ICD-10 Coordination and Maintenance Committee Meeting

Diagnosis Agenda

Zoom Webinar and Dial-In Information

•This meeting will be conducted via Zoom Webinar. The URL to join the Zoom Webinar, the password, and the call-in numbers are the same for both days of the meeting

•Day 1: March 9, 2021: The meeting will begin promptly at 9:00 AM ET and will end at 5:00 PM ET. Lunch will be held from 12:30 PM to 1:30 PM.

•Day 2: March 10, 2021: The meeting will begin promptly at 9:00 AM ET and will end at 5:00 PM ET. Lunch will be held from 12:30 PM to 1:15 PM.

To minimize feedback to the maximum extent possible, join the meeting using only **ONE** of the options listed below.

Option 1: Remote participants (attendees wishing to both view slides and ask questions during the Q&A portions of the meeting) must join the Zoom Webinar via the web. To join this Zoom Webinar conference from a PC, MAC, iPad, iPhone or Android device as well as, connect to the audio portion of the conference:

Click the following URL:
https://cms.zoomgov.com/j/1600784651?pwd=MEZLdWhJVGZLajV2eDF0ck4zbjBtdz09
Passcode: 798401

Option 2: Dial-in access is available for listen-only participants. Listen-only participants are participants who wish to only listen to the meeting and do not wish to comment or ask questions during the Q&A portions of the meeting.

1.From your phone, dial U.S.*: 669-254-5252 or 646-828-7666 or 833-568-8864 (Toll Free)

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   Passcode: 798401

If you experience technical difficulties during the meeting, please contact Theresa Eddins for assistance at theresa.eddins@cms.hhs.gov or 212-616-2527.

Those participating in the Zoom Webinar may ask questions during the Q&A portions of the meeting using the "Raise Your Hand" feature. If time does not permit you to comment or ask a question during the Q&A session, you may submit comments and questions at any time using the "Q&A" feature. All comments and questions submitted using the "Q&A" feature, along with CDC's responses to them, will be posted as soon as possible after the meeting on our web page located at: https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm. Remaining questions may be submitted via CDC/NCHS mailbox at NCHS ICD-10-CM nchsicd10CM@cdc.gov.

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 9-10, 2021	ICD-10 Coordination and Maintenance Committee Meeting.
March 2021	Recordings and slide presentations of the March 9-10, 2021 ICD-10 Coordination and Maintenance Committee Meeting will be posted on the following web pages: **Diagnosis code portion of the recording and related materials–** https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm **Procedure code portion of the recording and related materials–** https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html
April 1, 2021	There were no ICD-10 codes finalized to capture new diagnoses or new technology for implementation on April 1, 2021. Therefore, there will be no new ICD-10 diagnosis or procedure codes implemented on April 1, 2021.
April 9, 2021	**Deadline for receipt of public comments on proposed new procedure codes and revisions discussed at the March 9-10, 2021 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2021**.
April 2021	Notice of Proposed Rulemaking to be published in the Federal Register as mandated by Public Law 99-509. This notice will include references to the FY 2022 ICD-10-CM diagnosis and ICD-10-PCS procedure codes finalized to date. It will also include proposed revisions to the MS-DRG system based on ICD-10-CM/PCS codes on which the public may comment. The proposed rule can be accessed at: https://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/AcuteInpatientPPS/index.html?redirect=/AcuteInpatientPPS/IPPS/list.asp
May 10, 2021	**Deadline for receipt of public comments on proposed new diagnosis codes and revisions discussed at the March 9-10, 2021 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2022**.

May/June 2021	Final addendum for FY 2022 code updates posted on web pages as follows: **Diagnosis addendum -** https://www.cdc.gov/nchs/icd/icd10cm.htm **Procedure addendum -** https://www.cms.gov/Medicare/Coding/ICD10/index.html
June 11, 2021	**Deadline for requestors: Those members of the public requesting that topics be discussed at the September 14-15, 2021 ICD-10 Coordination and Maintenance Committee Meeting, must have their requests submitted to CMS for procedures and NCHS for diagnoses.**
July 2021	Federal Register notice for the September 14-15, 2021 ICD-10 Coordination and Maintenance Committee Meeting will be published. This will include the tentative agenda.
August 1, 2021	Hospital Inpatient Prospective Payment System final rule to be published in the Federal Register as mandated by Public Law 99-509. This rule will also include links to all the final codes to be implemented on October 1, 2021. This rule can be accessed at: https://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/AcuteInpatientPPS/index.html
August 2021	Tentative agenda for the Diagnosis portion of the September 15, 2021 ICD-10 Coordination and Maintenance Committee Meeting will be posted on the NCHS webpage at - https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm Tentative agenda for the Procedure portion of the September 14, 2021 ICD-10 Coordination and Maintenance Committee Meeting will be posted on the CMS webpage at – https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html
August 9, 2021	**On-line registration opens for the September 14-15, 2021 ICD-10 Coordination and Maintenance Committee Meeting at:** https://www.eventbrite.com/e/icd-10-coordination-and-maintenance-committee-meeting-tickets Please note that this meeting will be conducted virtually and registration is not required to attend. However, we are providing the

	ability to register on-line for those required to provide proof of attendance for continuing education purposes. The on-line registration will be available through September 9, 2021.
September 14-15, 2021	The September 2021 ICD-10 Coordination and Maintenance Committee Meeting will be held fully virtual, with no in-person audience. Those who wish to attend must participate via Zoom Webinar or by dialing in.
September 2021	Recordings and slide presentations of the September 14-15, 2021 ICD-10 Coordination and Maintenance Committee Meeting will be posted on the following web pages: **Diagnosis code portion of the recording and related materials–** https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm **Procedure code portion of the recording and related materials–** https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html
October 1, 2021	New and revised ICD-10-CM and ICD-10-PCS codes go into effect along with MS-DRG changes. Final addendum available on web pages as follows: **Diagnosis addendum –** https://www.cdc.gov/nchs/icd/icd10cm.htm **Procedure addendum –** https://www.cms.gov/Medicare/Coding/ICD10/
October 15, 2021	**Deadline for receipt of public comments on proposed new codes discussed at the September 14-15, 2021 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on April 1, 2022.**
November 2021	Any new ICD-10 codes required to capture new technology that will be implemented on the following April 1 will be announced. Information on any new codes to be implemented April 1, 2022 will be posted on the following websites: https://www.cdc.gov/nchs/icd/icd10cm.htm https://www.cms.gov/Medicare/Coding/ICD10/

November 15, 2021	**Deadline for receipt of public comments on proposed new codes and revisions discussed at the September 14-15, 2021 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2022.**

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

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Continuing Education Credits

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

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

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

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

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

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

Acute and Chronic Metabolic Acidosis

The Renal Physicians Association (RPA) is requesting a new ICD-10-CM code for chronic metabolic acidosis in chronic kidney disease (CKD).

Chronic metabolic acidosis is both a serious complication and an underlying cause of chronic kidney disease (CKD) progression. As kidney function deteriorates, patients cannot secrete adequate amounts of acid. The resulting acid-base imbalance leads to a reduction in serum bicarbonate. It is a clinically distinct disorder from acute metabolic acidosis, which either results from hypoperfusion, alterations in glucose metabolism (Diabetic or Starvation Ketoacidosis) or less commonly from ingestion of toxic substances. (1) Acute metabolic acidosis is typically associated with conditions that result in hospitalization and treatment is primarily aimed at correcting the underlying etiology (e.g., antimicrobial treatment of sepsis, volume resuscitation, control of hyperglycemia, etc.). In contrast, chronic metabolic acidosis is caused by a kidney-related pathology, most commonly CKD, and treatment is primarily aimed at increasing the serum bicarbonate level over the long term. Recognition and identification of acute versus chronic metabolic acidosis in CKD is therefore important to ensure appropriate clinical evaluation, treatment plans and optimal outcomes.

Chronic metabolic acidosis, a complication of CKD, is also associated with an increased risk of CKD progression and death. [(2)] Prospective, controlled studies and large retrospective cohort studies have shown an association between low serum bicarbonate levels and the progression of renal disease. [(3)] A 1mEq/L decline in serum bicarbonate is associated with a 6-9% increase in the risk of either end-stage renal disease (ESRD) or at least a 40% reduction in eGFR. The relationship between serum bicarbonate and CKD progression is linear and consistent with subgroups of patients who have reduced eGFR and chronic metabolic acidosis. [(4)]

Clinical evidence indicates that over time, metabolic acidosis may lead to adverse musculoskeletal effects including muscle wasting and loss of bone density as well as increased inflammation and kidney fibrosis. Left uncontrolled, a cycle of worsening acidosis and accelerated progression of kidney disease can result. Even mild metabolic acidosis may contribute to the development of bone disease and muscle degradation, a finding that has important implications for recognition and urgent treatment of chronic metabolic acidosis in CKD. [(5)] Other adverse effects of chronic metabolic acidosis in CKD include impaired cognitive, vascular and functional status. [(6)]

Chronic metabolic acidosis in CKD is undertreated in part due to lack of recognition of the condition. Addition of a new code for metabolic acidosis in CKD will enable better identification and tracking of this distinct set of patients, which is anticipated will advance the clinical understanding of the condition, and subsequently improve the diagnostic and treatment paradigms. This accurate identification is critical to facilitate research that will further elucidate the marked needs and characteristics of metabolic acidosis in CKD and aligns with the clinical consensus reached by clinical experts in the field of nephrology.

There is one ICD-10-CM code for non-diabetic acidosis, E87.2. E87.2 includes multiple forms of non-diabetic acidosis: lactic acidosis; respiratory acidosis; and metabolic acidosis. This proposal seeks to expand the E87.2 code to clarify acidosis and accommodate metabolic acidosis in CKD.

References

1. Kraut JA and Madia NE. Treatment of acute metabolic acidosis: a pathophysiologic approach. *Nature Reviews Nephrology* 8, 589-601 (2012).
2. Dubey AK, et al. Correction of metabolic acidosis improves muscle mass and renal function in chronic kidney disease stages 3 and 4: a randomized controlled trial. Nephrol Dial Transplant (2018) 1-9; 1. Kidney Disease: Improving Global Outcomes (KDIGO) CKD Work Group. KDIGO 2012 Clinical Practice Guideline for the Evaluation and Management of Chronic Kidney Disease. Kidney inter., Suppl. 2013; 3: 1–150; Kraut JA, Madias NE. Metabolic acidosis of CKD: an update. *Am J Kidney Dis.* 2016;67(2):307-317.
3. Goraya N, et al. A comparison of treating metabolic acidosis in CKD stage 4 hypertensive kidney disease with fruits and vegetables or sodium bicarbonate. *Clin J Am Soc Nephrol* 8: 371–381, 2013; Goraya N, et al. Treatment of metabolic acidosis in patients with stage 3 chronic kidney disease with fruits and vegetables or oral bicarbonate reduced urine angiotensinogen and preserves glomerular filtration rate. *Kidney International* advance online publication, 2 April 2014; doi:10.1038/ki.2014.83; Shah SN, et al. Serum bicarbonate levels and the progression of kidney disease: a cohort study. *Am J Kidney Dis.* 54(2): 270 - 277, 2009; Dobre M, et al. Association of serum bicarbonate with risk of renal and cardiovascular outcomes in CKD: a report from the chronic renal insufficiency cohort (CRIC) study. *Am J Kidney Dis*, 62(4): 670-678, 2013; Raphael K, et al. Higher serum bicarbonate levels with the normal range are associated with better survival and renal outcomes in African Americans. *Kidney Intl*, 2011;79(3):356-362; Tangri N, et al. A predictive model for progression of chronic kidney disease to kidney failure. *JAMA*, 305:1553-1559,2011; Kovesdy CP, et al. Association of serum bicarbonate levels with mortality in patients with non-dialysis-dependent CKD. *Nephrol Dial Transplant*, (2009) 24: 1232-1237; Navaneethan SD, et al. (2011) Serum bicarbonate and mortality in stage 3 and stage 4 chronic kidney disease. *CJASN*, 6: 2396 – 2402, 2011; Raphael K. Approach to the treatment of chronic metabolic acidosis in CKD. *Am J Kidney Dis* 67(4): 696 – 702, 2016.
4. Tangri N, et al. Use of the kidney failure risk equations to model clinical trial outcomes. Abstract and poster presented at the American Society of Nephrology (ASN)/Kidney Week 2018. October 2018.
5. Rajan, VR and Mitch, WE (2008). Muscle wasting in chronic kidney disease: the role of the ubiquitin proteasome system and its clinical impact. *Pediatr Nephrol* 23(4): 527-535; Kraut JA, Madias NE. Metabolic acidosis of CKD: an update. *Am J Kidney Dis.* 2016;67(2):307-317; Domrongkitchaiporn S, et al. Bone histology and bone mineral density after correction of acidosis in distal renal tubular acidosis. *Kidney Int.* 62: 2160 – 2166, 2002.
6. Dobre M, et al. (2018) Serum bicarbonate concentration and cognitive function in hypertensive adults. *CJASN* April 2018, 13 (4) 59-603; Kendrick J, et al. (2018). Effect of treatment of metabolic acidosis on vascular endothelial function in patients with CKD. A pilot randomized cross-over study. *CJASN* October 2018, 13 (10) 1463-1470.

TABULAR MODIFICATIONS

E87 Other disorders of fluid, electrolyte and acid-base balance

E87.2 Acidosis

Delete ~~Acidosis NOS~~
Delete ~~Lactic acidosis~~
Delete ~~Metabolic acidosis~~
Delete ~~Respiratory acidosis~~

Excludes1: diabetic acidosis - see categories E08-E10, E13 with ketoacidosis

New code E87.20 Acidosis unspecified
Add Lactic acidosis NOS
Add Metabolic acidosis NOS
Add Code also, if applicable, respiratory failure with hypercapnia (J96. with 5th character 2)

New code E87.21 Acute metabolic acidosis
Add Acute lactic acidosis

New code E87.22 Chronic metabolic acidosis
Add Chronic lactic acidosis

Add Code first underlying etiology, if applicable

New code E87.29 Other acidosis
Add Respiratory acidosis

J96 Respiratory failure, not elsewhere classified

J96.0 Acute respiratory failure
J96.02 Acute respiratory failure with hypercapnia
Add Acute respiratory acidosis

J96.1 Chronic respiratory failure
J96.12 Chronic respiratory failure with hypercapnia
Add Chronic respiratory acidosis

ANCA Vasculitis

Antineutrophilic cytoplasmic antibody (ANCA) associated vasculitis represents a group of autoimmune conditions that cause inflammation of blood vessels, and can affect multiple systems. It includes three main systemic vasculitides: granulomatosis with polyangiitis (GPA; or formerly Wegener granulomatosis), eosinophilic granulomatosis with polyangiitis (EGPA; or previously Churg-Strauss syndrome), and microscopic polyangiitis (MPA). In addition, other forms of ANCA vasculitis include drug-induced vasculitis and renal limited vasculitis.

ANCA vasculitis is rare, with incidence estimates ranging from 10 to 25 cases per million per year, and prevalence estimates ranging from 46 to 184 cases per million population. Although rare, it affects thousands of people annually in the U.S.

The antibodies in ANCA vasculitis generally may be further identified as perinuclear P-ANCA antibody, which is against neutrophil myeloperoxidase (MPO); or diffusely cytoplasmic, C-ANCA antibody, which is against neutrophil proteinase 3 (PR3).

Broad symptoms in different types of ANCA vasculitis include fatigue, fever, and weight loss. There are specific findings typical in the specific disorders. Renal involvement can range from none, to hematuria, or to rapidly progressive renal failure, in some cases with crescentic glomerulonephritis.

GPA typically involves the upper and lower respiratory tracts as well as the kidney. Upper respiratory tract manifestations may include bloody nasal discharge, nasal ulceration, sinusitis, and chronic otitis media. The nasal cartilage may be damaged, causing a characteristic saddle nose deformity. Lower respiratory tract involvement may include lung nodules, and alveolar hemorrhage in some cases can be severe and even fatal. Rarely GPA can cause tracheal stenosis. Renal involvement can cause rapidly progressive renal failure. The patient can present with high blood pressure, new-onset proteinuria, and active urinary sediments (hematuria, and leukocytes in urine). Around 90 percent of patients with multisystemic active GPA have ANCA positivity. Thus, absence of ANCA does not rule out the diagnosis of GPA. The antibodies in GPA may often be C-ANCA (anti-PR3).

MPA causes a necrotizing vasculitis of small vessels, without granuloma formation. It often can cause glomerulonephritis with acute renal failure. The kidney involvement is nearly always present in MPA. Lung involvement may occur but is less common than in GPA. MPA may cause inflamed capillaries, and this can lead to severe alveolar bleeding. It can also cause pulmonary fibrosis. Around 90 percent of those with MPA are ANCA positive. Sometimes MPA may be only affect the kidneys, and that may be referred to as renal limited MPA. The antibodies in MPA may often be P-ANCA (anti-MPO).

EGPA can cause eosinophilic granulomatous lesions which may involve the skin, heart, and gastrointestinal tract. It may also often affect the peripheral nervous system. The antibodies in EGPA can be either C-ANCA or P-ANCA (anti-PR3, or anti-MPO), but about 40% of those with EGPA are ANCA negative.

Drug-induced ANCA vasculitis has been linked to exposures to a number of different medications, including propylthiouracil, methimazole, carbimazole, hydralazine, and minocycline. Symptoms are often constitutional, such as arthralgias, fatigue, and skin rash. However, the full range of clinical features seen in other types of ANCA vasculitis can occur, including rapidly progressive renal failure and alveolar hemorrhage.

Early diagnosis and treatment of ANCA vasculitis is important, as if untreated, the two year survival is less than 10%, while with treatment, it is generally over 90%. The presence of ANCA antibodies in the specific disorders of GPA, MPA, and EGPA potentially can provide important prognostic information, and as noted previously, these antibodies are not universally present.

A proposal to add a specific code for ANCA vasculitis has been received from Mercy Coding Services.

References

Hunter RW, Welsh N, Farrah TE, Gallacher PJ, Dhaun N. ANCA associated vasculitis. BMJ 2020; 369:m1070. doi: https://doi.org/10.1136/bmj.m1070 (Published 14 April 2020) Retrieved from https://www.bmj.com/content/369/bmj.m1070

Qasim A, Patel J. ANCA Positive Vasculitis. StatPearls [Internet]. Dec 14, 2020. Retrieved from: https://www.ncbi.nlm.nih.gov/books/NBK554372/

Yates M, Watts R. ANCA-associated vasculitis. Clin Med (Lond). 2017 Feb; 17(1): 60–64. doi: https://doi.org/10.7861/clinmedicine.17-1-60 Retrieved from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6297586/

Murphy S. ANCA Vasculitis. Kidney Health Library, UNC Kidney Center. Sep 2018. Retrieved from https://unckidneycenter.org/kidneyhealthlibrary/glomerular-disease/anca-vasculitis/

TABULAR MODIFICATIONS

I77 Other disorders of arteries and arterioles

I77.8 Other specified disorders of arteries and arterioles

New code I77.82 Antineutrophilic cytoplasmic antibody [ANCA] vasculitis

Add ANCA associated vasculitis

Add ANCA positive vasculitis

Add Excludes2: eosinophilic granulomatosis with polyangiitis (M30.1)

Add granulomatosis with polyangiitis (M31.3-)

Add microscopic polyangiitis (M31.7)

Aortic Aneurysm and Dissection

It has been proposed by W. L. Gore & Associates, Inc., that ICD-10-CM be expanded to create specific codes identifying the anatomy involved, for aortic aneurysm and dissection. This would enable better capture of clinical presentation, and more utility for clinicians.

Patients who present with dissection or aneurysm formation within the aorta are treated on the basis of their anatomy. [(1)] The diagnosis, medical and surgical management, risk of adverse events, and overall resources involved with the patient's care are directly related to the site of disease. [(2-5)] For example, patients with an infrarenal abdominal aortic aneurysm may be treated via minimally invasive endovascular means and have a relatively short length of stay in the hospital, and lower risk of complications. [(6-7)] Conversely, patients who present with an aortic aneurysm near the mesenteric arteries often require extensive open surgical reconstruction, with length of stay of 5-7 days, risk of complications around 10%, and involving substantially more hospital resources. [(1-5)]

The aorta has three layers: from the innermost layer the intima, followed by the media, and the adventitia. An aortic dissection is a tear in the aorta that occurs between the intima and media. Expansion of this tear can block critical vessels branching from the aorta, leading to ischemia of the affected organ or extremity. The most feared aortic dissection is one that affects the ascending aorta, due to the potential for coronary ischemia.

A dissection that is restricted to the aortic arch may be treated in multiple different ways, depending on the exact location of the tear and the presentation of the patient. Use of either the Stanford or DeBakey classification may generate confusion, as neither differentiates whether an isolated dissection of the aortic arch is separate from a dissection of the ascending aorta. A focal dissection of the aortic arch could be treated with endovascular stent placement, which has a very different patient risk exposure compared to surgical reconstruction of the ascending aorta. It is proposed to create ICD-10-CM codes that specifically identify aneurysm of the aortic arch, and of the ascending aorta, and the descending thoracic aorta, to clearly identify the anatomy that is affected.

Similarly, a coding schema based on anatomy for the entire aorta would help differentiate the exact site of the dissection. New codes are proposed for dissections of the ascending aorta, aortic arch, and the descending aorta.

For aneurysms of the abdominal aorta, further granularity is proposed to accurately reflect the location of the aneurysm as pararenal, juxtarenal, and infrarenal; again, codes would be expanded reflecting the differentiation of ruptured and without rupture (based on the current codes). Similarly, codes for aneurysms of the thoracoabdominal aorta would be expanded, to create specific codes for supraceliac and paravisceral aneurysms (also rupture and without rupture, based on the current codes).

These additional codes will more accurately depict the pathology of disease, which in turn will have positive benefits to our public health, disease management, and resource allocation. This will enable improved insight into the incidence and prevalence of aortic dissection and aneurysm formation as it relates to the underlying anatomy [(8)], which in turn will drive innovation and appropriateness in the treatment of complex aortic pathologies [(9)], and enable establishment of a more direct relationship between the site of disease and the expected outcome, facilitating the management of patients from both a utilization management and resource planning perspective for hospitals. [(9)]

Footnotes

1. Crawford ES, Svensson LG, Coselli JS, Safi HJ, Hess KR. Aortic dissection and dissecting aortic aneurysms. Ann Surg. 1988 Sep;208(3):254-73.

2. Svensson LG, Crawford ES, Hess KR, Coselli JS, Safi HJ. Dissection of the aorta and dissecting aortic aneurysms. Improving early and long-term surgical results. Circulation. 1990 Nov;82(5 Suppl):IV, 24-38.

3. Svensson LG, Crawford ES, Hess KR, Coselli JS, Safi HJ. Thoracoabdominal aortic aneurysms associated with celiac, superior mesenteric, and renal artery occlusive disease: methods and analysis of results in 271 patients. J Vasc Surg. 1992 Sep;16(3):378-89; discussion 389-90.

4. Ham SW, Chong T, Moos J, Rowe VL, Cohen RG, Cunningham MJ, Wilcox A, Weaver FA. Arch and visceral/renal debranching combined with endovascular repair for thoracic and thoracoabdominal aortic aneurysms. J Vasc Surg. 2011 Jul;54(1):30-40; discussion 40-1. doi: 10.1016/j.jvs.2010.12.033. Epub 2011 Feb 21.

5. Coselli JS, LeMaire SA, Preventza O, de la Cruz KI, Cooley DA, Price MD, Stolz AP, Green SY, Arredondo CN, Rosengart TK. Outcomes of 3309 thoracoabdominal aortic aneurysm repairs. J Thorac Cardiovasc Surg. 2016 May;151(5):1323-37. doi: 10.1016/j.jtcvs.2015.12.050. Epub 2016 Jan 14.

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Additional References

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2. Silaschi M, Byrne J, Wendler O. Aortic dissection: medical, interventional and surgical management. Heart. 2017 Jan 1;103(1):78-87. doi: 10.1136/heartjnl-2015-308284. Epub 2016 Oct 12. Review.

TABULAR MODIFICATIONS

I71 Aortic aneurysm and dissection

Delete ~~Excludes1: aortic ectasia (I77.81-)~~

Delete ~~syphilitic aortic aneurysm (A52.01)~~

Delete ~~traumatic aortic aneurysm (S25.09, S35.09)~~

Add Code first, if applicable:

Add syphilitic aortic aneurysm (A52.01)

Add traumatic aortic aneurysm (S25.09, S35.09)

I71.0 Dissection

I71.01 Dissection of thoracic aorta

New code I71.010 Dissection of ascending aorta

New code I71.011 Dissection of aortic arch

New code I71.012 Dissection of descending thoracic aorta

New code I71.019 Dissection of thoracic aorta, unspecified

I71.1 Thoracic aortic aneurysm, ruptured

New code I71.10 Thoracic aortic aneurysm, ruptured, unspecified

New code I71.11 Aneurysm of the ascending aorta, ruptured

New code I71.12 Aneurysm of the aortic arch, ruptured

New code I71.13 Aneurysm of the descending thoracic aorta, ruptured

I71.2 Thoracic aortic aneurysm, without rupture

New code I71.20 Thoracic aortic aneurysm, without rupture, unspecified

New code I71.21 Aneurysm of the ascending aorta, without rupture

New code I71.22 Aneurysm of the aortic arch, without rupture

New code I71.23 Aneurysm of the descending thoracic aorta, without rupture

I71.3 Abdominal aortic aneurysm, ruptured

New code I71.30 Abdominal aortic aneurysm, ruptured, unspecified

New code I71.31 Pararenal abdominal aortic aneurysm, ruptured

New code I71.32 Juxtarenal abdominal aortic aneurysm, ruptured

New code I71.33 Infrarenal abdominal aortic aneurysm, ruptured

I71.4 Abdominal aortic aneurysm, without rupture

New code I71.40 Abdominal aortic aneurysm, without rupture, unspecified

New code I71.41 Pararenal abdominal aortic aneurysm, without rupture

New code I71.42 Juxtarenal abdominal aortic aneurysm, without rupture

New code I71.43 Infrarenal abdominal aortic aneurysm, without rupture

I71.5 Thoracoabdominal aortic aneurysm, ruptured

New code I71.50 Thoracoabdominal aortic aneurysm, ruptured, unspecified

New code I71.51 Supraceliac aneurysm of the abdominal aorta, ruptured

New code I71.52 Paravisceral aneurysm of the abdominal aorta, ruptured

I71.6 Thoracoabdominal aortic aneurysm, without rupture

New code I71.60 Thoracoabdominal aortic aneurysm, without rupture, unspecified

New code I71.61 Supraceliac aneurysm of the abdominal aorta, without rupture

New code I71.62 Paravisceral aneurysm of the abdominal aorta, without rupture

Apnea of Newborn and Related Issues

Apnea can occur in any newborn child. An apneic spell is generally defined as a cessation of breathing for 20 seconds or longer or a shorter pause accompanied by bradycardia (<100 beats per minute), cyanosis, and/or pallor. In practice, many apneic events, especially in preterm infants, are shorter than 20 seconds since these briefer pauses tend to result in bradycardia or hypoxemia.

On the basis of respiratory effort and airflow, apnea may be classified as central (cessation of breathing effort), obstructive (airflow obstruction usually at the pharyngeal level), or mixed. Apnea of prematurity is a developmental disorder caused by immaturity of neurologic and/or mechanical function of the respiratory system.

Central apnea is caused by immature medullary respiratory control centers. The specific pathophysiology is not understood completely but appears to involve a number of factors, including abnormal responses to hypoxia and hypercapnia. This is the most common type of apnea of prematurity.

Obstructive apnea is caused by obstructed airflow, neck flexion causing opposition of hypopharyngeal soft tissues, nasal occlusion, or reflex laryngospasm. Mixed apnea is a combination of central and obstructive apnea.

All types of apnea can cause hypoxemia, cyanosis, and bradycardia if the apnea is prolonged. Because bradycardia can also occur simultaneously with apnea, a central mechanism may be responsible for both. About 18% of infants who have died of sudden infant death syndrome (SIDS) had a history of prematurity, but apnea of prematurity is not a precursor to SIDS.

Apnea of prematurity is one of the most common diagnoses in the neonatal intensive care unit (NICU). This is distinct and separate condition from newborn sleep apnea. While apnea of prematurity can be diagnosed based on clinical findings, sleep apnea is diagnosed based on polysomnography. During this test at least three channels, chest wall movement, airflow documented by CO2 measurement, and oxygenation (generally measure as SpO2), are documented while the infant is awake and asleep.

Mixed and obstructive apnea can usually be managed with supplemental oxygen and continuous positive airway pressure (CPAP) ventilation. Occasionally surgical intervention, such as palatoplasty or in extreme cases tracheostomy, may be required. In addition, central may require medications to help stimulate the respiratory centers in the brain. Almost all of the children will be discharged with a home monitor, e.g. apnea/bradycardia monitor. Unfortunately, the sensitivity of these monitors may cause false positive alarms that result in the child's family to seek medical services.

As many of these babies will go home with monitoring devices after discharge from the hospital, it is also important to be able to identify encounters, often times in the acute care setting, when the parent presents with a newborn/infant after their home monitoring device goes off indicating a problem. These devices may vary, but typically detect apnea and bradycardia. After exam and review of the data, it is then discovered that there is nothing wrong with the baby. At that time there is no diagnosis to be made other than this was an observation after the home physiologic monitoring device went off, with no clinical findings.

In addition, bed sharing, i.e. co-sleeping, can increase the risk of apnea due to neurologic injury from smothering and suffocation of the infant by the adult. Lastly, related to that we are request the increased risk of suffocation to newborn/infant who share sleeping arrangements with an adult.

As a result of this complex issue facing neonates, particularly those who are premature, the American Academy of Pediatrics (AAP) are requesting additions to the ICD-10-CM code set to identify the specific types of sleep apnea and apnea (of prematurity) that occurs outside of sleep.

TABULAR MODIFICATIONS

	P28	Other respiratory conditions originating in the perinatal period
Delete		~~Excludes1: congenital malformations of the respiratory system (Q30-Q34)~~
Add		Code also, if applicable, congenital malformations of the respiratory system (Q30-Q34)
		P28.3 Primary sleep apnea of newborn
Delete		~~Central sleep apnea of newborn~~
Delete		~~Obstructive sleep apnea of newborn~~
Delete		~~Sleep apnea of newborn NOS~~
Add		Excludes2: apnea of prematurity (P28.40)
New code		P28.30 Sleep apnea of the newborn, unspecified
Add		Transient oxygen desaturation spells of newborn, sleep
New code		P28.31 Central sleep apnea of newborn
New code		P28.32 Obstructive sleep apnea of newborn

New code	P28.33 Mixed sleep apnea of the newborn
New code	P28.39 Other sleep apnea of newborn
	P28.4 Other apnea of newborn
Delete	~~Apnea of prematurity~~
Delete	~~Obstructive apnea of newborn~~
Delete	~~Excludes1:obstructive sleep apnea of newborn (P28.3)~~
Add	Excludes2:obstructive sleep apnea of newborn (P28.32)
New code	P28.40 Unspecified apnea (of prematurity) of newborn
Add	Apnea, NOS
Add	Transient oxygen desaturation spells of newborn
New code	P28.41 Central neonatal apnea (of prematurity)
New code	P28.42 Obstructive neonatal apnea (of prematurity)
New code	P28.43 Mixed neonatal apnea (of prematurity)
New code	P28.44 Apnea of prematurity
New code	P28.49 Other apnea of newborn

Z03 Encounter for medical observation for suspected diseases and conditions ruled out

Z03.8 Encounter for observation for other suspected diseases and conditions ruled out

New code	Z03.83 Encounter for observation for suspected condition related to home physiologic monitoring device
Add	Encounter for observation for apnea alarm without findings
Add	Encounter for observation for bradycardia alarm without findings
Add	Encounter for observation for malfunction of home cardiorespiratory monitor

Add	Encounter for observation for non-specific findings home physiologic monitoring device
Add	Encounter for observation for pulse oximeter alarm without findings
Add	Excludes1: apnea NOS (R06.81)
Add	neonatal bradycardia (P29.12)
Add	newborn apnea (P28.4)
Add	newborn sleep apnea (P28.3)
Add	sleep apnea (G47.3-)

Z72 Problems related to lifestyle

Z72.82 Problems related to sleep

New code	Z72.823 Risk of suffocation (smothering) under another while sleeping
Add	Child-caregiver co-sleeping
Add	Infant bed-sharing

Atrial Septal and Atrioventricular Septal Defect

For birth defects surveillance purposes, atrial septal defect (ASD) is considered a major malformation, whereas patent or persistent foramen ovale (PFO) is a normal finding in the immediate newborn period. Currently, these conditions are classified with in the same ICD-10-CM code.

NYS Birth Defects Registry routinely uses ICD codes to identify unreported cases in administrative hospital discharge data. In response to these audits, hospital staff review the medical records, and submit reports on those that are reportable defects and indicate which audited records are not reportable. A substantial number of notifications from hospitals about not reportable audited records are for children that were only diagnosed with a PFO. Therefore, hospitals must review that record twice and registry staff must spend time auditing records that are actually correct and corresponding with hospital staff. Separating the defects under different codes will drastically reduce this workload, as well as significantly improve data quality, collection and surveillance activities.

In a previous internal examination of New York State (NYS) Birth Defects Registry (BDR) data for 1990-1999 births in an 11-county surveillance region, we found that only about 5% of isolated ICD-9-CM 745.5 codes reported to the Registry were linked to a septal surgery within 5 years of birth (unpublished). Glidewell et al. conducted a small validation 745.5 study among 3 geographic locations in the United States, including NYS BDR data combined with other state-specific data ascertainment sources, and found only 24-59% of the time the code identified isolated ASD[5]. Having the two defects under the same code creates unnecessary additional workload for both hospital or healthcare provider reporters and internal surveillance staff, as well as challenges for researchers investigating risk factors for birth defects and the revalence, healthcare/service utilization, and outcomes in those living with birth defects.

The NYS BDR considers children to be eligible for surveillance it they have one or more major malformations. Thus, a child with only a PFO would not be reportable.

A 2018 publication in the journal Congenital Heart Disease indicates that “although the ICD-9-CM code 745.5 is widely used to indicate the presence of a secundum atrial septal defect (ASD), it is also used for patent foramen ovale (PFO) which is a normal variant and for "rule-out" congenital heart disease (CHD). The ICD-10-CM code Q21.1 perpetuates this issue”[1]. Studies support the use of separate diagnosis codes for component defects of Q21.1[1,2], highlight the inability to identify a specific defect using the ICD code[2], and acknowledge that the code cannot be relied upon to identify important conditions[3].

In addition to ICD codes, NYS BDR collects birth defect descriptions which helps tease out a portion of the PFO records that are submitted under code Q21.1. However, the assigned ICD-10 label for that code is “Atrial Septal Defect.” Some hospital reporters have reporting mechanisms that rely more heavily on those labels, leading to systematic misclassification.

Similarly, the ASD/PFO split is also important for the ability to do healthcare utilization research with administrative data sources. Since 2012, CDC has funded sites, including the NYS Department of Health, for surveillance projects to better understand the prevalence, healthcare utilization, and longer-term outcomes of adolescents and adults with CHD. For the pilot phase of the project, cases with isolated 745.5 codes were analyzed separately due to the potential for misclassification[5,6].

In addition, the different types of endocardial cushion defects share a common developmental process, but can have different clinical implications, severity, and treatments. Ostium primum atrial septal defect shares this developmental process but is generally a milder form clinically and considered a partial, rather than full, atrioventricular septal defect. It is important that the difference be reflected in the ICD-10-CM coding for both clinical and surveillance purposes.

Adding granularity to Q21.1 will drastically improve the data quality and accuracy of the component defects, allowing researchers and surveillance staff to analyze and understand the defects more efficiently. Using administrative data is a common practice and it is important to make improvements where feasible

This change would directly benefit all other birth defects registries across the nation, particularly those that only have access to ICD-10 codes and do not collect additional information, such as birth defect description. Active registries that use ICD codes to flag records to review would also greatly benefit from the ability to omit PFO records from their abstraction procedures.

This data is used to inform policies, guidelines, research, patient care recommendations, and more[1]. Medical professionals that have access to clearly distinguishable, accurate, and valid data on the different types of birth defects that currently comprise Q21.1 will be able to provide better patient care. They will better understand the prevalence of each defect and be able to reference more accurate research studies that summarize everything from best patient care practices to patient outcomes, ultimately impacting the day to day actions of medical providers, and experiences of patients.

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TABULAR MODIFICATIONS

Q21 Congenital malformations of cardiac septa
Excludes1: acquired cardiac septal defect (I51.0)

Q21.0 Ventricular septal defect
Roger's disease

New Subcategory Q21.1 Atrial septal defect
Delete ~~Coronary sinus defect~~
Delete ~~Patent or persistent foramen ovale~~
Delete ~~Patent or persistent ostium secundum defect (type II)~~
Delete ~~Patent or persistent sinus venosus defect~~

New Code Q21.10 Atrial septal defect, unspecified

New Code Q21.11 Atrial septal abnormality, of indeterminate type
Add Atrial septal defect or patent foramen ovale, exact type undetermined

New Code Q21.12 Secundum atrial septal defect
Add Fenestrated atrial septum
Add Patent or persistent ostium secundum defect (type II)

New Code Q21.13 Patent foramen ovale
Add Persistent foramen ovale

New Code Q21.14 Coronary sinus defect

New Code Q21.15 Sinus venosus defect

New Code Q21.19 Other specified atrial septal defect
Add Common atrium

New Subcategory	Q21.2 Atrioventricular septal defect
Delete	~~Common atrioventricular canal~~
Delete	~~Endocardial cushion defect~~
Delete	~~Ostium primum atrial septal defect (type I)~~
New Code	Q21.20 Atrioventricular septal defect, unspecified as to partial or complete
Add	Atrioventricular canal, NOS
Add	Endocardial cushion defect NOS
Add	Ostium primum atrial septal defect (type I) NOS
New Code	Q21.21 Partial atrioventricular septal defect
Add	Incomplete atrioventricular canal
Add	Incomplete atrioventricular septal defect
Add	Incomplete endocardial cushion defect
Add	Partial atrioventricular canal
Add	Partial endocardial cushion defect
Add	Ostium primum atrial septal defect (type I) with separate atrioventricular valves
New Code	Q21.22 Transitional atrioventricular septal defect
Add	Intermediate atrioventricular canal
Add	Intermediate atrioventricular septal defect
Add	Intermediate endocardial cushion defect
Add	Ostium primum atrial septal defect (type I) with separate atrioventricular valves and a small or restrictive inlet VSD
Add	Transitional atrioventricular canal
Add	Transitional endocardial cushion defect
New Code	Q21.23 Complete atrioventricular septal defect
Add	Common atrioventricular septal defect
Add	Common atrioventricular canal
Add	Common endocardial cushion defect
Add	Ostium primum atrial septal defect (type I) with common atrioventricular valve and a moderate or larger inlet VSD

Dementia: Stage of Severity, Behavioral and Psychological Symptoms

Dementia, also known as major neurocognitive disorder, is characterized by a significant decline in cognitive functions such as memory, problem-solving, attention, and language skills. It is generally due to an underlying disorder such as cerebrovascular disease or Alzheimer's disease, although a specific underlying disorder sometimes cannot be identified.

The burden for dementia is high to both patients, whose quality of life is greatly impacted, as well as society in terms of resources required. For example, among individuals age 65 or older, those with dementia have twice as many hospital stays per year and their rate of skilled nursing facility stays is almost four times higher.[1] In addition, patients with chronic conditions and dementia use more healthcare services than patients with chronic conditions who do not have dementia.[1]

Current codes for dementia do not identify the stage of severity and also do not fully identify behavioral and psychological symptoms of dementia (BPSD). Both of these clinical elements are major factors in patient management strategies. Particularly because dementia is progressive, there is a great need for the longitudinal clinical data to capture the stage of severity and the key associated disorders over time to move research and clinical studies forward.

Stage of Severity

The progression of dementia moves through three characteristic stages of cognitive impairment or neurobehavioral changes: mild dementia, moderate dementia, and severe dementia. In accordance with definitions for which there is broad consensus,[2] these stages are routinely used by clinicians working with dementia patients as well as professional societies and advocacy groups, including the American Academy of Neurology, the American Geriatrics Society, the Gerontological Society of America, the National Society on Aging, and the Alzheimer's Association.

Mild dementia : Clearly evident functional impact on daily life, affecting mainly instrumental activities. No longer fully independent/requires occasional assistance with daily life activities.
Moderate dementia : Extensive functional impact on daily life with impairment in basic activities. No longer independent and requires frequent assistance with daily life activities.

Severe dementia : Clinical interview may not be possible. Complete dependency due to severe functional impact on daily life with impairment in basic activities, including basic self-care.

In conjunction with the descriptive picture, the same stages are assigned though quantitative measures on various staging tests and instruments which have demonstrated reliability and validity in capturing disease progression.[3]

The precise scope of the stages may vary to some degree according to the type of dementia as well as patient education, age, and ethnicity. However, because the severity stages are based on changes in an individual's daily function, they are widely applied to dementia due to all underlying disorders as well as dementia of unknown etiology.

Management in earlier stages generally consists of establishing coping behaviors and managing symptoms with medications. Other medications are introduced in later stages when symptoms and associated conditions are more severe, and new environments or contracted caretakers often become necessary. Research suggests that the healthcare costs increase as the stage of severity does.[4]

It should be noted that a diagnosis of mild cognitive disorder, also known as mild cognitive impairment, has been recognized as preceding dementia in many cases. On the continuum, mild cognitive disorder is characterized by cognitive deficits that exceed those expected for a particular age but do not reach the level of clinical dementia. This pre-dementia state may be protracted but may also progress to dementia. A proposal to create new subcategory F06.7 with two new codes for mild cognitive disorder was presented at the September 2020 meeting of the ICD-10 Coordination and Maintenance Committee and is being re-presented at the March 2021 meeting. Related changes in the proposal for mild cognitive disorder are factored into the proposal for dementia.

Behavioral and Psychological Symptoms of Dementia

Although codes exist for dementia without and with behavioral disturbances, there is a need for additional detail on other key associated disorders, particularly psychotic disorders, mood disorders, and anxiety. Moreover, within behavioral disorders, there is a need to distinctly identify agitation. Associated disorders in dementia are variously referred to as behavioral and psychological symptoms of dementia (BPSD), noncognitive behavioral changes (NCBC), [5] and neuropsychiatric symptoms (NPS). These are broader than the current coding structure. BPSD can generally be grouped into three main categories: behavioral disturbances, psychotic disorders, and mood (affective) disorders.[6]

Anxiety is also a common BPSD. However, while some literature includes anxiety together with affective disorders, ICD-10-CM classifies anxiety separately. Patients may have predominantly one type of BPSD or they may have more than one. Reflecting the need to align management strategies with the type of BPSD displayed by the patient, consensus diagnostic criteria have developed over time for the main types. More recently, the International Psychogeriatric Association has developed and validated a provisional consensus definition of agitation in dementia, as distinct from how this disorder may present in other populations.[7]

The key associated disorders represent significant clinical problems in their own right and are actually responsible for driving the care provided to dementia patients. The associated disorders are what typically bring patients to the attention of clinicians. For example, agitation is the alarming factor which prompts visits to the Emergency Department. Importantly, to date, dementia itself is not directly treatable. What is being treated is actually the associated disorder. For example, dementia with delusions and hallucinations may result in psychosocial interventions or, failing that, treatment with anti-psychotic medication.

The presence of the key associated disorders also links to patient outcomes, impacting quality of life, cost of care, institutionalization, and accelerated mortality.[5]Agitation is generally considered the most disruptive of the behavioral disturbances because it is associated with increased rates of institutionalization.[7] Some studies have identified a correlation between psychoses and acceleration of cognitive decline and increased mortality,[8] as well as a correlation between mood disorders and lowered Quality of Life scores.[9]

Specific codes for the associated disorders will also support the National Partnership to Improve Dementia Care, a CMS priority to balance the use of pharmacologic approaches and to enhance patient-centered dementia care practices.

At some point, most patients with dementia are afflicted with some form of BPSD. There is an urgent need for the clinical data to identify the stages at which these disorders develop and how they present. This will help to enable recognition of the appropriate management strategies for interventions as well as development of new non-pharmacological and pharmacological approaches to improve the adverse outcomes.

The National Minority Quality Forum is the submitter for this proposal. The proposal was developed in collaboration with, and has the support of, the following clinical and scientific collaborators: Amita Patel, MD, CMD, MHA, CPE; David S. Geldmacher, MD, FANA, FACP; and Maureen Nash, MD, MS, FAPA, FACP. Additional advisors include Istvan Boksay, MD, PhD; Meenakshi Patel, MD, FACP, MMM, CMD; Sandra Swantek, MD, FAPA; and Ajanta S. Vinekar, MD, FAPA. The American Association for Geriatric Psychiatry has reviewed and supports this proposal.

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TABULAR MODIFICATIONS

A81 Atypical virus infections of central nervous system

Revise — Use additional code, if applicable, to identify:

Revise — dementia with behavioral disturbance (~~F02.81~~F02.11-, F02.21-, F02.31-, F02.81-)

Revise — dementia without behavioral disturbance (F02.10, F02.20, F02.30, F02.80)

Add — dementia with psychotic disorder (F02.12, F02.22, F02.32, F02.82)

Add — dementia with mood disorder (F02.13, F02.23, F02.33, F02.83)

F01 Vascular Dementia

New subcategory — F01.1 Vascular dementia, mild

New code — F01.10 Vascular dementia, mild, without behavioral disturbance, psychotic disorder, and mood disorder

Add — Major neurocognitive disorder due to vascular disease, mild, NOS

Add — Vascular dementia, mild, NOS

New sub-subcategory — F01.11 Vascular dementia, mild, with behavioral disturbance

Add — Use additional code, if applicable, to identify wandering in vascular dementia (Z91.83)

New code — F01.111 Vascular dementia, mild, with agitation

Add — Major neurocognitive disorder due to vascular disease, mild, with aberrant motor behavior such as rocking, pacing, restlessness, or exit-seeking

Add — Major neurocognitive disorder due to vascular disease, mild, with (physical, verbal) aggressive, combative, or violent behavior

Add — Vascular dementia, mild, with aberrant motor behavior such as rocking, pacing, restlessness, or exit-seeking

Add		Vascular dementia, mild, with (physical, verbal) aggressive, combative, or violent behavior
New code	F01.118	Vascular dementia, mild, with other behavioral disturbance
Add		Major neurocognitive disorder due to vascular disease, mild, with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition
Add		Vascular dementia, mild, with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition
New code	F01.12	Vascular dementia, mild, with psychotic disorder
Add		Major neurocognitive disorder due to vascular disease, mild, with psychotic disorder such as hallucinations, paranoia, suspiciousness, or delusional state
Add		Vascular dementia, mild, with psychotic disorder such as hallucinations, paranoia, suspiciousness, or delusional state
New code	F01.13	Vascular dementia, mild, with mood disorder
Add		Major neurocognitive disorder due to vascular disease, mild, with mood disorder such as depression, apathy, or anhedonia
Add		Vascular dementia, mild, with mood disorder such as depression, apathy, or anhedonia
New code	F01.14	Vascular dementia, mild, with anxiety
Add		Major neurocognitive disorder due to vascular disease, mild, with anxiety
New subcategory	F01.2	Vascular dementia, moderate
New code	F01.20	Vascular dementia, moderate, without behavioral disturbance, psychotic disorder, and mood disorder
Add		Major neurocognitive disorder due to vascular disease, moderate, NOS
Add		Vascular dementia, moderate, NOS
New sub-subcategory	F01.21	Vascular dementia, moderate, with behavioral disturbance
Add		Use additional code, if applicable, to identify wandering in vascular dementia (Z91.83)
New code	F01.211	Vascular dementia, moderate, with agitation
Add		Major neurocognitive disorder due to vascular disease, moderate, with aberrant motor behavior such as rocking, pacing, restlessness, or exit-seeking
Add		Major neurocognitive disorder due to vascular disease, moderate, with (physical, verbal) aggressive, combative, or violent behavior
Add		Vascular dementia, moderate, with aberrant motor behavior such as rocking, pacing, restlessness, or exit-seeking

Add — Vascular dementia, moderate, with (physical, verbal) aggressive, combative, or violent behavior

New code — F01.218 Vascular dementia, moderate, with other behavioral disturbance
Add — Major neurocognitive disorder due to vascular disease, moderate, with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition
Add — Vascular dementia, moderate, with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition

New code — F01.22 Vascular dementia, moderate, with psychotic disorder
Add — Major neurocognitive disorder due to vascular disease, moderate, with psychotic disorder such as hallucinations, paranoia, suspiciousness, or delusional state
Add — Vascular dementia, moderate, with psychotic disorder such as hallucinations, paranoia, suspiciousness, or delusional state

New code — F01.23 Vascular dementia, moderate, with mood disorder
Add — Major neurocognitive disorder due to vascular disease, moderate, with mood disorder such as depression, apathy, or anhedonia
Add — Vascular dementia, moderate, with mood disorder such as depression, apathy, or anhedonia

New code — F01.24 Vascular dementia, moderate, with anxiety
Add — Major neurocognitive disorder due to vascular disease, moderate, with anxiety

New subcategory — F01.3 Vascular dementia, severe
New code — F01.30 Vascular dementia, severe, without behavioral disturbance, psychotic disorder, and mood disorder
Add — Major neurocognitive disorder due to vascular disease, severe, NOS
Add — Vascular dementia, severe, NOS

New sub-subcategory — F01.31 Vascular dementia, severe, with behavioral disturbance
Add — Use additional code, if applicable, to identify wandering in vascular dementia (Z91.83)

New code	F01.311	Vascular dementia, severe, with agitation
Add		Major neurocognitive disorder due to vascular disease, severe, with aberrant motor behavior such as rocking, pacing, restlessness, or exit-seeking
Add		Major neurocognitive disorder due to vascular disease, severe, with (physical, verbal) aggressive, combative, or violent behavior
Add		Vascular dementia, severe, with aberrant motor behavior such as rocking, pacing, restlessness, or exit-seeking
Add		Vascular dementia, severe, with (physical, verbal) aggressive, combative, or violent behavior
New code	F01.318	Vascular dementia, severe, with other behavioral disturbance
Add		Major neurocognitive disorder due to vascular disease, severe, with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition
Add		Vascular dementia, severe, with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition
New code	F01.32	Vascular dementia, severe, with psychotic disorder
Add		Major neurocognitive disorder due to vascular disease, severe, with psychotic disorder such as hallucinations, paranoia, suspiciousness, or delusional state
Add		Vascular dementia, severe, with psychotic disorder such as hallucinations, paranoia, suspiciousness, or delusional state
New code	F01.33	Vascular dementia, severe, with mood disorder
Add		Major neurocognitive disorder due to vascular disease, severe, with mood disorder such as depression, apathy, or anhedonia
Add		Vascular dementia, severe, with mood disorder such as depression, apathy, or anhedonia
New code	F01.34	Vascular dementia, severe, with anxiety
Add		Major neurocognitive disorder due to vascular disease, severe, with anxiety
Revise	F01.5	Vascular dementia, <u>unspecified stage</u>
Revise	F01.50	Vascular dementia<u>, unspecified stage,</u> without behavioral disturbance, <u>psychotic disorder, and mood disorder</u>
Revise		Major neurocognitive disorder <u>due to vascular disease</u> ~~without behavioral disturbance~~ <u>NOS</u>
Add		<u>Vascular dementia NOS</u>

Revise	F01.51	Vascular dementia, unspecified stage, with behavioral disturbance
Delete		~~Major neurocognitive disorder due to vascular disease, with behavioral disturbance~~
Delete		~~Major neurocognitive disorder with aggressive behavior~~
Delete		~~Major neurocognitive disorder with combative behavior~~
Delete		~~Major neurocognitive disorder with violent behavior~~
Delete		~~Vascular dementia with aggressive behavior~~
Delete		~~Vascular dementia with combative behavior~~
Delete		~~Vascular dementia with violent behavior~~
New code	F01.511	Vascular dementia, unspecified stage, with agitation
Add		Major neurocognitive disorder due to vascular disease, unspecified stage, with aberrant motor behavior such as rocking, pacing, restlessness, or exit-seeking
Add		Major neurocognitive disorder due to vascular disease, unspecified stage, with (physical, verbal) aggressive, combative, or violent behavior
Add		Vascular dementia, unspecified stage, with aberrant motor behavior such as rocking, pacing, restlessness, or exit-seeking
Add		Vascular dementia, unspecified stage, with (physical, verbal) aggressive, combative, or violent behavior
New code	F01.518	Vascular dementia, unspecified stage, with other behavioral disturbance
Add		Major neurocognitive disorder due to vascular disease, unspecified stage, with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition
Add		Vascular dementia, unspecified stage, with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition
New code	F01.52	Vascular dementia, unspecified stage, with psychotic disorder
Add		Major neurocognitive disorder due to vascular disease, unspecified stage, with psychotic disorder such as hallucinations, paranoia, suspiciousness, or delusional state
Add		Vascular dementia, unspecified stage, with psychotic disorder such as hallucinations, paranoia, suspiciousness, or delusional state

New code F01.53 Vascular dementia, unspecified stage, with mood disorder
Add Major neurocognitive disorder due to vascular disease, unspecified stage, with mood disorder such as depression, apathy, or anhedonia
Add Vascular dementia, unspecified stage, with mood disorder such as depression, apathy, or anhedonia
New code F01.54 Vascular dementia, unspecified stage, with anxiety
Add Major neurocognitive disorder due to vascular disease, unspecified stage, with anxiety

F02 Dementia in other diseases classified elsewhere
Add **Excludes1: mild neurocognitive disorder due to known physiological condition with or without behavioral disturbance (F06.7-)**
New subcategory F02.1 Dementia in other diseases classified elsewhere, mild
New code F02.10 Dementia in other diseases classified elsewhere, mild, without behavioral disturbance, psychotic disorder, and mood disorder
Add Dementia in other diseases classified elsewhere, mild, NOS
Add Major neurocognitive disorder in other diseases classified elsewhere, mild, NOS

New sub-subcategory F02.11 Dementia in other diseases classified elsewhere, mild, with behavioral disturbance
Add Use additional code, if applicable, to identify wandering in dementia in conditions classified elsewhere (Z91.83)
New code F02.111Dementia in other diseases classified elsewhere, mild, with agitation
Add Dementia in other diseases classified elsewhere, mild, with aberrant motor behavior such as rocking, pacing, restlessness, or exit-seeking
Add Dementia in other diseases classified elsewhere, mild, with (physical, verbal) aggressive, combative, or violent behavior
Add Major neurocognitive disorder in other diseases classified elsewhere, mild, with aberrant motor behavior such as rocking, pacing, restlessness, or exit-seeking
Add Major neurocognitive disorder in other diseases classified elsewhere, mild, with (physical, verbal) aggressive, combative, or violent behavior

New code F02.118 Dementia in other diseases classified elsewhere, mild, with other behavioral disturbance
Add Dementia in other diseases classified elsewhere, mild, with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition
Add Major neurocognitive disorder in other diseases classified elsewhere, mild, with behavioral disturbances such as

sleep disturbance, social disinhibition, or sexual disinhibition

New code F02.12 Dementia in other diseases classified elsewhere, mild, with psychotic disorder
Add Dementia in other diseases classified elsewhere, mild, with psychotic disorder such as hallucinations, paranoia, suspiciousness, or delusional state
Add Major neurocognitive disorder in other diseases classified elsewhere, mild, with psychotic disorder such as hallucinations, paranoia, suspiciousness, or delusional state

New code F02.13 Dementia in other diseases classified elsewhere, mild, with mood disorder
Add Dementia in other diseases classified elsewhere, mild, with mood disorder such as depression, apathy, or anhedonia
Add Major neurocognitive disorder in other diseases classified elsewhere, mild, with mood disorder such as depression, apathy, or anhedonia

New code F02.14 Dementia in other diseases classified elsewhere, mild, with anxiety
Add Major neurocognitive disorder in other diseases classified elsewhere, mild, with anxiety

New subcategory F02.2 Dementia in other diseases classified elsewhere, moderate

New code F02.20 Dementia in other diseases classified elsewhere, moderate, without behavioral disturbance, psychotic disorder, and mood disorder
Add Dementia in other diseases classified elsewhere, moderate, NOS
Add Major neurocognitive disorder in other diseases classified elsewhere, moderate, NOS

New sub-category F02.21 Dementia in other diseases classified elsewhere, moderate, with behavioral disturbance
Add Use additional code, if applicable, to identify wandering in dementia in conditions classified elsewhere (Z91.83)
New code F02.211 Dementia in other diseases classified elsewhere, moderate, with agitation
Add Dementia in other diseases classified elsewhere, moderate, with aberrant motor behavior such as rocking, pacing, restlessness, or exit-seeking
Add Dementia in other diseases classified elsewhere, moderate, with (physical, verbal) aggressive, combative, or violent behavior
Add Major neurocognitive disorder in other diseases classified elsewhere, moderate, with aberrant motor behavior such as rocking, pacing, restlessness, or exit-seeking

Add		Major neurocognitive disorder in other diseases classified elsewhere, moderate, with (physical, verbal) aggressive, combative, or violent behavior
New code	F02.218	Dementia in other diseases classified elsewhere, moderate, with other behavioral disturbance
Add		Dementia in other diseases classified elsewhere, moderate, with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition
Add		Major neurocognitive disorder in other diseases classified elsewhere, moderate, with behavioral disturbance such as sleep disturbance, social disinhibition, or sexual disinhibition
New code	F02.22	Dementia in other diseases classified elsewhere, moderate, with psychotic disorder
Add		Dementia in other diseases classified elsewhere, moderate, with psychotic disorder such as hallucinations, paranoia, suspiciousness, or delusional state
Add		Major neurocognitive disorder in other diseases classified elsewhere, moderate, with psychotic disorder such as hallucinations, paranoia, suspiciousness, or delusional state
New code	F02.23	Dementia in other diseases classified elsewhere, moderate, with mood disorder
Add		Dementia in other diseases classified elsewhere, moderate, with mood disorder such as depression, apathy, or anhedonia
Add		Major neurocognitive disorder in other diseases classified elsewhere, moderate, with mood disorder such as depression, apathy, or anhedonia
New code	F02.24	Dementia in other diseases classified elsewhere, moderate, with anxiety
Add		Major neurocognitive disorder in other diseases classified elsewhere, moderate, with anxiety
New subcategory	F02.3	Dementia in other diseases classified elsewhere, severe
New code	F02.30	Dementia in other diseases classified elsewhere, severe, without behavioral disturbance, psychotic disorder, and mood disorder
Add		Dementia in other diseases classified elsewhere, severe, NOS
Add		Major neurocognitive disorder in other diseases classified elsewhere, severe, NOS
New sub-subcategory	F02.31	Dementia in other diseases classified elsewhere, severe, with behavioral disturbance
Add		Use additional code, if applicable, to identify wandering in dementia in conditions classified elsewhere (Z91.83)

New code	F02.311	Dementia in other diseases classified elsewhere, severe, with agitation
Add		Dementia in other diseases classified elsewhere, severe, with aberrant motor behavior such as rocking, pacing, restlessness, or exit-seeking
Add		Dementia in other diseases classified elsewhere, severe, with (physical, verbal) aggressive, combative, or violent behavior
Add		Major neurocognitive disorder in other diseases classified elsewhere, severe, with aberrant motor behavior such as rocking, pacing, restlessness, or exit-seeking
Add		Major neurocognitive disorder in other diseases classified elsewhere, severe, with (physical, verbal) aggressive, combative, or violent behavior
New code	F02.318	Dementia in other diseases classified elsewhere, severe, with other behavioral disturbance
Add		Dementia in other diseases classified elsewhere, severe, with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition
Add		Major neurocognitive disorder in other diseases classified elsewhere, severe, with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition
New code	F02.32	Dementia in other diseases classified elsewhere, severe, with psychotic disorder
Add		Dementia in other diseases classified elsewhere, severe, with psychotic disorder such as hallucinations, paranoia, suspiciousness, or delusional state
Add		Major neurocognitive disorder in other diseases classified elsewhere, severe, with psychotic disorder such as hallucinations, paranoia, suspiciousness, or delusional state
New code	F02.33	Dementia in other diseases classified elsewhere, severe, with mood disorder
Add		Dementia in other diseases classified elsewhere, severe, with mood disorder such as depression, apathy, or anhedonia
Add		Major neurocognitive disorder in other diseases classified elsewhere, severe, with mood disorder such as depression, apathy, or anhedonia
New code	F02.34	Dementia in other diseases classified elsewhere, severe, with anxiety
Add		Major neurocognitive disorder in other diseases classified elsewhere, severe, with anxiety
Revise	F02.8	Dementia in other diseases classified elsewhere, <u>unspecified stage</u>
Revise	F02.80	Dementia in other diseases classified elsewhere<u>, unspecified stage,</u> without behavioral disturbance, <u>psychotic disorder, and mood disorder</u>
		Dementia in other diseases classified elsewhere NOS
Revise		Major neurocognitive disorder in other diseases classified elsewhere <u>NOS</u>
Revise	F02.81	Dementia in other diseases classified elsewhere<u>, unspecified stage,</u> with behavioral disturbance
Delete		~~Dementia in other diseases classified elsewhere with aggressive behavior~~
Delete		~~Dementia in other diseases classified elsewhere with combative behavior~~

Delete		~~Dementia in other diseases classified elsewhere with violent behavior~~
Delete		~~Major neurocognitive disorder in other diseases classified elsewhere with aggressive behavior~~
Delete		~~Major neurocognitive disorder in other diseases classified elsewhere with combative behavior~~
Delete		~~Major neurocognitive disorder in other diseases classified elsewhere with violent behavior~~
New code	F02.811	Dementia in other diseases classified elsewhere, unspecified stage, with agitation
Add		Dementia in other diseases classified elsewhere, unspecified stage, with aberrant motor behavior such as rocking, pacing, restlessness, or exit-seeking
Add		Dementia in other diseases classified elsewhere, unspecified stage, with (physical, verbal) aggressive, combative, or violent behavior
Add		Major neurocognitive disorder in other diseases classified elsewhere, unspecified stage, with aberrant motor behavior such as rocking, pacing, restlessness, or exit-seeking
Add		Major neurocognitive disorder in other diseases classified elsewhere, unspecified stage, with (physical, verbal) aggressive, combative, or violent behavior
New code	F02.818	Dementia in other diseases classified elsewhere, unspecified stage, with other behavioral disturbance
Add		Dementia in other diseases classified elsewhere with sleep disturbance, social disinhibition, or sexual disinhibition
Add		Major neurocognitive disorder in other diseases classified elsewhere with sleep disturbance, social disinhibition, or sexual disinhibition
New code	F02.82	Dementia in other diseases classified elsewhere, unspecified stage, with psychotic disorder
Add		Dementia in other diseases classified elsewhere, unspecified stage, with psychotic disorder such as hallucinations, paranoia, suspiciousness, or delusional state
Add		Major neurocognitive disorder in other diseases classified elsewhere, unspecified, with psychotic disorder such as hallucinations, paranoia, suspiciousness, or delusional state
New code	F02.83	Dementia in other diseases classified elsewhere, unspecified stage, with mood disorder
Add		Dementia in other diseases classified elsewhere, unspecified stage, with mood disorder such as depression, apathy, or anhedonia
Add		Major neurocognitive disorder in other diseases classified elsewhere unspecified stage, with mood disorder such as with depression, apathy, or anhedonia

New code F02.84 Dementia in other diseases classified elsewhere, unspecified stage, with anxiety
Add Major neurocognitive disorder in other diseases classified elsewhere unspecified stage, with anxiety

F03 Unspecified Dementia
New subcategory F03.1 Unspecified dementia, mild
New code F03.10 Unspecified dementia, mild, without behavioral disturbance, psychotic disorder, and mood disorder
Add Dementia, mild, NOS
New sub-subcategory F03.11 Unspecified dementia, mild, with behavioral disturbance
Add Use additional code, if applicable, to identify wandering in unspecified dementia (Z91.83)
New code F03.111 Unspecified dementia, mild, with agitation
Add Unspecified dementia, mild, with aberrant motor behavior such as rocking, pacing, restlessness, or exit-seeking
Add Unspecified dementia, mild, with (physical, verbal) aggressive, combative, or violent behavior
New code F03.118 Unspecified dementia, mild, with other behavioral disturbance
Add Unspecified dementia, mild, with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition
New code F03.12 Unspecified dementia, mild, with psychotic disorder
Add Unspecified dementia, mild, with psychotic disorder such as hallucinations, paranoia, suspiciousness, or delusional state
New code F03.13 Unspecified dementia, mild, with mood disorder
Unspecified dementia, mild, with mood disorder such as depression, apathy, or anhedonia
New code F03.14 Unspecified dementia, mild, with anxiety

New subcategory F03.2 Unspecified dementia, moderate
New code F03.20 Unspecified dementia, moderate, without behavioral disturbance, psychotic disorder, and mood disorder
Unspecified dementia, moderate, NOS
New sub-subcategory F03.21 Unspecified dementia, moderate, with behavioral disturbance
New code F03.211 Unspecified dementia, moderate, with agitation
Add Unspecified dementia, moderate, with aberrant motor behavior such as rocking, pacing, restlessness, or exit-seeking
Add Unspecified dementia, moderate, with (physical, verbal) aggressive, combative, or violent behavior

New code F03.218 Unspecified dementia, moderate, with other behavioral disturbance
Add Unspecified dementia, moderate, with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition

New code F03.22 Unspecified dementia, moderate, with psychotic disorder
Add Unspecified dementia, moderate, with psychotic disorder such as hallucinations, paranoia, suspiciousness, or delusional state
New code F03.23 Unspecified dementia, moderate, with mood disorder
Add Unspecified dementia, moderate, with mood disorder such as depression, apathy, or anhedonia

New code F03.24 Unspecified dementia, moderate, with anxiety

New subcategory F03.3 Unspecified dementia, severe
New code F03.30 Unspecified dementia, severe, without behavioral disturbance, psychotic disorder, and mood disorder
Add Unspecified dementia, severe, NOS
New sub-subcategory F03.31 Unspecified dementia, severe, with behavioral disturbance
Add Use additional code, if applicable, to identify wandering in unspecified dementia (Z91.83)
New code F03.311 Unspecified dementia, severe, with agitation
Add Unspecified dementia, severe, with aberrant motor behavior such as rocking, pacing, restlessness, or exit-seeking
Add Unspecified dementia, severe, with (physical, verbal) aggressive, combative, or violent behavior
New code F03.318 Unspecified dementia, severe, with other behavioral disturbance
Add Unspecified dementia, severe, with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition

New code F03.32 Unspecified dementia, severe, with psychotic disorder
Add Unspecified dementia, severe, with psychotic disorder such as hallucinations, paranoia, suspiciousness, or delusional state
New code F03.33 Unspecified dementia, severe, with mood disorder
Add Unspecified dementia, severe, with mood disorder such as depression, apathy, or anhedonia
New code F03.34 Unspecified dementia, severe, with anxiety

Revise F03.9 Unspecified dementia, unspecified stage
Revise F03.90 Unspecified dementia, unspecified stage, without behavioral disturbance, psychotic disorder, and mood disorder
Revise F03.91 Unspecified dementia, unspecified stage, with behavioral disturbance

Delete ~~Unspecified dementia with aggressive behavior~~
Delete ~~Unspecified dementia with combative behavior~~
Delete ~~Unspecified dementia with violent behavior~~

New code F03.911 Unspecified dementia, unspecified stage, with agitation
Add Unspecified dementia, unspecified stage, with aberrant motor behavior such as rocking, pacing, restlessness, or exit-seeking
Add Unspecified dementia, unspecified stage, with (physical, verbal) aggressive, combative, or violent behavior
New code F03.918 Unspecified dementia, unspecified stage, with other behavioral disturbance
Add Unspecified dementia, unspecified stage, with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition

New code F03.92 Unspecified dementia, unspecified stage, with psychotic disorder
Add Unspecified dementia, unspecified stage, with psychotic disorder such as hallucinations, paranoia, suspiciousness, or delusional state
New code F03.93 Unspecified dementia, unspecified stage, with mood disorder
Add Unspecified dementia, unspecified stage, with mood disorder such as depression, apathy, or anhedonia
New code F03.94 Unspecified dementia, unspecified stage, with anxiety

G10 Huntington's disease
Huntington's chorea
Huntington's dementia
Delete ~~Code also dementia in other diseases classified elsewhere without behavioral disturbance (F02.80)~~
Revise Use additional code <u>if applicable</u> to identify:
Add dementia with behavioral disturbance (F02.11-, F02.21-, F02.31-, F02.81-)
Add dementia without behavioral disturbance (F02.10, F02.20, F02.30, F02.80)
Add dementia with psychotic disorder (F02.12, F02.22, F02.32, F02.82)
Add dementia with mood disorder (F02.13, F02.23, F02.33, F02.83)
Add mild cognitive disorder **due to known physiological condition** (F06.7-)

G20 Parkinson's disease
Revise Use additional code<u>, if applicable</u>, to identify:
Revise dementia with behavioral disturbance (~~F02.81~~<u>F02.11-, F02.21-, F02.31-, F02.81-</u>)
Revise dementia without behavioral disturbance (<u>F02.10, F02.20, F02.30,</u> F02.80)

Add dementia with psychotic disorder (F02.12, F02.22, F02.32, F02.82)
Add dementia with mood disorder (F02.13, F02.23, F02.33, F02.83)
Add mild cognitive disorder **due to known physiological condition** (F06.7-)

G30 Alzheimer's Disease
Revise Use additional code, if applicable, to identify:
delirium (F05)
Revise dementia with behavioral disturbance (~~F02.81~~F02.11-, F02.21-, F02.31-, F02.81-)
Revise dementia without behavioral disturbance (F02.10, F02.20, F02.30, F02.80)
Add dementia with psychotic disorder (F02.12, F02.22, F02.32, F02.82)
Add dementia with mood disorder (F02.13, F02.23, F02.33, F02.83)
Add mild cognitive disorder **due to known physiological condition** (F06.7-)

G31 Other degenerative diseases of nervous system, not elsewhere classified
Revise For codes G31.0-G31.83, G31.85-G31.9, use additional code, if applicable, to identify:
Revise dementia with behavioral disturbance (~~F02.81,~~F02.11-, F02.21-, F02.31-, F02.81-)
Revise dementia without behavioral disturbance (F02.10, F02.20, F02.30, F02.80)
Add dementia with psychotic disorder (F02.12, F02.22, F02.32, F02.82)
Add dementia with mood disorder (F02.13, F02.23, F02.33, F02.83)
Add mild cognitive disorder **due to known physiological condition** (F06.7-)

Encounter for Pediatric-to-Adult Transition Counseling

The American Academy of Pediatrics (AAP) is submitting the following proposal on behalf of *Got Transition (https://gottransition.org/about-us/*. Got Transition® is a national resource center on health care transition (HCT). Its aim is to improve the transition from pediatric to adult health care through the use of evidence-driven strategies for clinicians and other health care professionals; public health programs; payers and plans; youth and young adults; and parents and caregivers. Got Transition is a program of The National Alliance to Advance Adolescent Health and has a cooperative agreement from the federal Maternal and Child Health Bureau, Health Resources and Services Administration.

It is being requested to update to the ICD-10-CM code set to include a new code to capture encounters for pediatric-to-adult transition counseling. Current clinical recommendations jointly developed by the American Academy of Pediatrics, American Academy of Family Physicians, and American College of Physicians call for transitional counseling as part of routine primary and specialty care for youth and young adults with and without special health care needs between the ages of 14 and 25[1].

Currently, there is no way to monitor whether this reason for an encounter was accomplished, and more and more health systems are seeking to track provision of recommended pediatric-to-adult transition counseling in ambulatory care settings.

An encounter for transition counseling can involve counseling related to self-care skill attainment, building health literacy, education about one's own medical summary and emergency care plan, transfer planning, and guided integration into adult care.

Systematic reviews of transition evaluation studies have shown that when a structured transition process is in place, which includes transition counseling – improvements result in adherence to care, self-care skill-building, functional status, patient satisfaction, and ambulatory care use.[2,3,4]

References:

[1] White PH, Cooley WC, American Academy of Pediatrics, American Academy of Family Physicians. Supporting the health care transition from adolescence to adulthood in the medical home. Pediatrics. 2018;142(5):e20182587.

[2] Schmidt A, Ilango SM, McManus MA, Rogers KK, White PH. Outcomes of pediatric to adult health care transition interventions: An updated systematic review. Journal of Pediatric Nursing. 2020;51:92-107.

[3] Gabriel P, McManus M, Rogers K, White P. Outcome evidence for structured pediatric to adult health care transition interventions: a systematic review. The Journal of Pediatrics. 2017;188:263-269.

[4] Prior M, McManus M, White P, Davidson L. Measuring the "triple aim" in transition care: a systematic review. Pediatrics. 2014;134(6):e1648-61.

TABULAR MODIFICATIONS

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

Excludes2:contraceptive or procreation counseling (Z30-Z31)

sex counseling (Z70.-)

Z71.8 Other specified counseling

Excludes2:counseling for contraception (Z30.0-)

New code	Z71.87 Encounter for pediatric-to-adult transition counseling
Add	Code also chronic condition, if applicable, such as:
Add	autism spectrum disorder (F84.0)
Add	congenital malformations of the circulatory system (Q20-Q28)
Add	cystic fibrosis (E84-)
Add	sickle-cell disorder (D57-)

Encounter for PPD Test Reading and Medication Review

Health care encounters for PPD/Mantoux test reading and medication review are very common but there are no ICD-10-CM codes to identify these visits. Purified protein derivative (PPD) and Mantoux tuberculin skin test (TST) are commonly used skin tests to detect Tuberculosis. Millions of TST are ordered and administered every year. Positive PPD means infection with bacteria (mycobacterium tuberculosis) that causes the disease.

Medication review is an evaluation of a patient's medicine with the purpose of improving medication use and improving health outcomes. This involves examining drug-related issues and recommendations.

The Division of Healthcare Statistics of the National Center for Health Statistics, the federal agency that runs the National Ambulatory Medical Care Survey, is requesting the following new codes for these types of encounters.

TABULAR MODIFICATIONS

	Z11	Encounter for screening for infectious and parasitic diseases
New subcategory	Z11.1	Encounter for screening for respiratory tuberculosis
New code	Z11.11	Encounter for administration of screening test for respiratory tuberculosis
New code	Z11.12	Encounter for reading of screening test for respiratory tuberculosis
Add		Encounter for Mantoux reading
Add		Encounter for PPD reading
	Z71	Persons encountering health services for other counseling and medical advice, not elsewhere classified
	Z71.8	Other specified counseling
New code	Z71.86	Encounter for counseling related to medication management
Add		Encounter for medication advice NOS
Add		Encounter for medication review
Add		Excludes2: Encounter for therapeutic drug level monitoring (Z51.81)

Z76 Persons encountering health services in other circumstances

Z76.0 Encounter for issue of repeat prescription

Add Excludes2: Encounter for counseling related to medication management (Z71.86)
Encounter for therapeutic drug level monitoring (Z51.81)

Endometriosis

The American College of Obstetricians and Gynecologists (ACOG) and American Association of Gynecologic Laparoscopists (AAGL) are requesting expansion of the N80 code sections for endometriosis. Endometriosis is an often painful disorder in which tissue similar to the tissue that normally lines the inside of the uterus, the endometrium, grows outside the uterus. Endometriosis most commonly involves the ovaries, fallopian tubes and the tissue lining the pelvis. The primary symptom of endometriosis is pelvic pain, often associated with menstrual periods. Although many experience cramping during their menstrual periods, those with endometriosis typically describe menstrual pain that's far worse than usual. Pain usually increases over time.
This was previously presented at the September 2020 Coordination and Maintenance meeting and is being represented with changes suggested by comments.

The description of superficial and deep:

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

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

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

TABULAR MODIFICATIONS

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

New code	N80.01 Superficial endometriosis of the uterus
Add	Superficial endometriosis of the cervix
New code	N80.02 Deep endometriosis of the uterus
Add	Deep endometriosis of the cervix
Add	Deep retrocervical endometriosis
New code	N80.03 Adenomyosis of the uterus
Add	Adenomyosis NOS

N80.1 Endometriosis of ovary

New code	N80.10 Endometriosis of ovary, unspecified
Add	Endometriosis of ovary NOS
New sub-subcategory	N80.11 Superficial endometriosis of the ovary
New code	N80.111 Superficial endometriosis of right ovary
New code	N80.112 Superficial endometriosis of left ovary
New code	N80.113 Superficial endometriosis of bilateral ovaries
New code	N80.119 Superficial endometriosis of ovary, unspecified side
New sub-subcategory	N80.12 Deep endometriosis of ovary
Add	Deep ovarian endometriosis
Add	Endometrioma
New code	N80.121 Deep endometriosis of right ovary
New code	N80.122 Deep endometriosis of left ovary
New code	N80.123 Deep endometriosis of bilateral ovaries
New code	N80.129 Deep endometriosis of ovary, unspecified side

N80.2 Endometriosis of fallopian tube

New code	N80.20 Endometriosis of fallopian tube, unspecified
New sub-subcategory	N80.21 Superficial endometriosis of fallopian tube
New code	N80.211 Superficial endometriosis of right fallopian tube
New code	N80.212 Superficial endometriosis of left fallopian tube
New code	N80.213 Superficial endometriosis of bilateral fallopian tubes
New code	N80.219 Superficial endometriosis of unspecified fallopian tube

New sub-subcategory	N80.22 Deep endometriosis of the fallopian tube
Add	Deep endometriosis involving muscular wall of fallopian tube
New code	N80.221 Deep endometriosis of right fallopian tube
New code	N80.222 Deep endometriosis of left fallopian tube
New code	N80.223 Deep endometriosis of bilateral fallopian tubes
New code	N80.229 Deep endometriosis of unspecified fallopian tube
	N80.3 Endometriosis of pelvic peritoneum
Add	Endometriosis of the retroperitoneum
New code	N80.30 Endometriosis of pelvic peritoneum, unspecified
Add	Endometriosis of pelvic peritoneum, NOS
Add	Endometriosis of the retroperitoneum, NOS
New sub-subcategory	N80.31 Endometriosis of the anterior cul-de-sac
New code	N80.311 Endometriosis of the anterior cul-de-sac
Add	Endometriosis of the anterior cul-de-sac, NOS
New code	N80.312 Superficial endometriosis of the anterior cul-de-sac
New code	N80.313 Deep endometriosis of the anterior cul-de-sac
New sub-subcategory	N80.32 Endometriosis of the posterior cul-de-sac
New code	N80.321 Endometriosis of the posterior cul-de-sac
Add	Endometriosis of the posterior cul-de-sac, NOS
New code	N80.322 Superficial endometriosis of the posterior cul-de-sac
New code	N80.323 Deep endometriosis of the posterior cul-de-sac
New sub-subcategory	N80.33 Superficial endometriosis of the pelvic sidewall
Add	Endometriosis of the pelvic sidewall, NOS
New code	N80.331 Superficial endometriosis of the right pelvic sidewall

New code	N80.332 Superficial endometriosis of the left pelvic sidewall
New code	N80.333 Superficial endometriosis of bilateral pelvic sidewall
New code	N80.339 Superficial endometriosis of pelvic sidewall, unspecified
New sub-subcategory	N80.34 Deep endometriosis of the pelvic sidewall
New code	N80.341 Deep endometriosis of the right pelvic sidewall
New code	N80.342 Deep endometriosis of the left pelvic sidewall
New code	N80.343 Deep endometriosis of the bilateral pelvic sidewall
New code	N80.349 Deep endometriosis of the pelvic sidewall, unspecified
New sub-subcategory	N80.35 Superficial endometriosis of the pelvic brim
Add	Endometriosis of the pelvic brim, NOS
New code	N80.351 Superficial endometriosis of the right pelvic brim
New code	N80.352 Superficial endometriosis of the left pelvic brim
New code	N80.353 Superficial endometriosis of bilateral pelvic brim
New code	N80.359 Superficial endometriosis of the pelvic brim, unspecified
New sub subcategory	N80.36 Deep endometriosis of the pelvic brim
New code	N80.361 Deep endometriosis of the right pelvic brim
New code	N80.362 Deep endometriosis of the left pelvic brim
New code	N80.363 Deep endometriosis of bilateral pelvic brim
New code	N80.369 Deep endometriosis of the pelvic brim, unspecified
New subcategory	N80.37 Superficial endometriosis of the uterosacral ligament(s)
Add	Endometriosis of the uterosacral ligament(s), NOS
New code	N80.371 Superficial endometriosis of the right uterosacral ligament

New code	N80.372 Superficial endometriosis of the left uterosacral ligament
New code	N80.373 Superficial endometriosis of the bilateral uterosacral ligaments
New code	N80.379 Superficial endometriosis of the uterosacral ligament(s), unspecified
New subcategory	N80.38 Deep endometriosis of the uterosacral ligament(s)
New code	N80.381 Deep endometriosis of the right uterosacral ligament
New code	N80.382 Deep endometriosis of the left uterosacral ligament
New code	N80.383 Deep endometriosis of bilateral uterosacral ligament(s)
New code	N80.389 Deep endometriosis of the uterosacral ligament(s), unspecified
New sub subcategory	N80.39 Endometriosis of other pelvic peritoneum
New code	N80.390 Superficial endometriosis of the pelvic peritoneum, other specified sites
New code	N80.391 Deep endometriosis of the pelvic peritoneum, other specified sites
New code	N80.399 Endometriosis of the pelvic peritoneum, other specified sites, unspecified depth
	N80.4 Endometriosis of rectovaginal septum and vagina
New code	N80.41Endometriosis of rectovaginal septum without involvement of vagina
New code	N80.42 Endometriosis of rectovaginal septum with invasion of vagina
	N80.5 Endometriosis of intestine
	N80.50 Endometriosis of intestine, unspecified
New sub-subcategory	N80.51Endometriosis of the rectum
New code	N80.511 Superficial endometriosis of the rectum
New code	N80.512 Deep endometriosis of the rectum
Add	Deep endometriosis of the rectum, multifocal

New sub-subcategory	N80.52 Endometriosis of the sigmoid colon
New code	N80.521 Superficial endometriosis of the sigmoid colon
Add	Endometriosis of the sigmoid colon, NOS
New code	N80.522 Deep endometriosis of the sigmoid colon

New sub-subcategory	N80.53 Endometriosis of the cecum
New code	N80.531 Superficial endometriosis of the cecum
Add	Endometriosis of the cecum, NOS
Add	N80.532 Deep endometriosis of the cecum

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

New sub-subcategory	N80.55 Endometriosis of the colon
New code	N80.551 Superficial endometriosis of the colon
Add	Endometriosis of the colon, NOS
Add	Superficial endometriosis of the colon, NOS
New code	N80.552 Deep endometriosis of the colon

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

New subcategory	N80.A Endometriosis of bladder and ureters
New code	N80.A1 Superficial endometriosis of bladder
New code	N80.A2 Deep endometriosis of bladder

New sub-subcategory	N80.A3 Superficial endometriosis of ureter
Add	Extrinsic endometriosis of ureter
Add	Code also obstructive and reflux uropathy (N13.-)
New code	N80.A31 Superficial endometriosis of right ureter
New code	N80.A32 Superficial endometriosis of left ureter
New code	N80.A33 Superficial endometriosis of bilateral ureters
New code	N80.A39 Superficial endometriosis of unspecified ureter

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

New subcategory	N80.B Endometriosis of cardiothoracic space
Add	Endometriosis of thorax
Add	Code also, if applicable: catamenial pneumothorax (J93.83) catamenial hemothorax (J94.2)
New code	N80.B1 Endometriosis of pleura
New code	N80.B2 Endometriosis of lung
	N80.B3 Endometriosis of diaphragm
New sub subcategory	N80.B31 Superficial endometriosis of diaphragm
Add	Endometriosis of the diaphragm, NOS
New code	N80.B32 Deep endometriosis of diaphragm
New code	N80.B4 Endometriosis of the pericardial space
New code	N80.B5 Endometriosis of the mediastinal space
Add	Endometriosis of the mediastinal space, NOS
New code	N80.B6 Endometriosis of cardiothoracic space

New subcategory	N80.C Endometriosis of the abdomen
New code	N80.C0 Endometriosis of the abdomen, unspecified
New sub subcategory	N80.C1 Endometriosis of the anterior abdominal wall
New code	N80.C10 Endometriosis of the anterior abdominal wall, subcutaneous tissue
New code	N80.C11Endometriosis of the anterior abdominal wall, fascia and muscular layers
New code	N80.C19 Endometriosis of the anterior abdominal wall, unspecified depth
Add	Endometriosis of the anterior abdominal wall, NOS
New code	N80.C2 Endometriosis of the umbilicus
New code	N80.C3 Endometriosis of the inguinal canal
New code	N80.C4 Endometriosis of extra-pelvic abdominal peritoneum
New subcategory	N80.D Endometriosis of the pelvic nerves
Add	Endometriosis of the nerves of the retroperitoneum
New code	N80.D0 Endometriosis of the pelvic nerves, unspecified
Add	Endometriosis of nerve of the retroperitoneum, NOS
New Code	N80.D1 Endometriosis of the sacral splanchnic nerves
Add	Endometriosis of the pelvic splanchnic nerves
New Code	N80.D2 Endometriosis of the sacral nerve roots
New Code	N80.D3 Endometriosis of the obturator nerve
New Code	N80.D4 Endometriosis of the sciatic nerve
New Code	N80.D5 Endometriosis of the pudendal nerve
New Code	N80.D6 Endometriosis of the femoral nerve
New Code	N80.D9 Endometriosis of other pelvic nerve
Add	Endometriosis of the other nerves of the retroperitoneum
	N80.8 Other endometriosis
Delete	~~Endometriosis of thorax~~

Fetal Anomalies

The Society for Maternal Fetal Medicine (SMFM) and the American College of Obstetricians and Gynecologists (ACOG) are requesting that the O35 code sections for fetal anomalies (e.g. Central Nervous System Anomalies (CNS), Chromosomal Anomalies, and Fetal Abnormalities and Damage), be expanded to provide additional specificity for appropriate diagnosis coding and to assist in measuring the incidence of these specific anomalies, which is valuable from a public health perspective. This proposal will enable better tracking, measurement, and ultimately improved treatment modalities for identified fetal anomalies.

A proposal was presented at the September 2019 Coordination and Maintenance meeting. In response to public comments this proposal is being represented. This proposal will enable better tracking, measurement, and ultimately improved treatment modalities for identified fetal anomalies.

The proposed data set will be used primarily by physicians with specialized training and skill in assessing fetal anomalies during pregnancy. These physicians currently document these conditions during patient assessments but have no method of capturing the data with any reasonable specificity using the current code set. The expanded code sets would be reported once the condition has been confirmed. With the proposed expanded prenatal codes, the corresponding postnatal diagnosis will help assess the quality of prenatal care and diagnosis, and the allocation of public health resources as appropriate.

In the Unites States, 3% of all babies are born with a birth defect, or about 120,000 every year. According to the MMWR, birth defects are the leading cause of infant deaths, accounting for 20% of all infant deaths. Worldwide, about 3.2 million babies are born yearly with a congenital anomaly. Some congenital anomalies can be prevented, such as with vaccination, adequate supplements (e.g. folic acid, iodine), and adequate antenatal care. While the etiology of many birth defects is not clear, some have a clear etiology such as obesity, diabetes, and drug intake as well as some by race.

The proposed new codes represent a specific code assigned for the most common fetal abnormalities, classified by organ system. Their specificity provides guidance in reviewing and abstracting medical records.

The advantage of the expanded code set is that the additional codes will provide specificity for fetal conditions during the antepartum. Specific antenatal codes for fetal anomalies currently do not exist although most fetal anomalies are diagnosed during the antepartum with reasonable specificity. This information is documented and available in the patient records with no correspondingly specific ICD-10-CM code. The abnormalities are later captured in the neonatal record with postnatal or pediatric ICD-10-CM codes. The absence of antenatal ICD-10-CM codes to reflect many of the same

diagnoses limits the ability to assess the quality of the antenatal diagnosis as well as the evaluation of the different treatment modalities proposed for some of these diagnoses.

The specificity will help in assessing the quality of care for the different abnormalities if specified with the expanded code set. Some abnormalities will still be recognized even if the patient terminates her pregnancy. Clusters of teratogenic risk due to environmental or other exposure would be elucidated. Data would also be more accurate as many patients are mobile, meaning they deliver in a place other than where they were pregnant. Importantly, care disparities will be more correctly recognized. From a planning and monitoring perspectives, public health resources can be better allocated, and care quality can be assessed by matching prenatal and postnatal diagnoses.

The facial abnormalities series of codes are important because they identify relatively common abnormalities, some of which may be genetic in origin while others may be associated with drug intake (e.g. antiseizure medications). The correct and most specific diagnosis codes help in referring these patients to specialized centers where facial surgery is undertaken. The antenatal diagnosis is important because some of these babies may need specialized mouth suckling devices that are critical soon after delivery and a specific antenatal diagnosis would be helpful to avoid delays in providing proper care and nutrition.

With regards to specific chromosomal abnormality codes, the top trisomies are associated with different rates of multi-organ abnormalities and are managed in different ways. They also have varying and different implications as to pregnancy management. Some associated abnormalities may be lethal, others not, and a more efficient way to identify the associated abnormalities across a larger population will be helpful in counseling patients based on data to be developed with the new codes.

TABULAR MODIFICATIONS

O35 Maternal care for known or suspected fetal abnormality and damage
Includes: the listed conditions in the fetus as a reason for hospitalization or other obstetric care to the mother, or for termination of pregnancy
Code also any associated maternal condition
Excludes1: encounter for suspected maternal and fetal conditions ruled out (Z03.7-)

One of the following 7th characters is to be assigned to each code under category O35. 7th character 0 is for single gestations or multiple gestations where the fetus is unspecified. 7th characters 1 through 9 are for cases of multiple gestations to identify the fetus for which the code applies. The appropriate code from category O30, Multiple gestation, must also be assigned when assigning a code from category O35 that has a 7th character of 1 through 9.

0 not applicable or unspecified
1 fetus 1
2 fetus 2
3 fetus 3
4 fetus 4
5 fetus 5
9 other fetus

O35.0 Maternal care for (suspected) central nervous system malformation or damage in fetus

Delete ~~Maternal care for fetal anencephaly~~
Delete ~~Maternal care for fetal hydrocephalus~~
Delete ~~Maternal care for fetal spina bifida~~
Excludes2: chromosomal abnormality in fetus (O35.1)

New code O35.01 Maternal care for (suspected) central nervous system malformation or damage in fetus, agenesis of the corpus callosum

New code O35.02 Maternal care for (suspected) central nervous system malformation or damage in fetus, anencephaly

New code O35.03 Maternal care for (suspected) central nervous system malformation or damage in fetus, choroid plexus cysts

New code O35.04 Maternal care for (suspected) central nervous system malformation or damage in fetus, encephalocele

New code O35.05 Maternal care for (suspected) central nervous system malformation or damage in fetus, holoprosencephaly

New code O35.06 Maternal care for (suspected) central nervous system malformation or damage in fetus, hydrocephaly
Add Maternal care for fetal hydrocephalus

New code O35.07 Maternal care for (suspected) central nervous system malformation or damage in fetus, microcephaly

New code O35.08 Maternal care for (suspected) central nervous system malformation or damage in fetus, spina bifida

New code	O35.09 Maternal care for (suspected) other central nervous system malformation or damage in fetus
New subcategory	O35.1 Maternal care for (suspected) chromosomal abnormality in fetus
New code	O35.11 Maternal care for (suspected) chromosomal abnormality in fetus, Trisomy 13
New code	O35.12 Maternal care for (suspected) chromosomal abnormality in fetus, Trisomy 18
New code	O35.13 Maternal care for (suspected) chromosomal abnormality in fetus, Trisomy 21
New code	O35.14 Maternal care for (suspected) chromosomal abnormality in fetus, Turner Syndrome
New code	O35.15 Maternal care for (suspected) chromosomal abnormality in fetus, sex chromosome abnormality
New code	O35.19 Maternal care for (suspected) chromosomal abnormality in fetus, other chromosomal abnormality
New code	O35.A Maternal care for (suspected) fetal abnormality and damage, fetal facial anomalies
New code	O35.B Maternal care for other (suspected) fetal abnormality and damage, fetal cardiac anomalies
New code	O35.C Maternal care for other (suspected) fetal abnormality and damage, fetal pulmonary anomalies
New code	O35.D Maternal care for other (suspected) fetal abnormality and damage, fetal gastrointestinal anomalies
New code	O35.E Maternal care for other (suspected) fetal abnormality and damage, fetal genitourinary anomalies
New code	O35.F Maternal care for other (suspected) fetal abnormality and damage, fetal musculoskeletal anomalies

New code	O35.G Maternal care for other (suspected) fetal abnormality and damage, fetal upper extremities anomalies
New code	O35.H Maternal care for other (suspected) fetal abnormality and damage, fetal lower extremities anomalies

Flank Anatomical Specificity

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

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

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

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

TABULAR MODIFICATIONS

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

L02.21 Cutaneous abscess of trunk

New code L02.217 Cutaneous abscess of flank

L02.22 Furuncle of trunk
Boil of trunk
Folliculitis of trunk

New code L02.227 Furuncle of flank

L02.23 Carbuncle of trunk
New code L02.237 Carbuncle of flank

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

New subcategory R10.2 Pelvic and perineal pain
New code R10.20 Pelvic and perineal pain, unspecified
New code R10.21 Right pelvic pain
New code R10.22 Left pelvic pain
New code R10.23 Bilateral pelvic pain
New code R10.24 Perineal pain
New code R10.25 Suprapubic pain

New subcategory R10.4 Pain localized to lateral abdomen
Add Latus pain
New code R10.40 Flank pain, unspecified
New code R10.41 Right flank pain
New code R10.42 Left flank pain
New code R10.43 Bilateral flank pain

New subcategory S30.1 Contusion of abdominal wall and latus region
Delete ~~Contusion of flank~~
Delete ~~Contusion of groin~~
New code S30.10 Contusion of abdominal wall and latus region, unspecified
New code S30.11 Contusion of abdominal wall
New code S30.12 Contusion of flank

New code S30.13 Contusion of groin

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

S30.81 Abrasion of abdomen, lower back, pelvis, and external genitals

New code S30.81A Abrasion of flank

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

New code S30.82A Blister (nonthermal) of flank

S30.84 External constriction of abdomen, lower back, pelvis and external genitals

New code S30.84A External constriction of flank

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

New code S30.85A Superficial foreign body of flank

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

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

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

New code S30.87A Other superficial bite of flank

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

New code S30.9A Unspecified superficial injury of flank

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

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

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

New code	S31.107 Unspecified open wound of abdominal wall, left flank without penetration into peritoneal cavity
New code	S31.10A Unspecified open wound of abdominal wall, unspecified flank without penetration into peritoneal cavity
Add	Open wound of abdominal wall flank, NOS
	S31.11 Laceration without foreign body of abdominal wall without penetration into peritoneal cavity
New code	S31.116 Laceration without foreign body of abdominal wall, right flank without penetration into peritoneal cavity
New code	S31.117 Laceration without foreign body of abdominal wall, left flank without penetration into peritoneal cavity
New code	S31.11A Laceration without foreign body of abdominal wall, unspecified flank without penetration into peritoneal cavity
Add	Laceration without foreign body, flank NOS
	S31.12 Laceration with foreign body of abdominal wall without penetration into peritoneal cavity
New code	S31.126 Laceration with foreign body of abdominal wall, right flank without penetration into peritoneal cavity
New code	S31.127 Laceration with foreign body of abdominal wall, left flank without penetration into peritoneal cavity
New code	S31.12A Laceration with foreign body of abdominal wall unspecified flank without penetration into peritoneal cavity
Add	Laceration with foreign body of abdominal wall of flank NOS, without penetration into peritoneal cavity

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

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

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

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

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

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

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

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

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

Add — Puncture wound of abdominal wall with foreign body, flank NOS

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

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

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

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

Add — Open bite of abdominal wall, flank NOS

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

	S31.65 Open bite of abdominal wall with penetration into peritoneal cavity
New code	S31.656 Open bite of abdominal wall, right flank with penetration into peritoneal cavity
New code	S31.657 Open bite of abdominal wall, left flank with penetration into peritoneal cavity
New code	S31.65A Open bite of abdominal wall, unspecified flank with penetration into peritoneal cavity
Add	Open bite of abdominal wall, flank NOS, with penetration into peritoneal cavity

Fournier Disease of Vagina and Vulva

NCHS has received a proposal for a new code for Fournier disease or gangrene of the vagina and vulva. Fournier disease/gangrene is a severe infectious necrotizing condition. Currently this condition is coded to N76.89, Other specified inflammation of vagina and vulva. This code merely describes inflammation of the vagina and does not adequately reflect Fournier disease. Creation of a new code will more specifically classify this significant condition which is often a diabetic complication. This is a representation with changes incorporated in response to public comments received at the September 2020 Coordination and Maintenance meeting. Changes are noted in **bold**.

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

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

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

TABULAR MODIFICATIONS

N76 Other inflammation of vagina and vulva

Use additional code (B95-B97), to identify infectious agent

Excludes2: senile (atrophic)

vaginitis (N95.2)vulvar vestibulitis (N94.810)

N76.8 Other specified inflammation of vagina and vulva

New code N76.82 Fournier disease of vagina and vulva

Add Fournier gangrene of vagina and vulva

Add Code also, if applicable, diabetes mellitus (E08-E13 with .9)

Add Excludes1: gangrene in diabetes mellitus (E08-E13 with .52)

Immunoglobulin A Nephropathy (IgAN)

The Renal Physicians Association (RPA) is requesting a new ICD-10-CM code for Immunoglobulin A Nephropathy (IgAN), the most common form of glomerulonephropathy.[i]

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

IgAN is characterized by deposition of immune complexes containing Immunoglobulin A in the glomerulus and proliferation of mesangial cells.[v,vi] The course of disease progression in IgAN can usually be predicted by clinical signs (hypertension, proteinuria, impaired renal function) and histologic lesions (extent of sclerosis and tubulointerstitial damage).[10] Higher levels and longer duration of proteinuria are the strongest prognostic risk factors for disease progression.[vii,viii] There are a number of specific therapies that are used in the treatment of IgAN patients.[ix, x]

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

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

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

not be precise enough to indicate the severity or course of IgA nephropathy.

Currently, IgAN cases are commonly coded as N02.8, defined as “recurrent and persistent hematuria with ‘other’ morphologic changes.” RPA notes that N02.8 and N02.9 (“other” morphologic changes and “unspecified” morphologic changes, respectively) are both worded as “catch all” codes intended for vaguely defined cases. IgAN is a well-defined condition. Therefore, to avoid further confusion, RPA recommends adding a new code, N02.B, to specifically identify IgAN.

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TABULAR MODIFICATIONS

N02 Recurrent and Persistent Hematuria

New code N02.B Recurrent and persistent immunoglobulin A nephropathy

Limb Girdle Muscular Dystrophies

Muscular dystrophy has several major types and dozens of sub-types. (1) The five most common types of muscular dystrophy are: Becker, Duchenne, Facioscapulohumeral (FSH), Myotonic, and Limb Girdle. (2) There are specific ICD-10-CM codes for Myotonic (G71.11), Becker and Duchenne (G71.01), and FSH (G71.02) muscular dystrophies. It is proposed to add codes for limb girdle muscular dystrophy (LGMD) and selected LGMD subtypes. This proposal is based on a submission from and on behalf of a coalition of LGMD patient advocacy organizations and LGMD clinical experts, and reflects the input of clinicians, researchers, biopharmaceutical companies, physical therapists, coding experts, and other medical professionals familiar with LGMD.

Limb girdle muscular dystrophies are a group of genetically inherited conditions that primarily affect proximal skeletal muscle leading to loss of muscle fibers and progressive, predominantly proximal muscle weakness. (3) To be considered an LGMD, the condition must be described in at least two unrelated families, individuals must demonstrate degenerative changes on muscle imaging over the course of the disease, and have dystrophic changes on muscle histology, ultimately leading to end-stage pathology for the most affected muscles. Most affected individuals achieve independent walking, and most individuals have an elevated serum creatine kinase activity. (4)

There are currently 34 identified subtypes of LGMD, each with a unique genetic cause. (4) While clinical presentations can be similar (thus explaining the initial grouping) these differing genetic causes result in varying presentations and have variation in pathophysiology. Prevalence of subtypes can vary markedly in different subpopulations, due to founder effects. Some of the most prevalent LGMD subtypes are the autosomal recessive LGMDs caused by mutations in the genes that code for the proteins calpain-3, dysferlin, anoctamin5, and alpha-sarcoglycan. (6) Sarcoglycan is a tetramer, made up of four subunits, alpha-sarcoglycan, beta-sarcoglycan, gamma-sarcoglycan, and delta-sarcoglycan. Mutations in each of these can cause LGMD, with prevalence of dysfunction most common for alpha-sarcoglycan, followed in order by beta- sarcoglycan, gamma- sarcoglycan, and delta-sarcoglycan. There is ongoing work involving advanced clinical therapeutic programs that could potentially result in an FDA-approved treatment for a number of the LGMD subtypes within five years, including both beta sarcoglycanopathy and gamma sarcoglycanopathy. (5)

Similar to the rationale used to create ICD-10-CM codes for Duchenne, Becker, and FSH muscular dystrophies, creating specific codes for the LGMDs will provide more accurate diagnoses; increase access to targeted care management and treatment; and inform patient decision making on clinical trials and resources for subtype-specific patient communities. Specific codes will facilitate the surveillance of these diseases; will allow more accurate estimates of their incidence, prevalence, survivorship, mortality and its causes, injuries, symptoms, and health visits; will help to identify

factors that influence health status and secondary conditions, and will facilitate targeted therapeutic development and treatment at the LGMD subtype level. On a larger scale, ICD-10-CM codes can be used to compare health information across hospitals, regions, clinical settings, countries, and even across time in a given location and to facilitate the evaluation of clinical guidelines.

Conditions for which new specific codes have been proposed here have an estimated prevalence of at least one per million population in at least some available published information, with the caveats noted that such prevalence numbers can vary across different populations, and noting that there may be some uncertainty in these estimates, and different estimates from different studies. NCHS invites comments broadly related to creation of ICD-10-CM codes for rare conditions, as well as comments specific to this proposal.

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TABULAR MODIFICATIONS

	G71	Primary disorders of muscles		
		G71.0	Muscular dystrophy	
New sub-subcategory			G71.03	Limb girdle muscular dystrophies
New code				G71.031 Autosomal dominant limb girdle muscular dystrophy
Add				LGMD D4 calpain-3-related
Add				LGMD D5 collagen 6-related
Add				Limb girdle muscular dystrophy type 1

New code	G71.032	Autosomal recessive limb girdle muscular dystrophy due to calpain-3 dysfunction
Add		Limb girdle muscular dystrophy type 2A
Add		LGMD R1 calpain-3-related
Add		Primary calpainopathy
New code	G71.033	Limb girdle muscular dystrophy due to dysferlin dysfunction
Add		Dysferlinopathy
Add		LGMD R2 dysferlin-related
Add		Limb girdle muscular dystrophy type 2B
Add		Miyoshi Myopathy type 1
New subcategory	G71.034	Limb girdle muscular dystrophy due to sarcoglycan dysfunction
New code	G71.0340	Limb girdle muscular dystrophy due to sarcoglycan dysfunction, unspecified
Add		Sarcoglycanopathy, NOS
New code	G71.0341	Limb girdle muscular dystrophy due to alpha sarcoglycan dysfunction
Add		Alpha sarcoglycanopathy
Add		Limb-girdle muscular dystrophy due to alpha-sarcoglycan deficiency
Add		Limb girdle muscular dystrophy type 2D
New code	G71.0342	Limb girdle muscular dystrophy due to beta sarcoglycan dysfunction
Add		Beta sarcoglycanopathy
Add		Limb girdle muscular dystrophy due to beta-sarcoglycan deficiency
Add		Limb girdle muscular dystrophy type 2E

New code		G71.0349	Limb girdle muscular dystrophy due to other sarcoglycan dysfunction
Add			Delta sarcoglycanopathy
Add			Delta-sarcoglycan-related LGMD R6
Add			Gamma sarcoglycanopathy
Add			Gamma-sarcoglycan-related LGMD R5
Add			Limb girdle muscular dystrophy type 2C
Add			Limb girdle muscular dystrophy type 2F
New code		G71.035	Limb girdle muscular dystrophy due to anoctamin5 dysfunction
Add			Anoctamin-5-related LGMD R12
Add			Anoctaminopathy
Add			Autosomal recessive limb girdle muscular dystrophy type 2L
Add			Miyoshi Myopathy type 3
New code		G71.038	Other limb girdle muscular dystrophy
Add			LGMD R9 FKRP-related
Add			LGMD R22 collagen 6-related
Add			Limb girdle muscular dystrophy due to fukutin related protein dysfunction
Add			Limb girdle muscular dystrophy type 2I
Add			Other autosomal recessive limb girdle muscular dystrophy
New code		G71.039	Limb girdle muscular dystrophy, unspecified
	G71.09		Other specified muscular dystrophies
Delete			~~Limb-girdle muscular dystrophy~~

INDEX MODIFICATIONS

Add Calpainopathy (primary) G71.032
Add - autosomal dominant G71.031
Add - autosomal recessive G71.032

Cardiomyopathy (familial) (idiopathic) I42.9
- due to
Revise - - progressive muscular dystrophy (see also Dystrophy, muscular, by type) G71.09 [I43]

Dystrophy, dystrophia
Revise - Leyden-Möbius ~~G71.09~~ – see Dystrophy, muscular, limb-girdle
- muscular G71.00
Revise - - hereditary (progressive) (see also Dystrophy, muscular, by type) G71.09
Revise - - limb-girdle G71.039
Add - - - alpha-sarcoglycan-related G71.0341
Add - - - anoctamin-5-related autosomal recessive (R12) G71.035
Add - - - beta-sarcoglycan-related G71.0342
Add - - - calpain-3-related G71.032
Add - - - - autosomal dominant G71.031
Add - - - - autosomal recessive G71.032
Add - - - collagen VI related
Add - - - - autosomal dominant G71.031
Add - - - - autosomal recessive G71.038
Add - - - D1 (autosomal dominant) G71.031
Add - - - D2 (autosomal dominant) G71.031
Add - - - D3 (autosomal dominant) G71.031
Add - - - D4 (autosomal dominant) G71.031
Add - - - D5 (autosomal dominant) G71.031
Add - - - delta-sarcoglycan-related G71.0349
Add - - - FKRP-related autosomal recessive G71.038
Add - - - gamma-sarcoglycan-related G71.0349
Add - - - R1 (autosomal recessive) G71.032
Add - - - R2 (autosomal recessive) G71.033
Add - - - R3 (autosomal recessive) G71.0341
Add - - - R4 (autosomal recessive) G71.0342
Add - - - R5 (autosomal recessive) G71.0349
Add - - - R6 (autosomal recessive) G71.0349

Add - - - R7 (autosomal recessive) G71.038
Add - - - R8 (autosomal recessive) G71.038
Add - - - R9 (autosomal recessive) G71.038

Dystrophy, dystrophia…
- muscular … [continued]
Add - - - R10 (autosomal recessive) G71.038
Add - - - R11 (autosomal recessive) G71.038
Add - - - R12 (autosomal recessive) G71.035
Add - - - R13 (autosomal recessive) G71.038
Add - - - R14 (autosomal recessive) G71.038
Add - - - R15 (autosomal recessive) G71.038
Add - - - R16 (autosomal recessive) G71.038
Add - - - R17 (autosomal recessive) G71.038
Add - - - R18 (autosomal recessive) G71.038
Add - - - R19 (autosomal recessive) G71.038
Add - - - R20 (autosomal recessive) G71.038
Add - - - R21 (autosomal recessive) G71.038
Add - - - R22 (autosomal recessive) G71.038
Add - - - R23 (autosomal recessive) G71.038
Add - - - R24 (autosomal recessive) G71.038
Add - - - type 1 (autosomal dominant) G71.031
Add - - - type 1A (autosomal dominant) G71.031
Add - - - type 1B (autosomal dominant) G71.031
Add - - - type 1C (autosomal dominant) G71.031
Add - - - type 1E (autosomal dominant) G71.031
Add - - - type 1H (autosomal dominant) G71.031
Add - - - type 1I (autosomal dominant) G71.031
Add - - - type 2 (autosomal recessive) G71.038
Add - - - - specified NEC G71.038
Add - - - type 2A (autosomal recessive) G71.032
Add - - - type 2B (autosomal recessive) G71.033
Add - - - type 2C (autosomal recessive) G71.0349
Add - - - type 2D (autosomal recessive) G71.0341
Add - - - type 2E (autosomal recessive) G71.0342
Add - - - type 2F (autosomal recessive) G71.0349
Add - - - type 2I (autosomal recessive) G71.038
Add - - - type 2L (autosomal recessive) G71.035
Revise - - progressive (hereditary) (see also Dystrophy, muscular, by type) G71.09

Hypertrophy, hypertrophic
Revise - pseudomuscular (see also Dystrophy, muscular, by type, if applicable) G71.09

Revise Leyden-Möbius ~~Leyden-Moebius~~ dystrophy ~~G71.09~~ – see Dystrophy, muscular, limb-girdle

Add LGMD – see Dystrophy, muscular, limb-girdle

Myocardiopathy (congestive) (constrictive) (familial) (hypertrophic nonobstructive) (idiopathic) (infiltrative)(obstructive) (primary) (restrictive) (sporadic) -see also Cardiomyopathy I42.9

- in (due to)
Revise - - progressive muscular dystrophy (see also Dystrophy, muscular, by type) G71.09 [I43]

Myopathy G72.9
Revise - limb-girdle ~~G71.09~~ – see Dystrophy, muscular, limb-girdle

Paralysis, paralytic (complete) (incomplete) G83.9
Revise - pseudohypertrophic (muscle) (see also Dystrophy, muscular, by type, if applicable) G71.09

Paresis -see also Paralysis
Revise - pseudohypertrophic (see also Dystrophy, muscular, by type, if applicable) G71.09

Revise Pseudohypertrophy, muscle (see also Dystrophy, muscular, by type, if applicable) G71.09

Lumbar and Lumbosacral Intervertebral Annulus Fibrosus Defects

A previous proposal to create ICD-10-CM codes for lumbar and lumbosacral intervertebral annular fibrosis defects was presented in September, based on a request from Intrinsic Therapeutics. This is a repeat presentation, with revisions based on comments made from the prior presentation and further review. The proposal is described as consistent with policy guidance from the International Society for the Advancement of Spine Surgery (ISASS). Patient outcomes following lumbar and lumbosacral discectomy vary based on the presence and size of these defects.

The prior proposal was to create codes for small and large defects, involving the lumbar and the lumbosacral annulus fibrosus. For further detailed information and clinical references, please see the prior proposal. While the request identified small defects and being less than 6 mm wide and 4 mm high, and large being greater than or equal to 6 mm wide and greater than 4 mm high, there were concerns about using exact numbers in the classification. Also, there were concerns about whether the size would always be identified. Thus, the current proposal adds codes for cases where the size is unspecified. In addition, it was recommended that the Code first notes for lumbar or lumbosacral disc herniation be "if applicable."

Text which is new or modified from the prior proposal has been bolded in the proposed modifications shown below.

TABULAR MODIFICATIONS

M51 Thoracic, thoracolumbar, and lumbosacral intervertebral disc disorders

New subcategory	M51.A	**Other lumbar and lumbosacral annulus fibrosus disc defects**
New code	**M51.A0**	**Intervertebral annulus fibrosus defect, lumbar region, unspecified size**
Add		**Code first, if applicable, lumbar disc herniation (M51.06, M51.16, M51.26)**
New code	M51.A1	**Intervertebral annulus fibrosus defect, small, lumbar region**
Add		Code first, if applicable, lumbar disc herniation (M51.06, M51.16, M51.26)
New code	M51.A2	**Intervertebral annulus fibrosus defect, large, lumbar region**

Add		Code first, if applicable, lumbar disc herniation (M51.06, M51.16, M51.26)
New code	**M51.A3**	**Intervertebral annulus fibrosus defect, lumbosacral region, unspecified size**
Add		**Code first, if applicable, lumbosacral disc herniation (M51.17, M51.27)**
New code	**M51.A4**	**Intervertebral annulus fibrosus defect, small, lumbosacral region**
Add		Code first, if applicable, lumbosacral disc herniation (M51.17, M51.27)
New code	**M51.A5**	**Intervertebral annulus fibrosus defect, large, lumbosacral region**
Add		Code first, if applicable, lumbosacral disc herniation (M51.17, M51.27)

INDEX MODIFICATIONS

	Defect
Add	- intervertebral annulus fibrosus (see also Disease, intervertebral disc, by site) M51.9

Mild Cognitive Disorder Due to Known Physiological Conditions

The American Psychiatric Association (APA) presented this proposal at the September 2020 Coordination and Maintenance Meeting. Based on comments received, revisions **(noted in bold)** have been made for reconsideration.

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

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

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

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

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

This proposal is being modeled after F02.8, Dementia in diseases classified elsewhere, with a coding note instruction to "Code first the underlying physiological condition" in order to allow for the specification of the underlying pathologic condition. **A subset of the conditions listed under F02.8 have been included as well,** since mostly the same conditions that can cause dementia can also cause mild cognitive disorder. It is also being proposed to use a modified version of the excludes1 note that is currently under G31.84, Mild cognitive impairment, so stated, since most of these are also applicable to proposed new code (F06.7-).

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

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

References:

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TABULAR MODIFCATIONS

F02 Dementia in other diseases classified elsewhere

Includes: Major neurocognitive disorder in other diseases classified elsewhere

Add **Excludes1: mild neurocognitive disorder due to known physiological condition with or without behavioral disturbance (F06.7-)**

F06 Other mental disorders due to known physiological condition

New subcategory F06.7 Mild cognitive disorder due to known physiological condition

Add Mild cognitive impairment due to a known physiological condition

Add Mild neurocognitive disorder due to a known physiological condition

Add Code first the underlying physiological condition, such as:
Alzheimer's (G30.-)
frontotemporal dementia (G31.09)
human immunodeficiency virus [HIV] disease (B20)
Huntington's disease (G10)

Revise **Lewy body ~~disease~~ <u>dementia</u> (G31.83)**
Parkinson's disease (G20)
systemic lupus erythematosus (M32.-)
traumatic brain injury (S06.-)
vitamin B deficiency (E53.8)

Add

Excludes1: age related cognitive decline (R41.81)
altered mental status (R41.82)
cerebral degeneration (G31.9)
change in mental status (R41.82)
cognitive deficits following (sequelae of) cerebral hemorrhage or infarction (I69.01-I69.11-, I69.21-I69.31-, I69.81- I69.91-)
cognitive impairment due to intracranial or head injury (S06.-)
dementia (F01.-, F02.-, F03)
mild cognitive impairment due to unknown or unspecified etiology (G31.84)
neurologic neglect syndrome (R41.4)
personality change, nonpsychotic (F68.8)

New code

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

Add

Mild cognitive disorder due to known physiological condition, NOS

New code

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

F09 Unspecified mental disorder due to known physiological condition
Mental disorder NOS due to known physiological condition
Organic brain syndrome NOS
Organic mental disorder NOS
Organic psychosis NOS
Symptomatic psychosis NOS

Code first the underlying physiological condition

Add

Excludes1: Mild cognitive disorder due to known physiological condition (F06.7-)
psychosis NOS (F29)

G10 Huntington's disease
Huntington's chorea
Huntington's dementia

Code also dementia in other diseases classified elsewhere without behavioral disturbance (F02.80)

Add **Use additional code, if applicable, to identify mild cognitive disorders due to known physiological condition (F06.7-)**

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, if applicable, to identify mild cognitive disorders due to known physiological condition (F06.7-)**

G30 Alzheimer's disease
Includes: Alzheimer's dementia senile and presenile forms

Use additional code to identify:
delirium, if applicable (F05)
dementia with behavioral disturbance (F02.81)
dementia without behavioral disturbance (F02.80)

Add **mild cognitive disorders due to known physiological condition (F06.7-)**

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

Add **Use additional code, if applicable, to identify mild cognitive disorders due to known physiological condition (F06.7-)**

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

Revise Mild neurocognitive disorder of <u>uncertain or unknown etiology</u>

Add **Mild cognitive disorder NOS**

Add Excludes1:mild cognitive disorder due to a known physiological condition (F06.7-)

Delete ~~mild memory disturbance (F06.8)~~

S06 Intracranial injury
Includes: traumatic brain injury
Code also any associated:
open wound of head (S01.-)
skull fracture (S02.-)

Add **Use additional code, if applicable, to identify mild cognitive disorders due to known physiological condition (F06.7-)**

Poisoning by Methamphetamine

Methamphetamine is a powerful, highly addictive stimulant that affects the central nervous system. Crystal methamphetamine is a form of the drug that looks like glass fragments or shiny, bluish-white rocks. It is chemically similar to amphetamine, a drug used to treat attention-deficit hyperactivity disorder (ADHD) and narcolepsy, a sleep disorder.[1]

Methamphetamine increases the amount of the natural chemical dopamine in the brain. Dopamine is involved in body movement, motivation, and reinforcement of rewarding behaviors. The drug's ability to rapidly release high levels of dopamine in reward areas of the brain strongly reinforces drug-taking behavior, making the user want to repeat the experience.[1]

Methamphetamine is made illicitly and is illegal in the United States. Crystal methamphetamine is a Schedule II substance under the Controlled Substances Act. Schedule II drugs, which include cocaine and PCP have a high potential for abuse. Abuse of these drugs may lead to severe psychological or physical dependence.[2]

Currently, poisoning by, adverse effect of and underdosing of methamphetamine is classified under ICD-10-CM T43.62, Poisoning by, adverse effect of and underdosing of amphetamines. T43.62 is not specific to methamphetamine. Grouping methamphetamine with other amphetamines, such as prescription dextroamphetamine/amphetamine, results in difficulty tracking methamphetamine specifically. A new ICD-10-CM code specifically to track, trend, and research methamphetamine has been requested by the Arizona Medicaid Program, who also serves as the Single State Authority (SSA) for Substance Abuse Services.

Given the current trends related to methamphetamine related morbidity and mortality in the United States[3], including the report from CDC that provisional overdose deaths increased 10-fold by 2019[1], it is being requested that CDC implement a specific methamphetamine ICD-10 CM diagnosis code classification and remove methamphetamine as a type of amphetamine under the T43.62 classification. National Center for Health Statistics, Division of Analysis and Epidemiology supports the creation of a new ICD-10-CM code for Poisoning by Methamphetamine.

References:

1. Products - Vital Statistics Rapid Release - Provisional Drug Overdose Data (cdc.gov)
2. Crystal Methamphetamine Fast Facts - Department of Justice, (justice.gov/archive/ndic/pubs5/5049/5049p.pdf)
3. Patterns and Characteristics of Methamphetamine Use Among Adults — United States, 2015–2018 | MMWR (cdc.gov)

TABULAR MODIFICATION

	T43	Poisoning by, adverse effect of and underdosing of psychotropic drugs, not elsewhere classified
	T43.6	Poisoning by, adverse effect of and underdosing of psychostimulants
	T43.62	Poisoning by, adverse effect of and underdosing of amphetamines
Delete	~~Poisoning by, adverse effect of and underdosing of methamphetamines~~	
New subcategory	T43.65	Poisoning by, adverse effect of and underdosing of methamphetamines
New code	T43.651	Poisoning by methamphetamines, accidental (unintentional)
Add		Poisoning by methamphetamines NOS
New code	T43.652	Poisoning by methamphetamines, intentional self-harm
New code	T43.653	Poisoning by methamphetamines, assault
New code	T43.654	Poisoning by methamphetamines, undetermined
New code	T43.655	Adverse effect of methamphetamines
New code	T43.656	Underdosing of methamphetamines

Post COVID-19 Condition

The disease COVID-19, caused by the coronavirus SARS-CoV-2, is a significant public health issue, and for some people there can be long term effects following infection. These can range from symptoms such as loss of smell or taste, or can include chronic respiratory failure, in some cases particularly following COVID-19 pneumonia or Acute Respiratory Distress Syndrome (ARDS). WHO has added a new code to ICD-10 at U09.9, for Post COVID-19 condition, unspecified. It is proposed to add this code in ICD-10-CM. The implementation date is expected to be October 1, 2021. The comment deadline will be April 9, 2021.

TABULAR MODIFICATIONS

	B94	Sequelae of other and unspecified infectious and parasitic diseases
Add		Excludes2: Post COVID-19 condition (U09.9)
New category	U09	Post COVID-19 condition
New code	U09.9	Post COVID-19 condition, unspecified
Add		Note: This code enables establishment of a link with COVID-19.
Add		This code is not to be used in cases that still are presenting COVID-19.
Add		Code first the specific condition related to COVID-19 if known, such as:
Add		chronic respiratory failure (J96.1-)
Add		loss of smell (R43.8)
Add		loss of taste (R43.8)

Post Traumatic Visual Disturbance

Traumatic brain injury (TBI) is a serious public health problem in the United States and there is a need to better quantify the sequela to assist public health agencies and researchers gather additional data regarding these conditions and their impact. Currently, post traumatic visual disturbances may be reported using 7th character sequela code for TBI and the appropriate visual disturbance code.

Visual disturbance is also, in fact, a separate and distinct set of symptoms from TBI. Post traumatic visual disturbance can persist following the initial TBI and requires a separate code. Visual disturbances can develop as a result of several types of neurological events such as a traumatic brain injury, cerebrovascular accident, multiple sclerosis, or other neurologic impairments.

The American Optometric Association (AOA) is requesting a unique code specific to the visual effects of these injuries.

TABULAR MODIFICATIONS

	H53	Visual disturbances
	H53.1	Subjective visual disturbance
New code		H53.17 Post traumatic visual disturbance

Primary Blast Injury of Brain

This is a joint request from the Department of Defense and the Department of Veterans Affairs. ICD-10-CM diagnostic codes exist for primary blast injury to eight organs susceptible to primary blast injury from exposure to blast overpressure: colon, rectum, ear, lung, bronchus, small intestine, fallopian tube, and thoracic trachea. A diagnostic code does not exist for primary blast injury of the brain. Emerging clinical and experimental evidence supports the reality of this diagnosis absent impact acceleration. The injury can occur in the absence of head motion as clearly seen experimentally [(1)]. This injury can affect service members during training and combat operations.

An explosion generates a blast wave traveling faster than sound and creating a surge of high pressure followed by a vacuum. The primary blast-brain interaction includes two main mechanisms, which do not exclude each other; rather, they occur in parallel: 1) direct interaction with the head through direct passage of the blast wave through the skull (transcranial); and 2) kinetic energy transfer of the primary blast wave that compresses the torso, impacting blood vessels which send damaging energy pulses into the brain (transcorporal). [(2, 3)]

In 2014, the Institute of Medicine (IOM) evaluated health effects of exposure to blast, including the blast waves (the supersonic waves of intense air pressure that follow detonation of an explosive device). At that time, the IOM was not able to identify primary studies that focused exclusively on acute blast-related traumatic brain injury (TBI). However, since many of the studies cited in a previous IOM volume included both blast and non-blast TBI, the IOM concluded that it is likely that the injuries are at least as severe in blast TBI. The IOM noted that, although the clinical and pathologic syndromes of blast-induced TBI and other forms of TBI probably overlap extensively, there may be some differences that could potentially produce distinctive presentations and require different therapeutic strategies. For example, typical symptoms of concussion, such as seeing stars and experiencing a transient loss of consciousness (LOC), may be absent. The limited evidence at that time indicated that early malignant brain swelling may be more common in connection with blast than with other injuries. In addition, numerous studies suggested that blast TBI may confer distinctive neuroimaging patterns as measured by DTI (tractography). The IOM noted that blast-induced TBI may result in a diffuse bihemispheric pattern of disruption, unlike the more focal, often frontal and occipital (coup–contra coup) pattern classically observed in acceleration–deceleration concussive injury. That pattern could potentially result in a higher frequency of global cerebral complaints involving cognitive, visual, auditory, and other sensory modalities in those exposed to blast. [(4)]

In the intervening years since the 2014 IOM report, research continues to further elucidate the pathophysiological mechanisms and the distinct clinical features of primary blast-induced brain injury, together with potential prevention strategies and clinical management.

After the shock wave interacts with the body and head, a pressure wave passes through the body and head inducing complex response mechanisms, which can be divided into four main groups:

1. Primary tissue damage of the brain parenchyma caused by stretch, strain and/or rupture of parenchyma and blood vessels (initiating secondary brain injury mechanisms that lead to acute or chronic pathologic changes such as increased blood-brain barrier (BBB) permeability; compromised cerebrovascular and neuronal permeability; diffuse axonal injury; astrocyte and microglia activation; apoptotic cell death; Purkinje cell degeneration; and ultrastructural changes, such as increased vacuolization of cytoplasm, myelin sheet damage, and neurofilament abnormalities);
2. Changes triggered by the autonomic nervous system (ANS) that further contribute to cerebral hypoxia;
3. Consequences of increased vascular load;
4. Effects of locally synthesized and released mediators/modulators (so-called 'autacoids') and/or immune system activation. [2, 3]

Primary blast injury to the brain is a unique clinical entity with unique prevention and treatment ramifications. Data show distinct onset, duration, localization, and consequences that are unique to Blast Induced Neurotrauma (BINT). The features of cerebral edema, BBB dysfunction, and cerebral vasospasm in BINT differ significantly from changes seen after conventional TBI. Indeed, although traumatic cerebral vasospasm after BINT can develop early, often within 48 hours of injury, it can also present later, typically 10 days or more after initial injury. Although cerebral vasospasm is usually stimulated by subarachnoid hemorrhage (SAH), observations suggest that SAH is not necessary for vasospasm to occur in BINT. A recent experimental study using theoretical and in vitro models showed that a single rapid mechanical insult is capable of inducing vascular hypercontractility and remodeling, indicative of vasospasm initiation. The findings suggest a feasible scenario that the shock wave propagating through the vasculature interacts with cellular elements of vascular wall (endothelium, vascular smooth muscle). This interaction, in turn, leads to synthesis and release of various mediators and modulators, which initiate hypercontraction and subsequent genetic switch that potentiates vascular remodeling and cerebral vasospasm. Recent clinical studies imply that primary blast (i.e., blast forces alone) can cause negativistic behavioral changes when evaluated with selected measures of personality, and they may have greater post concussive sequelae, including deficits in attentional control and regional brain metabolism, compared with blunt mild TBI (mTBI). It has also been reported that blast-related injuries, specifically mild BINT (mBINT), during deployment has negative consequences on service members' perception of health. [2]

Recent studies **reveal** the effects of low-level occupational exposure to blast overpressure. Blast exposure and recurrent occupational overpressure exposure (ROPE) were independently associated

with mTBI, and Marines with both blast exposure during deployment and ROPE were especially likely to sustain mTBI [5]. Studies with military breachers are beginning to reveal the effects on the brain of occupational blast overpressure. [6, 7]

Given the number of service members, law enforcement officers, miners, and others that are routinely exposed to blast overpressure, primary blast injury to the brain is a significant public health problem. Primary blast injury to the brain is also an active and important area of research. Having codes for primary blast injury of brain has potential to help with public health prevention efforts and with future research that could advance the care of these incredibly ill patients.

The recommended tabular modification is proffered using similar placement of the other primary blast injury codes such as the Primary blast injury of thoracic trachea. Since we are not requesting that the primary blast injury codes for the brain have any greater specificity with respect to further localizing the injury (e.g. by lobe or hemisphere), the placement of the blast injury codes for the thoracic trachea and the brain are analogous. They are both specified injuries of an organ from an external cause without additional consideration to the anatomic reference (e.g., left, right, bilateral).

This proposal has been reviewed and supported by CDC/NCHS Division of Analysis & Epidemiology.

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TABULAR MODIFICATIONS

S06 Intracranial injury

The appropriate 7th character is to be added to each code from category S06

A initial encounter
D subsequent encounter
S sequela

S06.8 Other specified intracranial injuries

New subcategory — S06.8A Primary blast injury of brain, not elsewhere classified

New code — S06.8A0 Primary blast injury of brain, not elsewhere classified without loss of consciousness

New code — S06.8A1 Primary blast injury of brain, not elsewhere classified with loss of consciousness of 30 minutes or less

New code — S06.8A2 Primary blast injury of brain, not elsewhere classified with loss of consciousness of 31 minutes to 59 minutes

New code — S06.8A3 Primary blast injury of brain, not elsewhere classified with loss of consciousness of 1 hour to 5 hours 59 minutes

New code — S06.8A4 Primary blast injury of brain, not elsewhere classified with loss of consciousness of 6 hours to 24 hours

New code — S06.8A5 Primary blast injury of brain, not elsewhere classified with loss of consciousness greater than 24 hours with return to pre-existing conscious level

New code — S06.8A6 Primary blast injury of brain, not elsewhere classified with loss of consciousness greater than 24 hours without return to pre-existing conscious level with patient surviving

New code — S06.8A7 Primary blast injury of brain, not elsewhere classified with loss of consciousness of any duration with death due to brain injury prior to regaining consciousness

New code	S06.8A8	Primary blast injury of brain, not elsewhere classified with loss of consciousness of any duration with death due to other cause prior to regaining consciousness
New code	S06.8A9	Primary blast injury of brain, not elsewhere classified with loss of consciousness of unspecified duration

Prolonged Grief Disorder

Prolonged Grief Disorder (PGD) is a condition newly added to the 5th edition of the American Psychiatric Association (APA) *Diagnostic and Statistical Manual of Mental Disorders* (DSM-5). Over the past three decades, there has been increasing recognition and conclusive research demonstrating that prolonged grief disorder, which is characterized by intense, prolonged symptoms of grief coupled with clinically significant functional impairment that persists beyond twelve (12) months post-loss, constitutes a distinct mental disorder. It has been estimated that one out of ten bereaved adults following a non-violent loss is at risk for developing PGD [(1).]

The American Psychiatric Association is proposing a code for a new disorder, Prolonged Grief Disorder, to code category F43, Reaction to severe stress and adjustment disorders.
According to DSM-5, prolonged grief disorder involves the development of a prolonged grief response that persists for at least one year. This is characterized by intense yearning or longing for the deceased person (often with intense sorrow and frequent crying) and/or preoccupation with thoughts or memories of the deceased. Additional symptoms occurring since the death include identity disruption (e.g., feeling as though part of oneself has died), a marked sense of disbelief about the death, avoidance of reminders that the person is dead, at times intense emotional pain (e.g., anger, bitterness, guilt, worthlessness, self-pity) and emotional numbness at other times, intense loneliness, having problems engaging with friends, pursuing interests or planning for the future, and feeling that life is meaningless.

Numerous studies have demonstrated that prolonged grief disorder is distinct from other mental disorders, including major depressive disorder, generalized anxiety disorder, and posttraumatic stress disorder[(2-9)] and is associated with significant suffering and enduring functional impairments [(2,7-9,15)]. PGD has idiosyncratic neurobiological [(10)] and clinical [(7-9,11-13)] correlates. This disorder can persist unabated for months or even years [(9,14)]; is associated with marked increases in risks for serious medical conditions, including cardiac disease, hypertension, cancer, and immunological deficiency, as well as reduced quality of life [(15)] and may only respond to targeted interventions [(16,17).]

The American Psychiatric Association is requesting the following tabular modifications.

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TABULAR MODIFCATIONS

F43 Reaction to severe stress and adjustment disorders

New subcategory	F43.8	Other reactions to severe stress Other specified trauma and stressor-related disorder
New code		F43.81 Prolonged Grief Disorder
Add		Complicated grief
Add		Complicated grief disorder
Add		Persistent complex bereavement disorder
New code		F43.89 Other reactions to severe stress
Add		Other specified trauma and stressor-related disorder
	F43.9	Reaction to severe stress, unspecified
Add		Unspecified trauma and stressor-related disorder

Recurrent Vulvovaginal Candidiasis (RVVC)

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

This topic was presented at the September 2020, Coordination and Maintenance meeting and is being represented following changes proposed from public comments.

An estimated 75% of women[1] will have at least one episode of vulvovaginal candidiasis in their lifetime. However, most episodes of VVC are uncomplicated with mild to moderate symptoms that are quickly and successfully addressed via over-the-counter topical antifungal creams and/or a short course of oral fluconazole, an antifungal drug. Among uncomplicated cases of VVC, most are diagnosed on the basis of symptoms alone, and many are self-diagnosed and self-treated.

A smaller but significant subgroup of women develop a more complicated form of vulvovaginal candidiasis.[2] Complicated vulvovaginal candidiasis refers to severe disease, infection in an immune-compromised woman, or infection with a non-*C albicans* species. Most prominently, it refers to recurrent vulvovaginal candidiasis (RVVC), defined as 3-4 or more episodes of symptomatic infection within one year. [2,3,4,5] Prevalence of RVVC has been variously estimated in literature reviews and surveys at 5-9% of women.[3,4,6,7]

The population of women with recurrent vulvovaginal candidiasis is clinically distinct in multiple respects. While several risk factors such as antibiotic use, diabetes, or pregnancy are known, the vast majority of women with RVVC develop the infection without having any risk factor. This implies that a genetic component likely plays an important role in susceptibility to RVVC.[8]

An episode of uncomplicated VVC is often considered a nuisance that is easily resolved. However, women with RVVC typically endure multiple relapses and require months of treatment with a significant impact on their lives.[5] Although new drugs and regimens are being developed, current treatment for RVVC typically consists of topicals or oral fluconazole for 10 to 14 days, followed by a maintenance regimen of oral fluconazole once a week for at least 6 months. This controls symptoms in the great majority of patients, but cessation is followed by another episode of VVC in approximately 50% of women within three to four months, and likely a higher percentage over time.[2,3,5]

RVVC is debilitating for patients, both physically and in terms of their mental health. The physical symptoms interfere with normal elements of life, from urinating to sexual activity. Women with RVVC reported missing an average of about 6 hours of work for each recurrent episode. As surveyed, health-related quality of life is significantly worse for women with RVVC than in the

general population.[6] Over two-thirds of women with RVVC report depression and anxiety during recurrent episodes and over half report anxiety between episodes.[6] A sense of feeling "dirty" and suspecting sexually transmitted diseases from their partners is commonly reported.[7] Overall, women with RVVC ranked it similar to asthma and COPD and even higher than migraine for its negative impact on their quality of life.[6]

As often noted in the literature, the availability of over-the-counter treatment create difficulties in accurately determining the frequency of recurrent vulvovaginal candidiasis.[3,4] It is not currently possible to clearly differentiate recurrent vulvovaginal candidiasis from uncomplicated vulvovaginal candidiasis[7] or to track cases of this clinically significant population in the data.[5]

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

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TABULAR MODIFICATIONS

B37 Candidiasis

New subcategory	B37.3	Candidiasis of vulva and vagina Candidal vulvovaginitis Monilial vulvovaginitis Vaginal thrush
New code	B37.31	Acute candidiasis of vulva and vagina
Add		Candidiasis of vulva and vagina NOS

New code	B37.32	Chronic candidiasis of vulva and vagina
Add		Recurrent candidiasis of vulva and vagina

Refractory Angina Pectoris

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

At the September 9, 2020 ICD-10 Coordination and Maintenance Committee meeting, the creation of a specific code for refractory angina pectoris (I20.2 Refractory angina pectoris) and new codes in the subcategories of Chronic ischemic heart disease (I25.112) and Atherosclerosis of coronary artery bypass graft(s) and coronary artery of transplanted heart with angina pectoris (I25.702 and I25.712) were presented.

During the comment period, NCHS received a recommendation to create additional new codes in the Atherosclerosis of coronary artery bypass graft(s) and coronary artery of transplanted heart with angina pectoris subcategory. Neovasc is presenting this updated proposal to include those added new codes in **bold print.**

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

An increasing number of patients, particularly those with advanced, chronic coronary artery disease,[4] have severe symptoms of angina despite optimal medical therapy. However, refractory angina is common not only in patients who are not good candidates for revascularization, but also in patients following successful revascularization. Persistence or recurrence of angina after PCI or CABG surgery is well recognized and may affect 20–40% of patients during short and medium-term.[567891011].

When further revascularization options are limited, these patients are frequently described as having no option for treatment, and as having refractory angina. The care of these patients is challenging, and the guidance available from national practice guidelines is limited. In 2014, The American College of Cardiology/American Heart Association Task Force on Practice Guidelines, and the American Association for Thoracic Surgery, Preventive Cardiovascular Nurses Association, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons released guidelines for treatment of patients with refractory angina. The European Society of Cardiology (ESC) released guidelines in 2019 for treatment of patients with refractory angina.

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

Neovasc Inc. is submitting this proposal requesting the creation of a specific code for refractory angina pectoris. This will allow the ability to distinguish refractory angina pectoris from other angina diagnoses thus improving data collection and management of the disease.

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

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

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TABULAR MODIFICATIONS

I20 Angina pectoris
- I20.0 Unstable angina
- I20.1 Angina pectoris with documented spasm

New code
- I20.2 Refractory angina pectoris

I25 Chronic ischemic heart disease
- I25.1 Atherosclerotic heart disease of native coronary artery
  - I25.11 Atherosclerotic heart disease of native coronary artery with angina pectoris
    - I25.111 Atherosclerotic heart disease of native coronary artery with angina pectoris with documented spasm

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

I25.7 Atherosclerosis of coronary artery bypass graft(s) and coronary artery of transplanted heart with angina pectoris
I25.70 Atherosclerosis of coronary artery bypass graft(s), unspecified, with angina pectoris
I25.701 Atherosclerosis of coronary artery bypass graft(s), unspecified, with angina pectoris with documented spasm
New code I25.702 Atherosclerosis of coronary artery bypass graft(s), unspecified, with refractory angina pectoris

I25.71 Atherosclerosis of autologous vein coronary artery bypass graft(s) with angina pectoris
I25.710 Atherosclerosis of autologous vein coronary artery bypass graft(s) with unstable angina pectoris
I25.711 Atherosclerosis of autologous vein coronary artery bypass graft(s) with angina pectoris with documented spasm
New code **I25.712 Atherosclerosis of autologous vein coronary artery bypass graft(s) with refractory angina pectoris**

I25.72 Atherosclerosis of autologous artery coronary artery bypass graft(s) with angina pectoris Atherosclerosis of internal mammary artery graft with angina pectoris
I25.720 Atherosclerosis of autologous artery coronary artery bypass graft(s) with unstable angina pectoris
I25.721 Atherosclerosis of autologous artery coronary artery bypass graft(s) with angina pectoris with documented spasm
New code **I25.722 Atherosclerosis of autologous artery coronary artery bypass graft(s) with refractory angina pectoris**

	I25.73		Atherosclerosis of nonautologous biological coronary artery bypass graft(s) with angina pectoris
		I25.730	Atherosclerosis of nonautologous biological coronary artery bypass graft(s) with unstable angina pectoris
		I25.731	Atherosclerosis of nonautologous biological coronary artery bypass graft(s) with angina pectoris with documented spasm
New code		**I25.732**	**Atherosclerosis of nonautologous biological coronary artery bypass graft(s) with refractory angina pectoris**
	I25.75		Atherosclerosis of native coronary artery of transplanted heart with angina pectoris
		I25.750	Atherosclerosis of native coronary artery of transplanted heart with unstable angina
		I25.751	Atherosclerosis of native coronary artery of transplanted heart with angina pectoris with documented spasm
New Code		**I25.752**	**Atherosclerosis of native coronary artery of transplanted heart with refractory angina pectoris**
	I25.76		Atherosclerosis of bypass graft of coronary artery of transplanted heart with angina pectoris
		I25.760	Atherosclerosis of bypass graft of coronary artery of transplanted heart with unstable angina
		I25.761	Atherosclerosis of bypass graft of coronary artery of transplanted heart with angina pectoris with documented spasm
New Code		**I25.762**	**Atherosclerosis of bypass graft of coronary artery of transplanted heart with refractory angina pectoris**

	I25.79	Atherosclerosis of other coronary artery bypass graft(s) with angina pectoris
	I25.790	Atherosclerosis of other coronary artery bypass graft(s) with unstable angina pectoris
	I25.791	Atherosclerosis of other coronary artery bypass graft(s) with angina pectoris with documented spasm
New code	**I25.792**	**Atherosclerosis of other coronary artery bypass graft(s) with refractory angina pectoris**

Slipped Upper Femoral Epiphysis, Stable, Unstable

The American Academy of Orthopedic Surgeons (AAOS) is requesting expansion of code category M93.0, Slipped upper femoral epiphysis to add codes to slipped upper femoral epiphysis. This proposal was originally presented at the September 2018 and the March 2020, Coordination and Maintenance (C&M) meetings and is being represented following changes proposed from public comments. Those items are in **bold**.

AAOS clarified a question from the comment of "unspecified" in the M93.00 Unspecified slipped upper femoral epiphysis (nontraumatic) that this refers to the acuity (acute, chronic, acute on chronic) being unspecified and not to the stability being unspecified.

It is proposed to add new codes to reflect acute- and acute-on-chronic slips which reflect whether the hip is stable or unstable. Slipped capital femoral epiphysis (SCFE) is a failure through the growth plate (physis), which results in slippage of the overlying end of the proximal femur (epiphysis). Normally, the head of the femur (the capital femoral epiphysis) should sit squarely on the femoral neck. Abnormal shear failure through the growth plate results in the slip. The capital femoral epiphysis remains in the acetabulum (hip joint), while the metaphysis (upper end of the femur) moves in an anterior direction with external rotation. The condition usually develops gradually over time. Slips may present as stable or unstable:

> A stable SCFE causes some stiffness or pain in the knee or groin area, and possibly a limp that causes a child to walk with a foot outward. The pain and the limp usually tend to come and go, worsening with activity and getting better with rest. With a stable SCFE, a child still can walk, even if crutches are needed. The prognosis is relatively good for functional recovery.
>
> An unstable SCFE is a more severe slip that usually happens suddenly and is usually much more painful. A child will not be able to bear weight on the affected side. An unstable SCFE is also more serious because it can restrict blood flow to the hip joint, leading to tissue death in the head of the femur. For this reason, the prognosis is much more guarded.

Because the prognosis is strongly related to the stability of the slip (stable versus unstable) it should be reflected in the relevant diagnosis codes. Generally chronic slips are stable and only acute or acute-on-chronic slips can be unstable.

AAOS is requesting the following ICD-10-CM tabular modifications:

TABULAR MODIFICATIONS

M93 Other osteochondropathies

Excludes2: osteochondrosis of spine (M42.-)

M93.0 Slipped upper femoral epiphysis (nontraumatic)

Add **Slipped capital femoral epiphysis (SCFE)**

Add **SUFE**

Use additional code for associated chondrolysis (M94.3)

M93.00 Unspecified slipped upper femoral epiphysis (nontraumatic)

M93.001Unspecified slipped upper femoral epiphysis (nontraumatic), right hip

M93.002 Unspecified slipped upper femoral epiphysis (nontraumatic), left hip

M93.003 Unspecified slipped upper femoral epiphysis (nontraumatic), unspecified hip

New code M93.004 Unspecified slipped upper femoral epiphysis (nontraumatic), bilateral hips

Revise M93.01 Acute slipped upper femoral epiphysis stable (nontraumatic)

Revise M93.011 Acute slipped upper femoral epiphysis stable (nontraumatic), right hip

Revise M93.012 Acute slipped upper femoral epiphysis stable (nontraumatic), left hip

Revise M93.013 Acute slipped upper femoral epiphysis, **stable** (nontraumatic), unspecified hip

New code M93.014 Acute slipped upper femoral epiphysis stable (nontraumatic), bilateral hips

Revise M93.02 Chronic slipped upper femoral epiphysis **stable** (nontraumatic)

Revise M93.021 Chronic slipped upper femoral epiphysis stable (nontraumatic), right hip

Revise M93.022 Chronic slipped upper femoral epiphysis stable (nontraumatic), left hip

Revise M93.023 Chronic slipped upper femoral epiphysis stable (nontraumatic), unspecified hip

New code **M93.024 Chronic slipped upper femoral epiphysis stable (nontraumatic), bilateral hips**

Revise M93.03 Acute on chronic slipped upper femoral epiphysis **stable** (nontraumatic)

Revise M93.031 Acute on chronic slipped upper femoral epiphysis stable (nontraumatic), right hip

Revise	M93.032 Acute on chronic slipped upper femoral epiphysis stable (nontraumatic), left hip
Revise	M93.033 Acute on chronic slipped upper femoral epiphysis stable (nontraumatic), unspecified hip
New code	M93.034 Acute on chronic slipped upper femoral epiphysis stable (nontraumatic), bilateral hips
New subcategory	M93.04 Acute slipped upper femoral epiphysis, unstable (nontraumatic)
New code	M93.041 Acute slipped upper femoral epiphysis, unstable (nontraumatic), right hip
New code	M93.042 Acute slipped upper femoral epiphysis, unstable (nontraumatic), left hip
New code	M93.043 Acute slipped upper femoral epiphysis, unstable (nontraumatic), unspecified hip
New code	M93.044 Acute slipped upper femoral epiphysis, unstable (nontraumatic), bilateral hips
New subcategory	M93.05 Chronic slipped upper femoral epiphysis, unstable (nontraumatic)
New code	M93.051 Chronic slipped upper femoral epiphysis, unstable (nontraumatic), right hip
New code	M93.052 Chronic slipped upper femoral epiphysis, unstable (nontraumatic), left hip
New code	M93.053 Chronic slipped upper femoral epiphysis, unstable (nontraumatic), unspecified hip
New code	M93.054 Chronic slipped upper femoral epiphysis, unstable (nontraumatic), bilateral hips
New subcategory	M93.06 Acute on chronic slipped upper femoral epiphysis, unstable (nontraumatic)
New code	M93.061 Acute on chronic slipped upper femoral epiphysis, unstable (nontraumatic), right hip
New code	M93.062 Acute on chronic slipped upper femoral epiphysis, unstable (nontraumatic), left hip
New code	M93.063 Acute on chronic slipped upper femoral epiphysis, unstable (nontraumatic), unspecified hip
New code	M93.064 Acute on chronic slipped upper femoral epiphysis, unstable (nontraumatic), bilateral hips

New subcategory	**M93.07 Acute slipped upper femoral epiphysis, unspecified stability (nontraumatic)**
New code	**M93.071 Acute slipped upper femoral epiphysis, unspecified stability (nontraumatic), right hip**
New code	**M93.072 Acute slipped upper femoral epiphysis, unspecified stability (nontraumatic), left hip**
New code	**M93.073 Acute slipped upper femoral epiphysis, unspecified stability (nontraumatic), unspecified hip**
New code	**M93.074 Acute slipped upper femoral epiphysis, unspecified stability (nontraumatic), bilateral hips**

New subcategory	**M93.08 Chronic slipped upper femoral epiphysis, unspecified stability (nontraumatic)**
New code	**M93.081 Chronic slipped upper femoral epiphysis, unspecified stability (nontraumatic), right hip**
New code	**M93.082 Chronic slipped upper femoral epiphysis, unspecified stability (nontraumatic), left hip**
New code	**M93.083 Chronic slipped upper femoral epiphysis, unspecified stability (nontraumatic), unspecified hip**
New code	**M93.084 Chronic slipped upper femoral epiphysis, unspecified stability (nontraumatic), bilateral hips**

New subcategory	**M93.0A Acute on chronic slipped upper femoral epiphysis, unspecified stability (nontraumatic)**
New code	**M93.0A1 Acute on chronic slipped upper femoral epiphysis, unspecified stability (nontraumatic), right hip**
New code	**M93.0A2 Acute on chronic slipped upper femoral epiphysis, unspecified stability (nontraumatic), left hip**
New code	**M93.0A3 Acute on chronic slipped upper femoral epiphysis, unspecified stability (nontraumatic), unspecified hip**
New code	**M93.0A4 Acute on chronic slipped upper femoral epiphysis, unspecified stability (nontraumatic), bilateral hips**

Short Stature Due to Endocrine Disorder

This proposal submitted by Ipsen Biopharmaceuticals and supported by the Pediatric Endocrine Society was presented at the September 2020 Coordination and Maintenance meeting. Based on public comment, revisions have been made **(noted in bold)** and resubmitted for reconsideration.

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

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

Between birth and puberty, a normal growth rate depends on an adequate secretion and action of growth hormone, which is released from the pituitary gland in response to several factors: hypothalamic GH releasing hormone (GHRH), ghrelin, and somatostatin.[4] Growth hormone binds to GH receptors (GHR), mainly on cells in the liver, although most tissues contain GHRs.[5] The interaction between GH and the GHR induces formation and release of IGF-I. Both the circulating IGF-I, secreted from the liver into the circulation, and locally produced IGF-I, then exert the growth-promoting effects at the level of skeletal muscle, cartilage, bone, and other tissues.[4]
When these coordinated growth events are altered, short stature may occur. Short stature has a variety of causes and the first step in the diagnostic evaluation of growth impairment leading to short stature due to an endocrine disorder will be to rule out other causes of growth failure, including genetic syndromes such as Turner syndrome, and several other secondary causes like malnutrition and inflammatory disorders.[6]

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

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

hormone deficiency than laboratory assays.[9] GHD is treated with recombinant human GH (rhGH), also known as somatotropin.[7]

Growth hormone deficiency must also be ruled out in order to diagnose constitutional short stature, which, along with familial short stature, is a form of normal variant short stature often classified as idiopathic short stature (ISS).[8,10,11] Constitutional short stature or constitutional growth delay describes patients with an unknown cause of short stature. This diagnosis depends on ruling out other causes of short stature, and is further characterized by specific auxological characteristics.[11] Approximately 70% of children with a short stature diagnosis have some type of idiopathic short stature, including constitutional short stature, but also with other unknown etiologies.[3] In some situations of ISS (not constitutional short stature or benign familial short stature), use of supplemental rhGH can increase the growth potential despite normal endogenous GH production.[8]
For those children who do not have GHD despite having IGF-I deficiency, primary IGF-I deficiency (PIGFD) may be the underlying etiology.[8] Severe PIGFD (SPIGFD) is defined by height and circulating IGF-I concentrations below -3 SDS.[12,13] A subset of patients with SPIGFD have mutations in the GH receptor gene and have Laron-type short stature.[14] The prevalence rate of SPIGFD in children suspected of having a growth abnormality is approximately 1%.[15] In some situations, patients with GHD who develop GH inactivating antibodies are considered GH insensitive, also have IGFD, and could also benefit from treatment with rhIGF.[16,17]

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

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

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

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TABULAR MODIFICATIONS

E23 Hypofunction and other disorders of the pituitary gland

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

Excludes1:postprocedural hypopituitarism (E89.3)
Add short stature due to endocrine disorder (E34.3-)

E34 Other endocrine disorders

Excludes1: pseudohypoparathyroidism (E20.1)

New subcategory E34.3 Short stature due to endocrine disorder

Delete ~~Constitutional short stature~~
Delete ~~Laron-type short stature~~
Add **Excludes1: short stature (child) (R62.52)**

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

New code E34.3**1** Constitutional short stature

Add **Constitutional delay of growth, puberty, or maturation**

New code E34.3**2** Primary insulin-like growth factor-1 (IGF-1) deficiency
Add Acid-labile subunit gene (*IGFALS*) defect
Add Growth hormone gene 1 (*GH*1) defect with growth hormone neutralizing antibodies
Add Growth hormone insensitivity syndrome (GHIS)
Add Insulin-like growth factor 1 gene (*IGF1*) defect
Add Laron type short statue
Add Severe primary insulin-like growth factor-1 deficiency (SPIGFD)
Add Signal transducer and activator of transcription 5B gene (*STAT5b*) defect

New code E34.3**3** Insulin-like growth factor-1 (IGF-1) resistance

Add		Genetic syndrome with resistance to insulin-like growth factor-1
Add		Insulin-like growth factor-1 receptor (*IGF-1R*) defect
Add		Post-insulin-like growth factor-1 receptor signaling defect
New code	**E34.34**	**Other genetic causes of short stature**
New code	E34.39	Other short stature due to endocrine disorder

Social Determinants of Health

Over the last decades growing literature has clarified and further identified the social determinates of heath and the impact on health costs. This has sparked initiation and dissemination of national recommendations and projects. Advances have been made to collectively gain insight into social risks and social interventions; yet the terminology used to represent these concepts lags behind.

In 2017, national experts and thought leaders gathered in Washington, D.C. and identified a three step process to address terminology needs: collate existing terminology, assess the applicability of existing terms and collaboratively fill and address gaps, and craft a path for data standards to ground this work. Out of this, the Gravity Project was initiated.

The Gravity Project, convened in 2019, is a national, public, consensus-based community charged with developing data elements, and data standards for the social determinants of health by leveraging the insights of subject matter experts and key stakeholders across the medical and social care community (patients, providers, payers, community-based organizations, vendors, and government). The Project's terminology recommendations span all U.S. applicable coding systems: ICD-10-CM, SNOMED CT, LOINC, and CPTÒ/HCPCS when appropriate. (For review of the Gravity Project's process, principles, members, and full deliverables, please follow the link in "resources" below.)

In order to frame its work, the Gravity Project conceptualizes concentric rings of determinants. At the center are concerns driven by a person's own economic resources, or personal and social history. Next come risks of neighborhood resources and characteristics, including utilities, groceries, and neighborhood safety. The initial phase of Gravity's work focused principally on the risk imparted by lack of personal resources: food insecurity, homelessness, housing instability, inadequate housing, transportation insecurity, and general financial insecurity. Concerns of less than high school education and veterans were also addressed. In early 2021, the Gravity Project will focus on social connection and domains of interpersonal violence. In later 2021, the Project will focus on elements of digital equity and neighborhood/environmental factors.

This proposal is the Gravity Project's first ICD-10-CM code request submission. This proposal integrates the requests of two previous social risk submissions to the ICD-10-CM committee which was submitted by the American Medical Association/UnitedHealthcare (AMA/UHC) (multi-domain) and BlueCross BlueShield of Vermont (BCBS VT) (food insecurity).

The Gravity community has carefully considered the degree of risk associated with each domain, its subdomains, and calculations of domain severity (mild – severe) as presented in the peer reviewed literature. The reason for this is threefold.

- First, to aid on the ground workers in triaging resources to those most in need, anticipating the aim of analyzing the effects of interventions.

- Second, to anticipate the use of claims data to predict person-level risk within value based health care and risk adjustment.
- Third, to align with development and dissemination of national social risk quality metrics and Healthy People 2030 Objectives.

The Gravity community and collaboration with colleagues at the American Health Information Management Association (AHIMA) and the American Hospital Association (AHA), the Gravity Project *takes care to recommend* revisions to the classification that are *easily operationalized*. All ICD-10-CM recommendations are aligned with standardized screening questions and answers such as PRAPARE, the Accountable Health Screening Tool, or the Health Leads Screening Tools.

The Gravity Project is grounded in the criticality of having codes for the missing core domains to be able to capture the data at the highest level of specificity. Additionally, work is ongoing with SNOMED CT partners to build these concepts and further subdomains into SNOMED CT terminology.

Education (Less than a high school degree)- although current ICD-10-CM contains general concepts of literacy and underachievement there is, at present no way to distinctly represent the known risk imparted by inability to attain a high school diploma or equivalent, independent of literacy.

Homelessness- although current ICD-10-CM contains a code for homelessness, there is no distinction between sheltered and unsheltered homelessness. COVID discharge planning has given us a critical use case on why this distinction is necessary from both a treatment plan and risk perspective.

Housing Instability- there is a vast literature representing the health risks of economically driven housing instability for individuals and families. Yet, there are no specific codes to define this broad risk nor the specific risk of subtypes of housing instability that segue into homelessness.

Food insecurity- as stated in the previous VT BCBS submission, the health risks and health costs associated with food insecurity are vast. Furthermore, as evidenced by the research of the USDA, risk increases as severity of food insecurity increases. Yet, there is no specific code for food insecurity.

Inadequate drinking water supply- impact of drinking water supply and future neighborhood and environmental domains.

Transportation Insecurity- this domain represents both health risks and management complexities as systems consider transportation barriers to care.

Financial Insecurity and Material Hardship- ICD-10-CM currently has codes for low income and poverty. Financial insecurity (subjective evaluation of one's current financial situation) includes perceived inadequacy of financial resources and financial concerns including expectations regarding one's future economic situation. Material hardship is one's inability to obtain basic needs. It was determined that it is critical to define risk beyond low income and poverty thresholds.

Socioeconomic Risk Counseling- needed to represent the effort of assessing and patient centered goal setting required to address socioeconomic risks.

Non-compliance and financial hardship- to encompass the inability to follow nutritional recommendations.

Veterans- data element to capture personal history of military service.

Lastly, this submission is comprehensive, including new subcategories (example: Housing instability) and subclassifications (example: "Housing instability, housed, homelessness in past 12 months" respectively). This aligns with the peer-reviewed literature and reflects broad stakeholder requests.

Resources:

General-
• The Gravity Project- https://confluence.hl7.org/display/GRAV/The+Gravity+Project
• Arons, A., DeSilvey, S., Fichtenberg, C., & Gottlieb, L. (2019). Documenting social determinants of health-related clinical activities using standardized medical vocabularies. *JAMIA Open, 2*, 81-88. doi: https://doi.org/10.1093/jamiaopen/ooy051
• Screening Tools:
o Centers for Medicare & Medicad Services, "Accountable Health Communities Health Related Social Needs Screenign Tool"- https://innovation.cms.gov/files/worksheets/ahcm-screeningtool.pdf
o National Association of Community Health Centers, "Protocol for Responding and Assessing Patients' Assets, Risks, and Experiences (PRAPARE)"- https://www.nachc.org/research-and-data/prapare/
o Health Leads, "Health Leads Screening Toolkit" - https://healthleadsusa.org/resources/the-health-leads-screening-toolkit/
Education-
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Homelessness-
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Syed ST, Gerber BS, Sharp LK. Traveling towards disease: transportation barriers to health care access. *J Community Health*. 2013;38(5):976-993. doi:10.1007/s10900-013-9681-1
• AHA "Social Determinants of Health Series: Transportation and the Role of Hospitals"
Financial Strain-
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Material Hardship-
• Urban Institute, "Material Hardship among Nonelderly Adults and their Families in 2017: Implications for the safety Net," Retrieved from: https://www.urban.org/sites/default/files/publication/98918/material_hardship_among_nonelderly_adults_and_their_families_in_2017.pdf
• ASPE (2004) "Measures of Material Hardship" Retrieved from" https://aspe.hhs.gov/report/measures-material-hardship-final-report
Veterans-
• Veterans Choice Program- https://www.va.gov/health/newsfeatures/2017/july/things-toknow-about-the-veteran-choice-program.asp

TABULAR MODIFICATIONS

Z55 Problems related to education and literacy

Excludes1: disorder of psychological development (F80-F89)

New code Z55.5 Less than a high school diploma
Add No general equivalence degree (GED)

New category Z58 Problems related to physical environment
Excludes:2 occupational exposure (Z57.-)
New code Z58.6 Inadequate drinking-water supply
Add Lack of safe drinking water
Add ` Excludes2: deprivation of water (T73.1)

Z59 Problems related to housing and economic circumstances
Excludes2: problems related to upbringing (Z62.-)

New subcategory Z59.0 Homelessness

New code Z59.00 Homelessness unspecified

New code Z59.01 Sheltered homelessness

Add Doubled up

Add Living in a shelter such as: motel, temporary or transitional living situation, scattered site housing

New code Z59.02 Unsheltered homelessness

Add Residing in place not meant for human habitation such as: cars, parks, sidewalk, abandoned buildings

Add Residing on the street

Revise Z59.4 Lack of adequate food ~~and safe drinking water~~

Delete ~~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

Add Inadequate food

Add Lack of food

New code Z59.42 Food insecurity

Z59.8 Other problems related to housing and economic circumstances
Foreclosure on loan
Isolated dwelling
Problems with creditors

New sub subcategory Z59.81 Housing instability, housed

Add Past due on rent or mortgage

Add Unwanted multiple moves in the last 12 months

New code Z59.811 Housing instability, housed, with risk of homelessness

Add Imminent risk of homelessness

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

New code Z59.819 Housing instability, housed unspecified

New code Z59.82 Transportation insecurity
Add Excessive transportation time
Add Inaccessible transportation
Add Inadequate transportation
Add Lack of transportation
Add Unaffordable transportation
Add Unreliable transportation
Add Unsafe transportation

New code Z59.86 Financial insecurity, not elsewhere classified
Add Bankruptcy
Add Burdensome debt
Add Economic strain
Add Financial strain
Add Medical cost burden
Add Money problems
Add Running out of money
Add Unable to make ends meet
Add Excludes2: material hardship, not elsewhere classified (Z59.87)

New code Z59.87 Material hardship, not elsewhere classified
Add Material deprivation
Add Unable to obtain adequate clothing
Add Unable to obtain adequate utilities
Add Unable to obtain adequate childcare
Add Unable to obtain basic needs
Add Excludes2: financial insecurity, not elsewhere classified (Z59.86)

New code Z59.89 Other problems related to housing and economic circumstances
Add Isolated dwelling

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

Z71.8 Other specified counseling
New code Z71.88 Encounter for counseling for socioeconomic factors

Z91 Personal risk factors, not elsewhere classified
Z91.1 Patient’s noncompliance with medical treatment and regimen

New sub subcategory	Z91.11 Patient's noncompliance with dietary regimen
New code	Z91.110 Patient's noncompliance with dietary regimen due to financial hardship
New code	Z91.118 Patient's noncompliance with dietary regimen for other reason
New code	Z91.119 Patient's noncompliance with dietary regimen due to unspecified reason
New sub subcategory	Z91.19 Patient's noncompliance with other medical treatment and regimen nonadherence to medical treatment
New code	Z91.190 Patient's noncompliance with other medical treatment and regimen due to financial hardship
New code	Z91.198 Patient's noncompliance with other medical treatment and regimen for other reason
New code	Z91.199 Patient's noncompliance with other medical treatment and regimen due to unspecified reason
	Z91.8 Other specified personal risk factors, not elsewhere classified
New code	Z91.85 Personal history of military service
Add	Excludes2: Personal history of military deployment (Z91.82)

Substance Use Unspecified in Remission

In May 2013, the Fifth Edition of Diagnostic and Statistical Manual of Mental Disorders (DSM-5) was released by the American Psychiatric Association (APA). The DSM-5 provides substance use diagnostic criteria for ten different substances, including alcohol. The DSM-5 further categorizes the clinical diagnoses in terms of the current severity of the substance use disorder (mild, moderate or severe which correspond to abuse or dependence in ICD-10-CM) or, if not currently using the substance at a disordered level, whether the substance use disorder is in partial or full remission.

ICD-10-CM currently provides codes for differentiating levels of severity (e.g., F10.1 Alcohol abuse vs. Alcohol Dependence vs. F10.9 Alcohol use unspecified when the severity of use is not known) as well as codes for differentiating between current use and prior use but currently in remission (e.g., F10.10 Alcohol abuse uncomplicated vs. F10.11 Alcohol abuse in remission, and F10.20 Alcohol dependence, uncomplicated vs. F10.21 Alcohol dependence in remission).

At this time, the ICD-10-CM code set does not include a code for substance use, unspecified, in remission; one must know if a patient was most recently a mild, moderate, or severe user (abuse or dependent) to code the current remission status. Consequently, cases in which the patient is known to have been previously diagnosed with a substance use disorder and whose pattern of substance use currently meets the criteria for remission status, yet the severity of the substance use before achieving remission status is not known, cannot be coded.

It is the request of the submitter to create new ICD-10-CM codes for “unspecified use in remission” for the reporting of current remission status when previous severity is not known.

Moreover, in the course of preparing this submission, it was discovered that F10.90 (Alcohol use, unspecified, uncomplicated) which would be used in cases where the alcohol use pattern is unspecified, but it is known that the use pattern is not complicated by an alcohol-induced disorder such as alcohol-induced mood disorder. This contrasts with the existing F10.99 Alcohol use unspecified with unspecified alcohol-induced disorder, in which both the pattern of alcohol use and the possible presence of an alcohol-induced disorder are unspecified. Such a code exists for the other instances of F1x.90 (i.e., other drug classes). Given that alcohol is no different from the other substance classes with respect to these unspecified categories, the omission of F10.90 alcohol use, unspecified, uncomplicated is almost certainly an oversight and thus it is recommended that F10.90 also be added to ICD-10-CM.

This proposal is being submitted by a representative of the Kaiser Permanente Federation and has been reviewed and supported by the American Psychiatric Association.

TABULAR MODIFICATIONS

F10 Alcohol related disorders
F10.9 Alcohol use, unspecified
New code F10.90 Alcohol use, unspecified, uncomplicated
New code F10.91 Alcohol use, unspecified, in remission

F11 Opioid related disorders
F11.9 Opioid use, unspecified
New code F11.91 Opioid use, unspecified, in remission

F12 Cannabis related disorders
F12.9 Cannabis use, unspecified
New code F12.91 Cannabis use, unspecified, in remission

F13 Sedative, hypnotic, or anxiolytic related disorders
F13.9 Sedative, hypnotic or anxiolytic use, unspecified
New code F13.91 Sedative, hypnotic or anxiolytic use, unspecified, in remission

F14 Cocaine related disorders
F14.9 Cocaine use, unspecified
New code F14.91 Cocaine use, unspecified, in remission

F15 Other stimulant related disorders
F15.9 Other stimulant use, unspecified
New code F15.91 Other stimulant use, unspecified, in remission

F16 Hallucinogen related disorder
F16.9 Hallucinogen use, unspecified
New code F16.91 Hallucinogen use, unspecified, in remission

F18 Inhalant related disorders
F18.9 Inhalant use, unspecified
New code F18.91 Inhalant use, unspecified, in remission

F19 Other psychoactive substance related disorders
F19.9 Other psychoactive substance use, unspecified
New code F19.91 Other psychoactive substance use, unspecified, in remission

Torsades de Pointes

Torsades de pointes is a form of polymorphic ventricular tachycardia. It can be triggered by certain medications in susceptible individuals, and it can be fatal. Thus, identifying persons who might be at risk of torsades de pointes and managing their medications to reduce risk is a key medication safety initiative in many health care institutions. A proposal to create a specific ICD-10-CM code for torsades de pointes has been received from First Databank, a company that provides clinical decision support through electronic health records utilizing ICD-10-CM codes as diagnoses and problem list terms. It is thought that this will support initiatives identifying and managing patients at risk of torsade de pointes through proper medication use and safety.

Torsades de pointes can cause symptoms of palpitations, dizziness, and syncope, which are usually recurrent. It may be diagnosed based on EKG, where it has a distinct appearance, with the ventricular beats changing shape from one beat to the next (thus polymorphic). Sometimes longer term Holter monitoring may be necessary. Torsades is associated with long QT syndrome, which may be congenital, or acquired. The most common acquired causes are related to taking certain medications and related to electrolyte abnormalities.

It should be noted that torsades de pointes is different from other types of ventricular tachycardias. The current ICD-10-CM classification of torsades at code I47.2, Ventricular tachycardia, does not provide sufficient specificity, as it is grouped with other forms of ventricular tachycardia. It is anticipated that a new code for torsade de pointes has the potential to benefit many aspects of the healthcare industry, including research, reporting, and risk reduction strategies for drug-induced torsade de pointes. It will improve clarity and be of utility for health care providers.

References:

Dave J, Bessette MJ, Setnik G, et al. Torsade de Pointes. Medscape Cardiology. Updated: January 31, 2017. Retrieved from https://emedicine.medscape.com/article/1950863-overview

Schwartz PJ, Woosley RL. Predicting the Unpredictable: Drug-Induced QT Prolongation and Torsades de Pointes. JACC 67 (13), 1639-50. April 2016. Retrieved from https://www.onlinejacc.org/content/accj/67/13/1639.full.pdf

TABULAR MODIFICATIONS

	I47	Paroxysmal tachycardia	
		I47.2	Ventricular tachycardia
New code			I47.20 Ventricular tachycardia, unspecified
New code			I47.21 Torsades de pointes
Add			Code also, if applicable, long QT syndrome (I45.81)
New code			I47.29 Other ventricular tachycardia

Von Willebrand Disease Types

The American Society of Hematology (ASH) has proposed to establish new ICD-10-CM diagnosis codes for types of von Willebrand disease (VWD). ASH represents more than 18,000 clinicians and scientists worldwide who are committed to the study and treatment of blood and blood-related diseases.

Von Willebrand disease is the most common inherited bleeding disorder, in which the blood does not clot properly, with wide variability in clinical phenotype. According to the Centers for Disease Control and Prevention, about 3.2 million (or about 1 in every 100) people in the US have the disease.[1,2] More recent epidemiologic studies reference a prevalence of 1 in 1000.[3] People with VWD either have a low level of von Willebrand factor (VWF), a protein that helps the blood to clot, or the VWF protein does not work the way it should. Although VWD occurs among men and women equally, women are more likely to notice the symptoms because of heavy or abnormal bleeding during their menstrual periods and after childbirth.[1]

In 2006, the International Society of Thrombosis and Haemostasias (ISTH) classified VWD into six categories or subtypes, based on the difference in clinical features and therapeutic requirements. Recently, ASH in partnership with ISTH, the National Hemophilia Foundation (NHF) and the World Federation of Hemophilia (WFH), has developed clinical practice guidelines for the diagnosis and management of VWD. The guidelines were published in *Blood Advances* in Jan. 2021.[6] With these guidelines now available, it is critically important to update the ICD-10-CM classification for VWD to allow for the adoption of the guideline recommendations and to improve best practices for clinical care.

Currently, all the types of von Willebrand disease are coded to one ICD-10-CM code, D68.0 (Von Willebrand's disease). According to ASH, this makes making it difficult to accurately document, track, and in turn, appropriately treat the different subtypes of VWD. For this reason, ASH has requested the addition of new ICD-10-CM diagnosis codes for VWD to better track the disease and its subtypes.

Type 1 von Willebrand disease (VWD) is characterized by decreased levels (qualitative deficiency) of von Willebrand factor (VWF). The VWF that is made is functionally normal; however, lower circulating levels lead to an increased risk of bleeding. Options for treatment including VWF concentrate, desmopressin to stimulate release of stored VWF, and antifibrinolytic therapy.

Type 1C von Willebrand disease (VWD) is characterized by increased clearance of VWF, leading to decreased levels. As is the case with other type 1 VWD, the circulating VWF in type 1C is functionally normal, however the protein is degraded quickly and not available to participate in hemostasis, leading to increased risk of bleeding. Options for treatment include VWF concentrate in

conjunction with antifibrinolytic therapy. While desmopressin will lead to a transient release of stored endogenous VWF, the effect is transient with quick return to baseline levels and increased risk of bleeding if not maintained in high risk situations such as surgery.

All forms of type 2 VWD are characterized by functional defects in VWF with subtyping based on the specific functional defect. Treatment may involve desmopressin for most subtypes (except type 2B) along with VWF concentrate and adjunctive therapy with antifibrinolytics.

Type 2A von Willebrand disease (VWD) is characterized by abnormal platelet-dependent VWF function with loss of the most hemostatically active high-molecular weight multimers of VWF.

Type 2B von Willebrand disease (VWD) is characterized by abnormal function of VWF due to a gain of function mutation that increases binding of VWF to platelet glycoprotein 1b-alpha, often leading to thrombocytopenia. Desmopressin is contraindicated due to paradoxically worsened bleeding due to increased thrombocytopenia.

Type 2M von Willebrand disease (VWD) is characterized by abnormal platelet-dependent VWF function, however multimers are preserved, a major difference between type 2A and type 2M.

Type 2N von Willebrand disease (VWD) is characterized by abnormal factor VIII binding by VWF, leading to decreased factor VIII levels and varying degree of bleeding similar to hemophilia A.

Type 3 von Willebrand disease (VWD) is a qualitative form of VWD with near complete absence of circulating VWF. This is the most severe subtype, requiring use of VWF concentrate as desmopressin is not effective.[4,5,6,7]

Acquired von Willebrand disease syndrome (AVWS) is a deficiency in the amount or function of von Willebrand factor (VWF) that is due to acquired rather than inherited causes. Examples of causes for AVWS include shearing and subsequent degradation of VWF across stenotic heart valves or through mechanical circulatory support circuits such as left-ventricular assist devices or via extracorporeal membrane oxygenation. AVWS may arise due to autoantibody formation such as that seen in immune dysregulation disorders or VWF may be directly adsorbed onto malignant cells as observed in patients with Wilms tumors or Waldenstrom macroglobulinemia. Treatment consists of supportive therapy with VWF concentrate, desmopressin, and/or antifibrinolytic therapy along with correction of the underlying cause (e.g. valve replacement therapy or immunosuppression).[8,9]
Platelet-type von Willebrand disease is due to a functional defect in the platelet receptor for von Willebrand factor. Often misdiagnosed as type 2B von Willebrand disease, treatment consists of platelet transfusions in addition to standard VWD therapies such as VWF concentrate or antifibrinolytic therapy.[10]

This proposal is based on the original ASH request, but differs in proposing to combine type 1C VWD with other type 1 VWD, and also in proposing to include platelet-type von Willebrand disease within another VWD code. ASH has recommended the deletion of several terms currently listed under D68.0, which are no longer used in clinical practice. It is proposed that these and all index entries related to von Willebrand disease be directed to the entry at Disease, von Willebrand, with the types to be identified as subentries there (not shown). ASH has also proposed to add the term "Low von Willebrand factor" as an inclusion term for code R79.1, Abnormal Coagulation Profile. In addition, it has been recommended the apostrophe "s" be deleted (consistent with WHO ICD-10 updates), and also that the "v" in von Willebrand be made lowercase (consistent with the medical literature and usual practice).

References

1 Centers for Disease Control and Prevention. October 2020. What is von Willebrand Disease? November 23, 2020. https://www.cdc.gov/ncbddd/vwd/facts.html

2 Rodeghiero, F, Castaman, G, Dini, E. Epidemiological Investigation of the Prevalence of von Willebrand's Disease. Blood, Vol 69, No 2, 1987: pp 454-459.

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4 Sadler JE, et al. Update on the pathophysiology and classification of von Willebrand disease: a report of the Subcommittee on von Willebrand Factor. J Thromb Haemost. 2006 Oct;4(10):2103-14.

5 Ng C, Motto DG, Di Paola J. Diagnostic approach to von Willebrand disease. Blood. 2015 Mar 26;125(13):2029-37.

6 James P, et al. ASH ISTH NHF WFH 2021 Guidelines on the Diagnosis of von Willebrand Disease. Blood Adv. 2021 Jan 12;5(1):280-300. https://doi.org/10.1182/bloodadvances.2020003265.

7 Connell N and Flood VH, et al. ASH ISTH NHF WFH 2020 Guidelines on the Management of von Willebrand Disease. Blood Adv. In Press 2020.

8 Franchini M, Mannucci PM. The acquired von Willebrand syndrome focused for hematologists. Haematologica. 2020.

9 Nascimbene, A. et al. Acquired von Willebrand syndrome associated with left ventricular assist device. Blood, 23 June 2016, Vol 127 (25).

10 Othman M, Gresele P. Guidance on the diagnosis and management of Platelet- type von Willebrand Disease: a communication from the platelet physiology subcommittee of the ISTH. J Thromb Haemost. 2020.

TABULAR MODIFICATIONS

D68 Other coagulation defects

Revise Excludes1: abnormal coagulation profile NOS (R79.1)

Delete		~~coagulation defects complicating abortion or ectopic or molar pregnancy (O00-O07, O08.1)~~
Delete		~~coagulation defects complicating pregnancy, childbirth and the puerperium (O45.0, O46.0, O67.0, O72.3)~~
Add	Excludes2:	coagulation defects complicating abortion or ectopic or molar pregnancy (O00-O07, O08.1)
Add		coagulation defects complicating pregnancy, childbirth and the puerperium (O45.0, O46.0, O67.0, O72.3)

Revise	D68.0	Von Willebrand~~'s~~ disease
Delete		~~Angiohemophilia~~
Delete		~~Factor VIII deficiency with vascular defect~~
Delete		~~Vascular hemophilia~~

New code	D68.00	Von Willebrand disease, unspecified	
New code		D68.01	Von Willebrand disease, type 1
Add			Partial quantitative deficiency of von Willebrand factor
Add			Type 1C von Willebrand disease
New subcategory	D68.02	Von Willebrand disease, type 2	
Add		Qualitative defects of von Willebrand factor	
New code		D68.020	Von Willebrand disease, type 2A
Add			Qualitative defects of von Willebrand factor with decreased platelet adhesion and selective deficiency of high-molecular-weight multimers
New code		D68.021	Von Willebrand disease, type 2B
Add			Qualitative defects of von Willebrand factor with hyper-adhesive forms

Add		Qualitative defects of von Willebrand factor with high-molecular-weight von Willebrand factor loss
Add		Qualitative defects of von Willebrand factor with increased affinity for platelet glycoprotein lb
New code		D68.022 Von Willebrand disease, type 2M
Add		Qualitative defects of von Willebrand factor with defective platelet adhesion with a normal size distribution of von Willebrand factor multimers
New code		D68.023 Von Willebrand disease, type 2N
Add		Qualitative defects of von Willebrand factor with markedly decreased affinity for factor VIII
Add		Qualitative defects of von Willebrand factor with defective von Willebrand factor to factor VIII binding
New code		D68.029 Von Willebrand disease, type 2, unspecified
Add		Qualitative defect in von Willebrand factor function, with no further subtyping
New code	D68.03	Von Willebrand disease, type 3
Add		(Near) complete absence of von Willebrand factor
Add		Total quantitative deficiency of von Willebrand factor
New code	D68.04	Acquired von Willebrand disease
Add		Acquired von Willebrand syndrome
New code	D68.09	Other von Willebrand disease
Add		Platelet-type von Willebrand disease
Add		Pseudo-von Willebrand disease
Add		Code also, if applicable, qualitative platelet defects (D69.1)

R79 Other abnormal findings of blood chemistry

R79.1 Abnormal coagulation profile

Add Low von Willebrand factor

INDEX MODIFICATIONS

Revise Angiohemophilia (A) (B) ~~D68.0~~ – see Disease, von Willebrand

Defect, defective Q89.9
- platelets, qualitative D69.1
Revise - - constitutional ~~D68.0~~ – see Disease, von Willebrand

Deficiency, deficient
- factor -see also Deficiency, coagulation
- - VIII (congenital) (functional) (hereditary) (with functional defect) D66
Revise - - - with vascular defect ~~D68.0~~ – see Disease, von Willebrand
- platelet NEC D69.1
Revise - - constitutional ~~D68.0~~ – see Disease, von Willebrand

Disease
Revise - Minot-von Willebrand-Jürgens (angiohemophilia) D68.0

Hemophilia (classical) (familial) (hereditary) D66
Revise - vascular ~~D68.0~~ – see Disease, von Willebrand

Revise Minot-von Willebrand-Jurgens disease or syndrome (angiohemophilia) ~~D68.0~~ – see Disease, von Willebrand

Revise Pseudohemophilia (Bernuth's) (hereditary) (type B) ~~D68.0~~ – see Disease, von Willebrand

Syndrome -see also Disease
Revise - von Willebrand (-Jürgen) ~~D68.0~~ – see Disease, von Willebrand
Revise - Willebrand (-Jürgens) ~~D68.0~~ – see Disease, von Willebrand

Thrombopathy (Bernard-Soulier) D69.1
Revise - constitutional ~~D68.0~~ – see Disease, von Willebrand
Revise - Willebrand-Jurgens ~~D68.0~~ – see Disease, von Willebrand

Revise Von Willebrand (-Jurgens)(-Minot) disease or syndrome ~~D68.0~~ – see Disease, von Willebrand

Revise Willebrand (-Jürgens) thrombopathy ~~D68.0~~ – see Disease, von Willebrand

ICD-10-CM TABULAR OF DISEASES - PROPOSED ADDENDA

All proposed effective October 1, 2021

D64 Other anemias

Excludes1: refractory anemia (D46.-)
refractory anemia with excess blasts in transformation [RAEB T] (C92.0-)

D64.8 Other specified anemias

D64.81 Anemia due to antineoplastic chemotherapy
Antineoplastic chemotherapy induced anemia

Revise
Excludes~~1~~2: aplastic anemia due to antineoplastic chemotherapy (D61.1)

E63 Other nutritional deficiencies

Revise
Excludes~~1~~2: dehydration (E86.0)
failure to thrive, adult (R62.7)
failure to thrive, child (R62.51)
feeding problems in newborn (P92.-)
sequelae of malnutrition and other nutritional deficiencies (E64.-)

Ischemic heart diseases (I20-I25)

Revise
Use additional code, if applicable, to identify presence of hypertension (I10-I16)

I50 Heart failure

Revise
Excludes~~1~~2: neonatal cardiac failure (P29.0)

I51 Complications and ill-defined descriptions of heart disease

Revise
Excludes~~1~~2: any condition in I51.4-I51.9 due to hypertension (I11.-)
any condition in I51.4-I51.9 due to hypertension and chronic kidney disease (I13.-)
heart disease specified as rheumatic (I00-I09)

I83 Varicose veins of lower extremities

Revise
Excludes~~1~~2: varicose veins complicating pregnancy (O22.0-)
varicose veins complicating the puerperium (O87.4~~)~~

J47 Bronchiectasis

J47.0 Bronchiectasis with acute lower respiratory infection
Bronchiectasis with acute bronchitis

Revise ~~Use additional code~~ Code also to identify the infection

J96 Respiratory failure, not elsewhere classified

Revise Excludes~~1~~2: acute respiratory distress syndrome (J80)
cardiorespiratory failure (R09.2)
newborn respiratory distress syndrome (P22.0)
postprocedural respiratory failure (J95.82-)
respiratory arrest (R09.2)
respiratory arrest of newborn (P28.81)
respiratory failure of newborn (P28.5)

M41 Scoliosis

Includes: kyphoscoliosis
Excludes1: congenital scoliosis NOS (Q67.5)
congenital scoliosis due to bony malformation (Q76.3)
postural congenital scoliosis (Q67.5)
kyphoscoliotic heart disease (I27.1)
Delete ~~postprocedural scoliosis (M96.-)~~
Add Excludes2: postprocedural scoliosis (M96.-)

N97 Female infertility

Revise Excludes~~1~~2: female infertility associated with:
hypopituitarism (E23.0)
Stein-Leventhal syndrome (E28.2)

P29 Cardiovascular disorders originating in the perinatal period (458)

Revise Excludes~~1~~2: congenital malformations of the circulatory system (Q20-Q28)

Other diseases of the urinary system (N30-N39)

Revise Excludes~~1~~2: urinary infection (complicating):
abortion or ectopic or molar pregnancy (O00-O07, O08.8)
pregnancy, childbirth and the puerperium (O23.-, O75.3, O86.2-)

R26 Abnormalities of gait and mobility

Revise

Excludes~~1~~2: ataxia NOS (R27.0)
hereditary ataxia (G11.-)
locomotor (syphilitic) ataxia (A52.11)
immobility syndrome (paraplegic) (M62.3)

R27 Other lack of coordination

Revise

Excludes~~1~~2: ataxic gait (R26.0)
hereditary ataxia (G11.-)
vertigo NOS (R42)

T44 Poisoning by, adverse effect of and underdosing of drugs primarily affecting the autonomic nervous system

T44.8 Poisoning by, adverse effect of and underdosing of centrally-acting and adrenergic-neuron-blocking agents

Revise

Excludes~~1~~2: poisoning by, adverse effect of and underdosing of clonidine (T46.5)
poisoning by, adverse effect of and underdosing of guanethidine (T46.5)

S92 Fracture of foot and toe, except ankle

Delete

~~Excludes1: traumatic amputation of ankle and foot (S98.-)~~

Excludes2: fracture of ankle (S82.-)
fracture of malleolus (S82.-)

Add

traumatic amputation of ankle and foot (S98.-)

Z31 Encounter for procreative management

Revise

Excludes~~1~~2: complications associated with artificial fertilization (N98.-)
female infertility (N97.-)
male infertility (N46.-)

Z43 Encounter for attention to artificial openings

Revise

Excludes~~1~~2: complications of external stoma (J95.0-, K94.-, N99.5-)

Z93 Artificial opening status

Revise

Excludes~~1~~2: artificial openings requiring attention or management (Z43.- complications of external stoma (J95.0-, K94.-, N99.5-)