic or positional component, not elsewhere classified (R51.0)

	R51	Headache	
Delete		~~Facial pain NOS~~	
Delete		~~Excludes1:~~	~~atypical face pain (G50.1)~~
Delete			~~migraine and other headache syndromes (G43-G44)~~
Delete			~~trigeminal neuralgia (G50.0)~~
Add		Excludes2:	atypical face pain (G50.1)
Add			migraine and other headache syndromes (G43-G44)
Add			trigeminal neuralgia (G50.0)
New code		R51.0	Headache with orthostatic component, not elsewhere classified
Add			Headache with positional component, not elsewhere classified
New code		R51.9	Headache, unspecified
Add			Facial pain NOS

Other Eosinophil Diseases

Specific codes for certain other eosinophil diseases have been requested by the American Partnership for Eosinophilic Disorders (APFED), a 501(c)3 non-profit patient advocacy group, and the International Eosinophil Society, Inc. (IES), an organization of scientists and clinicians interested in the eosinophil and its roles in health and disease.

Eosinophilic Granulomatosis with Polyangiitis (EGPA) is a disorder previously known as Churg-Strauss, or polyarteritis with lung involvement. This is currently coded in ICD-10-CM to the code M30.1, with the title, "Polyarteritis with lung involvement [Churg-Strauss]." It also has the inclusion term, "Allergic granulomatous angiitis." EGPA is an autoimmune disorder that may affect multiple organ systems, especially the lungs, causing damage. It is characterized by the abnormal presence of high numbers of circulating (blood) and tissue eosinophils, inflammation of blood vessels (vasculitis), and the development of inflammatory nodular lesions called granulomas (granulomatosis). Without treatment, serious organ damage can occur and the disease may be fatal. Most affected individuals have a history of allergy and asthma. Other associated pulmonary abnormalities often precede the development of the generalized (systemic) symptoms and signs by as little as six months or as much as two decades. The peripheral nerves, kidneys, or gastrointestinal tract are often involved. The cause of EGPA is unknown. EGPA is treated similarly to other vasculitic syndromes with high doses of immunosuppressive agents including glucocorticoids and Cytoxan. Recently, mepolizumab has been FDA approved for EGPA.

The 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides revised the classification of vasculitides and approved Eosinophilic Granulomatosis with Polyangiitis (EGPA) as the official name, for the disease that had previously been referred to as Churg-Strauss Syndrome. It is being proposed to change the ICD-10-CM code title, to be consistent with the name that will now be in clinical use.

Drug rash with eosinophilia and systemic symptoms (DRESS) syndrome is a severe, life-threatening drug reaction characterized by widespread skin rash accompanied by marked systemic symptoms including fever, lymphadenopathy, facial edema, and maculopapular rash. Systemic involvement can also include hepatitis and interstitial pneumonia. Severe cardiac involvement may be present, with eosinophilic myocarditis. Less frequently, other organ manifestations such as nephritis or pancreatitis are present. Because DRESS is triggered by both long-term and short-term drug exposure, it is critical to seek and identify the culprit drugs in the time prior to eruption.

DRESS syndrome has been attributed to a synergistic interaction of lymphocyte activation, drug metabolic enzyme defects with accumulation of drug metabolites, eosinophil activation and viral infection reactivation (human herpesvirus-6, cytomegalovirus, Epstein–Barr virus) in persons with genetic susceptibility in association with certain human leukocyte antigen (HLA) class I alleles. Autoantibody formation and autoimmune diseases, including type I diabetes mellitus, autoimmune thyroid disease, sclerodermoid graft-versus-host disease, systemic lupus erythematosus and bullous pemphigoid may occur up to four years after resolution (and are attributed to depletion of regulatory T-cells upon recovery from DRESS syndrome).

Modifications of immune presentation of endogenous and self-proteins induce the inflammatory responses that triggers systemic clinical and biological signs. For example, abacavir (a reverse transcriptase inhibitor) binds non-covalently to the HLA groove modifying the self-peptide repertoire presented in the groove. Another drug is carbamazepine, an anticonvulsant medication, and susceptibility to carbamazepine reactivity is found in patients with HLA-B*15:02 variant. The mechanism of T-cell activation induced by carbamazepine is supposed to be the same as that described for abacavir. Allopurinol induces reactions in patients with HLA-B*58:01.These HLA associations probably share a similar T-cell immune activation mechanism that depends on the culprit drug/HLA interaction and HLA/peptide repertoire presentation. The identification of risk-associated HLA variants opens new avenues for physicians by using patient selection based on HLA typing.

In summary, DRESS is a systemic drug reaction in which eosinophil activation is driven by an immunological response directed against viral reactivation and a culprit drug. Eosinophils infiltrate organs in response to chemokines, in synergy with IL-5, and toxic granule protein release represents a key factor of tissue damage.

Prevalence of DRESS may be estimated based on the number of doses of agents administered, and has been estimated to have an overall population risk of between 1 in 1000 and 1 in 10,000 drug exposures. However, the total overall prevalence across all agents is not known, as some agents may only present with DRESS rarely. The prognosis and recovery are generally good, although death is possible in the acute phase of the disease (mortality rate of about 10% in some studies, but lower in others). While scoring systems exist for the diagnosis of DRESS, and international registries such as RegiSCAR were created to track cases, the creation of a specific ICD code for DRESS would enhance ability to track it, along with the ability to assess pharmacogenetics, detect new culprit agents, and assess important immunopathogenic questions regarding etiology, impact of genetic background, and potential for cross-reactive medications after the development of DRESS.

References

Cho YT, Yang CW, Chu CY. Drug Reaction with Eosinophilia and Systemic Symptoms (DRESS): An Interplay among Drugs, Viruses, and Immune System. Int J Mol Sci. 2017 Jun 9;18(6). pii: E1243. https://dx.doi.org/10.3390/ijms18061243

Jennette JC. Overview of the 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. Clin Exp Nephrol. 2013 Oct; 17(5): 603–606. https://dx.doi.org/10.1007/s10157-013-0869-6

Kardaun, SH, Sekula, P, Valeyrie-Allanore, L, et al. Drug reaction with eosinophilia and systemic symptoms (DRESS): an original multisystem adverse drug reaction. Results from the prospective RegiSCAR study. Br J Dermatol. 2013 Nov;169(5):1071-80. https://dx.doi.org/10.1111/bjd.12501

Peter, JG, Lehloenya, R, Dlamini, S, et al. Severe delayed cutaneous and systemic reactions to drugs: A global perspective on the science and art of current practice. J Allergy Clin Immunol Pract. 2017 May - Jun;5(3):547-563. https://dx.doi.org/10.1016%2Fj.jaip.2017.01.025

TABULAR MODIFICATIONS

D72 Other disorders of white blood cells

New code D72.B Drug rash with eosinophilia and systemic symptoms syndrome
Add DRESS syndrome
Add Use additional code for adverse effect, if applicable, to identify drug (T36-T50 with fifth or sixth character 5)

M30 Polyarteritis nodosa and related conditions

M30.1 Polyarteritis with lung involvement [Churg-Strauss]
Delete ~~Allergic granulomatous angiitis~~

New code M30.1X Eosinophilic granulomatosis with polyangiitis [EGPA]
Add Allergic granulomatous angiitis
Add Churg-Strauss
Add Polyarteritis with lung involvement

Pulmonary Eosinophilic Diseases

New codes for certain pulmonary eosinophilic diseases have been requested by the American Partnership for Eosinophilic Disorders (APFED), a 501(c)3 non-profit patient advocacy group, and the International Eosinophil Society, Inc. (IES), an organization of scientists and clinicians interested in the eosinophil and its roles in health and disease.

Eosinophilic Pneumonia
There are 2 types of eosinophilic pneumonia - acute and chronic. While both are characterized by eosinophil invasion of the lung tissue, they are quite different from one another and are described below.

Acute Eosinophilic Pneumonia

Acute eosinophilic pneumonia (AEP) is classified as a form of eosinophilic lung disease, one of a large group of interstitial lung diseases. AEP is different from chronic eosinophilic pneumonia (CEP), which is marked by slower progression, lack of progression to acute respiratory failure, frequent relapses and is often associated with asthma.

AEP is characterized by rapidly progressive respiratory failure, usually of less than one-month duration, often leading to the need for intensive care and mechanical ventilation. Approximately two-thirds of individuals may require mechanical ventilation. Chest imaging usually shows abnormalities throughout the lung fields. Finding eosinophils in lung washings from bronchoalveolar lavage (BAL) is key in the diagnosis of AEP. BAL fluid in individuals with AEP reveals abnormally high levels of eosinophils (greater than 25%). Associated symptoms are nonspecific and can include fever, cough, difficulty breathing (dyspnea) and chest pain. Less common symptoms include fatigue, muscle pain (myalgia), joint aches, and abdominal discomfort or pain.

While respiratory failure is a common feature of AEP, this disease is quite responsive to therapy with corticosteroids, making recognition and diagnosis critical. Within the medical literature, the dose and duration of corticosteroid therapy has varied greatly, with a recent series suggesting that just two-weeks of treatment is sufficient. Following treatment, long term prognosis is excellent.

The causes of AEP are not entirely clear, though it has been noted to be associated with new onset of cigarette smoking or an increase in smoking. Other case series of AEP in the literature include a series in U.S military personnel in Iraq and another in 9/11 rescue workers exposed to dust from the World Trade Center. Men are more commonly affected than women. The cause of AEP is unknown (idiopathic).

Researchers believe that AEP develops due to an unidentified triggering agent that causes the body to produce extra eosinophils and recruit them specifically to the lung. The exact reason for the overproduction and accumulation of eosinophils is unknown. Additional reports in the medical literature have linked some cases of AEP to the use of a number of medications. Drug-induced cases have been linked to minocycline, daptomycin, and velafaxine, an antidepressant, and others

(www.pneumotox.com). Several environmental factors including occupational exposures have been shown to trigger AEP including dust and smoke. It is unlikely that a single environmental factor causes AEP. Most likely, multiple factors are necessary for the development of the disorder, with association of a triggering condition in a predisposed individual. The triggering factor in AEP can be different from one individual to another.

Chronic Eosinophilic Pneumonia

Chronic Eonsinophilic Pneumonia (CEP) has been a well-established distinct entity in pulmonary medicine for decades, yet it does not have its own diagnostic ICD code. CEP is different from acute eosinophilic pneumonia (AEP), which is marked by rapid onset, the absence of asthma, a greater potential for acute respiratory failure and no relapse following treatment. As the name implies, CEP is a more chronic, indolent condition than AEP and is characterized by progressive shortness of breath and abnormalities on chest imaging (CT scan and chest x-ray) that often are located in the periphery of the lungs. Bronchoscopy with the finding of increased eosinophils on airway washings confirms the diagnosis. Eosinophils also increase in the bloodstream (peripheral blood eosinophilia). Common symptoms include shortness of breath (dyspnea), cough, fatigue, night sweats, low grade fevers, and unintended weight loss. Symptoms can be very similar to those seen in asthma, including the development of wheezing. In fact, a diagnosis of asthma may precede the development of CEP.

CEP is generally treated with a prolonged course of corticosteroids lasting several months or more, with relapses common during attempts to taper corticosteroids. Like many eosinophilic diseases, the exact inciting causes are unknown. Researchers believe that CEP may develop due to an unidentified, nonspecific triggering agent that causes the body to produce excess eosinophils. The exact reason for the overproduction and accumulation of eosinophils is unknown. CEP tends to recur and many individuals will relapse at some point, especially when therapy is not maintained. A relapse can occur as much as 10 years or more after the initial episode. Some individuals eventually develop severe asthma. In some cases, individuals with CEP have developed a related disorder known as Churg-Strauss syndrome, now renamed eosinophilic granulomatosis with polyangiitis, or EGPA, suggesting that there may be an overlap between these two disorders. The disorder can occur in individuals of any age, but is extremely rare in childhood. The peak incidence is during the fifth decade. CEP occurs twice as often in women than men.

Eosinophilic Asthma

Currently, diagnostic codes classify asthma based on severity, persistence, and the presence of exacerbation. However, our state of the art understanding of the immune mechanisms underlying asthma has defined a subset of asthma that is characterized by the presence of a certain level of eosinophils in the circulation and in the airways. In the United States, an estimated 25.7 million people have some form of asthma, with 15 percent of these people having severe asthma that is difficult to control with standard medications. Eosinophilic asthma is considered a leading cause of severe asthma, affecting 50 to 60 percent of people with the severe form of the disease.

Eosinophilic asthma can be difficult to treat and may have a detrimental effect on an individual's quality of life. It does not generally respond well to inhaled corticosteroids, even at high doses. Until recently, oral corticosteroids were the standard treatment for eosinophilic asthma. Although many

people respond well to these medications, they are not generally successful at controlling this disease long term. Some patients become dependent on the oral corticosteroids. A number of biologic treatments have been approved by the US FDA to treat eosinophilic asthma, and can reduce the frequency of eosinophilic asthma attacks. Patients with eosinophilic asthma have been found in several pivotal clinical trials to benefit from these therapies that specifically target eosinophils by blocking a molecule known as IL-5 or its receptor. In fact, these drugs reduce asthma exacerbations regardless of the cause and are oral steroid sparing. Their benefit on lung function is modest. With these circumstances shaping the current scientific, clinical, and pragmatic landscape, eosinophilic asthma has now become a well-accepted and widespread diagnostic entity.

The connection between eosinophils and asthma has been recognized and cited in research since 1889. Eosinophilic asthma is more prominent in adults. Adult asthma usually affects more women than men, but eosinophilic asthma affects men and women at about the same rate. As the level of eosinophils increases, inflammation and other symptoms of asthma become more severe. The primary symptom is shortness of breath rather than the more traditional wheezing in non-eosinophilic asthma. People with eosinophilic asthma may also experience chronic sinus infections, nasal polyps, eosinophilic otitis media, an inner ear infection, and aspirin-exacerbated respiratory disease. The symptoms of eosinophilic asthma differ from classic asthma and, in fact, more closely resemble those of chronic pulmonary obstructive disorder (COPD).

References

Akuthota P, Weller PF. Eosinophilic pneumonias. Clin Microbiol Rev. 2012 Oct;25(4):649-60. https://doi.org/10.1128/CMR.00025-12

Bel EH, Wenzel SE, Thompson PJ, et al. Oral glucocorticoid-sparing effect of mepolizumab in eosinophilic asthma. N Engl J Med 2014;371(13):1189–97. https://doi.org/10.1056/NEJMoa1403291

Bleecker ER, FitzGerald JM, Chanez P, et al. Efficacy and safety of benralizumab for patients with severe asthma uncontrolled with high-dosage inhaled corticosteroids and long-acting β2-agonists (SIROCCO): a randomised, multicentre, placebo-controlled phase 3 trial. The Lancet 2016;388(10056):2115–27. https://doi.org/10.1016/S0140-6736(16)31324-1

Bottrell J. Asthma Subgroups: What Is Persistent Eosinophilic Asthma? Asthma.net. November 29, 2016. Available at: https://asthma.net/living/subgroups-what-is-eosinophilic/

Burki NK. Chronic Eosinophilic Pneumonia. NORD - National Organization for Rare Disorders. Available at: https://rarediseases.org/rare-diseases/chronic-eosinophilic-pneumonia

Carrington CB, Addington WW, Goff AM, et al. Chronic eosinophilic pneumonia. N Engl J Med. 1969 Apr 10;280(15):787-98. https://doi.org/10.1056/NEJM196904102801501

Castro M, Zangrilli J, Wechsler ME, et al. Reslizumab for inadequately controlled asthma with elevated blood eosinophil counts: results from two multicentre, parallel, double-blind, randomised, placebo-controlled, phase 3 trials. Lancet Respir Med 2015;3(5):355–66. https://doi.org/10.1016/S2213-2600(15)00042-9

Cottin V. Acute Eosinophilic Pneumonia. NORD - National Organization for Rare Disorders. Available at: https://rarediseases.org/rare-diseases/acute-eosinophilic-pneumonia

de Groot JC, Ten Brinke A, Bel EH. Management of the patient with eosinophilic asthma: a new era begins. ERJ Open Res. 2015; 1: 00024-2016. https://doi.org/10.1183/23120541.00024-2015

FitzGerald JM, Bleecker ER, Nair P, et al. Benralizumab, an anti-interleukin-5 receptor α monoclonal antibody, as add-on treatment for patients with severe, uncontrolled, eosinophilic asthma (CALIMA): a randomised, double-blind, placebo-controlled phase 3 trial. The Lancet 2016;388(10056):2128–41. https://doi.org/10.1016/S0140-6736(16)31322-8

Jantina C. de Groot, Anneke ten Brinke and Elisabeth H.D. Bel. "Management of the patient with eosinophilic asthma: a new era begins".

Ortega HG, Liu MC, Pavord ID, et al. Mepolizumab treatment in patients with severe eosinophilic asthma. N Engl J Med 2014;371(13):1198–207. https://doi.org/10.1056/NEJMoa1403290

Yancey SW, Keene ON, Albers FC, et al. Biomarkers for severe eosinophilic asthma. J Allergy Clin Immunol 2017;140(6):1509–18. https://doi.org/10.1016/j.jaci.2017.10.005

TABULAR MODIFICATIONS

D72 Other disorders of white blood cells

D72.1 Eosinophilia

Delete ~~Excludes1: Löffler's syndrome (J82)~~
~~pulmonary eosinophilia (J82)~~

Add Excludes2: Löffler's syndrome (J82)
pulmonary eosinophilia (J82)

J82 Pulmonary eosinophilia, not elsewhere classified

Delete ~~Allergic pneumonia~~

Delete ~~Eosinophilic asthma~~

Delete ~~Eosinophilic pneumonia~~

Delete ~~Löffler's pneumonia~~

Delete ~~Tropical (pulmonary) eosinophilia NOS~~

Delete ~~Excludes1:pulmonary eosinophilia due to aspergillosis (B44.-)~~

Delete ~~pulmonary eosinophilia due to drugs (J70.2-J70.4)~~

Delete ~~pulmonary eosinophilia due to specified parasitic infection (B50-B83)~~

Delete ~~pulmonary eosinophilia due to systemic connective tissue disorders (M30-M36)~~

Delete ~~pulmonary infiltrate NOS (R91.8)~~

Add Excludes2:pulmonary eosinophilia due to aspergillosis (B44.-)

Add pulmonary eosinophilia due to drugs (J70.2-J70.4)

Add pulmonary eosinophilia due to specified parasitic infection (B50-B83)

Add pulmonary eosinophilia due to systemic connective tissue disorders (M30-M36)

Add pulmonary infiltrate NOS (R91.8)

New subcategory	J82.X	Pulmonary eosinophilia, not elsewhere classified
New code	J82.X1	Chronic eosinophilic pneumonia Eosinophilic pneumonia, NOS
New code	J82.X2	Acute eosinophilic pneumonia
New code	J82.X3	Eosinophilic asthma
New code	J82.X9	Other pulmonary eosinophilia, not elsewhere classified
Add		Allergic pneumonia
Add		Löffler's pneumonia
Ad		Tropical (pulmonary) eosinophilia NOS

INDEX MODIFICATIONS

	Eosinophilia …
Revise	- infiltrative – see Eosinophilia, pulmonary ~~J82~~
Revise	- Löffler's J82.X9
Revise	- pulmonary NEC J82.X9
Add	- - acute J82.X2
Add	- - asthmatic J82.X3
Add	- - chronic J82.X1
Revise	- tropical (pulmonary) J82.X9
	Infiltrate, infiltration
	- lung R91.8
Revise	- - eosinophilic – see Eosinophilia, pulmonary ~~J82~~
	- pulmonary R91.8
Revise	- - with eosinophilia – see Eosinophilia, pulmonary ~~J82~~
	Löffler's
Revise	- eosinophilia J82.X9
Revise	- pneumonia J82.X9
Revise	- syndrome (eosinophilic pneumonitis) J82.X9
	Pneumonia
Revise	- allergic ~~(eosinophilic)~~ (see also Pneumonitis, hypersensitivity) J82.X9
	- broncho-, bronchial …
Revise	- - allergic ~~(eosinophilic)~~ (see also Pneumonitis, hypersensitivity) J82.X9
Revise	- eosinophilic J82.X1
Add	- - acute J82.X2
Add	- - chronic J82.X1

	Pneumonitis …
Revise	- eosinophilic J82.X1
Add	- - acute J82.X2
Add	- - chronic J82.X1
	Syndrome …
Revise	- PIE (pulmonary infiltration with eosinophilia) (see also Eosinophilia, pulmonary) J82.X9
Revise	- Weingarten's (tropical eosinophilia) J82.X9

Social Determinants of Health

While there are a number of ICD-10-CM codes that identify a variety of Social Determinants of Health (SDOH), there are no unique ICD-10-CM codes that identify social diagnoses or barriers to health care. As such, United Healthcare (UHC) support new ICD-10-CM codes that expand the existing SDOH codes. The proposed expanded ICD-10-CM codes would capture these social diagnoses and barrier situations to assist providers and consumers in obtaining routine care, medications, and preventive services that are not captured today, thereby benefiting the industry as a whole in the management of patient care.

The Chronic Care Act of 2018 seeks to provide Medicare consumers more integrated care by integrating medical and non-medical care. Additionally, several of the Healthcare Effectiveness Data and Information Set (HEDIS) measures require to report an outpatient visit that may be difficult to complete due to social barriers that are unrelated to a consumer's health. For example, breast cancer screening must be completed in a location with mammography equipment. If a consumer has no means of transportation, or cannot afford to pay for transportation to a breast cancer screening center, the probability is high that this screening will not occur.

UHC believes expanding the ICD-10-CM code set would allow for population health improvement, along with the opportunity for National Committee for Quality Assurance (NCQA) to expand HEDIS measurements around social barrier identification and assistance in the future. Today, capturing SDOH barriers appear to vary widely throughout the industry, rendering it a fragmented, inconsistent way of both capturing and using this information.

Systematized Nomenclature of Medicine-Clinical Terms (SNOMED-CT) and Loginal Observation Identifiers Names and Codes (LOINC) are other coding standards that have also identified some SDOH codes, however for varying reasons, they would be a substandard solution.

UHC believes utilizing the ICD-10-CM codes is a logical choice, as it is the standard language between care providers and payers. The existing SDOH range Z55-Z65 in the ICD-10-CM has been labeled as "Persons with potential health hazards related to socioeconomics and psychosocial circumstances," validating that an expansion of these codes would be warranted within the ICD-10-CM classification.

In 2017, UHC began a national initiative to capture, code, and refer to social and governmental programs those members who self-identified a SDOH. The work began with data from UHC's Medicare Advantage members, but the model is applicable to data collection and use for any consumer. From 1/1/2016 – 10/30/18, UHC has received over 2 million coded claims for social barriers from its contracted and non-contracted providers when there is an available ICD-10-CM code. UHC has found that providers do submit codes when available. Additionally, much of this data exists in physician's electronic medical records as a result of health risk assessments, but without additional ICD-10-CM codes, SDOH cannot be coded or captured, to improve coordination of non-health and medical/behavioral services.

For example, if only the Z59.6 Low Income, code is used, there is virtually no way to assist the consumer without asking them for more specificity. Not only is this additional step time consuming

to the health care professional, it may be considered abrasive to the consumer, and it does not allow for any standardized assistance to be created for either the individual consumer or the broader population. Another example may be that solving for an inability to pay for prescriptions is a completely different action using different resources than solving for the inability to pay for transportation (or any of the other proposed Z59.61-68 codes listed below).

UHC is requesting further granularity to existing code categories Z55-Z65 to increase specificity needed to appropriately capture, analyze, and act on SDOH data to improve outcomes for both consumers and populations.

The National Committee for Quality Assurance (NCQA), the Arizona Health Care Cost Containment System (AHCCCS) and the Center for Healthcare Research & Transformation (CHRT) University of Michigan have endorsed the use and expansion of the ICD-10-CM codes as a standardized model for data collection and social barrier removal.

CDC Healthy People 2020 highlights the importance of addressing SDOH by including "create social and physical environments that promote good health for all" as one of the four overarching goals for the decade." As there is no single vocabulary that covers all aspects of SDOH, NCHS believes that a broader discussion with stakeholders and other code set developers should occur to ensure the goals of interoperability and harmonization are met and maintained.

While NCHS agrees that social determinants of health are important, some concepts may be considered for inclusion in ICD-10-CM, while others may be more appropriately placed in other code sets and vocabularies. We are interested in receiving input from the public on these proposed new codes.

TABULAR MODIFICATIONS

Z55 Problems related to education and literacy
Excludes1: disorders of psychological development (F80-F89)

New code Z55.5 Less than a high school degree
New code Z55.6 High school diploma or GED

Z56 Problems related to employment and unemployment
Excludes2: occupational exposure to risk factors (Z57.-)
problems related to housing and economic circumstances (Z59.-)

Z56.8 Other problems related to employment

New code Z56.83 Unemployed and seeking work
New code Z56.84 Unemployed but not seeking work
New code Z56.85 Employed part time or temporary

New code		Z56.86	Employed full time
	Z59	Problems related to housing and economic circumstances Excludes2: problems related to upbringing (Z62.-)	
New subcategory		Z59.6	Low Income
New code		Z59.61	Unable to pay for prescriptions
New code		Z59.62	Unable to pay for utilities
New code		Z59.63	Unable to pay for medical care
		Z59.64	Unable to pay for transportation for medical appointments or prescriptions
New code		Z59.65	Unable to pay for phone
New code		Z59.66	Unable to pay for adequate clothing
New code		Z59.67	Unable to find or pay for child care
New code		Z59.69	Unable to pay for other needed items
		Z59.9	Problem related to housing and economic circumstances, unspecified
New code		Z59.91	Worried about losing housing
	Z60	Problems related to social environment	
		Z60.8	Other problems related to social environment
New code		Z60.81	Unable to deal with stress
New code		Z60.82	Inadequate social interaction - limited to once or twice a week
New code		Z60.83	Can hardly ever count on family and friends in times of trouble
New code		Z60.84	Feeling unsafe in current location
New code		Z60.85	Stressed quite a bit or very much
New code		Z60.86	Stressed somewhat

Temperature-Sensitive Acquired Autoimmune Hemolytic Anemias

The acquired autoimmune hemolytic anemias (AIHAs) are a group of disorders characterized by production of autoantibodies that bind to red blood cells (RBCs), leading to their destruction (hemolysis). This proposal to update ICD-10-CM codes to expand D59.1, Other autoimmune hemolytic anemias, is based on a request by Sanofi (a pharmaceutical company), and is supported by the American Society of Hematology.

AIHA is classified into four types: drug-induced AIHA, and three temperature-sensitive AIHA types. These three are called warm, cold, and mixed type AIHA, which denote the temperatures at which the autoantibodies best react with RBCs. Warm-type AIHA is the most common form (about 70% of all AIHA cases), and includes autoantibody-RBC binding that occurs at 37°C. Cold-type AIHA has autoantibody-RBC binding that occurs below 37°C, and it accounts for about 15% of all AIHA cases. Finally mixed-type AIHA, combined cold and warm, occurs in less than 10% of AIHA, and it denotes patients with both warm and cold type AIHA.

Patients with AIHA experience symptoms specific to the type and degree of AIHA, that can include fatigue, jaundice, pallor, tachycardia, acrocyanosis, Raynaud's phenomenon (only cold type), dark urine, and splenomegaly. In addition, patients with AIHA also have an increased rate of thromboembolic events (TE) including pulmonary embolism, cerebral infarction, and myocardial infarction. The warm-type and cold-type AIHAs have significant differences in treament. Warm antibody disease is usually treated using steroids; however, these are less effective or ineffective for patients with cold-type disease.

Specific codes for the different types of AIHAs would help enable better tracking of each of these separate conditions, which would advance the clinical understanding of these conditions, and subsequently improve the diagnostic and treatment paradigms.

References

Bass GF, Tuscano ET, Tuscano JM. Diagnosis and classification of autoimmune hemolytic anemia. *Autoimmun Rev.* 2014;13(4-5):560-4.

Bylsma LC, Ording AG, Frøslev T, et al. Occurrence, survival, and thromboembolic risk in cold agglutinin disease: a population-based cohort analysis. HemaSphere, 2018;2:S1, Abstract n. PS1131. Proceedings from the 23rd European Hematology Association Congress; June 14–17, 2018; Stockholm, Sweden. Retrieved from https://learningcenter.ehaweb.org/eha/2018/stockholm/215445/sigbjrn.berentsen.the.occurrence.and.survival.of.cold.agglutinin.disease.in.html?f=topic=1574*media=3

Orphannet. Autoimmune hemolytic anemia. Orphanet website. 2010. Retrieved from https://www.orpha.net/consor/cgi-bin/OC_Exp.php?Expert=98375&lng=EN

TABULAR MODIFICATIONS

D59 Acquired hemolytic anemia

D59.1 Other autoimmune hemolytic anemias

Delete ~~Autoimmune hemolytic disease (cold type) (warm type)~~
Delete ~~Chronic cold hemagglutinin disease~~
Delete ~~Cold agglutinin disease~~
Delete ~~Cold agglutinin hemoglobinuria~~
Delete ~~Cold type (secondary) (symptomatic) hemolytic anemia~~
Delete ~~Warm type (secondary) (symptomatic) hemolytic anemia~~
Delete ~~Excludes1: Evans syndrome (D69.41)~~
Delete ~~hemolytic disease of newborn (P55.-)~~
Delete ~~paroxysmal cold hemoglobinuria (D59.6)~~
Add Excludes2: Evans syndrome (D69.41)
Add hemolytic disease of newborn (P55.-)
Add paroxysmal cold hemoglobinuria (D59.6)

New code D59.10 Autoimmune hemolytic anemia, unspecified

New code D59.11 Warm autoimmune hemolytic anemia
Add Warm type autoimmune hemolytic disease
Add Warm type (primary) (secondary) (symptomatic) autoimmune hemolytic anemia

New code D59.12 Cold autoimmune hemolytic anemia
Add Chronic cold hemagglutinin disease
Add Cold agglutinin disease
Add Cold agglutinin hemoglobinuria
Add Cold type (primary) (secondary) (symptomatic) autoimmune hemolytic anemia
Add Cold type autoimmune hemolytic disease

New code D59.13 Mixed type autoimmune hemolytic anemia
Add Mixed type autoimmune hemolytic disease
Add Mixed type, cold and warm, (primary) (secondary) (symptomatic) autoimmune hemolytic anemia

New code D59.19 Other autoimmune hemolytic anemia

ICD-10-CM TABULAR OF DISEASES - PROPOSED ADDENDA

All proposed effective October 1, 2020

A75 Typhus fever
Excludes1: rickettsiosis due to Ehrlichia sennetsu (A79.81)
A75.3 Typhus fever due to Rickettsia tsutsugamushi
Add Typhus fever due to Orientia Tsutsugamushi (scrub typhus)

A77 Spotted fever [tick-borne rickettsioses]
A77.4 Ehrlichiosis
Excludes1: Rickettsiosis due to Ehrlichia sennetsu (A79.81)
A77.40 Ehrlichiosis, unspecified
A77.41 Ehrlichiosis chafeensis [E. chafeensis]
A77.49 Other ehrlichiosis
Add Ehrlichiosis due to *E. ewingii*
Add Ehrlichiosis due to *E. muris euclairensis*

A77.8 Other spotted fevers
Add Spotted fever due to *Rickettsia parkeri*
Add Spotted fever due to *Rickettsia africae* (African Tick Bite Fever)
Add Rickettsia 364D/*R.philipii* (Pacific Coast tick fever)

A79 Other rickettsioses
A79.8 Other specified rickettsioses
A79.81 Rickettsiosis due to Ehrlichia sennetsu
Add Rickettsiosis due to Neorickettsia sennetsu

E86 Volume depletion
Use additional code(s) for any associated disorders of electrolyte and acid-base balance (E87.-)
Delete Excludes1: ~~hypovolemic shock NOS (R57.1)~~
Add Excludes2: hypovolemic shock NOS (R57.1)

G20 Parkinson's disease
Hemiparkinsonism
Idiopathic Parkinsonism or Parkinson's disease
Paralysis agitans
Parkinsonism or Parkinson's disease NOS
Primary Parkinsonism or Parkinson's disease
Add Use additional code to indentify:
Add dementia with behavioral disturbance (F02.81)
Add dementia without behavioral disturbance (F02.80)

G93 Other disorders of brain

G93.4 Other and unspecified encephalopathy
Delete Excludes1:~~toxic (metabolic) encephalopathy (G92)~~
Add Excludes2:toxic (metabolic) encephalopathy (G92)

I11 Hypertensive heart disease

Revise Includes: any condition in I50.- or I51.4-I51.7, I51.89, I51.9 due to hypertension

J43 Emphysema

Revise Excludes~~1~~ 2: traumatic subcutaneous emphysema (T79.7)

K56 Paralytic ileus and intestinal obstruction without hernia

Delete Excludes1: ~~stenosis of anus or rectum (K62.4)~~
Add Excludes2: stenosis of anus or rectum (K62.4)

M31 Other necrotizing vasculopathies

M31.3 Wegener's granulomatosis

Add Granulomatosis with polyangiitis
Necrotizing respiratory granulomatosis

N20 Calculus of kidney and ureter

N20.1 Calculus of ureter
Add Calculus of the ureterpelvic junction
Ureteric stone

O99 Other maternal diseases classifiable elsewhere but complicating pregnancy, childbirth and the puerperium

O99.2 Endocrine, nutritional and metabolic diseases complicating pregnancy, childbirth and the puerperium

Revise Conditions in E00-~~E88~~ E89

P00 Newborn affected by maternal conditions that may be unrelated to present pregnancy

P00.8 Newborn affected by other maternal conditions

P00.89 Newborn affected by other maternal conditions

Add Use additional code to dentify infectious agent, if known

Q51 Congenital malformations of uterus and cervix

Q51.2 Other doubling of uterus

Delete ~~Q51.20 Other doubling of uterus, unspecified~~

~~Septate uterus, unspecified~~

Q51.21 ~~Other~~ cComplete doubling of uterus

Complete septate uterus

Revise Q51.22 ~~Other~~ pPartial doubling of uterus

Partial septate uterus

Revise Q51.28 Other doubling of uterus~~, other specified~~

Revise Septate uterus~~, other specified~~ NOS

R44 Other symptoms and signs involving general sensations and perceptions

Revise Excludes1: alcoholic hallucinations (~~F1.5)~~ F10.151, F10.251, F10.951)

Revise hallucinations in drug psychosis ~~(F11-F19 with .5)~~ (F11-F19 with fifth to sixth characters 51)

R60 Edema, not elsewhere classified

Delete Excludes1: ~~hydrops fetalis NOS (P83.2)~~

S06 Intracranial injury

S06.3 Focal traumatic brain injury

Delete Excludes1: ~~focal cerebral edema (S06.1~~)
Add Excludes2: focal cerebral edema (S06.1)

T79 Certain early complications of trauma, not elsewhere classified
Excludes2: acute respiratory distress syndrome (J80)
complications occurring during or following medical procedures (T80-T88)
complications of surgical and medical care NEC (T80-T88)
newborn respiratory distress syndrome (P22.0)
The appropriate 7th character is to be added to each code from category T79
A initial encounter
D subsequent encounter
S sequela

T79.7 Traumatic subcutaneous emphysema
Revise Excludes~~1~~2: emphysema NOS (J43)
Revise emphysema (subcutaneous) resulting from a procedure (T81.82)

Z85 Personal history of malignant neoplasm

Z85.8 Personal history of malignant neoplasms of other organs and systems

Conditions classifiable to C00-C14, C40-C49, C69-C75, C7A.098, C76-C79

Z85.81 Personal history of malignant neoplasm of lip, oral cavity, and pharynx

Add Conditions classifiable to C00-C14

Z85.82 Personal history of malignant neoplasm of skin

Z85.820 Personal history of malignant melanoma of skin

Conditions classifiable to C43

Z85.821 Personal history of Merkel cell carcinoma

Conditions classifiable to C4A

Z85.828 Personal history of other malignant neoplasm of skin

Conditions classifiable to C44

Z85.83 Personal history of malignant neoplasm of bone and soft tissue

Add Conditions classifiable to C40-C41; C45-C49

Z85.84 Personal history of malignant neoplasm of eye and nervous tissue

Add		Conditions classifiable to C69-C72
	Z85.85	Personal history of malignant neoplasm of endocrine glands
Add		Conditions classifiable to C73-C75
	Z85.89	Personal history of malignant neoplasm of other organs and systems
Add		Conditions classifiable to C7A.098, C76, C77-C79

ICD-10-CM INDEX OF DISEASES - PROPOSED ADDENDA

All proposed effective October 1, 2020

Abscess
- peritoneum, peritoneal (perforated) (ruptured) K65.1
Add - presacral K65.1
Revise - - postoperative ~~T81.49~~ T81.43
Revise -wound ~~T81.49~~
Add - -postoperative T81.49

Arthritis, arthritic (acute) (chronic) (nonpyogenic) (subacute) M19.90
Add - cervical M47.812

Bronchitis (diffuse) (fibrinous) (hypostatic) (infective) (membranous) J40
Add - aspiration (due to food and vomit) J69.0
- aspiration (due to fumes or vapors) J68.0

Calculus, calculi, calculous
Add - ureteropelvic junction N20.1

Elevated, elevation
Add - troponin R77.8

Revise Emaciation (due to malnutrition) ~~E41~~ E43

Add Facet syndrome M47.89-

Fistula
- bile duct (common) (hepatic) K83.3
Revise - - with calculus, stones – see also Calculus, bile duct
Add - - -with cholecystitis
Add - - - - with obstruction K80.41

Fracture
Add -metaphyseal – see Fracture, shaft

Revise Gunshot wound - see also ~~Wound, open~~ Puncture

Hypertension, hypertensive (accelerated) (benign) (essential) (idiopathic) (malignant) (systemic) I10
Revise - intracranial, ~~(~~benign~~),~~ G93.2
- with
Revise - - heart involvement (conditions in I50.- or I51.4-I51.7, I51.89, I51.9, due to hypertension) - see Hypertension, heart

Infection, infected, infective (opportunistic) B99.9
-due to or resulting from
- - device, implant or graft (see also Complications, by site and type, infection or inflammation) T85.79
- - - catheter NEC T85.79
- - - - dialysis (renal) T82.7
Add - - - - -central line T80.211
Add - intrauterine inflammation O41.12

Inhalation
- steam -see Toxicity, vapors

Injury …
Revise - heart (meaning nontraumatic, see Injury, heart, nontraumatic) S26.90
Add - - nontraumatic I51.89

Ischemia, ischemic I99.8
Add -limb, critical – see Arteriosclerosis, extremities

Meningioma - see also Neoplasm, meninges, benign
Add -atypical – see Neoplasm, meninges, uncertain behavior

Myelitis
Add -flaccid G04.89

Pressure
- increased
Revise - - intracranial, ~~(~~benign~~)~~, G93.2

Schizophrenia, schizophrenic F20.9
Revise - chronic undifferentiated ~~F20.5~~ F20.9

Scoliosis (acquired) (postural) M41.9
Add - degenerative M41.8

Siderosis (lung) J63.4
Add - brain G93.89

Spider
- bite - see Toxicity, venom, spider
Add - - nonvenomous-see Bite, by site, superficial, insect

Add	Spondyloarthritis
Add	- axial (see also Spondlyitis, ankylosing)
Add	- - non-radiographic (M46.80)
Add	- - - cervical (M46.82)
Add	- - - cervicothoracic (M46.83)
Add	- - - lumbar (M46.84)
Add	- - - lumbosacral (M46.87)
Add	- - - multiple sites (M46.89)
Add	- - - occipito-atlanto-axial region (M46.81)
Add	- - - sacral and sacrococcygeal (M46.88)
Add	- - - thoracic (M46.84)
Add	- - - thoracolumbar (M46.85)
Revise	Spotted fever - see Fever, spotted ~~N92.3~~ A77.9
	Syndrome - see also Disease
Add	- facet M47.89-
Add	Triple I O41.12
	Version
Delete	~~- with extraction~~