



## ICD-10 Coordination and Maintenance Committee Meeting

### Diagnosis Agenda

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Donna Pickett, MPH, RHIA

Co-Chair, ICD-10 Coordination and Maintenance Committee

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**Instructions to join the virtual meeting on September 14-15, 2021 are as follows:**

**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. Meeting details for each day are as follows.
- Day 1: September 14, 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: September 15, 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.

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/join/1611807597?pwd=dnB1TWxkRW1HbDRWUytzTURrcUZAQT09>  
Passcode: 649118

**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)

2. Enter the webinar ID: 161 180 7597

\*If dialing in from outside of the U.S., visit <https://cms.zoomgov.com/u/abTTQHnQHa> for a list of Zoom International Dial-in Numbers.

**Option 3:** To join this Zoom Webinar conference from an H.323/SIP room system:

1. From your room system, dial 161.199.138.10 (US West) or 161.199.136.10 (US East)

2. Enter the webinar ID: 161 180 7597 Passcode: 649118

SIP: 1611807597 @sip.zoomgov.com Passcode: 649118

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 CDC's web page located at: [ICD - ICD-10-CM - Coordination and Maintenance Committee \(cdc.gov\)](https://www.cdc.gov/ncidod/dlnd/icd10cm/).

Remaining questions may be submitted via the ICD-10-CM mailbox at [nchsicd10cm@cdc.gov](mailto:nchsicd10cm@cdc.gov)

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**ICD-10 TIMELINE**

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

- September 14-15, 2021      The September 2021 ICD-10 Coordination and Maintenance Committee Meeting is fully virtual by zoom and dial-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](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 diseases or 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/>

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- 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.**
- December 3, 2021**      **Deadline for requestors: Those members of the public requesting that topics be discussed at the March 8-9, 2022 ICD-10 Coordination and Maintenance Committee Meeting must have their requests submitted to CMS for procedures and to NCHS for diagnoses by this date.**
- Requestors should indicate if they are submitting their code request for consideration for an October 1, 2022 implementation date, or an April 1, 2023 implementation date.**
- January 2022**              The ICD-10 Coordination and Maintenance Committee will make efforts to accommodate the requested implementation date for each request submitted, however, the Committee will determine which requests will be presented for consideration for an October 1, 2022 implementation date or an April 1, 2023 implementation date.
- Federal Register notice for the March 8-9, 2022 ICD-10 Coordination and Maintenance Committee Meeting will be published. This will include the tentative agenda.
- February 2022**              Tentative agenda for the Procedure portion of the March 8, 2022 ICD-10 Coordination and Maintenance Committee Meeting posted on CMS webpage as follows:  
<https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html>
- Tentative agenda for the Diagnosis portion of the March 9, 2022 ICD-10 Coordination and Maintenance Committee Meeting posted on NCHS homepage as follows:  
[https://www.cdc.gov/nchs/icd/icd10cm\\_maintenance.htm](https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm)
- February 1, 2022**              **On-line registration opens for the March 8-9, 2022 ICD-10 Coordination and Maintenance Committee Meeting at:**  
<https://www.cms.gov/events>  
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

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attendance for continuing education purposes. The on-line registration will be available through March 1, 2022.

March 8-9, 2022

ICD-10 Coordination and Maintenance Committee Meeting.

March 2022

Recordings and slide presentations of the March 8-9, 2022 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](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, 2022

Any new ICD-10 codes to capture new diseases or technology will be implemented on April 1, 2022.

**April 8, 2022**

**Deadline for receipt of public comments on proposed new procedure codes and revisions discussed at the March 8-9, 2022 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2022.**

April 2022

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 2023 ICD-10-CM diagnosis and ICD-10-PCS procedure codes finalized to date. It will also include proposed revisions to the MS-DRG system based on ICD-10-CM/PCS codes on which the public may comment. The proposed rule can be accessed at:

<https://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/AcuteInpatientPPS/index.html?redirect=/AcuteInpatientPPS/IPPS/list.asp>

May/June 2022

Final addendum posted on web pages as follows:

**Diagnosis addendum -**  
<https://www.cdc.gov/nchs/icd/icd10cm.htm>

**Procedure addendum -**  
<https://www.cms.gov/Medicare/Coding/ICD10/index.html>

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- June 10, 2022**                    **Deadline for requestors: Those members of the public requesting that topics be discussed at the September 13-14, 2022 ICD-10 Coordination and Maintenance Committee Meeting, must have their requests submitted to CMS for procedures and NCHS for diagnoses.**
- July 2022                        Federal Register notice for the September 13-14, 2022 ICD-10 Coordination and Maintenance Committee Meeting will be published. This will include the tentative agenda.
- August 1, 2022                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, 2022.  
This rule can be accessed at:  
<https://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/AcuteInpatientPPS/index.html>
- August 2022                    Tentative agenda for the Procedure portion of the September 13, 2022 ICD-10 Coordination and Maintenance Committee Meeting will be posted on the CMS webpage at –  
<https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html>
- Tentative agenda for the Diagnosis portion of the September 14, 2022 ICD-10 Coordination and Maintenance Committee Meeting will be posted on the NCHS webpage at -  
[https://www.cdc.gov/nchs/icd/icd10cm\\_maintenance.htm](https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm)
- August 12, 2022**                **On-line registration opens for the September 13-14, 2022 ICD-10 Coordination and Maintenance Committee Meeting at:**  
<https://www.cms.gov/events>  
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 12, 2022.
- September 13-14, 2022        The September 2022 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.

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September 2022 Recordings and slide presentations of the September 13-14, 2022 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](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, 2022 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 14, 2022 **Deadline for receipt of public comments on proposed new codes discussed at the September 13-14, 2021 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on April 1, 2023.**

November 2022 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, 2023 will be posted on the following websites:

<https://www.cdc.gov/nchs/icd/icd10cm.htm>

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

November 15, 2022 **Deadline for receipt of public comments on proposed new codes and revisions discussed at the September 13-14, 2021 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2023.**

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**Contact Information**

Mailing address:

National Center for Health Statistics  
ICD-9-CM Coordination and Maintenance Committee  
3311 Toledo Road  
Hyattsville, Maryland 20782

**During the COVID-19 Pandemic, fax and regular mail is not currently being monitored and all communication should be sent via e-mail.**

**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](mailto:nchsicd10CM@cdc.gov)**

Donna Pickett	(301) 458-4434
David Berglund	(301) 458-4095
Cheryl Bullock	(301) 458-4297
Shannon McConnell-Lamptey	(301) 458-4612
Traci Ramirez	(301) 458-4454
Herman Thurman	(301) 458-4282

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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 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, 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.

## **Activated Phosphoinositide 3-kinase Delta Syndrome (APDS)**

Activated Phosphoinositide 3-kinase Delta Syndrome (APDS) is a primary immune regulatory disorder (PIRD) recently identified in 2013. A proposal for a specific ICD-10-CM code for APDS has been received from Pharming Healthcare, Inc. This proposal has support from the American Academy of Allergy, Asthma & Immunology; Allergy & Asthma Network; American College of Allergy, Asthma & Immunology; Clinical Immunology Society; Immune Deficiency Foundation; and Jeffrey Modell Foundation.

APDS is a primary immune regulatory disorder (PIRD). It is clinically defined as a combined immunodeficiency, and impairs the immune system, such that individuals with this condition typically have low numbers of white blood cells, particularly certain types of B cells and T cells.<sup>1,2</sup> APDS is sometimes also called p110d-activating mutation causing senescent T cells, lymphadenopathy, and immunodeficiency (PASLI) disease. It results from an autosomal dominant mutation in either the catalytic p110 $\delta$  (PIK3CD) or regulatory p85 $\alpha$  (PIK3R1) subunits, resulting in gain-of-function in the phosphoinositide 3-kinase  $\delta$  (PI3K $\delta$ ) pathway.<sup>2</sup> However, patients with APDS as a PIRD can also develop autoimmune and inflammatory complications, and may thus in some cases need treatment with immunosuppressive therapy.<sup>2</sup>

Due to the effect on B and T cells, patients with APDS may develop immunodeficiencies and related problems, including but not limited to recurrent respiratory tract infections, bronchiectasis, herpes virus infections, autoimmunity, lymphoma, and neurodevelopment delay. Symptoms are highly variable, even within families carrying the same mutation. This has led to varied APDS treatments across patients.<sup>3</sup> Due to these multiple immunological complications, diagnosis of APDS can be difficult.

APDS typically manifests during early childhood. Recurrent respiratory infections are usually the first manifestation of APDS (occurring in infancy or childhood from 0-10 years of age), and bronchiectasis and autoimmunity occur later in childhood (4-6 years of age).<sup>1</sup> APDS is a rare disease; the submitter (Pharming) estimates incidence rates of a minimum of 1-2 cases per million worldwide, although there is some uncertainty in the prevalence and the condition may be underdiagnosed, as is the case with most rare diseases.<sup>5,6</sup>

Immunologists typically diagnose and treat APDS, although many different specialties may be involved in the care and management of patients with APDS. Over 40 clinically available tests can aid in diagnosis.<sup>4</sup> There are now commercially available genetic tests that can identify mutations in the genes (PIK3CD and PIK3R1) leading to a diagnosis of APDS 1 and APDS 2 respectively. Due to great clinical heterogeneity of APDS, treatment varies by immunologists by prescribing supportive therapies, such as prophylactic antibiotic therapy, immunoglobulin replacement therapy (IRT), and hematopoietic stem cell transplantation (HSCT), as well as sirolimus (rapamycin).<sup>5,7</sup>

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Pharming has a licensing agreement for leniolisib, in late stage development for the treatment of APDS.<sup>5</sup> Leniolisib is an oral, small molecule with immunomodulating and potentially antineoplastic activities, and is expected to treat overactivated PI3K $\delta$ , the root-cause of APDS.<sup>7</sup> While leniolisib is not yet Food and Drug Administration (FDA)-approved, and there is no current specific FDA-approved treatment for APDS, it is hoped that it will be approved, and reach the market in Q4 2022.<sup>8</sup>

Creation of a specific code for APDS will support appropriate research, and tracking of the disease, as well as alleviating any potential confusion among clinicians or others on the specific diagnosis of APDS, and ultimately enabling specific patients with APDS to receive much needed therapy. Because APDS is classified as a combined immunodeficiency, a specific code for APDS is proposed within category D81, Combined immunodeficiencies, at the code D81.82.

#### References

1. Coulter TI, Chandra A, Bacon CM, et al. Clinical spectrum and features of activated phosphoinositide 3-kinase  $\delta$  syndrome: A large patient cohort study. *J Allergy Clin Immunol.* 2017;139(2):597-606.e4. <https://doi.org/10.1016/j.jaci.2016.06.021>.
2. Coulter TI, Cant AJ. The Treatment of Activated PI3K $\delta$  Syndrome. *Front Immunol.* 2018;9:2043. Published 2018 Sep 7. <https://doi.org/10.3389/fimmu.2018.02043>.
3. Angulo I, Vadas O, Garçon F, et al. Phosphoinositide 3-kinase  $\delta$  gene mutation predisposes to respiratory infection and airway damage. *Science.* 2013;342(6160):866–871. <https://doi.org/10.1126/science.1243292>.
4. National Center for Biotechnology Information (NCBI), National Institutes of Health (NIH). Immunodeficiency 14. Genetic Testing Registry, available at: <https://www.ncbi.nlm.nih.gov/gtr/conditions/C3714976/>.
5. Pharming Healthcare, Inc. Pharming acquires exclusive license to CDZ173, a late stage drug for the treatment of APDS (Aug. 13, 2019). Available at: <https://www.pharming.com/sites/default/files/imce/Press%20releases/PR%20CDZ173%2013%20August%202019.pdf>.
6. National Library of Medicine (NLM), National Institutes of Health (NIH). Activated PI3K-delta syndrome. MedlinePlus. Available from <https://medlineplus.gov/genetics/condition/activated-pi3k-delta-syndrome/>.
7. Rao V, Koneti, Webster S, et al. Effective “activated PI3K $\delta$  syndrome”-targeted therapy with the PI3K $\delta$  inhibitor leniolisib. *Blood.* 2017: 130(21). <https://doi.org/10.1182/blood-2017-08-801191>
8. Pharming Healthcare, Inc., Pharming Group reports financial results for the first half of 2021 (Aug. 5, 2021), available at: <https://www.pharming.com/sites/default/files/imce/Press%20releases/H1%20PR%202021%2005AUG21%20PDF%20final...0.pdf>.

**TABULAR MODIFICATIONS**

Note that a different proposal is anticipated to also affect expansion of D81.8.

D81 Combined immunodeficiencies

D81.8 Other combined immunodeficiencies

New code	D81.82	Activated Phosphoinositide 3-kinase Delta Syndrome [APDS]
Add		p110d-activating mutation causing senescent T cells, lymphadenopathy, and immunodeficiency [PASLI] disease

## Angioectasia of small intestine

Angioectasia is an abnormal, tortuous, dilated small blood vessel in the mucosal and submucosal layers of the gastrointestinal (GI) tract. It is a common vascular abnormality in the GI tract. Patients with angioectasia may be asymptomatic or present with occult blood in stool, active GI bleeding (melena/hematochezia) and iron deficiency anemia. Small bowel angiodysplasia is a common cause of small bowel bleeding and can be diagnosed by EGD, enteroscopy (push or double balloon) and small bowel capsule endoscopy. Asymptomatic angioectasia is occasionally diagnosed as an incidental finding during colonoscopy or endoscopy performed for other reasons. It is a common cause of small bowel bleeds.

NCHS proposes the following new codes for clinical completeness to separately identify angioectasia of the small intestine. The American Gastroenterological Association (AGA) has reviewed and supports this proposal.

### TABULAR MODIFICATIONS

	K55	Vascular disorders of intestine
New subcategory Add	K55.A	Angioectasia of small intestine Angiodysplasia of small intestine
New code Add	K55.A0	Angioectasia of small intestine without bleeding Angioectasia of small intestine NOS
New code	K55.A1	Angioectasia of small intestine with bleeding

## 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.

A proposal was presented at the March 2021 Coordination and Maintenance meeting. In response to public comments, a revised proposal is being submitted for reconsideration. Changes are noted in **bold**.

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 a distinct and separate condition from newborn sleep apnea and typically first documented diagnosis in a premature baby who is apneic. This is different from the “unspecified” apnea in that for most premature babies, this is a specific diagnosis used when a premature baby suffers from (non-sleep) apnea as this is common until their lungs more fully mature. Testing for more specific apnea tends to not take place for a premature baby until the physician feels it is not resolving on its own and identification of the “type” is important. Unspecified newborn apnea is more relevant for a near full or full term baby whereby apnea spells are discovered, and further testing has yet to be conducted.

While apnea and 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

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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

Delete Add	P28	Other respiratory conditions originating in the perinatal period <del>Excludes1: congenital malformations of the respiratory system (Q30-Q34)</del> Code also, if applicable, congenital malformations of the respiratory system (Q30-Q34)
New sub-category Delete Delete Delete Add	P28.3	Primary sleep apnea of newborn <del>Central sleep apnea of newborn</del> <del>Obstructive sleep apnea of newborn</del> <del>Sleep apnea of newborn NOS</del> <b>Excludes2: other apnea of newborn (P28.4-)</b>

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New code	P28.30 <b>Primary</b> sleep apnea of newborn, unspecified
Add	Transient oxygen desaturation spells of newborn <b>during</b> sleep
New code	P28.31 <b>Primary</b> central sleep apnea of newborn
New code	P28.32 <b>Primary</b> obstructive sleep apnea of newborn
New code	P28.33 <b>Primary</b> mixed sleep apnea of the newborn
New code	P28.39 Other <b>primary</b> sleep apnea of newborn
New subcategory	P28.4 Other apnea of newborn
Delete	<del>Apnea of prematurity</del>
Delete	<del>Obstructive apnea of newborn</del>
Delete	<del>Excludes1: obstructive sleep apnea of newborn (P28.3)</del>
<b>Add</b>	<b>Excludes2: primary sleep apnea of newborn (P28.3-)</b>
New code	<b>P28.40 Unspecified apnea of newborn</b>
Add	Apnea of newborn, NOS
Add	Transient oxygen desaturation spells of newborn
New code	<b>P28.41 Central neonatal apnea of newborn</b>
New code	<b>P28.42 Obstructive apnea of newborn</b>
New code	<b>P28.43 Mixed neonatal apnea of newborn</b>
New code	P28.49 Other apnea of newborn
<b>Add</b>	<b>Apnea of Prematurity</b>
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

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Add	Excludes1: apnea NOS (R06.81)
Add	neonatal bradycardia (P29.12)
<b>Add</b>	<b>newborn apnea (P28.4-)</b>
Add	sleep apnea (G47.3-)
<b>Add</b>	<b>sleep apnea of newborn (P28.3-)</b>

Z72.8 Other problems related to lifestyle

Z72.82 Problems related to sleep

New code	Z72.823 Risk of suffocation (smothering) under another while sleeping
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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. This proposal was presented at the March 2021 Coordination and Maintenance Meeting. Based on public comments received, revisions have been made (**noted in bold**) and resubmitted for reconsideration.

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<sup>5</sup>. 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 prevalence, healthcare/service utilization, and outcomes in those living with birth defects.

The NYS BDR considers children to be eligible for surveillance if 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”<sup>1</sup>. Studies support the use of separate diagnosis codes for component defects of Q21.1<sup>1,2</sup>, highlight the inability to identify a specific defect using the ICD code<sup>2</sup>, and acknowledge that the code cannot be relied upon to identify important conditions<sup>3</sup>.

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.

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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<sup>5,6</sup>.

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<sup>1</sup>. 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.

This proposal has been reviewed and supported by the Centers for Disease Control and Prevention / National Center on Birth Defects and Developmental Disabilities/ Division of Birth Defects and Infant Disorders.

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### TABULAR MODIFICATIONS

Q21	Congenital malformations of cardiac septa Excludes 1: acquired cardiac septal defect (I51.0)
	Q21.0 Ventricular septal defect Roger's disease
New Subcategory	Q21.1 Atrial septal defect
Delete	<del>Coronary sinus defect</del>
Delete	<del>Patent or persistent foramen ovale</del>
Delete	<del>Patent or persistent ostium secundum defect (type II)</del>
Delete	<del>Patent or persistent sinus venosus defect</del>
<b>Add</b>	<b>Excludes 2: ostium primum atrial septal defect (type I) (Q21.20)</b>
<b>New Code</b>	<b>Q21.10 Atrial septal defect, unspecified</b>
<b>New Code</b>	<b>Q21.11 Atrial septal communication, type undetermined</b>
<b>Add</b>	<b>Atrial septal defect (ASD) versus patent foremen ovale (PFO)</b>
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

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New Code	Q21.14 Coronary sinus atrial septal defect
Add	Coronary sinus defect
Add	<b>Unroofed coronary sinus</b>
New Code	<b>Q21.15 Superior sinus venosus atrial septal defect</b>
	<b>Superior vena cava type atrial septal defect</b>
New code	<b>Q21.16 Inferior sinus venosus atrial septal defect</b>
Add	<b>Inferior vena cava type atrial septal defect</b>
New code	<b>Q21.17 Sinus venosus atrial septal defect, unspecified</b>
	<b>Sinus venosus defect, NOS</b>
New code	<b>Q21.19 Other specified atrial septal defect</b>
Add	<b>Common atrium</b>
Add	<b>Other specified atrial septal abnormality</b>
New Subcategory	Q21.2 Atrioventricular septal defect
Delete	<del>Common atrioventricular canal</del>
Delete	<del>Endocardial cushion defect</del>
Delete	<del>Ostium primum atrial septal defect (type I)</del>
Add	<b>Atrioventricular canal defect</b>
Add	<b>Endocardial cushion defect</b>
Add	<b>Ostium primum atrial septal defect (type I)</b>
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	Ostium primum atrial septal defect (type I) with separate atrioventricular valves
Add	Partial atrioventricular canal
Add	Partial endocardial cushion defect

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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 canal
Add	Common atrioventricular septal defect
Add	Common endocardial cushion defect
Add	Ostium primum atrial septal defect (type I) with common atrioventricular valve and a moderate or larger inlet VSD

## **Bronchiolitis Obliterans Syndrome and Bronchiolitis Obliterans**

Bronchiolitis obliterans syndrome (BOS) is a clinical syndrome characterized by airflow limitation not reversible with inhaled bronchodilators which may be associated with progressive dyspnea. It was first clearly described in early 1980s in the context of lung transplant as a rare fibrotic disorder involving terminal and respiratory bronchioles.<sup>1,2</sup> The histologic hallmark of BOS is obliterative bronchiolitis (OB), also called bronchiolitis obliterans, which consists of a fibrotic luminal obliteration of the respiratory and terminal bronchioles.

BOS is a rare disease (Orphanet code ORPHA: 1303).<sup>3</sup> Both immune-mediated and non-immune mediated factors can be responsible. In general, obliterative bronchiolitis may be associated with injury of small airways due to infections, systemic and autoimmune diseases, and certain inhaled agents, but it appears most frequently after lung transplantation as a manifestation of chronic lung allograft dysfunction (CLAD) or after allogeneic hematopoietic stem cell transplantation (alloHSCT) as the pulmonary manifestation of chronic graft-versus-host-disease (cGVHD). The incidence of BOS after lung transplantation and alloHSCT transplantation is of particular concern because it can adversely affect long-term outcomes and survival after these potentially life-saving procedures.

BOS is the most common manifestation (or phenotype) of CLAD, accounting for nearly 50% to 70% of the cases. Up to 30% of patients with CLAD develop a restrictive defect called restrictive allograft syndrome (RAS).<sup>4</sup> Based on this, the pulmonary council of the International Society for Heart and Lung Transplantation (ISHLT) classifies CLAD into four clinical sub-types: BOS, RAS, mixed and undefined, with the later being definite CLAD but with a combination of findings that does not fit into the other three defined types.<sup>4</sup> The exact incidence and prevalence of BOS after lung transplant are not known but data from the ISHLT registry suggests that nearly 50% of all lung transplant recipients are affected by BOS within 5 years and 75% within 10 years after transplantation, and it is the leading cause of death one year or more after lung transplant.<sup>5,6</sup>

In alloHSCT patients, BOS occurs as a pulmonary manifestation of cGVHD. In alloHSCT patients afflicted by cGVHD, BOS is an important contributor to morbidity and mortality.<sup>7</sup> It is the most common non-infectious pulmonary complication of alloHSCT, typically presenting after the first 100 days following transplantation, with most cases presenting between 12 to 18 months after transplantation.<sup>8</sup> The exact incidence and prevalence of BOS after alloHSCT are not known, but individual studies have reported that the prevalence of BOS in alloHSCT patients ranges from 3.4% to 10%.<sup>9</sup> It is associated with poor prognosis with a 5-year survival rate of 60%.<sup>7</sup>

A proposal to create specific codes for CLAD, including BOS, RAS, Mixed, and other, as well as BOS when due to alloHSCT, was received from Sajjad Raza, MD, PhD, MSM, Associate Director, Precision Medicine Group; with support reported from a number of international clinicians in this field. This proposal for completeness includes creation of a separate code for

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bronchiolitis obliterans, which may occur due to a number of different other causes as noted, and also certain other specific codes and index entries.

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**TABULAR MODIFICATIONS**

Revise	Chronic lower respiratory diseases ( <del>J40-J47</del> ) ( <u>J40-J4A</u> )
New	J44 Other chronic obstructive pulmonary disease
subcategory	J44.8 Other specified chronic obstructive pulmonary disease
New	J44.81 Bronchiolitis obliterans
Add	Excludes1: bronchiolitis obliterans syndrome following lung transplant (J4A.0)
Add	bronchiolitis obliterans syndrome, NOS (J4A.0)

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New code	J44.810	Bronchiolitis obliterans syndrome following hematopoietic stem cell transplant
Add		Bone marrow transplant-associated bronchiolitis obliterans (syndrome)
Add		Stem cell transplant-associated bronchiolitis obliterans (syndrome)
Add		Code first, if applicable:
Add		complication of bone marrow transplant (T86.09)
Add		complication of stem cell transplant (T86.5)
Add		Code also, if applicable, chronic graft-versus-host disease (D89.811)
New code	J44.818	Other obliterative bronchiolitis
Add		Bronchiolitis obliterans, not elsewhere classified
Add		Code also associated condition(s), if known
Add		Excludes1: Obliterative bronchiolitis (chronic) (subacute) due to inhalation of chemicals, gases, fumes and vapor (J68.41)
New code	J44.89	Other specified chronic obstructive pulmonary disease
Add		Chronic asthmatic (obstructive) bronchitis
Add		Chronic emphysematous bronchitis
	J4A	Chronic Lung Allograft Dysfunction
Add		Code first, if applicable:
Add		heart-lung transplant rejection (T86.31)
Add		lung transplant rejection (T86.810)
Add		other complications of heart-lung transplant (T86.39)
Add		other complications of lung transplant (T86.818)
New code	J4A.0	Bronchiolitis obliterans syndrome
Add		Bronchiolitis obliterans syndrome, NOS
Add		Bronchiolitis obliterans syndrome following lung transplant
Add		Transplant-associated bronchiolitis obliterans
New code	J4A.1	Restrictive allograft syndrome

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New code Add	J4A.2 Mixed chronic lung allograft dysfunction Bronchiolitis obliterans syndrome with restrictive allograft syndrome
New code Add	J4A.8 Other chronic lung allograft dysfunction Bronchiolitis obliterans syndrome with other measures of chronic lung allograft dysfunction
New code	J4A.9 Chronic lung allograft dysfunction, unspecified
	J68 Respiratory conditions due to inhalation of chemicals, gases, fumes and vapors
	J68.4 Chronic respiratory conditions due to chemicals, gases, fumes and vapors
Delete	<del>Emphysema (diffuse) (chronic) due to inhalation of chemicals, gases, fumes and vapors</del>
Delete	<del>Obliterative bronchiolitis (chronic) (subacute) due to inhalation of chemicals, gases, fumes and vapors</del>
Delete	<del>Pulmonary fibrosis (chronic) due to inhalation of chemicals, gases, fumes and vapors</del> <del>Excludes1: chronic pulmonary edema due to chemicals, gases, fumes and vapors (J68.1)</del>
New code	J68.41 Obliterative bronchiolitis (chronic) (subacute) due to inhalation of chemicals, gases, fumes and vapor
New code	J68.49 Other chronic respiratory conditions due to chemicals, gases, fumes and vapors
Add	Emphysema (diffuse) (chronic) due to inhalation of chemicals, gases, fumes and vapors
Add	Pulmonary fibrosis (chronic) due to inhalation of chemicals, gases, fumes and vapors Excludes1: chronic pulmonary edema due to chemicals, gases, fumes and vapors (J68.1)

## INDEX MODIFICATIONS

- Asthma, asthmatic (bronchial) (catarrh) (spasmodic) J45.909  
Revise - chronic obstructive J44.89  
Revise - with chronic obstructive bronchitis J44.89  
Revise - with chronic obstructive pulmonary disease J44.89
- Bronchiolitis (acute) (infective) (subacute) J21.9  
Revise - chemical (chronic) J68.41  
Revise - chronic (fibrosing) (~~obliterative~~) J44.89  
Add - - obliterative  
Revise - fibrosa obliterans ~~J44.9~~ J44.818  
Revise - obliterans ~~J42~~ (see also Bronchiolitis, obliterative) J44.818  
Add - - syndrome J4A.0  
Add - - - stem cell transplant associated J44.810  
Add - - transplant-associated – see Bronchiolitis, obliterans, syndrome  
Revise - obliterative (chronic) (subacute) (see also Bronchiolitis, obliterans) ~~J44.9~~  
J44.818  
Revise - - due to chemicals, gases, fumes or vapors (inhalation) J68.41  
Revise - - due to fumes or vapors J68.41
- Bronchitis (diffuse) (fibrinous) (hypostatic) (infective) (membranous) J40  
- asthmatic J45.9  
Revise - - chronic J44.89  
- chemical (acute) (subacute) J68.0  
Revise - - chronic J68.49  
- - due to fumes or vapors J68.0  
Revise - - - chronic J68.49  
- chronic J42  
Revise - - asthmatic (obstructive) J44.89  
Revise - - chemical (due to fumes or vapors) J68.49  
Revise - - due to chemicals, gases, fumes or vapors (inhalation) J68.49  
Revise - - emphysematous J44.89  
Revise - - obliterans ~~J44.9~~ see Bronchiolitis, obliterans  
Revise - - obstructive J44.89  
Revise - - with airways obstruction J44.89
- Revise - emphysematous (obstructive) J44.89  
Revise - obliterans (chronic) ~~J44.9~~ see Bronchiolitis, obliterans  
Revise - obstructive (chronic) (diffuse) J44.89  
Revise - with obstruction (airway) (lung) J44.89

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- Disease, diseased - see also Syndrome  
- lung J98.4  
- - obstructive (chronic) J44.9  
Revise - - - with bronchitis J44.89  
- respiratory (tract) J98.9  
- - chronic NOS J98.9  
Revise - - - due to chemicals, gases, fumes or vapors J68.49  
- - due to chemicals, gases, fumes or vapors J68.9  
Revise - - - chronic J68.49
- Dyspnea (nocturnal) (paroxysmal) R06.00  
- asthmatic (bronchial) J45.909  
- - with  
- - - bronchitis J45.909  
Revise - - - - chronic J44.89
- Obstruction, obstructed, obstructive  
- airway J98.8  
Revise - - with bronchitis (chronic) J44.89
- Pneumatocele (lung) J98.4  
Revise - tension ~~J44.9~~ J98.8
- Revise Vanishing lung J44.89

## **Caught, crushed, jammed, or pinched in or between objects**

The ICD-10-CM classification currently does not include an external cause code for a patient injured by a body part being crushed between a moving and a stationary object. Therefore, adding a unique code would contribute to the accuracy of healthcare statistics on external causes as reflected in provider clinical documentation.

An article by OSHAcademy states, “According to OSHA (Occupational Safety & Health Administration), caught-in or -between hazards are defined as: Injuries resulting from a person being squeezed, caught, crushed, pinched, or compressed between two or more objects, or between parts of an object. This includes individuals who get caught or crushed in operating equipment, between other mashing objects, between a moving and stationary object, or between two or more moving objects. Events that should be classified as caught-in or-between include being pulled into or caught in machinery and equipment (this includes strangulation as the result of clothing caught in running machinery and equipment), being compressed or crushed between rolling, sliding, or shifting objects such as semi-trailers and a dock wall, or between a truck frame and a hydraulic bed that is lowering.”

The submitter is requesting a new code to allow the ability to analyze this type of external cause. The data generated by a new code would also contribute to occupational health statistics gathered by organizations such as OSHA.

### **Reference:**

OSHAcademy. (2021). *What Are Caught-In or – Between Hazards?* Retrieved July 12, 2021, from: <https://www.oshatrain.org/courses/mods/807m1.html>

OSHA (Occupational Safety & Health Administration). (2020). *Fatality Inspection Data*. Retrieved July 12, 2021, from: <https://www.osha.gov/fatalities>

## **TABULAR MODIFICATIONS**

	W23	Caught, crushed, jammed or pinched in or between objects
New code	W23.2	Caught, crushed, jammed or pinched between a moving and stationary object

## Coma Not Elsewhere Classified

A propose creation of a new code for “Coma NEC” after the recent coding guideline change which limits Glasgow coma scale codes to traumatic brain injury. R40.20, Unspecified Coma, is the only code available for coma in patients who do not have TBI but have conditions without combination codes describing coma, for example, coma secondary to spontaneous brain hemorrhage. Because there is not a combination code describing brain hemorrhage with coma, an additional code must be assigned to reflect the coma. Unspecified coma does not seem appropriate because it is known/specified to be a non-TBI coma, making it coma NEC.

### TABULAR MODIFICATIONS

R40 Somnolence, stupor and coma

Excludes1: neonatal coma (P91.5)  
somnolence, stupor and coma in diabetes (E08-E13)  
somnolence, stupor and coma in hepatic failure (K72.-)  
somnolence, stupor and coma in hypoglycemia  
(nondiabetic) (E15)

R40.2 Coma

Code first any associated:

fracture of skull (S02.-)  
intracranial injury (S06.-)

Note: One code from each subcategory, R40.21-R40.23, is  
required to complete the coma scale

New code	R40.29	Other coma
Add		Secondary coma
Add		Code also underlying condition

## Contrast-Induced Nephropathy

There are two rare, but serious disorders associated with contrast dyes and the kidneys: contrast induced nephropathy (CIN) and nephrogenic systemic fibrosis (NSF).

Contrast-induced nephropathy (CIN) is defined as the impairment of kidney function—measured as either a 25% increase in serum creatinine (SCr) from baseline or a 0.5 mg/dL (44 µmol/L) increase in absolute SCr value—within 48-72 hours after intravenous contrast administration. CIN is a rare disorder and occurs when kidney problems are caused by the use of certain contrast dyes. In most cases contrast dyes used in tests, such as CT (computerized tomography) and angiograms, have no reported problems. About 2 percent of people receiving dyes can develop CIN. However, the risk for CIN can increase for people with diabetes, a history of heart and blood diseases, and chronic kidney disease (CKD). For example, the risk of CIN in people with advanced CKD (glomerular filtration rate (GFR) below 30 mL/min/1.73m<sup>2</sup>), increases to 30 to 40 percent. The risk of CIN in people with both CKD and diabetes is 20 to 50 percent.

Contrast-induced nephropathy (CIN) is the third leading cause of hospital acquired acute kidney injury and identifiable cause of iatrogenic acute kidney injury.

Cone Health is requesting a new ICD-10-CM code for contrast-induced nephropathy for coding specificity and research.

### References

<https://pubmed.ncbi.nlm.nih.gov/12441014/>  
<https://pubmed.ncbi.nlm.nih.gov/12629594/>  
<https://radiopaedia.org/articles/contrast-induced-nephropathy>  
<https://rebelem.com/contrast-induced-nephropathy-cin-really-not-thing/>  
<https://www.kidney.org/atoz/content/Contrast-Dye-and-Kidneys>

## TABULAR MODIFICATIONS

N14	Drug- and heavy-metal-induced tubulo-interstitial and tubular conditions Code first poisoning due to drug or toxin, if applicable (T36-T65 with fifth or sixth character 1-4 or 6) Use additional code for adverse effect, if applicable, to identify drug (T36-T50 with fifth or sixth character 5)
New subcategory	N14.1 Nephropathy induced by other drugs, medicaments and biological substances
New code Add	N14.11 Contrast induced nephropathy Contrast medium, radiography nephropathy

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New code

N14.19 Nephropathy induced by other drugs, medicaments and  
biological substances

## **Craniosynostosis and Other Congenital Deformities of Skull, Face and Jaw**

In a newborn, the bones of the cranium are separated by intervening sutures (i.e., gaps) that enable the infant's skull to pass through the birth canal and to allow for both growth of the skull and brain. Craniosynostosis is the premature closure of one or more cranial sutures. When one or more sutures closes prematurely, an abnormally shaped skull and also, in more severe cases, increased intracranial pressure can occur.

The prevalence of craniosynostosis is ~1 in 2000 births. Clinically, craniosynostosis is classified according to the suture involved. The most common sutures involved in craniosynostosis are sagittal (~60%), coronal (~25%), metopic (~15%), and lambdoid (~2%). Sagittal and metopic sutures are located midline. Coronal and lambdoid sutures extend laterally - left and right - on the skull. Therefore, coronal and lambdoid craniosynostosis can occur on one (i.e., unilateral) or both (i.e., bilateral) sides.

Pediatric clinicians routinely screen infants and children for abnormal shape of the cranium. These clinicians may suspect that craniosynostosis may be responsible for a particular head shape, and therefore pursue referral to craniosynostosis clinical specialists for further evaluation and treatment. Not all head shape findings (e.g., metopic ridge, sagittal crest) are abnormal or due to craniosynostosis. Definitive diagnosis of craniosynostosis is typically made with radiographic imaging of the skull (e.g., computerized tomography) and physical examination performed by a craniosynostosis clinical expert (e.g., neurosurgeon, plastic surgeon).

Currently, there is one ICD-10-CM code for craniosynostosis (Q75.0), for which acrocephaly, imperfect fusion of skull, oxycephaly, trigonocephaly are inclusion terms. These inclusion terms convey the subjective, phenotypic shape of the cranium that can occur as a result of craniosynostosis, but not the type/location of the craniosynostosis.

Classification of the type of the craniosynostosis is essential for several reasons, including (1) to accurately measure and assess worldwide trends in the epidemiology of craniosynostosis types, (2) outcomes and treatments vary by craniosynostosis type and (3) the removal of antiquated terms (acrocephaly, oxycephaly).

The revisions proposed are to achieve sufficient, clinical granularity of the type of craniosynostosis (i.e., sagittal, coronal, metopic, lambdoid, other, and not specified) and laterality (i.e., unilateral, bilateral, not specified). Sagittal and metopic craniosynostosis are midline, therefore the side is not applicable. The surgeons who diagnosis craniosynostosis most often, do not feel that knowing the actual side is more beneficial, but knowing if it was unilateral or bilateral is sufficient detail.

A related ICD-10-CM code for skull deformities is, Q67.4 Other congenital deformities of skull, face, and jaw. The terms in the Q67.4 convey important cranial findings and characteristics. However, the cranial terms are grouped with face, nose, and jaw deformities. The terms do not

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encompass all of the common cranial findings that typically prompt a primary care/general pediatric clinician to seek evaluation by a specialist for evaluation and treatment.

The following revisions are proposed to achieve sufficient, clinical granularity of the type of skull deformities. This granularity will significantly improve international classification, tracking, and surveillance of infants and children with craniosynostosis and skull characteristics that prompt evaluation for craniosynostosis.

Members of the American Society of Pediatric Neurosurgeons and the American Society of Craniofacial Surgeons have called for (a) revision of the current craniosynostosis ICD-10-CM diagnosis code (Q75.0) and (b) related revision of the current code for other congenital deformities of skull, face, and jaw (Q67.4) to provide more clinical granularity for the clinical modification.<sup>1,2</sup>

This proposal has been submitted by the following individuals and supported by the American Academy of Pediatrics:

Jay Berry, MD, MPH, Chief, Complex Care, Division of General Pediatrics, Boston Children's Hospital, Harvard Medical School; Chair, International Committee on Perioperative Care for Children with Medical Complexity

Michael Golinko, MD, Chief of Plastic Surgery, Director of the Cleft & Craniofacial Program, Monroe Carell Jr. Children's Hospital at Vanderbilt

John Meara, MD, DMD, MBA, Plastic Surgeon-in-Chief, Department of Plastic and Oral Surgery, Boston Children's Hospital; Kletjian Professor of Global Surgery, Harvard Medical School

Gareth Parry, PhD, Senior Staff Scientist, Department of Oral and Plastic Surgery, Boston Children's Hospital

Mark Proctor, MD, Neurosurgeon-in-Chief, Department of Neurosurgery, Boston Children's Hospital; Past Chairman, Pediatric Section of the American Association of Neurological Surgeons and Congress of Neurological Surgeons

#### References

<sup>1</sup>Gonzalez SR, Han A, Golinko MS. Shifting epidemiology of single-suture craniosynostosis and the need for a more granular ICD classification system: a national survey of members from the American Society of Pediatric Neurosurgeons (ASPN) and the American Society of Craniofacial Surgeons (ASCFS). *Childs Nerv Syst.* 2019;35(9):1443-1444.

<sup>2</sup>Gonzalez SR, Light JG, Golinko MS. Assessment of Epidemiological Trends in Craniosynostosis: Limitations of the Current Classification System. *Plast Reconstr Surg Glob Open.* 2020;8(3):e2597.

### TABULAR MODIFICATIONS

- Q67 Congenital musculoskeletal deformities of head, face, spine and chest  
Excludes1: congenital malformation syndromes classified to Q87.-  
Potter's syndrome (Q60.6)
- Q67.4 Other congenital deformities of skull, face and jaw
- Delete ~~Congenital depressions in skull~~  
Delete ~~Congenital hemifacial atrophy or hypertrophy~~  
Delete ~~Deviation of nasal septum, congenital~~  
Delete ~~Squashed or bent nose, congenital~~
- Excludes1: dentofacial anomalies [including malocclusion] (M26.-)  
syphilitic saddle nose (A50.5)
- New code Q67.40 Other congenital deformities of skull, face, and jaw unspecified
- New code Q67.41 Other congenital deformities of skull  
Add Congenital abnormal shape of skull  
Add Congenital asymmetry of forehead  
Add Congenital brow asymmetry  
Add Congenital depressions in skull  
Add Metopic ridge  
Add Sagittal crest
- New code Q67.42 Other congenital deformities of face  
Add Asymmetric crying face association  
Add Asymmetric crying facies  
Add Congenital hemifacial atrophy or hypertrophy  
Add Facial asymmetry, congenital
- New code Q67.43 Other congenital anomalies of nose  
Add Deviation of nasal septum, congenital  
Add Squashed or bent nose, congenital
- New code Q67.44 Other congenital deformities of jaw
- Q75 Other congenital malformations of skull and face bones  
Excludes1: congenital malformation of face NOS (Q18.-)  
congenital malformation syndromes classified to Q87.-  
dentofacial anomalies [including malocclusion] (M26.-)  
musculoskeletal deformities of head and face (Q67.0-Q67.4)  
skull defects associated with congenital anomalies of brain such as:

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anencephaly (Q00.0)  
encephalocele (Q01.-)  
hydrocephalus (Q03.-)  
microcephaly (Q02)

	Q75.0	Craniosynostosis
Delete		<del>Aerocephaly</del>
Delete		<del>Oxycephaly</del>
Delete		<del>Trigonocephaly</del>
New code	Q75.01	Sagittal craniosynostosis
Add		Scaphocephaly
New sub-category	Q75.02	Coronal craniosynostosis
Add		Anterior plagiocephaly
New code	Q75.021	Coronal craniosynostosis unilateral
New code	Q75.022	Coronal craniosynostosis bilateral
Add		Brachycephaly
New code	Q75.029	Coronal craniosynostosis unspecified
New code	Q75.03	Metopic craniosynostosis
Add		Trigonocephaly
New sub-category	Q75.04	Lambdoid craniosynostosis
Add		Posterior plagiocephaly
New code	Q75.041	Lambdoid craniosynostosis unilateral
New code	Q75.042	Lambdoid craniosynostosis bilateral
New code	Q75.049	Lambdoid craniosynostosis unspecified
New subcategory	Q75.05	Other craniosynostosis
New code	Q75.051	Other craniosynostosis unilateral
New code	Q75.052	Other craniosynostosis bilateral
New code	Q75.059	Other craniosynostosis unspecified
New subcategory	Q75.06	Unspecific Craniosynostosis
Add		Craniosynostosis, NOS
New code	Q75.061	Unspecified Craniosynostosis, unilateral
New code	Q75.062	Unspecified Craniosynostosis, bilateral
New code	Q75.063	Unspecified Craniosynostosis, multiple sites
New Code	Q75.069	Unspecified Craniosynostosis, unspecified
Add		Imperfect fusion of skull

## **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 submitter of this proposal, The National Minority Quality Forum presented this proposal at the March 2021 Coordination and Maintenance Meeting. Based on comments received, the proposal is being resubmitted for reconsideration. Changes are noted in **bold**.

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.<sup>1</sup> In addition, patients with chronic conditions and dementia use more healthcare services than patients with chronic conditions who do not have dementia.<sup>1</sup>

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,<sup>2</sup> 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.

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*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.<sup>3</sup>

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.<sup>4</sup>

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 new codes for mild cognitive disorder was presented at the September 2020 and March 2021 Coordination and Maintenance Committee Meeting. **Additional revisions have been made to that proposal and is being re-presented at the September 2021 meeting.** Related changes in the proposal for mild **neurocognitive 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),<sup>5</sup> 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.<sup>6</sup>

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

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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.<sup>7</sup>

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.<sup>5</sup> Agitation is generally considered the most disruptive of the behavioral disturbances because it is associated with increased rates of institutionalization.<sup>7</sup> Some studies have identified a correlation between psychoses and acceleration of cognitive decline and increased mortality,<sup>8</sup> as well as a correlation between mood disorders and lowered Quality of Life scores.<sup>9</sup>

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 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.

### **References**

<sup>1</sup>Alzheimer's Association, "2020 Alzheimer's Disease Facts and Figures," *Alzheimers Dement*, 2020; 16(3): 391-460. doi.org/10.1002/alz.12068.

<sup>2</sup>Jack, C., Bennett, D., Blennow, K., Carrillo, M., Dunn, B., Haeblerlein, S., et al. "NIA-AA Research Framework: Toward a Biological Definition of Alzheimer's Disease," *Alzheimers Dement*, 2018; 14(4): 535-62.

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<sup>3</sup>Rikkert, M.G., Tona, K.D., Janssen, L., et al., "Validity, Reliability, and Feasibility of Clinical Staging Scales in Dementia: A Systematic Review," *Am J Alzheimers Dis Other Demen.*, 2011; 26(5):357–365.

<sup>4</sup>Leibson, CL, Long, KH, Ransom, JE, Roberts, RO, Hass, SL, et al., "Direct Medical Costs and Source of Cost Differences Across the Spectrum of Cognitive Decline: A Population-based Study," *Alzheimers Dement*, 2015; 11(8):917–32.

<sup>5</sup>Victoroff J, Lin FV, Coburn KL, Shillcut SD, et al, "Noncognitive Behavioral Changes Associated With Alzheimer's Disease: Implications of Neuroimaging Findings" *J Neuropsychiatry Clin Neurosci* 2018; 30:14–21.

## TABULAR MODIFICATIONS

### A81 Atypical virus infections of central nervous system

Revise Use additional code, if applicable, to identify:  
**Add dementia with anxiety (F02.84, F02.A4, F02.B4, F02.C4)**  
Revise dementia with behavioral disturbance (F02.81, F02.A1-, F02.B1-, F02.C1-)  
Add dementia with mood **disturbance** (F02.83, F02.A3, F02.B3, F02.C3)  
Add dementia with psychotic **disturbance** (F02.82, F02.A2, F02.B2, F02.C2)  
Revise dementia without behavioral disturbance (F02.80, F02.A0, F02.B0, F02.C0)  
**Add mild neurocognitive disorder due to known physiological condition (F06.7-)**

### F01 Vascular Dementia

Vascular dementia as a result of infarction of the brain due to vascular disease, including hypertensive cerebrovascular disease.

Includes: arteriosclerotic dementia  
**Add major neurocognitive disorder due to vascular disease**  
**Add multi-infarct dementia**

Code first the underlying physiological condition or sequelae of cerebrovascular disease.

Revise F01.5 Vascular dementia, unspecified severity

Revise F01.50 Vascular dementia, unspecified severity, without behavioral disturbance, psychotic disturbance, mood disturbance, and anxiety

Revise Major neurocognitive disorder due to vascular disease ~~without behavioral disturbance-NOS~~

Add Vascular dementia NOS

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Revise	F01.51 Vascular dementia, <del>unspecified severity</del> , with behavioral disturbance
Delete	<del>Major neurocognitive disorder due to vascular disease, with behavioral disturbance</del>
Delete	<del>Major neurocognitive disorder with aggressive behavior</del>
Delete	<del>Major neurocognitive disorder with combative behavior</del>
Delete	<del>Major neurocognitive disorder with violent behavior</del>
Delete	<del>Vascular dementia with aggressive behavior</del>
Delete	<del>Vascular dementia with combative behavior</del>
Delete	<del>Vascular dementia with violent behavior</del>
Delete	<b>Use additional code, if applicable, to identify wandering in vascular dementia (Z91.83)</b>
New code	F01.511 Vascular dementia, unspecified <b>severity</b> , with agitation
Add	Major neurocognitive disorder due to vascular disease, unspecified <b>severity</b> , with aberrant motor behavior such as restlessness, rocking, pacing, or exit-seeking
Add	Major neurocognitive disorder due to vascular disease, unspecified <b>severity</b> , with <b>verbal or physical behaviors such as profanity, shouting, threatening, anger, aggression, combativeness, or violence</b>
Add	Vascular dementia, unspecified <b>severity</b> , with aberrant motor behavior such as restlessness, rocking, pacing, or exit-seeking
Add	Vascular dementia, unspecified <b>severity</b> , with <b>verbal or physical behaviors such as profanity, shouting, threatening, anger, aggression, combativeness, or violence</b>
New code	F01.518 Vascular dementia, unspecified <b>severity</b> , with other behavioral disturbance
Add	Major neurocognitive disorder due to vascular disease, unspecified <b>severity</b> , with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition
Add	Vascular dementia, unspecified <b>severity</b> , with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition
Add	<b>Use additional code, if applicable, to identify wandering in vascular dementia (Z91.83)</b>

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- New code F01.52 Vascular dementia, unspecified **severity**, with psychotic **disturbance**  
Add Major neurocognitive disorder due to vascular disease, unspecified **severity**, with psychotic **disturbance** such as hallucinations, paranoia, suspiciousness, or delusional state  
Add Vascular dementia, unspecified **severity**, with psychotic **disturbance** such as hallucinations, paranoia, suspiciousness, or delusional state
- New code F01.53 Vascular dementia, unspecified **severity**, with mood **disturbance**  
Add Major neurocognitive disorder due to vascular disease, unspecified **severity**, with mood **disturbance** such as depression, apathy, or anhedonia  
Add Vascular dementia, unspecified **severity**, with mood **disturbance** such as depression, apathy, or anhedonia
- New code F01.54 Vascular dementia, unspecified **severity**, with anxiety  
Add Major neurocognitive disorder due to vascular disease, unspecified **severity**, with anxiety
- New sub-category **F01.A** Vascular dementia, mild  
**Add Excludes1: mild neurocognitive disorder due to known physiological condition with or without behavioral disturbance (F06.7-)**
- New code F01.A0 Vascular dementia, mild, without behavioral disturbance, psychotic **disturbance, mood disturbance, and anxiety**  
Add Major neurocognitive disorder due to vascular disease, mild, NOS  
Add Vascular dementia, mild, NOS
- New sub-sub-category F01.A1 Vascular dementia, mild, with behavioral disturbance
- New code F01.A11 Vascular dementia, mild, with agitation  
Add Major neurocognitive disorder due to vascular disease, mild, with aberrant motor behavior such as restlessness, rocking, pacing, or exit-seeking  
Add Major neurocognitive disorder due to vascular disease, mild, with **verbal or physical behaviors such as profanity, shouting, threatening, anger, aggression, combativeness, or violence**  
Add Vascular dementia, mild, with aberrant motor behavior such as restlessness, rocking, pacing, or exit-seeking

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- Add Vascular dementia, mild, with **verbal or physical behaviors such as profanity, shouting, threatening, anger, aggression, combativeness, or violence**
- New code F01.A18 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
- Add **Use additional code, if applicable, to identify wandering in vascular dementia (Z91.83)**
- New code F01.A2 Vascular dementia, mild, with psychotic **disturbance**  
Add Major neurocognitive disorder due to vascular disease, mild, with psychotic **disturbance** such as hallucinations, paranoia, suspiciousness, or delusional state  
Add Vascular dementia, mild, with psychotic **disturbance** such as hallucinations, paranoia, suspiciousness, or delusional state
- New code F01.A3 Vascular dementia, mild, with mood **disturbance**  
Add Major neurocognitive disorder due to vascular disease, mild, with mood **disturbance** such as depression, apathy, or anhedonia  
Add Vascular dementia, mild, with mood **disturbance** such as depression, apathy, or anhedonia
- New code F01.A4 Vascular dementia, mild, with anxiety  
Add Major neurocognitive disorder due to vascular disease, mild, with anxiety
- New sub-category **F01.B** Vascular dementia, moderate
- New code F01.B0 Vascular dementia, moderate, without behavioral disturbance, psychotic **disturbance, mood disturbance, and anxiety**  
Add Major neurocognitive disorder due to vascular disease, moderate, NOS  
Add Vascular dementia, moderate, NOS
- New sub-sub-category F01.B1 Vascular dementia, moderate, with behavioral disturbance

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New code	F01.B11 Vascular dementia, moderate, with agitation
Add	Major neurocognitive disorder due to vascular disease, moderate, with aberrant motor behavior such as restlessness, rocking, pacing, or exit-seeking
Add	Major neurocognitive disorder due to vascular disease, moderate, with <b>verbal or physical behaviors such as profanity, shouting, threatening, anger, aggression, combativeness, or violence</b>
Add	Vascular dementia, moderate, with aberrant motor behavior such as restlessness, rocking, pacing, or exit-seeking
Add	Vascular dementia, moderate, with <b>verbal or physical behaviors such as profanity, shouting, threatening, anger, aggression, combativeness, or violence</b>
New code	F01.B18 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
Add	<b>Use additional code, if applicable, to identify wandering in vascular dementia (Z91.83)</b>
New code	F01.B2 Vascular dementia, moderate, with psychotic <b>disturbance</b>
Add	Major neurocognitive disorder due to vascular disease, moderate, with psychotic <b>disturbance</b> such as hallucinations, paranoia, suspiciousness, or delusional state
Add	Vascular dementia, moderate, with psychotic <b>disturbance</b> such as hallucinations, paranoia, suspiciousness, or delusional state
New code	F01.B3 Vascular dementia, moderate, with mood <b>disturbance</b>
Add	Major neurocognitive disorder due to vascular disease, moderate, with mood <b>disturbance</b> such as depression, apathy, or anhedonia
Add	Vascular dementia, moderate, with mood <b>disturbance</b> such as depression, apathy, or anhedonia
New code	F01.B4 Vascular dementia, moderate, with anxiety
Add	Major neurocognitive disorder due to vascular disease, moderate, with anxiety

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New sub-category **F01.C** Vascular dementia, severe

New code F01.C0 Vascular dementia, severe, without behavioral disturbance, psychotic  
**disturbance, mood disturbance, and anxiety**

Add Major neurocognitive disorder due to vascular disease, severe, NOS

Add Vascular dementia, severe, NOS

New sub-sub-category F01.C1 Vascular dementia, severe, with behavioral disturbance

New code F01.C11 Vascular dementia, severe, with agitation

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

Add Major neurocognitive disorder due to vascular disease, severe,  
with **verbal or physical behaviors such as profanity,  
shouting, threatening, anger, aggression, combativeness, or  
violence**

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

Add Vascular dementia, severe, with **verbal or physical behaviors  
such as profanity, shouting, threatening, anger,  
aggression, combativeness, or violence**

New code F01.C18 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

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

New code F01.C2 Vascular dementia, severe, with psychotic **disturbance**

Add Major neurocognitive disorder due to vascular disease, severe, with  
psychotic **disturbance** such as hallucinations, paranoia,  
suspiciousness, or delusional state

Add Vascular dementia, severe, with psychotic **disturbance** such as  
hallucinations, paranoia, suspiciousness, or delusional state

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New code	F01.C3 Vascular dementia, severe, with mood <b>disturbance</b>
Add	Major neurocognitive disorder due to vascular disease, severe, with mood <b>disturbance</b> such as depression, apathy, or anhedonia
Add	Vascular dementia, severe, with mood <b>disturbance</b> such as depression, apathy, or anhedonia
New code	F01.C4 Vascular dementia, severe, with anxiety
Add	Major neurocognitive disorder due to vascular disease, severe, with anxiety
	F02 Dementia in other diseases classified elsewhere
	Includes: Major neurocognitive disorder in other disease classified elsewhere
Add	Excludes1: mild neurocognitive disorder due to known physiological condition with or without behavioral disturbance (F06.7-)
Revise	F02.8 Dementia in other diseases classified elsewhere, <u>unspecified severity</u>
Revise	F02.80 Dementia in other diseases classified elsewhere, <u>unspecified severity</u> , without behavioral disturbance, <u>psychotic disturbance, mood disturbance, and anxiety</u>
Revise	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 severity</u> , with behavioral disturbance
Delete	<del>Dementia in other diseases classified elsewhere with aggressive behavior</del>
Delete	<del>Dementia in other diseases classified elsewhere with combative behavior</del>
Delete	<del>Dementia in other diseases classified elsewhere with violent behavior</del>
Delete	<del>Major neurocognitive disorder in other diseases classified elsewhere with aggressive behavior</del>
Delete	<del>Major neurocognitive disorder in other diseases classified elsewhere with combative behavior</del>
Delete	<del>Major neurocognitive disorder in other diseases classified elsewhere with violent behavior</del>
Delete	<del>Use additional code, if applicable, to identify wandering in dementia in conditions classified elsewhere (Z91.83)</del>

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New code	F02.811 Dementia in other diseases classified elsewhere, unspecified <b>severity</b> , with agitation
Add	Dementia in other diseases classified elsewhere, unspecified <b>severity</b> , with aberrant motor behavior such as restlessness, rocking, pacing, or exit-seeking
Add	Dementia in other diseases classified elsewhere, unspecified <b>severity</b> , with <b>verbal or physical behaviors such as profanity, shouting, threatening, anger, aggression, combativeness, or violence</b>
Add	Major neurocognitive disorder in other diseases classified elsewhere, unspecified <b>severity</b> , with aberrant motor behavior such as restlessness, rocking, pacing, or exit-seeking
Add	Major neurocognitive disorder in other diseases classified elsewhere, unspecified <b>severity</b> , with <b>verbal or physical behaviors such as profanity, shouting, threatening, anger, aggression, combativeness, or violence</b>
New code	F02.818 Dementia in other diseases classified elsewhere, unspecified <b>severity</b> , 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
Add	<b>Use additional code, if applicable, to identify wandering in dementia in conditions classified elsewhere (Z91.83)</b>
New code	F02.82 Dementia in other diseases classified elsewhere, unspecified <b>severity</b> , with psychotic <b>disturbance</b>
Add	Dementia in other diseases classified elsewhere, unspecified <b>severity</b> , with psychotic <b>disturbance</b> such as hallucinations, paranoia, suspiciousness, or delusional state
Add	Major neurocognitive disorder in other diseases classified elsewhere, unspecified, with psychotic <b>disturbance</b> such as hallucinations, paranoia, suspiciousness, or delusional state
New code	F02.83 Dementia in other diseases classified elsewhere, unspecified <b>severity</b> , with mood <b>disturbance</b>
Add	Dementia in other diseases classified elsewhere, unspecified <b>severity</b> , with mood <b>disturbance</b> such as depression, apathy, or anhedonia

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Add	Major neurocognitive disorder in other diseases classified elsewhere unspecified <b>severity</b> , with mood <b>disturbance</b> such as with depression, apathy, or anhedonia
New code	F02.84 Dementia in other diseases classified elsewhere, unspecified <b>severity</b> , with anxiety
Add	Major neurocognitive disorder in other diseases classified elsewhere unspecified <b>severity</b> , with anxiety
New subcategory	<b>F02.A</b> Dementia in other diseases classified elsewhere, mild
New code	F02.A0 Dementia in other diseases classified elsewhere, mild, without behavioral disturbance, psychotic <b>disturbance</b> , mood <b>disturbance</b> , and anxiety
Add	Dementia in other diseases classified elsewhere, mild, NOS
Add	Major neurocognitive disorder in other diseases classified elsewhere, mild, NOS
New sub-sub-category	F02.A1 Dementia in other diseases classified elsewhere, mild, with behavioral disturbance
New code	F02.A11 Dementia in other diseases classified elsewhere, mild, with agitation
Add	Dementia in other diseases classified elsewhere, mild, with aberrant motor behavior such as restlessness, rocking, pacing, or exit-seeking
Add	Dementia in other diseases classified elsewhere, mild, with <b>verbal or physical behaviors such as profanity, shouting, threatening, anger, aggression, combativeness, or violence</b>
Add	Major neurocognitive disorder in other diseases classified elsewhere, mild, with aberrant motor behavior such as restlessness, rocking, pacing, or exit-seeking
	Major neurocognitive disorder in other diseases classified elsewhere, mild, with <b>verbal or physical behaviors such as profanity, shouting, threatening, anger, aggression, combativeness, or violence</b>

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New code	F02.A18 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
<b>Add</b>	<b>Use additional code, if applicable, to identify wandering in dementia in conditions classified elsewhere (Z91.83)</b>
New code	F02.A2 Dementia in other diseases classified elsewhere, mild, with psychotic <b>disturbance</b>
Add	Dementia in other diseases classified elsewhere, mild, with psychotic <b>disturbance</b> such as hallucinations, paranoia, suspiciousness, or delusional state
Add	Major neurocognitive disorder in other diseases classified elsewhere, mild, with psychotic <b>disturbance</b> such as hallucinations, paranoia, suspiciousness, or delusional state
New code	F02.A3 Dementia in other diseases classified elsewhere, mild, with mood <b>disturbance</b>
Add	Dementia in other diseases classified elsewhere, mild, with mood <b>disturbance</b> such as depression, apathy, or anhedonia
Add	Major neurocognitive disorder in other diseases classified elsewhere, mild, with mood <b>disturbance</b> such as depression, apathy, or anhedonia
New code	F02.A4 Dementia in other diseases classified elsewhere, mild, with anxiety
Add	Major neurocognitive disorder in other diseases classified elsewhere, mild, with anxiety
New sub-category	<b>F02.B</b> Dementia in other diseases classified elsewhere, moderate
New code	F02.B0 Dementia in other diseases classified elsewhere, moderate, without behavioral disturbance, psychotic <b>disturbance</b> , mood <b>disturbance</b> , and anxiety
Add	Dementia in other diseases classified elsewhere, moderate, NOS
Add	Major neurocognitive disorder in other diseases classified elsewhere, moderate, NOS

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New sub-category	F02.B1 Dementia in other diseases classified elsewhere, moderate, with behavioral disturbance
New code	F02.B11 Dementia in other diseases classified elsewhere, moderate, with agitation
Add	Dementia in other diseases classified elsewhere, moderate, with aberrant motor behavior such as restlessness, rocking, pacing, or exit-seeking
Add	Dementia in other diseases classified elsewhere, moderate, with <b>verbal or physical behaviors such as profanity, shouting, threatening, anger, aggression, combativeness, or violence</b>
Add	Major neurocognitive disorder in other diseases classified elsewhere, moderate, with aberrant motor behavior such as restlessness, rocking, pacing, or exit-seeking
Add	Major neurocognitive disorder in other diseases classified elsewhere, moderate, with <b>verbal or physical behaviors such as profanity, shouting, threatening, anger, aggression, combativeness, or violence</b>
New code	F02.B18 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
Add	<b>Use additional code, if applicable, to identify wandering in dementia in conditions classified elsewhere (Z91.83)</b>
New code	F02.B2 Dementia in other diseases classified elsewhere, moderate, with psychotic <b>disturbance</b>
Add	Dementia in other diseases classified elsewhere, moderate, with psychotic <b>disturbance</b> such as hallucinations, paranoia, suspiciousness, or delusional state
Add	Major neurocognitive disorder in other diseases classified elsewhere, moderate, with psychotic <b>disturbance</b> such as hallucinations, paranoia, suspiciousness, or delusional state

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New code	F02.B3 Dementia in other diseases classified elsewhere, moderate, with mood <b>disturbance</b>
Add	Dementia in other diseases classified elsewhere, moderate, with mood <b>disturbance</b> such as depression, apathy, or anhedonia
Add	Major neurocognitive disorder in other diseases classified elsewhere, moderate, with mood <b>disturbance</b> such as depression, apathy, or anhedonia
New code	F02.B4 Dementia in other diseases classified elsewhere, moderate, with anxiety
Add	Major neurocognitive disorder in other diseases classified elsewhere, moderate, with anxiety
New sub-category	<b>F02.C</b> Dementia in other diseases classified elsewhere, severe
New code	F02.C0 Dementia in other diseases classified elsewhere, severe, without behavioral disturbance, psychotic <b>disturbance</b> , mood <b>disturbance</b> , and <b>anxiety</b>
Add	Dementia in other diseases classified elsewhere, severe, NOS
Add	Major neurocognitive disorder in other diseases classified elsewhere, severe, NOS
New sub-sub-category	F02.C1 Dementia in other diseases classified elsewhere, severe, with behavioral disturbance
New code	F02.C11 Dementia in other diseases classified elsewhere, severe, with agitation
Add	Dementia in other diseases classified elsewhere, severe, with aberrant motor behavior such as restlessness, rocking, pacing, or exit-seeking
Add	Dementia in other diseases classified elsewhere, severe, with <b>verbal or physical behaviors such as profanity, shouting, threatening, anger, aggression, combativeness, or violence</b>
Add	Major neurocognitive disorder in other diseases classified elsewhere, severe, with aberrant motor behavior such as restlessness, rocking, pacing, or exit-seeking
Add	Major neurocognitive disorder in other diseases classified elsewhere, severe, with <b>verbal or physical behaviors such as profanity, shouting, threatening, anger, aggression, combativeness, or violence</b>

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New code	F02.C18 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
Add	<b>Use additional code, if applicable, to identify wandering in dementia in conditions classified elsewhere (Z91.83)</b>
New code	F02.C2 Dementia in other diseases classified elsewhere, severe, with psychotic <b>disturbance</b>
Add	Dementia in other diseases classified elsewhere, severe, with psychotic <b>disturbance</b> such as hallucinations, paranoia, suspiciousness, or delusional state
Add	Major neurocognitive disorder in other diseases classified elsewhere, severe, with psychotic <b>disturbance</b> such as hallucinations, paranoia, suspiciousness, or delusional state
New code	F02.C3 Dementia in other diseases classified elsewhere, severe, with mood <b>disturbance</b>
Add	Dementia in other diseases classified elsewhere, severe, with mood <b>disturbance</b> such as depression, apathy, or anhedonia
Add	Major neurocognitive disorder in other diseases classified elsewhere, severe, with mood <b>disturbance</b> such as depression, apathy, or anhedonia
New code	F02.C4 Dementia in other diseases classified elsewhere, severe, with anxiety
Add	Major neurocognitive disorder in other diseases classified elsewhere, severe, with anxiety
Add	F03 Unspecified Dementia <b>Major neurocognitive disorder NOS</b> Presenile dementia NOS Presenile psychosis NOS Primary degenerative dementia NOS Senile dementia NOS Senile dementia depressed or paranoid type Senile psychosis NOS

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Revise	F03.9 Unspecified dementia, <u>unspecified severity</u>
Revise	F03.90 Unspecified dementia, <u>unspecified severity</u> , without behavioral disturbance, <u>psychotic disturbance, mood disturbance, and anxiety</u> Dementia NOS
Revise	F03.91 Unspecified dementia, <u>unspecified severity</u> , with behavioral disturbance
Delete	<del>Unspecified dementia with aggressive behavior</del>
Delete	<del>Unspecified dementia with combative behavior</del>
Delete	<del>Unspecified dementia with violent behavior</del>
Delete	<del>Use additional code, if applicable, to identify wandering in unspecified dementia (Z91.83)</del>
New code	F03.911 Unspecified dementia, unspecified severity, with agitation
Add	Unspecified dementia, unspecified severity, with aberrant motor behavior such as restlessness, rocking, pacing, or exit-seeking
Add	Unspecified dementia, unspecified severity, with <b>verbal or physical behaviors such as profanity, shouting, threatening, anger, aggression, combativeness, or violence</b>
New code	F03.918 Unspecified dementia, unspecified severity, with other behavioral disturbance
Add	Unspecified dementia, unspecified severity, with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition
Add	<b>Use additional code, if applicable, to identify wandering in unspecified dementia (Z91.83)</b>
New code	F03.92 Unspecified dementia, unspecified severity, with psychotic <b>disturbance</b>
Add	Unspecified dementia, unspecified severity, with psychotic <b>disturbance</b> such as hallucinations, paranoia, suspiciousness, or delusional state

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New code	F03.93 Unspecified dementia, unspecified <b>severity</b> , with mood <b>disturbance</b>
Add	Unspecified dementia, unspecified <b>severity</b> , with mood <b>disturbance</b> such as depression, apathy, or anhedonia
New code	F03.94 Unspecified dementia, unspecified <b>severity</b> , with anxiety
New sub-category	<b>F03.A</b> Unspecified dementia, mild
Add	<b>Excludes1: mild neurocognitive disorder due to known physiological condition with or without behavioral disturbance (F06.7-)</b>
New code	F03.A0 Unspecified dementia, mild, without behavioral disturbance, psychotic <b>disturbance</b> , mood <b>disturbance</b> , and <b>anxiety</b>
Add	Dementia, mild, NOS
New sub-sub-category	F03.A1 Unspecified dementia, mild, with behavioral disturbance
New code	F03.A11 Unspecified dementia, mild, with agitation
Add	Unspecified dementia, mild, with aberrant motor behavior such as restlessness, rocking, pacing, or exit-seeking
Add	Unspecified dementia, mild, with <b>verbal or physical behaviors such as profanity, shouting, threatening, anger, aggression, combativeness, or violence</b>
New code	F03.A18 Unspecified dementia, mild, with other behavioral disturbance
Add	Unspecified dementia, mild, with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition
Add	<b>Use additional code, if applicable, to identify wandering in unspecified dementia (Z91.83)</b>
New code	F03.A2 Unspecified dementia, mild, with psychotic <b>disturbance</b>
Add	Unspecified dementia, mild, with psychotic <b>disturbance</b> such as hallucinations, paranoia, suspiciousness, or delusional state
New code	F03.A3 Unspecified dementia, mild, with mood <b>disturbance</b>
	Unspecified dementia, mild, with mood <b>disturbance</b> such as depression, apathy, or anhedonia

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New code	F03.A4 Unspecified dementia, mild, with anxiety
New sub-category	<b>F03.B</b> Unspecified dementia, moderate
New code	F03.B0 Unspecified dementia, moderate, without behavioral disturbance, psychotic <b>disturbance</b> , mood <b>disturbance</b> , and anxiety <b>Dementia</b> , moderate, NOS
New sub-subcategory	F03.B1 Unspecified dementia, moderate, with behavioral disturbance
New code	F03.B11 Unspecified dementia, moderate, with agitation
Add	Unspecified dementia, moderate, with aberrant motor behavior such as restlessness, rocking, pacing, or exit-seeking
Add	Unspecified dementia, moderate, with <b>verbal or physical behaviors such as profanity, shouting, threatening, anger, aggression, combativeness</b> , or violence
New code	F03.B18 Unspecified dementia, moderate, with other behavioral disturbance
Add	Unspecified dementia, moderate, with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition
Add	<b>Use additional code, if applicable, to identify wandering in unspecified dementia (Z91.83)</b>
New code	F03.B2 Unspecified dementia, moderate, with psychotic <b>disturbance</b>
Add	Unspecified dementia, moderate, with psychotic <b>disturbance</b> such as hallucinations, paranoia, suspiciousness, or delusional state
New code	F03.B3 Unspecified dementia, moderate, with mood <b>disturbance</b>
Add	Unspecified dementia, moderate, with mood <b>disturbance</b> such as depression, apathy, or anhedonia
New code	F03.B4 Unspecified dementia, moderate, with anxiety
New sub-category	<b>F03.C</b> Unspecified dementia, severe

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New code	F03.C0 Unspecified dementia, severe, without behavioral disturbance, psychotic <b>disturbance</b> , mood <b>disturbance</b> , and <b>anxiety</b>
Add	<b>Dementia</b> , severe, NOS
New sub-sub-category	F03.C1 Unspecified dementia, severe, with behavioral disturbance
New code	F03.C11 Unspecified dementia, severe, with agitation
Add	Unspecified dementia, severe, with aberrant motor behavior such as restlessness, rocking, pacing, or exit-seeking
Add	Unspecified dementia, severe, with <b>verbal or physical behaviors such as profanity, shouting, threatening, anger, aggression, combativeness</b> , or violence
New code	F03.C18 Unspecified dementia, severe, with other behavioral disturbance
Add	Unspecified dementia, severe, with behavioral disturbances such as sleep disturbance, social disinhibition, or sexual disinhibition
Add	<b>Use additional code, if applicable, to identify wandering in unspecified dementia (Z91.83)</b>
New code	F03.C2 Unspecified dementia, severe, with psychotic <b>disturbance</b>
Add	Unspecified dementia, severe, with psychotic <b>disturbance</b> such as hallucinations, paranoia, suspiciousness, or delusional state
New code	F03.C3 Unspecified dementia, severe, with mood <b>disturbance</b>
Add	Unspecified dementia, severe, with mood <b>disturbance</b> such as depression, apathy, or anhedonia
New code	F03.C4 Unspecified dementia, severe, with anxiety
	G10 Huntington's disease
	Huntington's chorea
	Huntington's dementia
Delete	<del>Code also dementia in other diseases classified elsewhere without behavioral disturbance (F02.80)</del>

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Revise Use additional code, if applicable, to identify:  
Add **dementia with anxiety (F02.84, F02.A4, F02.B4, F02.C4)**  
Add dementia with behavioral disturbance (F02.81-, F02.A1-, F02.B1-,  
F02.C1-)  
Add dementia with mood **disturbance** (F02.83, F02.A3, F02.B3, F02.C3)  
Add dementia with psychotic **disturbance** (F02.82, F02.A2, F02.B2, F02.C2)  
Revise dementia without behavioral disturbance (F02.80, F02.A0, F02.B0,  
F02.C0)  
Add mild **neurocognitive** disorder due to known physiological condition  
(F06.7-)

G20 Parkinson's disease

Revise Use additional code, if applicable, to identify:  
Add **dementia with anxiety (F02.84, F02.A4, F02.B4, F02.C4)**  
Revise dementia with behavioral disturbance (F02.81-, F02.A1-, F02.B1-, F02.C1-)  
Add dementia with mood **disturbance** (F02.83, F02.A3, F02.B3, F02.C3)  
Add dementia with psychotic **disturbance** (F02.82, F02.A2, F02.B2, F02.C2)  
Revise dementia without behavioral disturbance (F02.80, F02.A0, F02.B0,  
F02.C0)  
Add mild **neurocognitive** disorder due to known physiological condition  
(F06.7-)

G30 Alzheimer's Disease

Revise Use additional code, if applicable, to identify:  
delirium (F05)  
Add **dementia with anxiety (F02.84, F02.A4, F02.B4, F02.C4)**  
Revise dementia with behavioral disturbance (F02.81-, F02.A1-, F02.B1-,  
F02.C1-)  
Add dementia with mood **disturbance** (F02.83, F02.A3, F02.B3, F02.C3)  
Add dementia with psychotic **disturbance** (F02.82, F02.A2, F02.B2, F02.C2)  
Revise dementia without behavioral disturbance (F02.80, F02.A0, F02.B0,  
F02.C0)  
Add mild **neurocognitive** 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:  
Add **dementia with anxiety (F02.84, F02.A4, F02.B4, F02.C4)**  
Revise dementia with behavioral disturbance (F02.81-, F02.A1-, F02.B1-,  
F02.C1-)  
Add dementia with mood **disturbance** (F02.83, F02.A3, F02.B3, F02.C3)

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Add	dementia with psychotic <b>disturbance</b> (F02.82, F02.A2, F02.B2, F02.C2)
Revise	dementia without behavioral disturbance (F02.80, <u>F02.A0, F02.B0,</u> <u>F02.C0</u> )
Add	mild <b>neuro</b> cognitive disorder due to known physiological condition (F06.7-)

## Desmoid Tumors

Desmoid tumors are a rare type of tumor arising in deep connective and soft tissues which often have a variable and unpredictable course. Because desmoid tumors do not metastasize, they are not classified as malignant. However, desmoid tumors tend to be locally aggressive, infiltrative, and destructive, such that the condition is also known as aggressive fibromatosis.

Desmoid tumors constitute 0.03% of all tumors. The estimated incidence in the general population is 2-4 per million people per year. Desmoid tumors are observed to be more common in persons aged 10-40 years but can occur in other age groups. Desmoid tumors can commonly occur in women after childbirth. The female: male gender ratio is 2:1. In children, the gender incidence is the same.

In the US, it is estimated that about 900 to 1,500 people are diagnosed with desmoid tumors each year, although this may be significantly understated because of the challenges in diagnosis and reporting. The diagnosis is typically made via biopsy. It is about twice as common in women as men and tends to peak between the ages of 30 to 40 years old, although it may occur in anyone including infants, young children, and teenagers. The cause of desmoid tumors is generally unknown but up to 90% are associated with mutations of the  $\beta$ -catenin protein, potentially derived from trauma and inappropriate wound healing.

Desmoid tumors can occur in any soft or connective tissue throughout the body. In practice, the locations are typically categorized into four general areas:

- abdominal wall
- extremities/shoulder and pelvic girdles/chest wall
- intraabdominal/retroperitoneal/pelvic cavity
- head and neck/intrathoracic

This categorization is useful clinically because, in addition to tumor size and infiltration, location generally determines symptoms, is strongly linked to morbidity and mortality, and influences the treatment.

Abdominal wall tumors may present as a noticeable mass, which is sometimes revealed as pregnancy stretches the wall. Extremity tumors often present with significant pain and restricted mobility. Intraabdominal/retroperitoneal/pelvic cavity desmoid tumors can be asymptomatic, or they may present as weight loss or with significant comorbidities such as bowel obstruction or renal failure. Head and neck/intrathoracic tumors may present with symptoms such as dysphagia or shortness of breath.

The more serious desmoid tumors appear in the intraabdominal/retroperitoneal/pelvic cavity area and in the head and neck/intrathoracic area. Although desmoid tumors do not occur within vital organs themselves, these locations often involve desmoid tumors attaching to and/or compressing vital organs. For example, intraabdominal desmoid tumors may compress the

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intestines and kidneys, and intrathoracic desmoids may compress the lungs. Similarly, critical blood vessels such as the vena cava and the mesenteric arteries may also be compressed. Compression of organs or vessels can be life-threatening and increased mortality is associated with desmoid tumors in these areas.

Desmoid tumors are often excised and may also be ablated. However, they frequently prove difficult to completely remove, especially when nearby tissues are infiltrated. Moreover, even after apparent complete removal, desmoid tumors quite commonly recur locally. For this reason, medical treatments are heavily used. These include chemotherapy, either systemic or via isolated limb perfusion; hormone-blocking agents such as tamoxifen; kinase inhibitors to arrest tumor progression; and radiation therapy.

Because the behavior of desmoid tumors is unpredictable, active surveillance is recommended as the frontline approach. When progression occurs, the course of treatment is then influenced by the anatomic location of the tumor. For example, surgical removal is favored as the first-line treatment for abdominal wall desmoid tumors, with medical treatment such as chemotherapy as a second-line. For the other areas, medical treatments are usually first-line. Second-line treatment of extremities/shoulder and pelvic girdles/chest wall includes surgery and isolated limb perfusion. Intraabdominal/retroperitoneal/pelvic cavity desmoid tumors are treated with surgery, radiation therapy, or both as second-line. For head and neck/intrathoracic desmoid tumors, second-line treatment is radiation or radiation with surgery. Because of the number of vital organs in the neck, first line treatment may proceed directly to radiation therapy or surgery with radiation.

The Desmoid Tumor Research Foundation is requesting the creation of ICD-10-CM codes for coding specificity and research.

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4. See also: <https://dtrf.org/published-research-articles/>

### TABULAR MODIFICATIONS

- D48 Neoplasm of uncertain behavior of other and unspecified sites
  - D48.1 Neoplasm of uncertain behavior of connective and other soft tissue
    - Neoplasm of uncertain behavior of connective tissue of ear

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Neoplasm of uncertain behavior of connective tissue of eyelid  
Stromal tumors of uncertain behavior of digestive system

Excludes1: neoplasm of uncertain behavior of articular  
cartilage (D48.0)  
neoplasm of uncertain behavior of cartilage of  
larynx (D38.0)  
neoplasm of uncertain behavior of cartilage of  
nose (D38.5)  
neoplasm of uncertain behavior of connective tissue  
of breast (D48.6-)

New subcategory	D48.11	Desmoid tumor
New code	D48.110	Desmoid tumor of head and neck
New code	D48.111	Desmoid tumor of chest wall
New code	D48.112	Desmoid tumor, intrathoracic
New code	D48.113	Desmoid tumor of abdominal wall
New code	D48.114	Desmoid tumor, intraabdominal
Add		Desmoid tumor, peritoneal, retroperitoneal
Add		Desmoid tumor of pelvic cavity
New code	D48.115	Desmoid tumor of limbs and girdles
Add		Desmoid tumor of buttock
Add		Desmoid tumor of lower extremity, and pelvic girdle
Add		Desmoid tumor of upper extremity and shoulder girdle
New code	D48.118	Desmoid tumor of other sites
New code	D48.119	Desmoid tumor of unspecified site Desmoid tumor NOS
New code	D48.19	Other specified neoplasm of uncertain behavior of connective and other soft tissue

## **Electric assisted bicycles**

In the last decade electric assisted bicycles, commonly referred to as e-bicycles, have rapidly proliferated as popular rental and personal devices worldwide. Despite rising ridership, this mode lacks reliable injury epidemiology data. Currently, ICD-10-CM External Cause of Morbidity codes do not distinguish e-bicycle injuries from those sustained on motorcycles. Given that these forms of transportation are regulated differently, draw significantly different ridership, commonly operate on different parts of the public right of way, and travel at different average speeds, it is imperative that distinct e-bicycle injury codes be adopted to facilitate comprehensive, nation-wide e-bicycle injury surveillance.

Electric bicycles are similar in form to conventional bicycles, with two or three wheels propelled partially or entirely by electric power and equipped with a battery. In the United States, e-bicycles are regulated under the Consumer Product Safety Act. Regulation of e-bicycles varies according to state motor vehicle codes or traffic laws, which most commonly regulate e-bicycles as bicycles and clearly distinct from motor vehicles or motorcycles. International literature on e-bicycles does not share a single definition of e-bicycle, and sometimes includes higher-speed vehicle types, such as mopeds, in the same vehicle classification. Currently, ICD-10-CM is not suitable for e-bicycle injury surveillance at the local, state, or national level. Introduction of e-bicycle-specific ICD-10-CM External Cause of Morbidity codes would reduce the likelihood of misclassification.

While e-bicycles are similar in form to motorized bicycles, they should have a distinct classification within the “motorcycle” section of the ICD-10-CM for the following reasons: e-bicycles travel at slower speeds and in a variety of spaces compared to mopeds and motorcycles (sidewalks, bike lanes, shared use pathways, streets versus in-street only); e-bicycles are subject to different regulations than mopeds and motorcycles including helmet use and licensing. Without separate codes, it is impossible to use ICD-10-CM to extract the particular epidemiology of e-bicycle injury. Additionally, to further reduce the likelihood of misclassification, the term “electric assisted bicycle” should be added to the list of “Includes” modes of transport for section V20-V29 and the list of “Excludes” modes of transport for section V10-V19.

Accurate injury surveillance is critical to the successful development, implementation, and evaluation of prevention initiatives. Urban transportation’s rapidly expanding last-mile transit movement and escalating competition between rideshare companies for market share continues to result in increased use of e-bicycles.

The following organizations are requesting new ICD-10-CM External Cause of Morbidity codes to differentiate e-bicycle injuries: 1) UNC Highway Safety Research Center, 2) Vision Zero SF Injury Prevention Research Collaborative (VZIPR): Zuckerberg San Francisco General Hospital and Trauma Center & San Francisco Department of Public Health, 3) University of Tennessee, Knoxville - Department of Civil and Environmental Engineering, 4) University of California, Davis - Institute of Transportation Studies, 5) American College of Surgeons, Committee on

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Trauma, Injury Prevention and Control committee, 6) Portland State University - Transportation Research and Education Center.

**TABULAR MODIFICATIONS**

**Motorcycle rider injured in transport accident (V20-V29)**

Includes: moped  
motorcycle with sidecar  
motorized bicycle  
motor scooter

**V20 Motorcycle rider injured in collision with pedestrian or animal**

New subcategory	V20.0 Motorcycle driver injured in collision with pedestrian or animal in nontraffic accident
New code	V20.01 Electric (assisted) bicycle driver injured in collision with pedestrian or animal in nontraffic accident
New code	V20.09 Other motorcycle driver injured in collision with pedestrian or animal in nontraffic accident
New subcategory	V20.1 Motorcycle passenger injured in collision with pedestrian or animal in nontraffic accident
New code	V20.11 Electric (assisted) bicycle passenger injured in collision with pedestrian or animal in nontraffic accident
New code	V20.19 Other motorcycle passenger injured in collision with pedestrian or animal in nontraffic accident
New subcategory	V20.2 Unspecified motorcycle rider injured in collision with pedestrian or animal in nontraffic accident
New code	V20.21 Unspecified electric (assisted) bicycle rider injured in collision with pedestrian or animal in nontraffic accident
New code	V20.29 Unspecified rider of other motorcycle injured in collision with pedestrian or animal in nontraffic accident
New subcategory	V20.3 Person boarding or alighting a motorcycle injured in collision with pedestrian or animal

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New code	V20.31	Person boarding or alighting an electric (assisted) bicycle injured in collision with pedestrian or animal
New code	V20.39	Person boarding or alighting of other motorcycle injured in collision with pedestrian or animal
New subcategory	V20.4	Motorcycle driver injured in collision with pedestrian or animal in traffic accident
New code	V20.41	Electric (assisted) bicycle driver injured in collision with pedestrian or animal in traffic accident
New code	V20.49	Other motorcycle driver injured in collision with pedestrian or animal in traffic accident
New subcategory	V20.5	Motorcycle passenger injured in collision with pedestrian or animal in traffic accident
New code	V20.51	Electric (assisted) bicycle passenger injured in collision with pedestrian or animal in traffic accident
New code	V20.59	Other motorcycle passenger injured in collision with pedestrian or animal in traffic accident
New subcategory	V20.9	Unspecified motorcycle rider injured in collision with pedestrian or animal in traffic accident
New code	V20.91	Unspecified electric (assisted) bicycle rider injured in collision with pedestrian or animal in traffic accident
New code	V20.99	Unspecified rider of other motorcycle injured in collision with pedestrian or animal in traffic accident
	V21	Motorcycle rider injured in collision with pedal cycle
New subcategory	V21.0	Motorcycle driver injured in collision with pedal cycle in nontraffic accident
New code	V21.01	Electric (assisted) bicycle driver injured in collision with pedal cycle in nontraffic accident
New code	V21.09	Other motorcycle driver injured in collision with pedal cycle in nontraffic accident

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New subcategory	V21.1 Motorcycle passenger injured in collision with pedal cycle in nontraffic accident
New code	V21.11 Electric (assisted) bicycle passenger injured in collision with pedal cycle in nontraffic accident
New code	V21.19 Other motorcycle passenger injured in collision with pedal cycle in nontraffic accident
New subcategory	V21.2 Unspecified motorcycle rider injured in collision with pedal cycle in nontraffic accident
New code	V21.21 Unspecified electric (assisted) bicycle rider injured in collision with pedal cycle in nontraffic accident
New code	V21.29 Unspecified rider of other motorcycle injured in collision with pedal cycle in nontraffic accident
New subcategory	V21.3 Person boarding or alighting a motorcycle injured in collision with pedal cycle
New code	V21.31 Person boarding or alighting an electric (assisted) bicycle injured in collision with pedal cycle
New code	V21.39 Person boarding or alighting of other motorcycle injured in collision with pedal cycle
New subcategory	V21.4 Motorcycle driver injured in collision with pedal cycle in traffic accident
New code	V21.41 Electric (assisted) bicycle driver injured in collision with pedal cycle in traffic accident
New code	V21.49 Other motorcycle driver injured in collision with pedal cycle in traffic accident
New subcategory	V21.5 Motorcycle passenger injured in collision with pedal cycle in traffic accident

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New code	V21.51	Electric (assisted) bicycle passenger injured in collision with pedal cycle in traffic accident
New code	V21.59	Other motorcycle passenger injured in collision with pedal cycle in traffic accident
New subcategory	V21.9	Unspecified motorcycle rider injured in collision with pedal cycle in traffic accident
New code	V21.91	Unspecified electric (assisted) bicycle rider injured in collision with pedal cycle in traffic accident
New code	V21.99	Unspecified rider of other motorcycle injured in collision with pedal cycle in traffic accident
	V22	Motorcycle rider injured in collision with two-or three-wheeled motor vehicle
New subcategory	V22.0	Motorcycle driver injured in collision with two-or three-wheeled motor vehicle in nontraffic accident
New code	V22.01	Electric (assisted) bicycle driver injured in collision with two-or three-wheeled motor vehicle in nontraffic accident
New code	V22.09	Other motorcycle driver injured in collision with two-or three-wheeled motor vehicle in nontraffic accident
New subcategory	V22.1	Motorcycle passenger injured in collision with two-or three-wheeled motor vehicle in nontraffic accident
New code	V22.11	Electric (assisted) bicycle passenger injured in collision with two-or three-wheeled motor vehicle in nontraffic accident
New code	V22.19	Other motorcycle passenger injured in collision with two-or three-wheeled motor vehicle in nontraffic accident
New subcategory	V22.2	Unspecified motorcycle rider injured in collision with two-or three-wheeled motor in nontraffic accident

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New code	V22.21 Unspecified electric (assisted) bicycle rider injured in collision with two-or three-wheeled motor vehicle in nontraffic accident
New code	V22.29 Unspecified rider of other motorcycle injured in collision with two-or three-wheeled motor vehicle in nontraffic accident
New subcategory	V22.3 Person boarding or alighting a motorcycle injured in collision with two-or three-wheeled motor vehicle
New code	V22.31 Person boarding or alighting an electric (assisted) bicycle injured in collision with two-or three-wheeled motor vehicle
New code	V22.39 Person boarding or alighting of other motorcycle injured in collision with two-or three-wheeled motor vehicle
New subcategory	V22.4 Motorcycle driver injured in collision with two-or three-wheeled motor vehicle in traffic accident
New code	V22.41 Electric (assisted) bicycle driver injured in collision with two-or three-wheeled motor vehicle in traffic accident
New code	V22.49 Other motorcycle driver injured in collision with two-or three-wheeled motor vehicle in traffic accident
New subcategory	V22.5 Motorcycle passenger injured in collision with two-or three-wheeled motor vehicle in traffic accident
New code	V22.51 Electric (assisted) bicycle passenger injured in collision with two-or three-wheeled motor vehicle in traffic accident
New code	V22.59 Other motorcycle passenger injured in collision with two-or three-wheeled motor vehicle in traffic accident
New subcategory	V22.9 Unspecified motorcycle rider injured in collision with two-or three-wheeled motor vehicle in traffic accident
New code	V22.91 Unspecified electric (assisted) bicycle rider injured in collision with two-or three-wheeled motor vehicle in traffic accident

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New code	V22.99 Unspecified rider of other motorcycle injured in collision with two-or three-wheeled motor vehicle in traffic accident
	V23 Motorcycle rider injured in collision with car, pick-up truck or van
New subcategory	V23.0 Motorcycle driver injured in collision with car, pick-up truck or van in nontraffic accident
New code	V23.01 Electric (assisted) bicycle driver injured in collision with car, pick-up truck or van in nontraffic accident
New code	V23.09 Other motorcycle driver injured in collision with car, pick-up truck or van in nontraffic accident
New subcategory	V23.1 Motorcycle passenger injured in collision with car, pick-up truck or van in nontraffic accident
New code	V23.11 Electric (assisted) bicycle passenger injured in collision with car, pick-up truck or van in nontraffic accident
New code	V23.19 Other motorcycle passenger injured in collision with car, pick-up truck or van in nontraffic accident
New subcategory	V23.2 Unspecified motorcycle rider injured in collision with car, pick-up truck or van in nontraffic accident
New code	V23.21 Unspecified electric (assisted) bicycle rider injured in collision with car, pick-up truck or van in nontraffic accident
New code	V23.29 Unspecified rider of other motorcycle injured in collision with car, pick-up truck or van in nontraffic accident
New subcategory	V23.3 Person boarding or alighting a motorcycle injured in collision with car, pick-up truck or van
New code	V23.31 Person boarding or alighting an electric (assisted) bicycle injured in collision with car, pick-up truck or van

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New code V23.39 Person boarding or alighting of other motorcycle injured in collision with car, pick-up truck or van

New subcategory V23.4 Motorcycle driver injured in collision with car, pick-up truck or van in traffic accident

New code V23.41 Electric (assisted) bicycle driver injured in collision with car, pick-up truck or van in traffic accident

New code V23.49 Other motorcycle driver injured in collision with car, pick-up truck or van in traffic accident

New subcategory V23.5 Motorcycle passenger injured in collision with car, pick-up truck or van in traffic accident

New code V23.51 Electric (assisted) bicycle passenger injured in collision with car, pick-up truck or van in traffic accident

New code V23.59 Other motorcycle passenger injured in collision with car, pick-up truck or van in traffic accident

New subcategory V23.9 Unspecified motorcycle rider injured in collision with car, pick-up truck or van in traffic accident

New code V23.91 Unspecified electric (assisted) bicycle rider injured in collision with car, pick-up truck or van in traffic accident

New code V23.99 Unspecified rider of other motorcycle injured in collision with car, pick-up truck or van in traffic accident

V24 Motorcycle rider injured in collision with heavy transport vehicle or bus

New subcategory V24.0 Motorcycle driver injured in collision with heavy transport vehicle or bus in nontraffic accident

New code V24.01 Electric (assisted) bicycle driver injured in collision with heavy transport vehicle or bus in nontraffic accident

New code V24.09 Other motorcycle driver injured in collision with heavy transport vehicle or bus in nontraffic accident

New subcategory V24.1 Motorcycle passenger injured in collision with heavy transport vehicle or bus in nontraffic accident

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New code	V24.11 Electric (assisted) bicycle passenger injured in collision with heavy transport vehicle or bus in nontraffic accident
New code	V24.19 Other motorcycle passenger injured in collision with heavy transport vehicle or bus in nontraffic accident
New subcategory	V24.2 Unspecified motorcycle rider injured in collision with heavy transport vehicle or bus in nontraffic accident
New code	V24.21 Unspecified electric (assisted) bicycle rider injured in collision with heavy transport vehicle or bus in nontraffic accident
New code	V24.29 Unspecified rider of other motorcycle injured in collision with heavy transport vehicle or bus in nontraffic accident
New subcategory	V24.3 Person boarding or alighting a motorcycle injured in collision with heavy transport vehicle or bus
New code	V24.31 Person boarding or alighting an electric (assisted) bicycle injured in collision with heavy transport vehicle or bus
New code	V24.39 Person boarding or alighting of other motorcycle injured in collision with heavy transport vehicle or bus
New subcategory	V24.4 Motorcycle driver injured in collision with heavy transport vehicle or bus in traffic accident
New code	V24.41 Electric (assisted) bicycle driver injured in collision with heavy transport vehicle or bus in traffic accident
New code	V24.49 Other motorcycle driver injured in collision with heavy transport vehicle or bus in traffic accident
New subcategory	V24.5 Motorcycle passenger injured in collision with heavy transport vehicle or bus in traffic accident
New code	V24.51 Electric (assisted) bicycle passenger injured in collision with heavy transport vehicle or bus in traffic accident
New code	V24.59 Other motorcycle passenger injured in collision with heavy transport vehicle or bus in traffic accident

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New subcategory	V24.9 Unspecified motorcycle rider injured in collision with heavy transport vehicle or bus in traffic accident
New code	V24.91 Unspecified electric (assisted) bicycle rider injured in heavy transport vehicle or bus in traffic accident
New code	V24.99 Unspecified rider of other motorcycle injured in collision heavy transport vehicle or bus in traffic accident
V25 Motorcycle rider injured in collision with railway train or railway vehicle	
New subcategory	V25.0 Motorcycle driver injured in collision with railway train or railway vehicle in nontraffic accident
New code	V25.01 Electric (assisted) bicycle driver injured in collision with railway train or railway vehicle in nontraffic accident
New code	V25.09 Other motorcycle driver injured in collision with railway train or railway vehicle in nontraffic accident
New subcategory	V25.1 Motorcycle passenger injured in collision with railway train or railway vehicle in nontraffic accident
New code	V25.11 Electric (assisted) bicycle passenger injured in collision with railway train or railway vehicle in nontraffic accident
New code	V25.19 Other motorcycle passenger injured in collision with railway train or railway vehicle in nontraffic accident
New subcategory	V25.2 Unspecified motorcycle rider injured in collision with railway train or railway vehicle in nontraffic accident
New code	V25.21 Unspecified electric (assisted) bicycle rider injured in collision with railway train or railway vehicle in nontraffic accident
New code	V25.29 Unspecified rider of other motorcycle injured in collision with railway train or railway vehicle in nontraffic accident
New subcategory	V25.3 Person boarding or alighting a motorcycle injured in collision with railway train or railway vehicle

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New code	V25.31	Person boarding or alighting an electric (assisted) bicycle injured in collision with railway train or railway vehicle
New code	V25.39	Person boarding or alighting of other motorcycle injured in collision with railway train or railway vehicle
New subcategory	V25.4	Motorcycle driver injured in collision with railway train or railway vehicle in traffic accident
New code	V25.41	Electric (assisted) bicycle driver injured in collision with railway train or railway vehicle in traffic accident
New code	V25.49	Other motorcycle driver injured in collision with railway train or railway vehicle in traffic accident
New subcategory	V25.5	Motorcycle passenger injured in collision with railway train or railway vehicle in traffic accident
New code	V25.51	Electric (assisted) bicycle passenger injured in collision with railway train or railway vehicle in traffic accident
New code	V25.99	Other motorcycle passenger injured in collision with railway train or railway vehicle in traffic accident
New subcategory	V25.9	Unspecified motorcycle rider injured in collision with railway train or railway vehicle in traffic accident
New code	V25.91	Unspecified electric (assisted) bicycle rider injured in collision with railway train or railway vehicle in traffic accident
New code	V25.99	Unspecified rider of other motorcycle injured in collision with railway train or railway vehicle in traffic accident
	V26	Motorcycle rider injured in collision with other nonmotor vehicle
New subcategory	V26.0	Motorcycle driver injured in collision with other nonmotor vehicle in nontraffic accident
New code	V26.01	Electric (assisted) bicycle driver injured in collision with other nonmotor vehicle in nontraffic accident
New code	V26.09	Other motorcycle driver injured in collision with other nonmotor vehicle in nontraffic accident

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New subcategory	V26.1 Motorcycle passenger injured in collision other nonmotor vehicle in nontraffic accident
New code	V26.11 Electric (assisted) bicycle passenger injured in collision with other nonmotor vehicle in nontraffic accident
New code	V26.19 Other motorcycle passenger injured in collision with other nonmotor vehicle in nontraffic accident
New subcategory	V26.2 Unspecified motorcycle rider injured in collision with other nonmotor vehicle in nontraffic accident
New code	V26.21 Unspecified electric (assisted) bicycle rider injured in collision with other nonmotor vehicle in nontraffic accident
New code	V26.29 Unspecified rider of other motorcycle injured in collision with other nonmotor vehicle in nontraffic accident
New subcategory	V26.3 Person boarding or alighting a motorcycle injured in collision with other nonmotor vehicle
New code	V26.31 Person boarding or alighting an electric (assisted) bicycle injured in collision with other nonmotor vehicle
New code	V26.39 Person boarding or alighting of other motorcycle injured in collision with other nonmotor vehicle
New subcategory	V26.4 Motorcycle driver injured in collision with other nonmotor vehicle in traffic accident
New code	V26.41 Electric (assisted) bicycle driver injured in collision with other nonmotor vehicle in traffic accident
New code	V26.49 Other motorcycle driver injured in collision with other nonmotor vehicle in traffic accident
New subcategory	V26.5 Motorcycle passenger injured in collision with other nonmotor vehicle in traffic accident
New code	V26.51 Electric (assisted) bicycle passenger injured in collision with other nonmotor vehicle in traffic accident
New code	V26.59 Other motorcycle passenger injured in collision with other nonmotor vehicle in traffic accident

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New subcategory	V26.9 Unspecified motorcycle rider injured in collision with other nonmotor vehicle in traffic accident
New code	V26.91 Unspecified electric (assisted) bicycle rider injured in collision with nonmotor vehicle in traffic accident
New code	V26.99 Unspecified rider of other motorcycle injured in collision with other nonmotor vehicle in traffic accident
V27 Motorcycle rider injured in collision with fixed or stationary object	
New subcategory	V27.0 Motorcycle driver injured in collision with fixed or stationary object in nontraffic accident
New code	V27.01 Electric (assisted) bicycle driver injured in collision with fixed or stationary object in nontraffic accident
New code	V27.09 Other motorcycle driver injured in collision with fixed or stationary object in nontraffic accident
New subcategory	V27.1 Motorcycle passenger injured in collision with fixed or stationary object in nontraffic accident
New code	V27.11 Electric (assisted) bicycle passenger injured in collision with fixed or stationary object in nontraffic accident
New code	V27.19 Other motorcycle passenger injured in collision with fixed or stationary object in nontraffic accident
New subcategory	V27.2 Unspecified motorcycle rider injured in collision with fixed or stationary object in nontraffic accident
New code	V27.21 Unspecified electric (assisted) bicycle rider injured in collision with fixed or stationary object in nontraffic accident
New code	V27.29 Unspecified rider of other motorcycle injured in collision with fixed or stationary object in nontraffic accident
New subcategory	V27.3 Person boarding or alighting a motorcycle injured in collision with fixed or stationary object

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New code	V27.31	Person boarding or alighting an electric (assisted) bicycle injured in collision with fixed or stationary object
New code	V27.39	Person boarding or alighting of other motorcycle injured in collision with fixed or stationary object
New subcategory	V27.4	Motorcycle driver injured in collision with fixed or stationary object in traffic accident
New code	V27.41	Electric (assisted) bicycle driver injured in collision with fixed or stationary object in traffic accident
New code	V27.49	Other motorcycle driver injured in collision with fixed or stationary object in traffic accident
New subcategory	V27.5	Motorcycle passenger injured in collision with fixed or stationary object in traffic accident
New code	V27.51	Electric (assisted) bicycle passenger injured in collision with fixed or stationary object in traffic accident
New code	V27.99	Other motorcycle passenger injured in collision with fixed or stationary object in traffic accident
New subcategory	V27.9	Unspecified motorcycle rider injured in collision with fixed or stationary object in traffic accident
New code	V27.91	Unspecified electric (assisted) bicycle rider injured in collision with fixed or stationary object in traffic accident
New code	V27.99	Unspecified rider of other motorcycle injured in collision with fixed or stationary object in traffic accident
	V28	Motorcycle rider injured in noncollision transport accident
New subcategory	V28.0	Motorcycle driver injured in noncollision transport accident in nontraffic accident
New code	V28.01	Electric (assisted) bicycle driver injured in noncollision transport accident in nontraffic accident

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New code	V28.09 Other motorcycle driver injured in noncollision transport accident in nontraffic accident
New subcategory	V28.1 Motorcycle passenger injured in noncollision transport accident in nontraffic accident
New code	V28.11 Electric (assisted) bicycle passenger injured in noncollision transport accident in nontraffic accident
New code	V28.19 Other motorcycle passenger injured in noncollision transport accident in nontraffic accident
New subcategory	V28.2 Unspecified motorcycle rider injured in noncollision transport accident in nontraffic accident
New code	V28.21 Unspecified electric (assisted) bicycle rider injured in noncollision transport accident in nontraffic accident
New code	V28.29 Unspecified rider of other motorcycle injured in noncollision transport accident in nontraffic accident
New subcategory	V28.3 Person boarding or alighting a motorcycle injured in noncollision transport accident
New code	V28.31 Person boarding or alighting an electric (assisted) bicycle injured in noncollision with transport accident
New code	V28.39 Person boarding or alighting of other motorcycle injured in noncollision with transport accident
New subcategory	V28.4 Motorcycle driver injured in noncollision transport accident in traffic accident
New code	V28.41 Electric (assisted) bicycle driver injured in noncollision transport accident in traffic accident
New code	V28.49 Other motorcycle driver injured in noncollision transport accident in traffic accident
New subcategory	V28.5 Motorcycle passenger injured in noncollision transport accident in traffic accident
New code	V28.51 Electric (assisted) bicycle passenger injured in noncollision transport accident in traffic accident

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New code	V28.59 Other motorcycle passenger injured in noncollision transport accident in traffic accident
New subcategory	V28.9 Unspecified motorcycle rider injured in noncollision transport accident in traffic accident
New code	V28.91 Unspecified electric (assisted) bicycle rider injured in noncollision transport accident in traffic accident
New code	V28.99 Unspecified rider of other motorcycle injured in noncollision transport accident in traffic accident
	V29 Motorcycle rider injured in other and unspecified transport accident
	V29.0 Motorcycle driver injured in collision with other and unspecified motor vehicles in nontraffic accident
New subcategory	V29.00 Motorcycle driver injured in collision with unspecified motor vehicles in nontraffic accident
New code	V29.001 Electric (assisted) bicycle driver injured in collision with unspecified motor vehicles in nontraffic accident
New code	V29.008 Other motorcycle driver injured in collision with unspecified motor vehicles in nontraffic accident
New subcategory	V29.09 Motorcycle driver injured in collision with other motor vehicles in nontraffic accident
New code	V29.091 Electric (assisted) bicycle driver injured in collision with other motor vehicles in nontraffic accident
New code	V29.098 Other motorcycle driver injured in collision with other motor vehicles in nontraffic accident
	V29.1 Motorcycle passenger injured in collision with other and unspecified motor vehicles in nontraffic accident
New subcategory	V29.10 Motorcycle passenger injured in collision with unspecified motor vehicles in nontraffic accident

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New code	V29.101 Electric (assisted) bicycle passenger injured in collision with unspecified motor vehicles in nontraffic accident
New code	V29.198 Other motorcycle passenger injured in collision with unspecified motor vehicles in nontraffic accident
New subcategory	V29.19 Motorcycle passenger injured in collision with other motor vehicles in nontraffic accident
New code	V29.191 Electric (assisted) bicycle passenger injured in collision with other motor vehicles in nontraffic accident
New code	V29.198 Other motorcycle passenger injured in collision with other motor vehicles in nontraffic accident
New subcategory	V29.2 Unspecified motorcycle rider injured in collision with other and unspecified motor vehicles in nontraffic accident
New subcategory	V29.20 Unspecified motorcycle rider injured in collision with unspecified motor vehicles in nontraffic accident
Delete	<del>Motorcycle collision NOS, nontraffic</del>
New code	V29.201 Unspecified electric (assisted) bicycle rider injured in collision with unspecified motor vehicles in nontraffic accident
New code	V29.208 Unspecified rider of other motorcycle injured in collision with unspecified motor vehicle in nontraffic accident
Add	Motorcycle collision NOS, nontraffic
New subcategory	V29.29 Unspecified motorcycle rider injured in collision with other motor vehicles in nontraffic accident
New code	V29.291 Unspecified electric (assisted) bicycle rider injured in collision with other motor vehicles in nontraffic accident
New code	V29.298 Unspecified rider of other motorcycle rider injured in collision with other motor vehicles in nontraffic accident

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New subcategory	V29.3 Motorcycle rider (driver) (passenger) injured in unspecified nontraffic accident
Delete	<del>Motorcycle accident NOS, nontraffic</del>
Delete	<del>Motorcycle rider injured in nontraffic accident NOS</del>
New code	V29.31 Electric (assisted) bicycle (driver) (passenger) injured in unspecified nontraffic accident
New code	V29.39 Other motorcycle (driver) (passenger) injured in unspecified nontraffic accident
Add	Motorcycle accident NOS, nontraffic
Add	Motorcycle rider injured in nontraffic accident NOS
	V29.4 Motorcycle driver injured in collision with other and unspecified motor vehicles in traffic accident
New subcategory	V29.40 Motorcycle driver injured in collision with unspecified motor vehicles in traffic accident
New code	V29.401 Electric (assisted) bicycle driver injured in collision with unspecified motor vehicles in traffic accident
New code	V29.408 Other motorcycle driver injured in collision with unspecified motor vehicles in traffic accident
New subcategory	V29.49 Motorcycle driver injured in collision with other motor vehicles in traffic accident
New code	V29.491 Electric (assisted) bicycle driver injured in collision with other motor vehicles in traffic accident
New code	V29.498 Other motorcycle driver injured in collision with other motor vehicles in traffic accident
	V29.5 Motorcycle passenger injured in collision with other and unspecified motor vehicles in traffic accident
New subcategory	V29.50 Motorcycle passenger injured in collision with unspecified motor vehicles in traffic accident

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New code	V29.501 Electric (assisted) bicycle passenger injured in collision with unspecified motor vehicles in traffic accident
New code	V29.508 Other motorcycle passenger injured in collision with unspecified motor vehicles in traffic accident
New subcategory	V29.59 Motorcycle passenger injured in collision with other motor vehicles in traffic accident
New code	V29.591 Electric (assisted) bicycle passenger injured in collision with other motor vehicles in traffic accident
New code	V29.598 Other motorcycle passenger injured in collision with other motor vehicles in traffic accident
	V29.6 Unspecified motorcycle rider injured in collision with other and unspecified motor vehicles in traffic accident
New subcategory	V29.60 Unspecified motorcycle rider injured in collision with unspecified motor vehicles in traffic accident
Delete	<del>Motorcycle collision NOS (traffic)</del>
New code	V29.601 Unspecified electric (assisted) bicycle rider injured in collision with unspecified motor vehicles in traffic accident
New code	V29.608 Unspecified rider of other motorcycle injured in collision with unspecified motor vehicle in traffic accident
Add	Motorcycle collision NOS (traffic)
New subcategory	V29.69 Unspecified motorcycle rider injured in collision with other motor vehicles in traffic accident
New code	V29.691 Unspecified electric (assisted) bicycle rider injured in collision with other motor vehicles in traffic accident
New code	V29.698 Unspecified rider of other motorcycle rider injured in collision with other motor vehicle in traffic accident

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V29.8 Motorcycle rider (driver) (passenger) injured in other specified  
transport accidents

New subcategory	V29.81 Motorcycle rider (driver) (passenger) injured in transport accident with military vehicle
New code	V29.811 Electric (assisted) bicycle rider (driver) (passenger) injured in transport accident with military vehicle
New code	V29.818 Rider (driver) (passenger) of other motorcycle injured in transport accident with military vehicle
New subcategory	V29.88 Motorcycle rider (driver) (passenger) injured in other specified transport accidents
New code	V29.881 Electric (assisted) bicycle rider (driver) (passenger) injured in other specified transport accidents
New code	V29.888 Rider (driver) (passenger) of other motorcycle injured in other specified transport accidents
New subcategory	V29.9 Motorcycle rider (driver) (passenger) injured in unspecified traffic accident
Delete	<del>Motorcycle accident NOS</del>
New code	V29.91 Electric (assisted) bicycle (driver) (passenger) injured in unspecified traffic accident
New code	V29.99 Other motorcycle (driver) (passenger) injured in unspecified traffic accident
Add	Motorcycle accident NOS

## **Encounter for follow-up examination after completed treatment for malignant neoplasm**

Following diagnosis and treatment of malignant tumors, regular patient follow-up is essential. This is consequent to known risks of tumor recurrences, risk of metastasis of select tumors, and the enhanced probability of generating additional primary cutaneous cancers following an initial diagnosis.

Consequent to these known risks, the National Comprehensive Cancer Network (NCCN) guidelines recommend continued lifetime surveillance following a diagnosis of malignant neoplasm, with follow-up intervals guided by the type(s) of treated tumors. In relation to skin cancers, the American Academy of Dermatology Association (AADA) published Guidelines of Care stipulating continued evaluations for new primary skin cancers on at least an annual basis following a diagnosis of squamous cell or basal cell carcinoma.

The American Academy of Dermatology/Association now seeks to bring over the list of codes from WHO ICD-10 that did not transition to ICD-10-CM for code category Z08 to include follow-up encounters after completed treatment for malignant neoplasms using other treatment modalities, including surgical.

### **TABULAR MODIFICATIONS**

	Z08	Encounter for follow-up examination after completed treatment for malignant neoplasm
New code	Z08.0	Encounter for follow-up examination after surgery for malignant neoplasm
Add		Medical surveillance following surgery
New code	Z08.1	Encounter for follow-up examination after radiotherapy for malignant neoplasm
Add		Medical surveillance following completed treatment, radiotherapy
New code	Z08.2	Encounter for follow-up examination after chemotherapy for malignant neoplasm
Add		Medical surveillance following completed treatment, chemotherapy
New code	Z08.8	Encounter for follow-up examination after other treatment for malignant neoplasm
Add		Medical surveillance following completed treatment, hormone therapy
Add		Medical surveillance following completed treatment, immunotherapy
Add		Medical surveillance following completed treatment, targeted therapy



## **Encounter for Vaginal Delivery Requiring Assistance**

The American College of Obstetricians and Gynecologists (ACOG) is proposing to add a new code to specify encounters for full term requiring assistance in vaginal deliveries to identify this condition when found, to track the frequency at which it occurs, as well as to allow mechanized tracking of the most effective treatment modalities through diagnosis code searches in EMR datasets.

The rates of cesarean births have gained national and international attention as efforts to reduce maternal mortality have identified unnecessary cesarean births as a confounder. The use of vacuum, obstetric forceps, though decreased in use, continues to be an important tool for labor management and avoiding cesarean birth. In 2013, 3.3% of vaginal births utilized instruments, referred to as operative vaginal birth. Indications for operative vaginal birth includes a prolonged second stage of labor, potential fetal compromise, and/or maternal issues (exhaustion, medical issues) that warrant shortening the second stage of labor.

Currently, ICD-10-CM does not include a code for successful instrumentation assisted vaginal delivery. The codes for documenting delivery are limited to O80, Encounter for full-term uncomplicated delivery, and O82, Encounter for cesarean delivery without indication. Additionally, the ICD-10 Code O80 instructional notes state, “Delivery requiring minimal or no assistance, with or without episiotomy, without fetal manipulation [e.g., rotation version] or instrumentation [forceps] of a spontaneous, cephalic, vaginal, full-term, single, live-born infant.”

This definition limits the ability to capture successful assistance via instrumentation vaginal birth.

ACOG requests the following modifications:

### References

<sup>[i]</sup> World Health Organization. WHO Statement on Cesarean Section Rates. 2015. Available at: [https://www.who.int/reproductivehealth/publications/maternal\\_perinatal\\_health/cs-statement/en/](https://www.who.int/reproductivehealth/publications/maternal_perinatal_health/cs-statement/en/)

<sup>[ii]</sup> Martin JA, Hamilton BE, Osterman MJ, Curtin SC, Matthews TJ. Births: final data for 2013. Natl Vital Stat Rep 2015;64:1–65

<sup>[iii]</sup> Operative vaginal birth. ACOG Practice Bulletin No. 219. American College of Obstetricians and Gynecologists. Obstet Gynecol 2020;135:e149–59.

**TABULAR MODIFICATIONS**

Revise	O80 Encounter for full-term uncomplicated <u>vaginal</u> delivery
Revise	<u>Vaginal</u> delivery requiring minimal or no assistance, with or without episiotomy, without instrumentation [forceps] of a spontaneous, cephalic, vaginal, full-term, single, live-born infant Use additional code to indicate outcome of delivery (Z37.0)
New code	O81 Encounter for vaginal delivery requiring assistance via instrumentation [forceps or vacuum], with or without episiotomy, of a spontaneous, cephalic, full-term, single, live-born infant
Add	Use additional code to indicate outcome of delivery (Z37.0)

## 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. This was previously presented at the September 2020 and the March 2021 Coordination and Maintenance meetings and is being represented with changes. These changes are in bold.

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.

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

#### **J93 Pneumothorax and air leak**

##### **J93.1 Other spontaneous pneumothorax**

##### **J93.12 Secondary spontaneous pneumothorax**

**Code first underlying condition, such as:**

**catamenial pneumothorax due to endometriosis**

**(~~N80.8~~N80.B-)**

Revise

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N80 Endometriosis

N80.0 Endometriosis of uterus

Delete	<del>Adenomyosis</del>
Add	Endometriosis of the cervix
	Excludes1: stromal endometriosis (D39.0)
New code	N80.00 Endometriosis of the uterus, unspecified
New code	N80.01 Superficial endometriosis of the uterus
New code	N80.02 Deep endometriosis of the uterus
Add	Deep retrocervical endometriosis
New code	N80.03 Adenomyosis of the uterus
Add	Adenomyosis NOS

N80.1 Endometriosis of ovary

<b>New</b>	
<b>sub-subcategory</b>	<b>N80.10 Endometriosis of ovary, unspecified depth</b>
<b>New code</b>	<b>N80.101 Endometriosis of right ovary, unspecified depth</b>
<b>New code</b>	<b>N80.102 Endometriosis of left ovary, unspecified depth</b>
<b>New code</b>	<b>N80.103 Endometriosis of bilateral ovaries, unspecified depth</b>
<b>New code</b>	<b>N80.109 Endometriosis of unspecified ovary, unspecified depth</b>
<b>Add</b>	<b>Endometriosis of ovary NOS</b>

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

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N80.2 Endometriosis of fallopian tube

<b>New</b>	
<b>sub-subcategory</b>	<b>N80.20 Endometriosis of fallopian tube, unspecified depth</b>
<b>New code</b>	<b>N80.201 Endometriosis of right fallopian tube, unspecified depth</b>
<b>New code</b>	<b>N80.202 Endometriosis of left fallopian tube, unspecified depth</b>
<b>New code</b>	<b>N80.203 Endometriosis of bilateral fallopian tubes, unspecified depth</b>
<b>New code</b>	<b>N80.209 Endometriosis of unspecified fallopian tube, unspecified depth</b>
<b>Add</b>	<b>Endometriosis fallopian tube NOS</b>

New	N80.21 Superficial endometriosis of fallopian tube
sub-subcategory	
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	N80.22 Deep endometriosis of the fallopian tube
sub-subcategory	Deep endometriosis involving muscular wall of fallopian tube
Add	
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

New code	N80.30 Endometriosis of pelvic peritoneum, unspecified
<b>Add</b>	<b>Endometriosis of the retroperitoneum NOS</b>

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<b>New</b>	
<b>sub-subcategory</b>	<b>N80.31 Endometriosis of the anterior cul-de-sac</b>
<b>New code</b>	<b>N80.311 Superficial endometriosis of the anterior cul-de-sac</b>
<b>New code</b>	<b>N80.312 Deep endometriosis of the anterior cul-de-sac</b>
<b>New code</b>	<b>N80.319 Endometriosis of the anterior cul-de-sac, unspecified depth</b>
<b>Add</b>	<b>Endometriosis of the anterior cul-de-sac NOS</b>
<b>New</b>	
<b>sub-subcategory</b>	<b>N80.32 Endometriosis of the posterior cul-de-sac</b>
<b>New code</b>	<b>N80.321 Superficial endometriosis of the posterior cul-de-sac</b>
<b>New code</b>	<b>N80.322 Deep endometriosis of the posterior cul-de-sac</b>
<b>New code</b>	<b>N80.329 Endometriosis of the posterior cul-de-sac, unspecified depth</b>
<b>Add</b>	<b>Endometriosis of the posterior cul-de-sac NOS</b>
<b>New</b>	
<b>sub-subcategory</b>	<b>N80.33 Superficial endometriosis of the pelvic sidewall</b>
<b>New code</b>	<b>N80.331 Superficial endometriosis of the right pelvic sidewall</b>
<b>New code</b>	<b>N80.332 Superficial endometriosis of the left pelvic sidewall</b>
<b>New code</b>	<b>N80.333 Superficial endometriosis of bilateral pelvic sidewall</b>
<b>New code</b>	<b>N80.339 Superficial endometriosis of pelvic sidewall, unspecified</b>
<b>New</b>	
<b>sub-subcategory</b>	<b>N80.34 Deep endometriosis of the pelvic sidewall</b>
<b>New code</b>	<b>N80.341 Deep endometriosis of the right pelvic sidewall</b>
<b>New code</b>	<b>N80.342 Deep endometriosis of the left pelvic sidewall</b>
<b>New code</b>	<b>N80.343 Deep endometriosis of the bilateral pelvic sidewall</b>
<b>New code</b>	<b>N80.349 Deep endometriosis of the pelvic sidewall, unspecified</b>

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<b>New sub-subcategory</b>	<b>N80.35 Endometriosis of the pelvic sidewall, unspecified depth</b>
<b>New code</b>	<b>N80.351 Endometriosis of the right pelvic sidewall, unspecified depth</b>
<b>New code</b>	<b>N80.352 Endometriosis of the left pelvic sidewall, unspecified depth</b>
<b>New code</b>	<b>N80.353 Endometriosis of bilateral pelvic sidewall, unspecified depth</b>
<b>New code</b>	<b>N80.359 Endometriosis of pelvic sidewall, unspecified side, unspecified depth</b>
<b>Add</b>	<b>Endometriosis of the pelvic sidewall NOS</b>
<b>New sub subcategory</b>	<b>N80.36 Superficial endometriosis of the pelvic brim</b>
<b>New code</b>	<b>N80.361 Superficial endometriosis of the right pelvic brim</b>
<b>New code</b>	<b>N80.362 Superficial endometriosis of the left pelvic brim</b>
<b>New code</b>	<b>N80.363 Superficial endometriosis of bilateral pelvic brim</b>
<b>New code</b>	<b>N80.369 Superficial endometriosis of the pelvic brim, unspecified</b>
<b>New sub subcategory</b>	<b>N80.37 Deep endometriosis of the pelvic brim</b>
<b>New code</b>	<b>N80.371 Deep endometriosis of the right pelvic brim</b>
<b>New code</b>	<b>N80.372 Deep endometriosis of the left pelvic brim</b>
<b>New code</b>	<b>N80.373 Deep endometriosis of bilateral pelvic brim</b>
<b>New code</b>	<b>N80.379 Deep endometriosis of the pelvic brim, unspecified</b>
<b>New sub subcategory</b>	<b>N80.38 Endometriosis of the pelvic brim, unspecified depth</b>
<b>New code</b>	<b>N80.381 Endometriosis of the right pelvic brim, unspecified depth</b>
<b>New code</b>	<b>N80.382 Endometriosis of the left pelvic brim, unspecified depth</b>
<b>New code</b>	<b>N80.383 Endometriosis of bilateral pelvic brim, unspecified depth</b>

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<b>New code</b>	<b>N80.389 Endometriosis of the pelvic brim, unspecified side, unspecified depth</b>
<b>Add</b>	<b>Endometriosis of the pelvic brim NOS</b>
<b>New subcategory</b>	<b>N80.3A Superficial endometriosis of the uterosacral ligament(s)</b>
<b>New code</b>	<b>N80.3A1 Superficial endometriosis of the right uterosacral ligament</b>
<b>New code</b>	<b>N80.3A2 Superficial endometriosis of the left uterosacral ligament</b>
<b>New code</b>	<b>N80.3A3 Superficial endometriosis of the bilateral uterosacral ligaments</b>
<b>New code</b>	<b>N80.3A9 Superficial endometriosis of the uterosacral ligament(s), unspecified</b>
<b>New subcategory</b>	<b>N80.3B Deep endometriosis of the uterosacral ligament(s)</b>
<b>New code</b>	<b>N80.3B1 Deep endometriosis of the right uterosacral ligament</b>
<b>New code</b>	<b>N80.3B2 Deep endometriosis of the left uterosacral ligament</b>
<b>New code</b>	<b>N80.3B3 Deep endometriosis of bilateral uterosacral ligament(s)</b>
<b>New code</b>	<b>N80.3B9 Deep endometriosis of the uterosacral ligament(s), unspecified</b>
<b>New Subcategory</b>	<b>N80.3C Endometriosis of the uterosacral ligament(s), unspecified depth</b>
<b>New code</b>	<b>N80.3C1 Endometriosis of the right uterosacral ligament, unspecified depth</b>
<b>New code</b>	<b>N80.3C2 Endometriosis of the left uterosacral ligament, unspecified depth</b>
<b>New code</b>	<b>N80.3C3 Endometriosis of bilateral uterosacral ligament(s), unspecified depth</b>
<b>New code</b>	<b>N80.3C9 Endometriosis of the uterosacral ligament(s), unspecified side, unspecified depth</b>
<b>Add</b>	<b>Endometriosis of the uterosacral ligament(s) NOS</b>
<b>New sub-subcategory</b>	<b>N80.39 Endometriosis of other pelvic peritoneum</b>
<b>New code</b>	<b>N80.391 Superficial endometriosis of the pelvic peritoneum, other specified sites</b>

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New code	N80.392 Deep endometriosis of the pelvic peritoneum, other specified sites
New code	N80.399 Endometriosis of the pelvic peritoneum, other specified sites, unspecified depth
<b>New code</b>	<b>N80.40 Endometriosis of the rectovaginal septum unspecified involvement of vagina</b>
<b>Add</b>	<b>Endometriosis of the rectovaginal septum, NOS</b>
New code	N80.41 Endometriosis of rectovaginal septum without involvement of vagina
New code	N80.42 Endometriosis of rectovaginal septum with involvement of vagina
	N80.5 Endometriosis of intestine
	N80.50 Endometriosis of intestine, unspecified
New sub-subcategory	N80.51 Endometriosis 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
<b>New code</b>	<b>N80.519 Endometriosis of the rectum, unspecified depth</b>
<b>Add</b>	<b>Endometriosis of the rectum NOS</b>
New sub-subcategory	N80.52 Endometriosis of the sigmoid colon
New code	N80.521 Superficial endometriosis of the sigmoid colon
New code	N80.522 Deep endometriosis of the sigmoid colon
<b>New code</b>	<b>N80.529 Endometriosis of the sigmoid colon, unspecified depth</b>
<b>Add</b>	<b>Endometriosis of the sigmoid colon NOS</b>
New sub-subcategory	N80.53 Endometriosis of the cecum
New code	N80.531 Superficial endometriosis of the cecum
Add	N80.532 Deep endometriosis of the cecum
<b>New code</b>	<b>N80.539 Endometriosis of the cecum, unspecified depth</b>
<b>Add</b>	<b>Endometriosis of the cecum NOS</b>

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New sub-subcategory	N80.54 Endometriosis of the appendix
New code	N80.541 Superficial endometriosis of the appendix
New code	N80.542 Deep endometriosis of the appendix
<b>New code</b>	<b>N80.549 Endometriosis of the appendix, unspecified depth</b>
<b>Add</b>	<b>Endometriosis of the appendix NOS</b>
New sub-subcategory	N80.55 Endometriosis <b>of other parts</b> of the colon
<b>Add</b>	<b>Endometriosis of descending colon</b>
<b>Add</b>	<b>Endometriosis of transverse colon</b>
New code	N80.551 Superficial endometriosis <b>of other parts</b> of the colon
New code	N80.552 Deep endometriosis <b>of other parts</b> of the colon
New code	N80.559 Endometriosis <b>of other parts</b> of the colon, unspecified depth
<b>Add</b>	<b>Endometriosis of colon NOS</b>
New sub-subcategory	N80.56 Endometriosis of the small intestine
New code	N80.561 Superficial endometriosis of the small intestine
New code	N80.562 Deep endometriosis of the small intestine
Add	Deep endometriosis of the small intestine, multifocal
<b>New code</b>	<b>N80.569 Endometriosis of the small intestine, unspecified depth</b>
<b>Add</b>	<b>Endometriosis of the small intestine NOS</b>
New subcategory	N80.A Endometriosis of bladder and ureters
<b>New code</b>	<b>N80.A0 Endometriosis of bladder, unspecified depth</b>
<b>Add</b>	<b>Endometriosis of bladder NOS</b>
New code	N80.A1 Superficial endometriosis of bladder
New code	N80.A2 Deep endometriosis of bladder
<b>New sub-subcategory</b>	<b>N80.A4 Superficial endometriosis of ureter</b>
<b>Add</b>	<b>Extrinsic endometriosis of ureter</b>
<b>Add</b>	<b>Code also obstructive and reflux uropathy (N13.-)</b>

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New code	<b>N80.A41 Superficial endometriosis of right ureter</b>
New code	<b>N80.A42 Superficial endometriosis of left ureter</b>
New code	<b>N80.A43 Superficial endometriosis of bilateral ureters</b>
New code	<b>N80.A49 Superficial endometriosis of unspecified ureter</b>
New sub-subcategory	<b>N80.A5 Deep endometriosis of ureter</b>
Add	<b>Intrinsic endometriosis of ureter</b>
Add	<b>Code also obstructive and reflux uropathy (N13.-)</b>
New code	<b>N80.A51 Deep endometriosis of right ureter</b>
New code	<b>N80.A52 Deep endometriosis of left ureter</b>
New code	<b>N80.A53 Deep endometriosis of bilateral ureters</b>
New code	<b>N80.A59 Deep endometriosis of unspecified ureter</b>
New code	<b>N80.A6 Endometriosis of ureter, unspecified depth</b>
Add	<b>Code also obstructive and reflux uropathy (N13.-)</b>
New code	<b>N80.A61 Endometriosis of right ureter, unspecified depth</b>
New code	<b>N80.A62 Endometriosis of left ureter, unspecified depth</b>
New code	<b>N80.A63 Endometriosis of bilateral ureters, unspecified depth</b>
New code	<b>N80.A69 Endometriosis of unspecified ureter, unspecified depth</b>
New subcategory	<b>N80.B Endometriosis of cardiothoracic space</b>
Add	<b>Endometriosis of thorax</b>
Add	<b>Code also, if applicable:</b> <b>catamenial pneumothorax (J93.12)</b> <b>catamenial hemothorax (J94.2)</b>
New code	<b>N80.B1 Endometriosis of pleura</b>
New code	<b>N80.B2 Endometriosis of lung</b>
	<b>N80.B3 Endometriosis of diaphragm</b>
New sub-subcategory	<b>N80.B31 Superficial endometriosis of diaphragm</b>
New code	<b>N80.B32 Deep endometriosis of diaphragm</b>

<b>New code</b>	<b>N80.B39 Endometriosis of diaphragm, unspecified depth</b>
<b>Add</b>	<b>Endometriosis of the diaphragm NOS</b>
New code	N80.B4 Endometriosis of the pericardial space
New code	N80.B5 Endometriosis of the mediastinal space
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
<b>Add</b>	<b>Endometriosis of the abdomen NOS</b>
New sub-subcategory	N80.C1 Endometriosis of the anterior abdominal wall
New code wall,	N80.C10 Endometriosis of the anterior abdominal subcutaneous tissue
New code wall,	N80.C11 Endometriosis of the anterior abdominal 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
<b>New code</b>	<b>N80.C9 Endometriosis of other site of abdomen</b>
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

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New Code  
Add

N80.D9 Endometriosis of other pelvic nerve  
Endometriosis of the other nerves of the  
retroperitoneum

**Add**  
Delete

N80.8 Other endometriosis  
**Endometriosis of other site**  
~~Endometriosis of thorax~~

## Extraocular Muscle Entrapment

Extraocular muscle entrapment in a nondisplaced orbital fracture, although a well-known entity in pediatric trauma, is atypical in adults. It can present with a triad of bradycardia, nausea, and in rare cases, syncope, and result in severe fibrosis of damaged and incarcerated muscle<sup>1</sup>.

An article published by AO Surgery Reference, “The inferior rectus muscle is the most common ocular muscle to become entrapped with an orbital floor fracture (trap-door phenomenon) and this may not be visible on conventional x-rays. Entrapment requires urgent freeing of the muscle to prevent necrosis of the incarcerated muscle. Clinical examination should give evidence on impaired ocular muscle function. Entrapment is often associated with severe ocular pain on attempted range of motion, as well as nausea and vomiting, especially in children”<sup>2</sup>.

NCHS received a proposal requesting the creation of new ICD-10-CM codes for extraocular muscle entrapment for coding specificity and research.

American Academy of Ophthalmology (AAO) and American Association of Oral and Maxillofacial Surgeons (AAOMS) supports this proposal.

### References

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2. Cornelius, C.-P., Gellrich, N., Hillerup, S., Kusumoto, K., & Schubert, W. (n.d.). Emergency treatment. Retrieved from Surgery Reference: <https://surgeryreference.aofoundation.org/cmft/trauma/midface/further-reading/emergency-treatment#>

## TABULAR MODIFICATIONS

	H50	Other strabismus	
		H50.6	Mechanical strabismus
New subcategory		H50.62	Inferior oblique muscle entrapment
New code		H50.621	Inferior oblique muscle entrapment, right
New code		H50.622	Inferior oblique muscle entrapment, left
New code		H50.629	Inferior oblique muscle entrapment, unspecified
New sub-subcategory		H50.63	Inferior rectus muscle entrapment
New code		H50.631	Inferior rectus muscle entrapment, right
New code		H50.632	Inferior rectus muscle entrapment, left
New code		H50.639	Inferior rectus muscle entrapment, unspecified
New sub-subcategory		H50.64	Lateral rectus muscle entrapment
New code		H50.641	Lateral rectus muscle entrapment, right

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New code		H50.642	Lateral rectus muscle entrapment, left
New code		H50.649	Lateral rectus muscle entrapment, unspecified
New sub-subcategory	H50.65		Medial rectus muscle entrapment
New code		H50.651	Medial rectus muscle entrapment, right
New code		H50.652	Medial rectus muscle entrapment, left
New code		H50.659	Medial rectus muscle entrapment, unspecified
New sub-subcategory	H50.66		Superior oblique muscle entrapment
New code		H50.661	Superior oblique muscle entrapment, right
New code		H50.662	Superior oblique muscle entrapment, left
New code		H50.669	Superior oblique muscle entrapment, unspecified
New sub-subcategory	H50.67		Superior rectus muscle entrapment
New code		H50.671	Superior rectus muscle entrapment, right
New code		H50.672	Superior rectus muscle entrapment, left
New code		H50.679	Superior rectus muscle entrapment, unspecified

## Foreign Body Sensation

Foreign body or globus sensation is the persistent feeling of a lump in the throat or that something is stuck in the throat. Some patients describe it as throat fullness. It is usually not painful but described as annoying. It is a very common condition<sup>1</sup>.

Providers' documentation of foreign body sensation in throat, currently codes to R09.89, Other specified symptoms and signs involving the circulatory and respiratory systems. This is a broad code that includes the following: bruit (arterial), abnormal chest percussion, feeling of foreign body in throat, friction sounds in chest, chest tympany, choking sensation, rales, and weak pulse.

The submitters are requesting the creation of ICD-10-CM codes to provide coding specificity that accurately reflects providers' documentation.

American Gastroenterological Association (AGA) supports this proposal.

### References

Foreign Body Sensation. (n.d.). Retrieved from San Diego ENT: <https://sandiegoent.com/throat/voice-swallowing/foreign-body-sensation/>

## TABULAR MODIFICATIONS

	R09	Other symptoms and signs involving the circulatory and respiratory system
		Excludes1: acute respiratory distress syndrome (J80) respiratory arrest of newborn (P28.81) respiratory distress syndrome of newborn (P22.0) respiratory failure (J96.-) respiratory failure of newborn (P28.5)
	R09.8	Other specified symptoms and signs involving the circulatory and respiratory systems
	R09.89	Other specified symptoms and signs involving the circulatory and respiratory systems
Delete		<del>Bruit (arterial)</del>
		Abnormal chest percussion
Add		Bruit (arterial)
Add		Chest tympany
Add		Choking sensation
Delete		<del>Feeling of foreign body in throat</del>
		Friction sounds in chest
Delete		<del>Chest tympany</del>
Delete		<del>Choking sensation</del>

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New subcategory	R09.A Foreign body sensation
New code	R09.A0 Foreign body sensation, unspecified
New code	R09.A1 Foreign body sensation, globus
New code	R09.A9 Other foreign body sensation

## Hemolytic-Uremic Syndrome

There was a previous proposal for expansion of the hemolytic-uremic syndrome (HUS) code in September 2020. This is now being brought back with revisions based on previous comments, and further input from multiple parties. Further expansion is being proposed that differs from the original proposal.

Hemolytic-uremic syndrome (HUS) most often occurs after a gastrointestinal infection with E coli bacteria (*Escherichia coli* O157:H7); this is called typical HUS. However, the condition has also been linked to other gastrointestinal infections, including shigella and salmonella. It has also been linked to nongastrointestinal infections, particularly with pneumococcus. One broad approach to classifying HUS groups cases that are associated with infections separately.

There are a number of causes of atypical HUS (aHUS). Certain genetic disorders may lead to aHUS, particularly related to the complement system. However, these generally give a genetic predisposition to aHUS, and in most cases, there also needs to be a triggering event. In such cases, aHUS can develop following a trigger by an acute infection, such as chicken pox or influenza. Pregnancy is another potential trigger. HUS due to genetic disorders would be considered hereditary HUS. There are also a number of other causes of secondary atypical HUS, including exposure to certain drugs (e.g., quinine, cancer chemotherapy, and oral contraceptives, among others).

### References

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Kavanagh D, Goodship TH, Richards A. Atypical Hemolytic Uremic Syndrome. *Semin Nephrol.* 2013 Nov; 33(6): 508–530.

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MedlinePlus. Hemolytic-uremic syndrome. National Library of Medicine.

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Lieberman K. Atypical Hemolytic Uremic Syndrome. Rare Disease Database. National Organization for Rare Disorders.

<https://rarediseases.org/rare-diseases/atypical-hemolytic-uremic-syndrome/>

**TABULAR MODIFICATIONS**

D59 Acquired hemolytic anemia

D59.3 Hemolytic-uremic syndrome

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

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

New code D59.30 Hemolytic-uremic syndrome, unspecified  
Add Hemolytic-uremic syndrome NOS

New code D59.31 Infection-associated hemolytic-uremic syndrome  
Add Shiga toxin-producing *E. coli* [STEC] related hemolytic  
uremic syndrome  
Add Typical hemolytic uremic syndrome

Add Use additional code to identify associated infection,  
such as :  
Add *E. coli* infection (B96.2-)  
Add Human immunodeficiency virus [HIV] disease  
(B20)  
Add Pneumococcal meningitis (G00.1)  
Add Pneumococcal pneumonia (J13)  
Add Sepsis due to *Streptococcus pneumoniae*  
(A40.3)  
Add *Shigella dysenteriae* (A03.9)  
Add *Streptococcus pneumoniae* as the cause of  
diseases classified elsewhere (B95.3)

New code D59.32 Hereditary hemolytic-uremic syndrome  
Add Atypical hemolytic uremic syndrome with an identified  
genetic cause  
Add Code also, if applicable:  
Add defects in the complement system (D84.1)  
Add methylmalonic acidemia (E71.120)



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Syndrome - see also Disease  
Revise - hemolytic-uremic D59.30  
Add - - atypical D59.39  
Add - - - genetic D59.32  
Add - - - hereditary D59.32  
Add - - - infection-associated D59.31  
Add - - - secondary D59.39  
Add - - - specified NEC D59.39  
Add - - due to genetic disorder D59.32  
Add - - familial D59.32  
Add - - hereditary D59.32  
Add - - infection-associated D59.31  
Add - - secondary D59.39  
Add - - Shiga toxin-producing E. coli [STEC] related D59.31  
Add - - specified NEC D59.39  
Add - - typical D59.31

## **Heparin-Induced Thrombocytopenia (HIT)**

Heparin-induced thrombocytopenia (HIT) is a decrease in platelet count that occurs during or shortly after exposure to heparin. With the wide usage of heparin for treating or preventing blood clots, this is a common problem, which can be serious and life-threatening.<sup>1</sup> For a number of years there have been two types of HIT recognized, identified as type 1 HIT and type 2 HIT. There has also more recently been a type of HIT identified as autoimmune HIT, or spontaneous HIT, which may occur without the patient previously receiving heparin. Currently there is only one code for Heparin Induced Thrombocytopenia (HIT), D75.82. It has been requested by the Agency for Healthcare Research and Quality (AHRQ) to create specific codes for Type 1 HIT and Type 2 HIT, and also to differentiate cases of autoimmune HIT. It is also proposed to separate other clinically similar conditions that occur unrelated to heparin, including spontaneous HIT syndrome.

Type 1 HIT (also known as “non-immune heparin-induced thrombocytopenia”) is a non-immunologic response to heparin treatment, mediated by a direct interaction between heparin and circulating platelets causing platelet clumping or sequestration. It has also been called heparin-associated thrombocytopenia in the past.<sup>1</sup> HIT type I affects up to 10% of patients being treated with heparin. It usually occurs within the first 48 to 72 hours after initiation of heparin treatment, and is characterized by a mild and transient thrombocytopenia, often returning to normal within four days once the heparin is withdrawn.<sup>1</sup> It is not associated with an increased risk of thrombosis or any significant complication. However, it may be coded if it leads to follow-up laboratory testing or another clinical decision, such as temporarily withholding or stopping unfractionated heparin.

In Type 2 HIT, there is an immune, antibody-mediated reaction that causes thrombocytopenia starting 5-10 days after initiation of heparin treatment<sup>2</sup>, and which may be associated with a hypercoagulable state.<sup>3</sup> The principal antigen is a complex of heparin and platelet factor 4 (PF4), a small positively charged molecule found in  $\alpha$ -granules of platelets. When heparin binds with PF4, it undergoes a conformational change and becomes immunogenic, leading to the production of heparin–PF4 antibodies (HIT antibodies) and heparin–PF4–IgG multimolecular immune complexes. These immune complexes activate platelets, causing the release of prothrombotic platelet-derived microparticles, platelet consumption, and thrombocytopenia. These microparticles in turn promote excessive thrombin generation, frequently resulting in arterial thromboses, deep vein thromboses (DVT), and/or pulmonary emboli (PE).<sup>4</sup> This condition occurs in 3% or so of patients receiving intravenous unfractionated heparin, and perhaps about 0.5% of patients receiving subcutaneous low molecular-weight heparins.

A number of patients have atypical presentations, and may have factors that cause their platelet count to continue to decrease even after heparin is stopped, or if there is a mix of antibodies that are heparin-dependent and heparin-independent. This condition has been described as “autoimmune heparin-induced thrombocytopenia (aHIT) syndrome.” These patients tend to have unusual HIT syndromes such as delayed-onset HIT, persisting HIT, fondaparinux-associated

HIT, and HIT induced by exposure to heparin ‘flushes.’ Such patients often show unusual clinical features, such as severe and persistent thrombocytopenia, often accompanied by disseminated intravascular coagulation (DIC) and microvascular thrombosis.<sup>5</sup> It has also been recognized that some affected patients may have no known history of heparin exposure, and other factors may trigger the same clinical syndrome. When patients present with clinical symptoms and laboratory features of HIT despite not having previously received heparin, either in the recent past or at all, this is referred to as “spontaneous HIT syndrome.” Sera from these patients contain antibodies that activate platelets strongly even in the absence of heparin. Certain triggers and types may be referred to as thrombosis with thrombocytopenia syndrome, or when related to certain vaccines (usually with adenoviral vectors), vaccine-induced thrombotic thrombocytopenia. Detecting and differentiating these types may require specific testing for HIT antibodies both with and without heparin present, and clinically these may be associated with slower recovery from thrombocytopenia.<sup>6</sup>

A DVT or PE resulting from immune-mediated HIT currently triggers a numerator event for the Agency for Healthcare Research and Quality’s Patient Safety Indicator (PSI) 12 (Perioperative Pulmonary Embolism or Deep Vein Thrombosis Rate), as well as CMS’ implementation of PSI 12. It is not the intent of PSI 12 to capture HIT-related events, because these events cannot be predicted or avoided in patients who have no prior history of receiving heparin. Having separate codes for immune-mediated HIT would allow these events to be excluded from the denominator of PSI 12, without inappropriately excluding cases involving non-immune HIT.

It has been requested to add sixth digits to D75.82 to specify these three types of heparin-induced or heparin-associated thrombocytopenia, and it is also being proposed to add a new code D75.83, to include cases of other platelet-activating anti-PF4 disorders, that do not involve exposure to heparin.

## References

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2. Ahmed I, Majeed A, and Powell R, Heparin induced thrombocytopenia: diagnosis and management update. *Postgrad Med J.* 2007 Sep; 83(983): 575–582.  
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<https://doi.org/10.1111/jth.13813>
6. Ning S, Warkentin TE. IV Immunoglobulin for Autoimmune Heparin-Induced Thrombocytopenia. *Chest* 2017 152(3):453-455.

**TABULAR MODIFICATIONS**

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

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

D75.82 Heparin induced thrombocytopenia (HIT)

New code	D75.821	Non-immune heparin-induced thrombocytopenia
Add		Heparin-associated thrombocytopenia
Add		Non-immune HIT
Add		Type 1 heparin-induced thrombocytopenia
New code	D75.822	Immune-mediated heparin-induced thrombocytopenia
Add		Immune-mediated HIT
Add		Type 2 heparin-induced thrombocytopenia
New code	D75.828	Other heparin-induced thrombocytopenia syndrome
Add		Autoimmune heparin-induced thrombocytopenia syndrome
Add		Delayed-onset heparin-induced thrombocytopenia
Add		Persisting heparin-induced thrombocytopenia
New code	D75.829	Heparin-induced thrombocytopenia, unspecified
New code	D75.83	Other platelet-activating anti-PF4 disorders
Add		Spontaneous heparin-induced thrombocytopenia syndrome (without heparin exposure)
Add		Thrombosis with thrombocytopenia syndrome
Add		Vaccine-induced thrombotic thrombocytopenia

## Hepatic encephalopathy

This topic was presented at the September 2015, March 2016 and September 2016 Coordination and Maintenance meeting and based on comments received during each public comment period it is being represented for consideration. World Health Organization (WHO) made a change in ICD-10 by including the manifestation of hepatic coma to various causes of hepatic failure. In ICD-9-CM, hepatic encephalopathy had a unique code with hepatic coma, portal-systemic encephalopathy and hepatocerebral intoxication as inclusion terms.

Hepatic encephalopathy (HE) involves altered consciousness and behavior related to insufficient liver function. HE is the loss of brain function that occurs when the liver is unable to remove toxins from the blood. Ammonia, which is produced by your body when proteins are digested, is one of the toxins that's normally made harmless by your liver. When ammonia or other toxic substances build up in the body when your liver isn't working well, it may affect your brain and cause HE.

The most commonly used staging scale of hepatic encephalopathy is the West Haven Grading System. The stages of HE span from minimal changes in memory and coordination in stage 0; sleep disruptions, and forgetfulness in stage 1; lethargy and mild disorientation in stage 2; amnesia and profound confusion in stage 3; to coma in stage 4.

NCHS proposes the following tabular changes to capture acute HE up to the hepatic coma and to harmonize with ICD-11 reporting of hepatic encephalopathy for research and clinical purposes.

### TABULAR MODIFICATIONS

	K72 Hepatic failure, not elsewhere classified
Delete	Includes: <del>hepatic encephalopathy NOS</del>
	K76 Other diseases of liver
	K76.8 Other specified diseases of liver
New code	K76.82 Hepatic encephalopathy
Add	Hepatic encephalopathy, NOS
Add	Hepatic encephalopathy, West Haven Criteria Grade 0
Add	Hepatic encephalopathy, West Haven Criteria Grade 1
Add	Hepatic encephalopathy, West Haven Criteria Grade 2
Add	Hepatic encephalopathy, West Haven Criteria Grade 3
Add	Hepatocerebral intoxication
Add	Portal-systemic encephalopathy
Add	Code also underlying liver disease, such as:
Add	acute and subacute hepatic failure without coma (K72.00)
Add	alcoholic hepatic failure without coma (K70.40)
Add	chronic hepatic failure without coma (K72.10)
Add	hepatic failure without coma (K72.90)
Add	hepatic failure with toxic liver disease without coma (K71.10)

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Add  
Add  
Add  
Add

icterus of newborn (P55-P59)  
postprocedural hepatic failure (K91.82)  
viral hepatitis without hepatic coma (B15.9,  
(B16.1, B16.9, B17.10, B19.10, B19.20, B19.9)

## **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.<sup>i</sup> The proposal was originally presented at the March 2021 Coordination and Maintenance meeting. Additions to the original proposal to expand IgAN to specify the type of glomerulonephritis. These additions have been added from comments received at the March meeting. The changes are bolded. RPA supports the additional codes.

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.<sup>ii</sup> It is estimated that IgAN accounts for up to 10% of all patients in need of renal replacement therapy for ESRD in western countries.<sup>iii</sup> 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.<sup>iv</sup>

IgAN is characterized by deposition of immune complexes containing Immunoglobulin A in the glomerulus and proliferation of mesangial cells.<sup>v,vi</sup> 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).<sup>1</sup> Higher levels and longer duration of proteinuria are the strongest prognostic risk factors for disease progression.<sup>vii,viii</sup> There are a number of specific therapies that are used in the treatment of IgAN patients.<sup>ix, x</sup>

IgAN is diagnosed by renal biopsy.<sup>xi</sup> 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.<sup>xii</sup> 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

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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.” We note 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, we recommend adding a new code, N02.B, to specifically identify IgAN. We recommend that ICD-10-CM continue to use N02.8 and N02.9 for other or unspecified morphologic changes, respectively.

<sup>i</sup> Kidney Disease Improving Global Outcomes. KDIGO Clinical Practice Guideline for Glomerulonephritis. 2012. *Kidney International Supplements* 2: 139. <https://kdigo.org/wp-content/uploads/2017/02/KDIGO-2012-GN-Guideline-English.pdf>.

<sup>ii</sup> Liu M, Chen Y, Zhou J et al. Implication of Urinary Complement Factor H in the Progression of Immunoglobulin A Nephropathy. 2015. *PLoS One*. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4452759/>.

<sup>iii</sup> Schena FP. Epidemiology of End-Stage Renal Disease: International Comparisons of Renal Replacement Therapy. *Kidney International* 57: S39-S45. [https://www.kidney-international.org/article/S0085-2538\(15\)47039-5/fulltext](https://www.kidney-international.org/article/S0085-2538(15)47039-5/fulltext).

<sup>iv</sup> Yuzawa, et al. Evidence-based Clinical Practice Guidelines for IgA Nephropathy 2014. 2016. *Clin Exp Nephrol* 20: 511-535.

<sup>v</sup> Sethi S, Haas M, Markowitz GS, et al. Mayo Clinic/Renal Pathology Society Consensus Report on Pathologic Classification, Diagnosis, and Reporting of GN. 2016. *J Am Soc Nephrol* 27: 1278-1287. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4849835/>.

<sup>vi</sup> Yuzawa, et al. Evidence-Based Clinical Practice Guidelines for IgA Nephropathy 2014. 2016. *Clin Exp Nephrol* 20: 511-535. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4956709/>.

<sup>vii</sup> Edstrom Halling S, Soderberg MP, Berg UB. Predictors of Outcome in Paediatric IgA Nephropathy With Regard to Clinical and Histopathological Variables (Oxford Classification). 2012. 27: 715-722. <https://pubmed.ncbi.nlm.nih.gov/21750154/>.

<sup>viii</sup> Suzuki H, Kiryluk K, Novak J, Moldoveanu Z, Herr AB, Renfrow MB, Wyatt RJ, Scolari F, Mestecky J, Gharavi AG, Julian BA. The Pathophysiology of IgA Nephropathy. 2011. *J Am Soc Nephrol* 22: 1795-1803. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3892742/>

<sup>ix</sup> Floege J, Barbour SJ, Cattran DC, et al. Management and treatment of glomerular diseases (part 1): Conclusions from a Kidney Disease: Improving Global Outcomes (KDIGO) Controversies Conference. 2019, *Kidney International*, 95:268–280; <https://doi.org/10.1016/j.kint.2018.10.018>.

<sup>x</sup> Cambier A, Rabant M, Peuchmaur M, et al. Immunosuppressive Treatment in Children with IgA Nephropathy and the Clinical Value of Podocytopathic Features. 2018, *Kidney Int Rep*, 3:916–925; <https://doi.org/10.1016/j.ekir.2018.03.013>.

<sup>xi</sup> Lai KN, Leung JC, Tang SC. The Treatment of IgA Nephropathy. 2015: *Kidney Dis (Basel)*. 1:19-26. <https://pubmed.ncbi.nlm.nih.gov/27536661/>

<sup>xii</sup> A Working Group of the International IgA Nephropathy Network and the Renal Pathology Society, Roberts ISD, Cook HT, et al. The Oxford classification of IgA nephropathy: pathology definitions, correlations, and reproducibility. 2009. *Kidney International*. 76: 546-556. [https://www.kidney-international.org/article/S0085-2538\(15\)53999-9/fulltext](https://www.kidney-international.org/article/S0085-2538(15)53999-9/fulltext).

<sup>xiii</sup> Fogo AB. Decade in Review—Glomerular Disease: The Glomerulus Reveals Some Secrets. 2015. *Nat Rev Nephrol* 11:63-64. <https://pubmed.ncbi.nlm.nih.gov/26369392/>.

The combination of the MEST-C score (a classification system inclusive of mesangial hypercellularity (M), endocapillary hypercellularity (E), segmental sclerosis of the capillary tuft (S), tubular atrophy/interstitial fibrosis (T) and crescent formation (C) with blood pressure, proteinuria and eGFR at the time of biopsy are helpful in predicting prognostic outcome.

## TABULAR MODIFICATIONS

	N02	Recurrent and Persistent Hematuria
New sub category		N02.B Recurrent and persistent immunoglobulin A nephropathy
New code		<b>N02.B1 Recurrent and persistent immunoglobulin A nephropathy with glomerular lesion</b>
New code		<b>N02.B2 Recurrent and persistent immunoglobulin A nephropathy with focal and segmental hyalinosis or sclerosis</b>
New code		<b>N02.B3 Recurrent and persistent immunoglobulin A nephropathy with membranoproliferative (diffuse)</b>
New code		<b>N02.B4 Recurrent and persistent immunoglobulin A nephropathy with membranous (diffuse)</b>
New code		<b>N02.B5 Recurrent and persistent immunoglobulin A nephropathy with mesangial proliferative (diffuse)</b>
New code		<b>N02.B6 Recurrent and persistent immunoglobulin A nephropathy with mesangiocapillary (diffuse)</b>
New code		<b>N02.B9 Other recurrent and persistent immunoglobulin A nephropathy</b>

## INDEX MODIFICATIONS

	<b>Nephropathy</b>
Revise	- IgA <del>N02.8</del> <u>N02.B-</u>
Revise	- - with glomerular lesion <del>N02.9</del> <u>N02.B1</u>
Revise	- - - focal and segmental hyalinosis or sclerosis <del>N02.1</del> <u>N02.B2</u>
Revise	- - - membranoproliferative (diffuse) <del>N02.5</del> <u>N02.B3</u>
Revise	- - - membranous (diffuse) <del>N02.2</del> <u>N02.B4</u>
Revise	- - - mesangial proliferative (diffuse) <del>N02.3</del> <u>N02.B5</u>
Revise	- - - mesangiocapillary (diffuse) <del>N02.5</del> <u>N02.B6</u>
Revise	- - - proliferative NEC <del>N02.8</del> <u>N02.B9</u>
Revise	- - - specified pathology NEC <del>N02.8</del> <u>N02.B9</u>

## **Insulin Resistant Syndrome**

There is no unanimous definition for metabolic syndrome globally<sup>1</sup> but there is an agreement on the following criteria. The National Institute of Health defines metabolic syndrome as the presence of at least 3 of the following traits (including the ones that are controlled by medication): large waist, elevated triglyceride level, reduced HDL cholesterol, increased blood pressure and elevated fasting blood glucose. Other names for metabolic syndrome are: Dysmetabolic syndrome, Hypertriglyceridemic waist, Insulin resistance syndrome, Obesity syndrome or Syndrome X.

The National Heart, Lung and Blood Institute states the following: Insulin resistance also may increase your risk for metabolic syndrome. Insulin resistance is a condition in which the body cannot use its insulin properly. Insulin is a hormone that helps move blood sugar into cells where it is used for energy. Insulin resistance can lead to high blood sugar levels, and it is intricately linked to overweight and obesity. Genetics and aging may also contribute to the development of this syndrome.

Type A insulin-resistance syndrome belongs to the group of extreme insulin-resistance syndromes (which includes leprechaunism, the lipodystrophies, Rabson-Mendenhall syndrome) (characterized by the triad of hyperinsulinemia, acanthosis nigricans (skin lesions associated with insulin resistance), and signs of hyperandrogenism in females without lipodystrophy and who are not overweight<sup>2</sup>).

The submitter is requesting the new ICD-10-CM codes for coding specificity.

This proposal is supported by Office of Genomics Precision Public Health, American College of Medical Genetics and Genomics, and PTEN Research

### References

1. Changing definitions of metabolic syndrome. Parikh and Mohan, <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3263200/>
2. Orphanet: Insulin resistance syndrome type A. INSERM US14 -- ALL RIGHTS RESERVED [https://www.orpha.net/consor/cgi-bin/OC\\_Exp.php?Expert=2297](https://www.orpha.net/consor/cgi-bin/OC_Exp.php?Expert=2297)

## **TABULAR MODIFICATIONS**

- E88 Other and unspecified metabolic disorders  
Use additional codes for associated conditions  
Excludes1: histiocytosis X (chronic) (C96.6)  
other specified metabolic disorders (E88.8)

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Delete	E88.81	Metabolic syndrome <del>Dysmetabolic syndrome X</del> Use additional codes for associated manifestations, such as: obesity (E66.-)
New code	E88.810	Insulin resistance syndrome, Type A
New code	E88.818	Other metabolic syndrome
Add		Insulin resistance syndrome, Type B
Add		Other insulin resistance syndrome
New code	E88.819	Metabolic syndrome, unspecified
Add		Dysmetabolic syndrome X
Add		Insulin resistance syndrome, unspecified New code

## **Intracranial Injury with Unknown Loss of Consciousness**

Often patients will present with injuries that are coded to S06, Intracranial Injury, who present without a clear history of loss of consciousness (LOC). The current default, “with loss of consciousness of unspecified duration” implies the patient had a LOC, which may not be the case.

In order to better track this group of patients, the American Academy of Pediatrics (AAP) proposes adding a unique code to code category S06, Intracranial Injury for patients in which it is unclear whether there was an actual loss of consciousness or not.

It is being proposed to expand the loss of consciousness periods in codes found under S06.0, Concussion, to be more in line with the rest of the category. There are patients who may have a LOC greater than 30 minutes who have not been diagnosed with a specific traumatic brain injury as the cause.

Under the current coding options, patients who have only been diagnosed with a concussion and who have an LOC greater than 30 minutes would be coded to either S06.0X9, Concussion with loss of consciousness of unspecified duration or S06.9X, Unspecified intracranial injury. This proposal will allow for better tracking and clinical information. Also, AAP is asking to add inclusion term of brief loss of consciousness to specified codes with loss of consciousness 30 minute or less.

The American Academy of Pediatrics (AAP) are requesting the following tabular modifications:

### **TABULAR MODIFICATIONS**

S06 Intracranial injury  
Includes: traumatic brain injury  
Code also any associated:  
    open wound of head (S01.-)  
    skull fracture (S02.-)  
Excludes1: head injury NOS (S09.90)

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

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The appropriate 7th character is to be added to each code from category S06  
 A initial encounter  
 D subsequent encounter  
 S sequela

Note: 7th characters D and S do not apply to codes in category S06 with 6th  
 character 7 - death due to brain injury prior to regaining consciousness, or  
 8 - death due to other cause

S06.0 Concussion  
 Comotio cerebri  
 Excludes1: concussion with other intracranial injuries classified in  
 subcategories S06.1- to S06.6-, S06.81-and S06.82-  
 code to specified intracranial injury

S06.0X Concussion

New sub-subcategory	S06.0X1 Concussion with loss of consciousness of 30 minutes or less
Add	Concussion with brief loss of consciousness
New code	S06.0XA Concussion with loss of consciousness status unknown
Add	Concussion NOS
	S06.0X9 Concussion with loss of consciousness of unspecified duration
Delete	<del>Concussion NOS</del>

S06.1 Traumatic cerebral edema  
 Diffuse traumatic cerebral edema  
 Focal traumatic cerebral edema

S06.1X Traumatic cerebral edema

	S06.1X1 Traumatic cerebral edema with loss of consciousness of 30 minutes or less
Add	Traumatic cerebral edema with brief loss of consciousness
New code	S06.1XA Traumatic cerebral edema with loss of consciousness status unknown
Add	Traumatic cerebral edema NOS

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Delete	S06.1X9 Traumatic cerebral edema with loss of consciousness of unspecified duration <del>Traumatic cerebral edema NOS</del>
	S06.2 Diffuse traumatic brain injury Diffuse axonal brain injury
Add	S06.2X Diffuse traumatic brain injury S06.2X1 Diffuse traumatic brain injury with loss of consciousness of 30 minutes or less Diffuse traumatic brain injury with brief loss of consciousness
New code	S06.2XA Diffuse traumatic brain injury with loss of consciousness status unknown
Add	Diffuse traumatic brain injury NOS
Delete	S06.2X9 Diffuse traumatic brain injury with loss of consciousness of unspecified duration <del>Diffuse traumatic brain injury NOS</del>
	S06.3 Focal traumatic brain injury
Add	S06.30 Unspecified focal traumatic brain injury S06.301 Unspecified focal traumatic brain injury with loss of consciousness of 30 minutes or less Unspecified focal traumatic brain injury with brief loss of consciousness
New code	S06.30A Unspecified focal traumatic brain injury with loss of consciousness status unknown
Add	Unspecified focal traumatic brain injury NOS
Delete	S06.309 Unspecified focal traumatic brain injury with loss of consciousness of unspecified duration <del>Unspecified focal traumatic brain injury NOS</del>
Add	S06.31 Contusion and laceration of right cerebrum S06.311 Contusion and laceration of right cerebrum with loss of consciousness of 30 minutes or less Contusion and laceration of right cerebrum with brief loss of consciousness

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New code	S06.31A Contusion and laceration of right cerebrum with loss of consciousness status unknown
Add	Contusion and laceration of right cerebrum NOS
	S06.319 Contusion and laceration of right cerebrum with loss of consciousness of unspecified duration
Delete	<del>Contusion and laceration of right cerebrum NOS</del>
	S06.32 Contusion and laceration of left cerebrum
	S06.321 Contusion and laceration of left cerebrum with loss of consciousness of 30 minutes or less
Add	Contusion and laceration of left cerebrum with brief loss of consciousness
	S06.32A Contusion and laceration of left cerebrum with loss of consciousness status unknown
Add	Contusion and laceration of left cerebrum NOS
	S06.329 Contusion and laceration of left cerebrum with loss of consciousness of unspecified duration
Delete	<del>Contusion and laceration of left cerebrum NOS</del>
	S06.33 Contusion and laceration of cerebrum, unspecified
	S06.331 Contusion and laceration of cerebrum, unspecified, with loss of consciousness of 30 minutes or less
Add	Contusion and laceration of cerebrum, unspecified, with brief loss of consciousness
	S06.33A Contusion and laceration of cerebrum, unspecified with loss of consciousness status unknown
Add	Contusion and laceration of cerebrum NOS
	S06.339 Contusion and laceration of cerebrum, unspecified, with loss of consciousness of unspecified duration
Delete	<del>Contusion and laceration of cerebrum NOS</del>
	S06.34 Traumatic hemorrhage of right cerebrum

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Traumatic intracerebral hemorrhage and hematoma of right  
cerebrum

Add S06.341 Traumatic hemorrhage of right cerebrum with loss  
of consciousness of 30 minutes or less  
Traumatic hemorrhage of right cerebrum with loss  
of consciousness

New code S06.34A Traumatic hemorrhage of right cerebrum with  
Add brief loss of consciousness status unknown  
Traumatic hemorrhage of right cerebrum NOS

Delete S06.349 Traumatic hemorrhage of right cerebrum with  
loss of consciousness of unspecified duration  
~~Traumatic hemorrhage of right cerebrum NOS~~

S06.35 Traumatic hemorrhage of left cerebrum  
Traumatic intracerebral hemorrhage and hematoma of left  
cerebrum

Add S06.351 Traumatic hemorrhage of left cerebrum with loss  
of consciousness of 30 minutes or less  
Traumatic hemorrhage of left cerebrum with brief  
loss of consciousness

New code S06.35A Traumatic hemorrhage of left cerebrum with loss  
Add of consciousness status unknown  
Traumatic hemorrhage of left cerebrum NOS

Delete S06.359 Traumatic hemorrhage of left cerebrum with loss  
of consciousness of unspecified duration  
~~Traumatic hemorrhage of left cerebrum NOS~~

S06.36 Traumatic hemorrhage of cerebrum, unspecified  
Traumatic intracerebral hemorrhage and hematoma,  
unspecified

Add S06.361 Traumatic hemorrhage of cerebrum, unspecified,  
with loss of consciousness of 30 minutes or less  
Traumatic hemorrhage of cerebrum, unspecified,  
with brief loss of consciousness

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New code S06.36A Traumatic hemorrhage of cerebrum, unspecified  
with loss of consciousness status unknown  
Add Traumatic hemorrhage of cerebrum NOS

S06.369 Traumatic hemorrhage of cerebrum, unspecified,  
with loss of consciousness of unspecified  
duration  
Delete ~~Traumatic hemorrhage of cerebrum NOS~~

S06.37 Contusion, laceration, and hemorrhage of cerebellum

S06.371 Contusion, laceration, and hemorrhage of  
cerebellum with loss of consciousness of 30  
minutes or less  
Add Contusion, laceration, and hemorrhage of  
cerebellum with brief loss of consciousness

New code S06.37A Contusion, laceration, and hemorrhage of  
cerebellum with loss of consciousness status  
unknown  
Add Contusion, laceration, and hemorrhage of  
cerebellum NOS

S06.379 Contusion, laceration, and hemorrhage of  
cerebellum with loss of consciousness of  
unspecified duration  
Delete ~~Contusion, laceration, and hemorrhage of  
cerebellum NOS~~

S06.38 Contusion, laceration, and hemorrhage of brainstem

S06.381 Contusion, laceration, and hemorrhage of  
brainstem with loss of consciousness of 30  
minutes or less  
Add Contusion, laceration, and hemorrhage of  
brainstem with brief loss of consciousness

New code S06.38A Contusion, laceration, and hemorrhage of  
brainstem with loss of consciousness status  
unknown  
Add Contusion, laceration, and hemorrhage of  
brainstem NOS

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Delete	S06.389 Contusion, laceration, and hemorrhage of brainstem with loss of consciousness of unspecified duration <del>Contusion, laceration, and hemorrhage of brainstem NOS</del>
	S06.4 Epidural hemorrhage Extradural hemorrhage NOS Extradural hemorrhage (traumatic)
	S06.4X Epidural hemorrhage S06.4X1 Epidural hemorrhage with loss of consciousness of 30 minutes or less
Add	Epidural hemorrhage with brief loss of consciousness
New code	S06.4XA Epidural hemorrhage with loss of consciousness status unknown
Add	Epidural hemorrhage NOS
	S06.4X9 Epidural hemorrhage with loss of consciousness of unspecified duration
Delete	<del>Epidural hemorrhage NOS</del>
	S06.5 Traumatic subdural hemorrhage S06.5X Traumatic subdural hemorrhage S06.5X1 Traumatic subdural hemorrhage with loss of consciousness of 30 minutes or less
Add	Traumatic subdural hemorrhage with brief loss of consciousness
New code	S06.5XA Traumatic subdural hemorrhage with loss of consciousness status unknown
Add	Traumatic subdural hemorrhage NOS
	S06.5X9 Traumatic subdural hemorrhage with loss of consciousness of unspecified duration
Delete	<del>Traumatic subdural hemorrhage NOS</del>

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S06.6 Traumatic subarachnoid hemorrhage

S06.6X Traumatic subarachnoid hemorrhage

S06.6X1 Traumatic subarachnoid hemorrhage with  
loss of consciousness of 30 minutes  
or less

Add Traumatic subarachnoid hemorrhage with  
brief loss of consciousness

New code S06.6XA Traumatic subarachnoid hemorrhage with  
loss of consciousness status unknown

Add Traumatic subarachnoid hemorrhage NOS

S06.6X9 Traumatic subarachnoid hemorrhage with  
loss of consciousness of unspecified  
duration

Delete ~~Traumatic subarachnoid hemorrhage NOS~~

S06.8 Other specified intracranial injuries

S06.81 Injury of right internal carotid artery, intracranial  
portion, not elsewhere classified

S06.811 Injury of right internal carotid artery,  
intracranial portion, not elsewhere  
classified with loss of consciousness of  
30 minutes or less

Add Injury of right internal carotid artery,  
intracranial portion, not elsewhere  
classified with brief loss of  
consciousness

New code S06.81A Injury of right internal carotid artery,  
intracranial portion, not elsewhere  
classified with loss of consciousness  
status unknown

Add Injury of right internal carotid artery,  
intracranial portion, not elsewhere  
classified NOS

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Delete	<p>S06.819 Injury of right internal carotid artery, intracranial portion, not elsewhere classified with loss of consciousness of unspecified duration</p> <p><del>Injury of right internal carotid artery, intracranial portion, not elsewhere classified NOS</del></p>
	<p>S06.82 Injury of left internal carotid artery, intracranial portion, not elsewhere classified</p>
Add	<p>S06.821 Injury of left internal carotid artery, intracranial portion, not elsewhere classified with loss of consciousness of 30 minutes or less</p> <p>Injury of left internal carotid artery, intracranial portion, not elsewhere classified with brief loss of consciousness</p>
New code	<p>S06.82A Injury of left internal carotid artery, intracranial portion, not elsewhere classified with loss of consciousness status unknown</p>
Add	<p>Injury of left internal carotid artery, intracranial portion, not elsewhere classified NOS</p>
Delete	<p>S06.829 Injury of left internal carotid artery, intracranial portion, not elsewhere classified with loss of consciousness of unspecified duration</p> <p><del>Injury of left internal carotid artery, intracranial portion, not elsewhere classified NOS</del></p>
	<p>S06.8A Primary blast injury of brain, not elsewhere classified Code also, if applicable, focal traumatic brain injury (S06.3-)</p> <p>Excludes2: traumatic cerebral edema (S06.1)</p>
Add	<p>S06.8A1 Primary blast injury of brain, not elsewhere classified with loss of consciousness of 30 minutes or less</p> <p>Primary blast injury of brain, not elsewhere classified with brief loss of consciousness</p>

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New code S06.8AA Primary blast injury of brain, not elsewhere  
classified with loss of consciousness status  
unknown

Add Primary blast injury of brain NOS

Delete S06.8A9 Primary blast injury of brain, not elsewhere  
classified with loss of consciousness of  
unspecified duration

~~Primary blast injury of brain NOS~~

S06.89 Other specified intracranial injury

Add S06.891 Other specified intracranial injury with loss of  
consciousness of 30 minutes or less  
Other specified intracranial injury with brief loss  
of consciousness

New code S06.89A Other specified intracranial injury with loss of  
consciousness status unknown

S06.9 Unspecified intracranial injury

Brain injury NOS

Head injury NOS with loss of consciousness

Traumatic brain injury NOS

Excludes1: conditions classifiable to S06.0- to S06.8-code to  
specified intracranial injury head injury NOS (S09.90)

S06.9X Unspecified intracranial injury

Add S06.9X1 Unspecified intracranial injury with loss of  
consciousness of 30 minutes or less  
Unspecified intracranial injury with brief loss of  
consciousness

New code S06.9XA Unspecified intracranial injury with loss of  
consciousness status unknown

## **Isthmocele**

The American College of Obstetricians and Gynecologists (ACOG), The American Society for Reproductive Medicine (ASRM) and the American Association of Gynecologic Laparoscopists (AAGL) are proposing a new diagnosis code to specify the presence of a defect in the myometrium from a cesarean section for the non-pregnant patient. The term isthmocele (also known as cesarean scar defect or niche) describes a dehiscence at the incision site of a previous cesarean section. A proposal for isthmocele scarring at a different code category was presented at the September 2019 Coordination and Maintenance meeting, but comments did not support the request for a new code at that code category.

Isthmocele can complicate pregnancy in many ways. It may cause abnormal placentation, interstitial pregnancy, cesarean scar dehiscence and uterine rupture. ICD-10-CM has codes that describe the isthmocele in the obstetric setting at subcategory O34.2-, Maternal care due to uterine scar from previous surgery. There is currently no diagnosis code for isthmocele in the gynecologic patient population.

In the non-pregnant patient, isthmocele may be the cause of pelvic pain, abnormal uterine bleeding, secondary infertility, vaginal discharge, postmenstrual spotting, dyspareunia and dysmenorrhea. Many patients are asymptomatic and the isthmocele is found incidentally on imaging. The prevalence of isthmocele in randomly selected women of reproductive age is 24-70% using transvaginal ultrasound, and 56-84% by saline sonohysterogram. Depending on symptoms, the treatment can be medical or surgical. Hysteroscopy and laparoscopy are the minimally invasive approaches mainly used to repair the defect.

ACOG is requesting new ICD-10-CM codes to identify this condition when found in the non-pregnant patient. This will allow the ability to track the frequency at which this condition occurs, as well as to facilitate tracking of the most effective treatment modalities through diagnosis code searches in EMT datasets. The following tabular modifications are being proposed.

### References

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- Van der Voet LF, Vervoort AJ, Veersema S, et al. Minimally Invasive Therapy for Gynecological Symptoms related to a Niche in the Cesarean Scar: a systematic review. *BJOG* 121:145-156, 2014
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- Raimondo G., Grifone G., Raimondo D., et al. Hysteroscopic Treatment of Cesarean-induced Isthmocele: A Prospective Study. *J Minim Invasive Gynecol.* 22:297-301, 2015
- Roberge S., Boutin A., Chailier N., Moore L., Jastrow N., Demers S., et al. Systematic Review of Cesarean Scar Assessment in the Nonpregnant State: Imaging techniques and Uterine Scar Defect. *Am J Perinatol.* 29:465-471, 2012

**TABULAR MODIFICATIONS**

N85 Other noninflammatory disorders of uterus, except cervix

Excludes1: endometriosis (N80.-)  
inflammatory diseases of uterus (N71.-)  
noninflammatory disorders of cervix, except malposition  
(N86-N88)  
polyp of corpus uteri (N84.0)  
uterine prolapse (N81.-)

New code	N85.A Isthmocele
Add	Isthmocele (non-pregnant state)
Add	Code also any associated conditions such as:
Add	abnormal uterine and vaginal bleeding, unspecified (N93.9)
	female infertility of uterine origin (N97.2)
	pelvic and perineal pain (R10.2)
Add	Excludes1: maternal care for cesarean scar defect (isthmocele) (O34.22)

## Long Term (current) Drug Therapy

The American Academy of Pediatrics (AAP) presented this proposal at the March 2020 Coordination and Maintenance Committee Meeting. This proposal has been revised to address comments received during the public comment period. Changes for reconsideration are shown in **bold**.

The number and types of medications that patients are taking daily seems to be increasing almost exponentially. Some of these medications carry longer term risks and should be identified so they can be more closely monitored and tracked.

As these medications are becoming more common in healthcare, it is of key importance to identify the class of drug the patient is using, especially since currently all such medications fall under the nonspecific code Z79.8, Other long term (current) drug therapy. The American Academy of Pediatrics (AAP) is requesting expansion of this code set to capture more of these medications to better identify and monitor the risk and long term outcomes.

As was pointed out by one of the commentors at the previous presentation, a single drug may have multiple applications. It is of the opinion of the Academy that it is best to identify the drugs based on its principal activity, e.g., antimetabolite agent, then as to its clinical application. In order to help clarify which medications fall into which of the proposed categories, the Academy has reviewed a number of sources, including the National Cancer Institute and the American Cancer Society.

**NCHS also received a separate request for an additional new code to capture long term (current ) use of injectable non-insulin antidiabetic drugs. The requestor's justification is that the existing code, Z79.899, Other long term (current) drug therapy, does not adequately capture the medication that is being used. This code request has been incorporated in this proposal. The American Academy of Pediatrics supports this additional request.**

### TABULAR MODIFICATONS

#### Z79 Long term (current) drug therapy

New subcategory	Z79.6 Long term (current) use of immunomodulators and immunosuppressants
<b>Add</b>	<b>Use additional code, if applicable, to identify:</b>
<b>Add</b>	<b>neoplasms (C00-D49)</b>
<b>Add</b>	<b>sickle cell disorders (D57)</b>
<b>Add</b>	<b>transplanted organ and tissue status (Z94)</b>
Add	Excludes2: long term (current) use of steroids (Z79.5-)

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Add	long term (current) use of agents affecting estrogen receptors and estrogen levels (Z79.81-)
New code	<b>Z79.60 Long term (current) use of unspecified immunomodulators and immunosuppressants</b>
New code	Z79.61 Long term (current) use of immunomodulator
Add	<b>Long term (current) use of apremilast (Otezla)</b>
Add	<b>Long term (current) use of Immunomodulatory imide drug</b>
Add	<b>Long term (current) use of lenalidomide (Revlimid)</b>
Add	<b>Long term (current) use of pomalidomide (Pomalyst)</b>
New sub-subcategory	<b>Z79.62 Long term (current) use of immunosuppressant</b>
New code	<b>Z79.620 Long term (current) use of immunosuppressive biologic</b>
Add	<b>Long term (current) use of adalimumab (Humira)</b>
Add	<b>Long term (current) use of etanercept (Enbrel)</b>
Add	<b>Long term (current) use of infliximab (Remicade)</b>
Add	<b>Long term (current) use of monoclonal antibodies</b>
New code	<b>Z79.621 Long term (current) use of calcineurin inhibitor</b>
Add	<b>Long term (current) use of cyclosporine</b>
Add	<b>Long term (current) use of tacrolimus</b>
New code	<b>Z79.622 Long term (current) use of Janus kinase inhibitor</b>
Add	<b>Long term (current) use of tofacitinib (Xeljanz)</b>
New code	<b>Z79.623 Long term (current) use of mammalian target of rapamycin (mTOR) inhibitor</b>
Add	<b>Long term (current) use of sirolimus (Rapamune)</b>

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<b>New code</b>	<b>Z79.624 Long term (current) use of inhibitors of nucleotide synthesis</b>
<b>Add</b>	<b>Long term (current) use of azathioprine (Imuran)</b>
<b>Add</b>	<b>Long term (current) use of mycophenolate (CellCept)</b>
<b>Add</b>	<b>Long term (current) use of Purine synthesis (IMDH) inhibitors</b>
<b>New sub-subcategory</b>	<b>Z79.63 Long term (current) use of chemotherapeutic agent</b>
<b>New code</b>	<b>Z79.630 Long term (current) use of alkylating agent</b>
<b>Add</b>	<b>Long term (current) use of chlorambucil (Leukeran)</b>
<b>Add</b>	<b>Long term (current) use of cisplatin (Platinol)</b>
<b>Add</b>	<b>Long term (current) use of cyclophosphamide (Cytosan)</b>
<b>New code</b>	<b>Z79.631 Long term (current) use of antimetabolite agent</b>
<b>Add</b>	<b>Long term (current) use of 5-fluorouracil (5-FU)</b>
<b>Add</b>	<b>Long term (current) use of 6-mercaptopurine (6-MP)</b>
<b>Add</b>	<b>Long term (current) use of cytarabine (Ara-C)</b>
<b>Add</b>	<b>Long term (current) use of methotrexate</b>
<b>New code</b>	<b>Z79.632 Long term (current) use of antitumor antibiotic</b>
<b>Add</b>	<b>Long term (current) use of bleomycin</b>
<b>Add</b>	<b>Long term (current) use of doxorubicin (Adriamycin)</b>
<b>Add</b>	<b>Long term (current) use of mitomycin C</b>
<b>New code</b>	<b>Z79.633 Long term (current) use of mitotic inhibitor</b>
<b>Add</b>	<b>Long term (current) use of paclitaxel (Taxol)</b>
<b>Add</b>	<b>Long term (current) use of plant alkaloids</b>

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**Add** **Long term (current) use of vinblastine**  
**Add** **Long term (current) use of vincristine**

**New code** **Z79.634 Long term (current) use of  
topoisomerase inhibitor**  
**Add** **Long term (current) use of etoposide  
(Vepesid)**  
**Add** **Long term (current) use of irinotecan  
(Camptosar)**  
**Add** **Long term (current) use of topotecan  
(Hycamtin)**

**New code** **Z79.64 Long term (current) use of myelosuppressive agent**  
**Add** **Long term (current) use of hydroxyurea**

**New code** **Z79.69 Long term (current) use of other immunomodulators  
and immunosuppressants**

**Z79.8 Other long term (current) drug therapy**

**Z79.84 Long term (current) use of oral hypoglycemic drugs**  
**Long term (current) use of oral antidiabetic drugs**  
**Excludes2: long term (current) use of insulin (Z79.4)**

**New code** **Z79.85 Long-term (current) use of injectable non-insulin  
antidiabetic drugs**

## **Malignant pericardial effusion**

Malignant pericardial effusion (MPE) is a complication of neoplastic disease. Approximately 10% of cancer patients develop cardiac metastases, based on autopsy studies.<sup>1,2</sup> Many may be asymptomatic. Of these metastatic lesions, about three quarters involve the epicardium, the innermost layer of the pericardium, and among those with epicardial metastases, about one third had a pericardial effusion.<sup>1</sup>

The most common etiologies of malignant disease of the pericardium include cancers of the lung and breast, while a number of other cancers may also produce malignant neoplasms, including malignant melanoma and leukemia or lymphoma.<sup>2</sup> The presence of symptomatic pericardial effusion in a patient with a malignancy suggests a poor prognosis, with a median survival time of 2 to 5 months after diagnosis.<sup>3</sup>

It is important to differentiate malignant pericardial effusion from the broader category of pericardial diagnoses related to neoplastic disease. Pericardial effusion in patients with malignancies may result from primary or metastatic involvement of cardiac structures, but may also be seen with radiation-induced pericarditis, opportunistic infection, or toxicity of chemotherapeutic agents. The malignant pericardial effusion occupies a smaller, more definitive niche as a secondary process due to metastatic disease of cardiac and pericardiac tissues.<sup>2,3</sup>

Malignant pericardial effusion is one of the most common types of pericardial effusion, and its presence is of clinical and prognostic importance. A proposal was received from Howard Rodenberg, MD, MPH, of the Association of Clinical Documentation Integrity Specialists (ACDIS), to expand and create a specific ICD-10-CM code for malignant pericardial effusion.

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**TABULAR MODIFICATIONS**

	I31	Other diseases of pericardium	
		I31.2 Hemopericardium, not elsewhere classified	
Add		Excludes1: ...	malignant pericardial effusion (I31.31)
		I31.3 Pericardial effusion (noninflammatory)	
Delete		<del>Chylopericardium</del>	
New code		I31.31 Malignant pericardial effusion in disease classified elsewhere	
Add			Code first underlying neoplasm
New code		I31.39 Other pericardial effusion (noninflammatory)	
Add			Chylopericardium

## **Mild Neurocognitive Disorder Due to Known Physiological Conditions**

The American Psychiatric Association (APA) presented this proposal at the September 2020 and March 2021 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 Psychiatric Association is 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 <sup>(1)</sup> 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 <sup>(2)</sup>. 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 <sup>(3)</sup>.

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 <sup>(4)</sup>, due to frontotemporal degeneration <sup>(5)</sup>, due to HIV disease <sup>(6)</sup>, due to Lewy body disease <sup>(7)</sup>, due to traumatic brain injury <sup>(8)</sup>, due to Parkinson's disease <sup>(9)</sup>, and due to Huntington's disease <sup>(10)</sup>. However, there is currently no ICD-10-CM for cases of mild cognitive disorder due to other medical conditions.

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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 5<sup>th</sup> 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.<sup>(11,12)</sup>

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 of uncertain or unknown etiology.”.

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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 sub-category

F06.7 Mild **neurocognitive** disorder due to known physiological condition

Add

Mild **neurocognitive** 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 **Disease** (G30.-)  
frontotemporal dementia (G31.09)  
human immunodeficiency virus [HIV] disease (B20)  
Huntington's disease (G10)

**Revise**

**Neurocognitive disorder Dementia with Lewy bodies (G31.83)**  
Parkinson's disease (G20)  
systemic lupus erythematosus (M32.-)  
traumatic brain injury (S06.-)  
vitamin B deficiency (E53-)

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Add	<p>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)</p>
New code	<p>F06.70 Mild <b>neurocognitive</b> disorder due to known physiological condition without behavioral disturbance</p>
Add	<p>Mild <b>neurocognitive</b> disorder due to known physiological condition, NOS</p>
New code	<p>F06.71 Mild <b>neurocognitive</b> disorder due to known physiological condition with behavioral disturbance</p>
	<p>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</p> <p>Code first the underlying physiological condition</p>
Add	<p>Excludes1:mild <b>neurocognitive</b> disorder due to known physiological condition (F06.7-) psychosis NOS (F29)</p>
	<p>G10 Huntington's disease Huntington's chorea Huntington's dementia</p>
Delete	<p><del>Code also dementia in other diseases classified elsewhere without behavioral disturbance (F02.80)</del></p>

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**Revise** Use additional code, if applicable, to identify:  
**Add** dementia with anxiety (F02.84, F02.A4, F02.B4, F02.C4)  
**Add** dementia with behavioral disturbance (F02.81-, F02.A1-  
F02.B1-, F02.C1-)  
**Add** dementia with mood disturbance (F02.83, F02.A3,  
F02.B3, F02.C3)  
**Add** dementia with psychotic disturbance (F02.82, F02.A2,  
F02.B2, F02.C2)  
**Add** dementia without behavioral disturbance (F02.80,  
F02.A0, F02.B0, F02.C0)  
**Add** mild neurocognitive disorder 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

**Revise** Use additional code, if applicable, to identify:  
**Add** dementia with anxiety (F02.84, F02.A4, F02.B4, F02.C4)  
**Revise** dementia with behavioral disturbance (F02.81-, F02.A1-,  
F02.B1-, F02.C1-)  
**Add** dementia with psychotic disturbance (F02.82, F02.A2,  
F02.B2, F02.C2)  
**Add** dementia with mood disturbance (F02.83, F02.A3,  
F02.B3, F02.C3)  
**Revise** dementia without behavioral disturbance (F02.80,  
F02.A0, F02.B0, F02.C0)  
**Add** mild neurocognitive disorder due to known physiological  
condition (F06.7-)

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

**Revise** Use additional code, if applicable, to identify:  
**Add** dementia with anxiety (F02.84, F02.A4, F02.B4, F02.C4)  
**Add** dementia with behavioral disturbance (F02.81-, F02.A1-  
F02.B1-, F02.C1-)

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- Add**                    **dementia with mood disturbance (F02.83, F02.A3, F02.B3, F02.C3)**
- Add**                    **dementia with psychotic disturbance (F02.82, F02.A2, F02.B2, F02.C2)**
- Add**                    **dementia without behavioral disturbance (F02.80, F02.A0, F02.B0, F02.C0)**
- Add**                    **mild neurocognitive 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:**
- Add**                    **dementia with anxiety (F02.84, F02.A4, F02.B4, F02.C4)**
- Revise**                **dementia with behavioral disturbance (F02.81-F02.A1-F02.B1-, F02.C1-)**
- Add**                    **dementia with psychotic disturbance (F02.82, F02.A2, F02.B2, F02.C2)**
- Add**                    **dementia with mood disturbance (F02.83, F02.A3, F02.B3, F02.C3)**
- Revise**                **dementia without behavioral disturbance (F02.80, F02.A0, F02.B0, F02.C0)**
- Add**                    **mild neurocognitive disorder due to known physiological condition (F06.7-)**
- Revise**                G31.09 Other frontotemporal neurocognitive-~~dementia~~ **disorder**  
Frontal dementia
- Add**                    Use additional code, if applicable, to identify mild **neurocognitive** disorders due to known physiological condition (F06.7-)
- G31.8 Other specified degenerative diseases of nervous system
- Revise**                **G31.83 Neurocognitive disorder ~~Dementia~~-with **Lewy bodies** /  
Dementia with Parkinsonism  
Lewy body dementia  
Lewy body disease**
- Add**                    Use additional code, if applicable, to identify mild **neurocognitive** disorders due to known physiological condition (F06.7-)
- Revise**                G31.84 Mild cognitive impairment of uncertain or unknown etiology, ~~so stated~~

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Revise	Mild neurocognitive disorder of <u>uncertain or unknown etiology</u>
Add	Mild cognitive disorder NOS
Add	Excludes1:mild <b>neurocognitive</b> disorder due to a known physiological condition (F06.7-)
<b>Add</b>	<b>cerebrovascular diseases (I60-I69)</b>
Delete	<del>mild memory disturbance (F06.8)</del>
<b>Add</b>	<b>Use additional code to identify presence of:</b> <b>alcohol abuse and dependence (F10.-)</b> <b>exposure to environmental tobacco smoke (Z77.22)</b> <b>history of tobacco dependence (Z87.891)</b> <b>hypertension (I10-I16)</b> <b>mild neurocognitive disorders due to known physiological condition (F06.7-)</b> <b>occupational exposure to environmental tobacco smoke (Z57.31)</b> <b>tobacco dependence (F17.-)</b>  <b>tobacco use (Z72.0)</b>
S06	Intracranial injury Includes: traumatic brain injury  Code also any associated: open wound of head (S01.-) skull fracture (S02.-)
<b>Add</b>	<b>Use additional code, if applicable, to identify mild neurocognitive disorders due to known physiological condition (F06.7-)</b>

## Mitral Annulus Calcification

The mitral annulus separates the left atrium from the left ventricle. It has a complex saddle shape that is divided into anterior and posterior portions. Mitral annulus calcification is a chronic, degenerative process of the fibrous support structure of the mitral valve.<sup>1</sup>

The reported incidence is between 8% and 15%, but it significantly increases with age and in patients with multiple cardiovascular risk factors or chronic kidney disease (CKD). Its clinical relevance comes from MAC's association with an increased rate of mortality and cardiovascular disease. MAC has also been found to increase the incidence of mitral valve disease and arrhythmias and to influence the outcome of cardiac surgery.<sup>1</sup> In fact, the risk of surgical mitral valve replacement in patient with severe MAC is high due to comorbidities and technical challenges related to calcium burden.

Surgical management of severe MAC is associated with significant risks and complexity.<sup>1,2</sup> Transcatheter mitral valve replacement (TMVR) is being studied as an alternative to surgery in patients with native mitral valve disease with severe MAC who are poor candidates for surgery.<sup>35</sup>

There are currently no ICD-10-CM codes describing the presence of MAC or MAC as a mitral valve disease.

This proposal is based on a request received from Mayra Guerrero, MD, FACC, FSCAI, of the Mayo Clinic College of Medicine, Department of Cardiovascular Medicine, and Mayo Clinic Hospital, the Heart Valve Collaborative, and Abbott Laboratories, Inc.; for new ICD-10-CM codes to specifically identify mitral annulus calcification to help identify patients with this condition, and to correlate with potential response to various treatments of mitral valve disease.

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**TABULAR MODIFICATIONS**

	I34	Nonrheumatic mitral valve disorders
		I34.0 Nonrheumatic mitral (valve) insufficiency
Add		Code also, if applicable:
Add		nonrheumatic mitral (valve) annulus calcification (I34.81)
		I34.2 Nonrheumatic mitral (valve) stenosis
Add		Code also, if applicable:
Add		nonrheumatic mitral (valve) annulus calcification (I34.81)
		I34.8 Other nonrheumatic mitral valve disorders
New code		I34.81 Nonrheumatic mitral (valve) annulus calcification
Add		Nonrheumatic mitral (valve) annular calcification
Add		Code also, if applicable:
Add		nonrheumatic mitral (valve) insufficiency (I34.0)
Add		nonrheumatic mitral (valve) stenosis (I34.2)
New code		I34.89 Other nonrheumatic mitral valve disorders

**INDEX MODIFICATIONS**

	Calcification
Revise	- heart valve - see <u>also</u> Endocarditis
Add	- - mitral – see Calcification, mitral
Add	- mitral (valve)
Add	- - annular I34.81
Add	- - - nonrheumatic I34.81
Add	- - - rheumatic I05.8
Add	- - annulus I34.81
Add	- - - nonrheumatic I34.81
Add	- - - rheumatic I05.8

## **Muscle Wasting and Atrophy of the Back**

The two main muscle groups of the back that experience primary muscle wasting and atrophy are multifidus muscle and paraspinal muscles. The multifidus muscle is a deep muscle of the back and part of the transversospinales muscle group<sup>1</sup>. Anatomically, the multifidus muscle is deep to the semispinalis muscle and superficial to the rotatores muscles<sup>1</sup>. The multifidus muscle runs the entire length of the vertebral column and on both sides of the vertebral column<sup>1</sup>. Its origin is the posterior sacrum, posterior superior iliac spin, mammillary process located on the superior articular process of the lumbar vertebrae, transverse process of the thoracic vertebrae and articular process of the lower cervical vertebra C4 – C7<sup>1</sup>. Its insertion is the base of each spinous process from L5 to C2. At each level, the insertion is 2 – 4 levels of origin<sup>1</sup>. The actions of the multifidus muscle include extension of the vertebral column and rotation of the vertebral column<sup>1</sup>. Along with other muscles of the back, the multifidus muscle plays an important role in the stabilization of the back.

The paraspinal muscles are sometimes referred to as the erector spinae. They are also part of the deep musculature of the back. They run the base of the cranium all the way to the pelvis<sup>2</sup> and are comprised of the spinalis muscles, the longissimus muscles, and iliocostalis muscles. Their function includes extension and lateral flexion of the spine and they play a role in maintaining posture.

The muscles of the back may experience degeneration<sup>3</sup>. Signs of this degeneration include decreased muscle size. Decreased radiographic density, and increased fat deposits within the muscles. For example, fatty infiltration of the lumbar multifidus is common among adults and is strongly associated to low back pain<sup>1</sup>. Magnetic resonance spectroscopy has concluded that the lumbar multifidus may experience fat deposition<sup>5</sup>. Furthermore, peer-reviewed literature tells us that people who experience low back pain have a significantly higher fat content within the muscles of the back compared to asymptomatic controls<sup>5</sup>. These are all forms of muscle wasting and atrophy of the back that can occur in the cervical, thoracic, and lumbosacral regions of the back.

There are newer treatment options successfully addressing muscle wasting and atrophy of the back, including implantable neurostimulation. This approach is designed to overcome arthrogenic muscle inhibition and elicit episodic contraction of the lumbar multifidus and resulting in pain and disability scores reported by patients suffering from chronic low back pain<sup>1,2</sup>.

With low back pain being incredibly common<sup>1</sup> and a frequent cause of visits to healthcare providers, it is useful to identify its cause. Peer-reviewed literature is replete with evidence that this very common low back pain is often caused by atrophy of the muscles of the back, typically involving fat infiltration<sup>2,3,4,5,6,7,8</sup>. Peer-reviewed literature also identifies that paraspinal muscles are significantly smaller than normal when fat infiltration occurs and patients experience instability and low back pain<sup>1</sup>. This decrease in size typifies atrophy of the muscles of the back. In recent years, the medical community has enjoyed the introduction of new imaging modalities, new research, more clinical experience, and new literature related to low back pain, all of which,

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considered together, makes it clear the this very common complaint is often caused by muscles wasting and atrophy of the muscles of the back.

It is proposed that new codes for muscle wasting and atrophy of the back will be beneficial because ICD-10-CM is an instrument in conducting research, such as epidemiology studies, tracking public health, and measuring quality, safety, and efficacy of care<sup>2</sup>.

Brigham and Women's Healthcare, Harvard Medical School, Center for Pain Management is requesting the creation of ICD-10-CM codes for muscle wasting and atrophy of the back.

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- <sup>17</sup> CMS Basic Introduction to ICD-10-CM

**TABULAR MODIFICATIONS**

M62 Other disorders of muscle

Excludes1: alcoholic myopathy (G72.1)  
cramp and spasm (R25.2)  
drug-induced myopathy (G72.0)  
myalgia (M79.1-)  
stiff-man syndrome (G25.82)

Excludes2: nontraumatic hematoma of muscle (M79.81)

M62.5 Muscle wasting and atrophy, not elsewhere classified

Disuse atrophy NEC

Excludes1: neuralgic amyotrophy (G54.5)  
progressive muscular atrophy (G12.21)  
sarcopenia (M62.84)

Excludes2: pelvic muscle wasting (N81.84)

New subcategory

M62.5A Muscle wasting and atrophy, not elsewhere  
classified, back

New code

M62.5A0 Muscle wasting and atrophy, not  
elsewhere classified, back, cervical

New code

M62.5A1 Muscle wasting and atrophy, not  
elsewhere classified, back, thoracic

New code

M62.5A2 Muscle wasting and atrophy, not  
elsewhere classified, back, lumbosacral

New code

M62.5A9 Muscle wasting and atrophy, not  
elsewhere classified, back, unspecified  
level

## Non-Traumatic Peritoneal Hemorrhage

Retroperitoneal hemorrhage is a particularly important site of occult or concealed hemorrhage.<sup>1</sup> In one series, for example, 66% of patients were anticoagulated (42% on warfarin, 30% on heparin, and 11% on low-molecular-weight heparin); 30% were on antiplatelet therapy; 16% were taking both anticoagulant and antiplatelet medications; and 15% were taking neither.<sup>1</sup> The most common symptom was pain: abdominal (67%), leg (24%), hip (22%), and back (21%); 10.1% were misdiagnosed upon their initial encounter. Mortality in this series was 6% within 7 days, 10% within 30 days, and 19% within 6 months. In another series, 82% of patients were on therapeutic anticoagulation, overall mortality was 22%, but hemorrhage-related mortality was 6%.<sup>2</sup> A recent review identifies other risk factors for spontaneous retroperitoneal hemorrhage, including strenuous exercise, coughing, coagulation disorders, and invasive procedures on or through the abdominal wall. The management of retroperitoneal hemorrhage or hematoma is largely supportive, with the reversal of anticoagulation, transfusions if needed, and angioembolization if bleeding continues in the setting of hemorrhagic shock.<sup>3</sup>

Retroperitoneal fibrosis is a slowly progressive disorder in which the ureters and other abdominal organs or vessels may become blocked by a fibrous mass and inflammation in the back of the abdomen.

University of California, UC Davis Division of General Medicine is requesting the creation of ICD-10-CM codes for non-traumatic peritoneal hemorrhage and retroperitoneal fibrosis for coding specificity.

### References

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4. Kasotakis G. Retroperitoneal and rectus sheath hematomas. *Surg Clin North Am*. 2014 Feb;94(1):71-6.

## TABULAR MODIFICATIONS

K66	Other disorders of peritoneum
	K66.1 Hemoperitoneum
Add	Excludes2: abdominal hemorrhage (R58.81)
Add	retroperitoneal hematoma (K68.3)
Add	retroperitoneal hemorrhage (R58.82)

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	K68	Disorders of retroperitoneum
New code	K68.2	Retroperitoneal fibrosis
Add		Code also, if applicable, associated obstruction of ureter (N13.5)
New code	K68.3	Retroperitoneal hematoma
Add	Excludes2:	retroperitoneal hemorrhage (R58.82)
	R58	Hemorrhage, not elsewhere classified
Delete		<del>Hemorrhage NOS</del>
New subcategory	R58.8	Other hemorrhage, not elsewhere classified
New code	R58.81	Abdominal hemorrhage
Add		Intra-abdominal hemorrhage
Add		Subdiaphragmatic hemorrhage
New code	R58.82	Retroperitoneal hemorrhage
New code	R58.83	Ruptured vessel (blood)
New code	R58.89	Other hemorrhage, not elsewhere classified
New code	R58.9	Hemorrhage, unspecified

**INDEX MODIFICATIONS**

Revise	Ecchymosis R58. <u>89</u>
	Extravasation
Revise	- blood R58. <u>9</u>
	Hematoma (traumatic) (skin surface intact) -see also Contusion
Revise	- retroperitoneal (nontraumatic) <del>K66.4</del> <u>K68.3</u>
Revise	Hemorrhage, hemorrhagic (concealed) R58. <u>9</u>
Revise	- abdomen R58. <u>81</u>
Revise	- artery R58. <u>83</u>
Revise	- internal (organs) NEC R58. <u>81</u>
Revise	- intra-abdominal R58. <u>81</u>
Revise	- mucous membrane NEC R58. <u>89</u>
Revise	- retroperitoneal R58. <u>82</u>
Revise	- scalp R58. <u>89</u>
Revise	- secondary (nontraumatic) R58. <u>89</u>
Revise	- subdiaphragmatic R58. <u>81</u>
Revise	- viscera NEC R58. <u>81</u>
	Rupture, ruptured
Revise	- splenic vein R58. <u>83</u>
Revise	- vena cava R58. <u>83</u>

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Revise - vessel (blood) R58.83

Syndrome -see also Disease

Revise - retroperitoneal fibrosis ~~N13.5~~ K68.2

## **Onset (spontaneous) of labor after 37 completed weeks of gestation but before 39 completed weeks gestation, with vaginal delivery**

The American College of Obstetricians and Gynecologists (ACOG) is proposing to add new code(s) that allow for reporting of the onset of labor after 37 completed weeks of gestation but before 39 completed weeks of gestation, with vaginal delivery. The current code set does not include codes that allow for the reporting of vaginal delivery within the early term period of 37.0 gestational weeks and 38.6 gestational weeks. This proposal seeks to fill this coding gap.

Some patients will present in spontaneous labor in the early term period they may also subsequently need augmentation with induction agents such as Pitocin to progress to vaginal delivery. To better describe these deliveries that progressed from spontaneous labor there is a need for an additional ICD-10-CM code description.

Early term delivery, between 37 0/7 and 38 6/7 weeks, can lead to associated neonatal morbidities. In ACOG Committee Opinion 765 ACOG and SMFM recommends against non-medically indicated planned C-section, inductions of labor and cervical ripening prior to 39 0/7 weeks and as this can lead to neonatal pulmonary complications, increased need for admission to neonatal intensive care, hypoglycemia and neonatal mortality. Because of the known morbidity and mortality concerns quality metrics for early term deliveries are now closely monitored yielding a need for proper code description to allow for quality metric clarification. Better understanding of spontaneous labor versus improper scheduled delivery in early term period can lead to better patient outcomes as well as improved accuracy of quality metrics.

ACOG is requesting the following modifications:

### **References**

American College of Obstetricians and Gynecologists. (2013, April). Avoidance of Nonmedically Indicated Early-Term Deliveries and Associated Neonatal Morbidities. *ACOG Committee Opinion Number 765, 133(2)*, e156-163.  
Clark, S. L., Meyers, J. A., Milton, C. G., Frye, D. R., Horner, S., Baker, A., & Perlin, J. B. (2014, January). Validation of The Joint Commission Exclusion Criteria for Elective Early-Term Delivery. *Obstetrics & Gynecology, 123(1)*, 29-33.

### **TABULAR MODIFICATIONS**

O75 Other complications of labor and delivery, not elsewhere classified

Excludes2: puerperal (postpartum) infection (O86.-)  
puerperal (postpartum) sepsis (O85)

O75.8 Other specified complications of labor and delivery

O75.81 Maternal exhaustion complicating labor and delivery

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O75.82 Onset (spontaneous) of labor after 37 completed weeks of gestation but before 39 completed weeks gestation, with delivery by (planned) cesarean section

Delivery by (planned) cesarean section occurring after 37 completed weeks of gestation but before 39 completed weeks gestation due to (spontaneous) onset of labor

Code first to specify reason for planned cesarean section such as:  
cephalopelvic disproportion (normally formed fetus) (O33.9)  
previous cesarean delivery (O34.21)

New code O75.83 Onset (spontaneous) of labor after 37 completed weeks of gestation but before 39 completed weeks gestation, with vaginal delivery  
O75.89 Other specified complications of labor and delivery

## **Parkinson's Disease with OFF episodes**

Parkinson's disease (PD) is a progressive neurodegenerative disease that presents with motor symptoms such as bradykinesia with muscle rigidity, tremor, and/or postural instability, as well as non-motor symptoms such as anxiety/panic attacks, problems with executive function, and pain.<sup>1</sup> Normally, neurons in the substantia nigra produce the neurotransmitter dopamine, which helps to regulate movement. In patients with PD, these neurons (among others) begin to die and less dopamine is produced, resulting in PD symptoms.<sup>2</sup>

It is estimated that approximately 1.04 million people in the United States had PD in 2017 and 1.2 million are estimated to have PD by 2030.<sup>3,4</sup> Currently, no cure or disease-modifying therapies exist and treatment relies mainly upon levodopa to relieve motor and nonmotor symptoms.<sup>5</sup> As PD is a progressive disease, patients receiving standard maintenance treatment with levodopa will experience a narrowing duration of effect, leading to complications/fluctuations (dyskinesias/OFF episodes) that become difficult to control.<sup>2,6,7</sup> Each patient's experience with PD is unique with some patients experiencing dyskinesias, OFF episodes, or both.<sup>7-9</sup>

Motor fluctuations are inherent to PD and are likely to occur in 50% of patients in 5 years and 100% of patients within 10 years of treatment initiation.<sup>10,11</sup> Based on our epidemiology model using the 1 million people in the United States with PD (2020), it is estimated there are 375,000 PD patients experiencing OFF episodes.<sup>11-13</sup> Motor fluctuations are typically described as periods of good motor function (ON state) followed by periods when PD symptoms reemerge (OFF state) or when uncontrollable hyperkinetic movements are present.<sup>14-15</sup> The occurrence of motor fluctuations (OFF episodes/dyskinesias) are important signs/symptoms to monitor in the management of PD because it can be an indication that therapy may need to be optimized to control baseline symptoms.<sup>15</sup>

A wide range of symptoms have been observed during OFF states such as tremor, rigidity, bradykinesia, difficulty with speech and balance, weakness, and reduced dexterity.<sup>15,16</sup> Response fluctuations may also present as nonmotor symptoms.<sup>15,16</sup> Non-motor symptoms that have been reported to occur during fluctuations include apathy, anxiety, irritability, mood changes, cognitive changes, fatigue, pain, and drenching sweats.<sup>15,16</sup>

Fluctuations may have a significant impact on patients.<sup>17</sup> Fluctuations such as OFF episodes may also increase hospitalizations and emergency department (ED) visits, as well as increasing intensive care unit (ICU) admission and prolonging the length of stay.<sup>18</sup> In a recent real world analysis of PD patients (N=1409), patients who reported experiencing "OFF" episodes were associated with three times higher number of emergency room visits and hospitalizations compared to those without "OFF" episodes. The study also demonstrated that each incremental OFF-hour/day may also result in 60-70% greater ICU admission and length of hospital stays.<sup>18</sup> Interventions specifically targeting the reduction of OFF-time may help reduce the number of OFF-episode related ER visits, hospitalizations and subsequent health care resource utilization.

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Sunovion Pharmaceuticals Incorporated with 39 Movement Disorder Specialists (MDS) and the Unified Parkinson's Advocacy Council (UPAC) are requesting the following new codes to enhance the tracking and the progression of Parkinson's disease.

References:

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18. Rajagopalan K, Barton J, Pike J. Real-world assessment of "OFF" episode-related health resource use among patients with Parkinson's disease. Poster presented: 2nd Pan American Parkinson's Disease and Movement Disorders Congress; June 22-24, 2018; Miami, FL.

**TABULAR MODIFICATIONS**

	G20	Parkinson's disease
		Hemiparkinsonism
		Idiopathic Parkinsonism or Parkinson's disease
		Paralysis agitans
Delete		<del>Parkinsonism or Parkinson's disease NOS</del>
		Primary Parkinsonism or Parkinson's disease
New subcategory		G20.A Parkinson's disease without dyskinesia
New code		G20.A1 Parkinson's disease without dyskinesia, without fluctuations
Add		Parkinson's disease without dyskinesia, without OFF episodes
New code		G20.A2 Parkinson's disease without dyskinesia, with fluctuations
Add		Parkinson's disease without dyskinesia, with OFF episodes
New subcategory		G20.B Parkinson's disease with dyskinesia
Add		Excludes1: Drug induced dystonia (G24.0-)
New code		G20.B1 Parkinson's disease with dyskinesia, without fluctuations
Add		Parkinson's disease with dyskinesia, without OFF episodes
New code		G20.B2 Parkinson's disease with dyskinesia, with fluctuations
Add		Parkinson's disease with dyskinesia, with OFF episodes
New code		G20.C Parkinsonism, unspecified
Add		Parkinsonism, NOS

## **Perpetrator of assault, maltreatment and neglect**

The ICD-10-CM classification currently does not include codes for ex-spouse, ex-partner, friend, or acquaintance type perpetrators. Adding codes for these specific perpetrators would provide more detail for accurate coding and reporting on assault and abuse cases. Per the *ICD-10-CM Official Guidelines for Coding and Reporting FY 2021* on page 88, “A perpetrator code (Y07) should be added when the perpetrator of the abuse is known.”

The proposed codes Y07.2, Acquaintance or friend, perpetrator of maltreatment and neglect and Y07.3, Official authority, perpetrator of maltreatment and neglect, are existing codes under the World Health Organization (WHO) ICD-10 category Y07, Perpetrator of assault, maltreatment and neglect.

The submitter is requesting new codes to increase the accuracy of statistics for healthcare research and decision making.

### **TABULAR MODIFICATIONS**

	Y07	Perpetrator of assault, maltreatment and neglect
New code	Y07.2	Acquaintance or friend, perpetrator of maltreatment and neglect
New code	Y07.3	Official authority, perpetrator of maltreatment and neglect
New code	Y07.A	Ex-spouse or ex-partner, perpetrator of maltreatment and neglect
Add		Ex-husband, perpetrator of maltreatment and neglect
Add		Ex-wife, perpetrator of maltreatment and neglect

## **Personal History of (Corrected) Congenital Malformations and Personal History of (Corrected) Certain Conditions Arising in the Perinatal Period**

The Children's Hospital Association (CHA) is requesting new ICD-10-CM codes for selected digestive and musculoskeletal system congenital conditions. These conditions continue to cause morbidity throughout the lifetime of patients who were born with these congenital conditions even though they were corrected or palliated by surgical means.

These select conditions are commonly repaired when the child is young, either through a single definitive procedure or by performing multiple staged procedures to accomplish the repair. Once the repair is complete, the patient may continue to require medical care and/or surgical treatment of new or acquired conditions that are related to the original surgical repair of the congenital condition or the remaining abnormal anatomy that these patients have throughout their lives.

Currently, there are no "sequelae of congenital condition" codes to help describe these issues in ICD-10-CM. Also, in these instances, the original congenital condition is no longer present once the repair has been completed. Therefore, the Q category code would not be assigned such as when codes for other perinatal or congenital conditions could be assigned when the congenital condition is still present and requiring treatment.

To provide a more specific method of tracking patients who have a history of repaired congenital conditions but still require related treatment over their lifetime due to this history, The Children's Hospital Association (CHA) would like to expand two sections of code category Z87, Personal history of other diseases and conditions.

The specific digestive system conditions (and rationale) requested to be captured through the expansion of Z87.73 include:

- Tracheoesophageal fistula or atresia (Q39.0, Q39.1, Q39.2): these patients frequently have tracheal or esophageal stenoses at the area of the previous fistula or repair that require dilation or further surgical repairs.
- Persistent cloaca (cloacal anomaly) (Q43.7): These patients frequently have lifelong genitourinary tract and intestinal tract difficulties that may require further treatment or surgical repairs.

The specific musculoskeletal system conditions (and rationale) requested to be captured through the expansion of Z87.76 include:

Congenital diaphragmatic hernia (Q79.0) and other congenital diaphragm malformations (Q79.1)

- Gastroschisis (Q79.3)
- Prune Belly Syndrome (Q79.4)
- Other abdominal wall congenital malformations (Q79.2, Q79.51, Q79.59): Includes omphalocele, congenital hernia of bladder, and all other congenital abdominal wall malformations.

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These patients can have many problems throughout their life such as hernias, abdominal scarring, bowel obstructions, dysmotility/dysfunction of bowel or bladder, bowel incontinence, bladder incontinence, etc. that require further treatment or surgical repair.

There are currently specific ICD-10-CM codes under Z87.7 for a few congenital conditions [i.e., Z87.710, Personal history of (corrected) hypospadias and Z87.730 Personal history of (corrected) cleft lip and palate].

This proposal is requesting similar codes to those for personal history of hypospadias and cleft lip/palate. There is already a precedence in the code set for these types of codes and this proposal would be an expansion of the current ICD-10-CM codes to provide more patient detail.

The Children's Hospital Association (CHA) is also requesting new codes for Personal history of certain conditions arising in the perinatal period, Z87.6- so that a code can be created for Personal history of (corrected) necrotizing enterocolitis of newborn. This is another condition that can be surgically corrected but the patient may commonly have sequela or related conditions after treatment that may require further surgical treatment or hospitalization.

A personal history code for this condition will help tell the complete story of the patient's illness that cannot be captured by current available codes. Code Category Z87.6 is in the WHO's ICD-10 classification system but was not brought forward into the United States' ICD-10- Clinical Modification.

This proposal is supported by the American Academy of Pediatrics.

### **TABULAR MODIFICATIONS**

Z87	Personal history of other diseases and conditions Code first any follow-up examination after treatment (Z09)
New subcategory	Z87.6 Personal history of certain (corrected) conditions arising in the perinatal period Conditions classifiable to P00-P96
New code	Z87.61 Personal history of (corrected) necrotizing enterocolitis of newborn
New code	Z87.68 Personal history of other (corrected) conditions arising in the perinatal period
	Z87.7 Personal history of (corrected) congenital malformations

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Conditions classifiable to Q00-Q89 that have been repaired  
or corrected

Z87.73 Personal history of (corrected) congenital malformations  
of digestive system

Z87.730 Personal history of (corrected) cleft lip and  
palate

New code Z87.731 Personal history of (corrected)  
tracheoesophageal fistula or atresia

New code Z87.732 Personal history of (corrected) persistent  
cloaca or cloacal anomaly

Z87.738 Personal history of other specified  
(corrected) congenital malformations  
of digestive system

New subcategory Z87.76 Personal history of (corrected) congenital malformations of  
integument, limbs and musculoskeletal system

New code Z87.760 Personal history of (corrected) congenital  
diaphragmatic hernia or other congenital  
diaphragm malformations

New code Z87.761 Personal history of (corrected) gastroschisis

New code Z87.762 Personal history of (corrected) prune belly  
malformation

New code Z87.763 Personal history of other (corrected) congenital  
abdominal wall malformations

New code Z87.768 Personal history of other specified (corrected)  
congenital malformations of integument,  
limbs and musculoskeletal system

## **PIK3CA-related Overgrowth Spectrum and Related Disorders**

PIK3CA (Phosphatidylinositol-4,5-Bisphosphate 3-Kinase Catalytic Subunit Alpha) is a gene that is involved with cell growth among other things. It is associated with a number of disorders, certain of these grouped under the general description “PIK3CA-related overgrowth spectrum” (PROS). There are additional non-malignant PIK3CA-related disorders, also encompassing PIK3CA-related congenital anomalies without overgrowth. PROS is a group of rare disorders most often caused by post-zygotic somatic mutations in the *PIK3CA* gene that can result in asymmetric overgrowth of parts of the body.<sup>1,2</sup> Unique to PROS and unlike in other congenital anomalies, the somatic *PIK3CA* mutation tends to manifest in certain cells or certain locations of the body, also known as somatic mosaicism, and rarely occurs through a *de novo* germline mutation.<sup>1-3</sup> Specific disorders that fall within PROS include but are not limited to<sup>1,4,5</sup>:

- Fibroadipose hyperplasia
- Congenital lipomatous overgrowth, vascular malformations, epidermal nevi and scoliosis/skeletal/spinal anomalies (CLOVES) syndrome
- Megalencephaly-capillary malformation syndrome
- Hemihyperplasia-multiple lipomatosis (HHML) syndrome
- Hemimegalencephaly
- Facial infiltrating lipomatosis
- Klippel-Trenaunay syndrome

Patients with PROS may develop a variety of signs and symptoms because overgrowth occurs in different parts of the body depending on the specific disorder.<sup>6</sup> Some examples of clinical symptoms associated with individual manifestations of PROS include having a larger-than-normal brain (megalencephaly), lipomatous (fatty) growths on the arms or face, seizures, intellectual disability, vascular malformations, and overgrowth of one or more areas of the body with normal growth elsewhere.<sup>1</sup> Providers tend to suspect PROS from the exhibited phenotype, and may confirm diagnosis through genetic testing for the *PIK3CA* gene.<sup>2,7</sup> Although there is no cure for PROS, patients can receive treatments to help manage symptoms of the disorder through a multidisciplinary team of clinicians.<sup>1</sup>

Study of the role of *PIK3CA* mutations in these congenital malformation syndromes also aligns with the work of the NIH’s National Center for Advancing Translational Sciences (NCATS) in its SaME concept initiative (SaME Therapeutics—Clinical Trials of Drugs Targeting Shared Molecular Etiologies in Rare Diseases)<sup>9</sup> in identifying drugs with potential benefit for affected patients. Notably, alpelisib, a therapeutic already approved and in use for HR+, HER2-, *PIK3CA*-mutated advanced or metastatic breast cancer, is currently under clinical investigation<sup>10</sup> for treatment of certain manifestations of *PIK3CA*-related Congenital Anomaly Spectrum, and in fact is currently available on a Compassionate Use basis<sup>11</sup> for treatment of those manifestations.

Parkes Weber Syndrome is a disorder causing overgrowth of one limb, usually a leg, along with capillary malformations or arteriovenous malformations. Some cases of Parkes Weber syndrome result from mutations in the *RASA1* gene. It is proposed to index Parkes Weber syndrome to code Q87.2, Congenital malformation syndromes predominantly involving limbs; along with a note to see also Malformation, arteriovenous.

In a different aspect of association, germline mutations in PIK3CA have been reported in cases of Cowden syndrome.<sup>13</sup> It thus also appears that PIK3CA may be a Cowden syndrome susceptibility gene.<sup>13</sup> The phosphoinositide 3-kinases are a family of proteins involved in regulation of cell growth, metabolism, and proliferation, among other things.<sup>14</sup> The PTEN gene is a tumor suppressor, that is a negative regulator of the phosphoinositide 3-kinase (PI3K) signaling.<sup>14</sup> PIK3CA mutations has also been found in a number of types of cancer, but these have been somatic mutations in cancer tissue, and conversely, the PIK3CA-related overgrowth syndromes (generally involving somatic mosaicism) are not associated with malignancies.

More specific ICD-10-CM coding describing both the molecular etiology, when available, and the clinical phenotype (i.e., syndrome or anomaly) would facilitate identifying those who have non-malignant PIK3CA-related disorders (including PROS), and differentiate them from patients with other types of overgrowth and non-overgrowth congenital syndromes, and vascular anomalies. In addition, such specific ICD-10-CM coding would enable providers and researchers to track and measure outcomes from clinical interventions, which could ultimately result in improved treatment modalities and the development of standard of care protocols for patients suffering from non-malignant PIK3CA-related congenital disorders.

This proposal has been based on a request for new specific ICD-10-CM coding related to a number of specific non-malignant PIK3CA-related disorders, but with some simplification and modifications to follow ICD clinical structure. This request was received from parties noted below, including clinicians who are specialists in the diagnosis and treatment of non-malignant PIK3CA-related disorders, Novartis Corporation, and the executive director of a major patient advocacy group, who all support this proposal and provided clinical input and coding recommendations during its development.

- Denise M. Adams, MD – Children’s Hospital of Philadelphia (current); Co-Director, Vascular Anomalies Center, Boston Children’s Hospital (former)
- Ilona Frieden, MD – Director, University of California San Francisco Birthmarks and Vascular Anomalies Center, UCSF Benioff Children's Hospital
- Adrienne M. Hammill, MD, Ph.D. – Research Director, Hemangioma & Vascular Malformation Program, Cincinnati Children’s Hospital
- Taizo Nakano, MD – Medical Director, Vascular Anomalies Center, Children's Hospital Colorado Center for Cancer and Blood Disorders
- Jonathan A. Perkins, DO – Head, Vascular Anomalies Program, Seattle Children’s Hospital
- Kristen Davis, Executive Director of CLOVES Syndrome Community

The International Society for the Study of Vascular Anomalies (ISSVA) also provided clinical input and coding recommendations during the development of this proposal.

#### References

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## TABULAR MODIFICATIONS

Q87 Other specified congenital malformation syndromes affecting multiple systems

Q87.2 Congenital malformation syndromes predominantly involving limbs

Delete	<del>Holt Oram syndrome</del>
Delete	<del>Klippel-Trenaunay-Weber syndrome</del>
Delete	<del>Nail patella syndrome</del>
Delete	<del>Rubinstein-Taybi syndrome</del>

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Delete	<del>Sirenomelia syndrome</del>
Delete	<del>Thrombocytopenia with absent radius [TAR] syndrome</del>
Delete	<del>VATER syndrome</del>
New code	Q87.21 Klippel-Trenaunay syndrome
New code	Q87.210 PIK3CA-related Klippel-Trenaunay syndrome
New code	Q87.218 Other Klippel-Trenaunay syndrome
New code	Q87.219 Klippel-Trenaunay syndrome, unspecified Klippel-Trenaunay-Weber syndrome, NOS
New code	Q87.29 Other congenital malformation syndromes predominantly involving limbs
Add	Holt-Oram syndrome
Add	Nail patella syndrome
Add	Parkes-Weber syndrome
Add	Rubinstein-Taybi syndrome
Add	Sirenomelia syndrome
Add	Thrombocytopenia with absent radius [TAR] syndrome
Add	VATER syndrome
Add	Q87.3 Congenital malformation syndromes involving early overgrowth Excludes1: PIK3CA-related overgrowth syndrome (Q87.A)
New subcategory	Q87.A PIK3CA-related congenital anomaly disorder
Add	Non-malignant PIK3CA-related congenital anomaly spectrum
Add	PIK3CA-related capillary malformation of the lower lip, lymphatic malformation, and partial or generalized overgrowth (CLAPO) syndrome
Add	PIK3CA-related congenital lipomatous overgrowth, vascular malformations, epidermal nevi and scoliosis/skeletal/spinal anomalies (CLOVES) syndrome
Add	PIK3CA-related epidermal nevus, benign lichenoid keratosis, seborrheic keratosis
Add	PIK3CA-related facial infiltrating lipomatosis
Add	PIK3CA-related fibroadipose hyperplasia
Add	PIK3CA-related fibroadipose vascular anomaly
Add	PIK3CA-related hemihyperplasia-multiple lipomatosis (HHML)
Add	PIK3CA-related hemimegalencephaly
Add	PIK3CA-related isolated lymphatic malformation

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Add PIK3CA-related macrodactyly  
Add PIK3CA-related megalencephaly-capillary malformation  
syndrome  
Add PIK3CA-related muscular hemihyperplasia  
Add PIK3CA-related overgrowth spectrum disorder  
Add PIK3CA-related segmental overgrowth  
Add Code also, if applicable, specific manifestations such as:  
Add hemimegalencephaly (Q04.8)  
Add megalencephaly (Q04.5)  
Add scoliosis (M41.-)

**INDEX MODIFICATIONS**

Add Hemimegalencephaly Q04.8

## **Postural orthostatic tachycardia syndrome (POTS)**

Postural orthostatic tachycardia syndrome (POTS) is a chronic autonomic nervous system disorder that can cause severe disability, and impaired quality of life.<sup>1</sup> POTS is estimated to affect as many as 500,000 to 3 million people in the U.S.<sup>2</sup>, although precise epidemiological studies have not been conducted to date.

Although POTS was given its modern definition in 1993<sup>3</sup>, it has been described in the medical literature since the Civil War.<sup>4</sup> At that time, a military physician, J. M. DaCosta, described a post-infectious syndrome in soldiers resulting in severe lightheadedness, tachycardia, dyspnea, headache, abdominal distension, and fatigue. This has also been known as DaCosta syndrome, Irritable Heart, Solider's Heart, Civil War Syndrome, Effort Syndrome, and many other terms throughout history. Infectious agents are one of the most common triggers for the onset of POTS.<sup>1,5</sup>

There is not a specific ICD-10-CM code for POTS, nor is there an index entry for it. One code that has been recommended and used is I49.8, Other specified cardiac arrhythmias. However, this is misleading. The tachycardia in POTS is sinus tachycardia that is noted upon standing which returns to either normal sinus rhythm or persistent, albeit lower rate of, sinus tachycardia upon assuming a recumbent position.

Creation of a specific code for POTS will support research, including clinical, epidemiological, medical utilization and economic impact, and other POTS research. There is growing interest in POTS research from government agencies and academic medical centers around the world. The US Congress has recognized POTS and the need for improved clinical care and research.<sup>6</sup> The U.S. National Institutes of Health recently issued its first Notice of Special Interest to Stimulate Research on the Diagnosis, Treatment, and Mechanistic Understanding of Postural Orthostatic Tachycardia Syndrome.<sup>7</sup>

A request for creation of a specific code for POTS within the category G90 has been received from Jeffrey R. Boris, MD (Jeffrey R. Boris, MD LLC), and Lauren Stiles, JD (Dysautonomia International and Research Assistant Professor of Neurology at Stony Brook University).

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**TABULAR MODIFICATIONS**

G90 Disorders of autonomic nervous system

New code	G90.A Postural orthostatic tachycardia syndrome
Add	Chronic orthostatic intolerance
Add	Postural tachycardia syndrome

## Postviral and Related Fatigue Syndromes

In 2015, the Institute of Medicine (IOM), now called the National Academy of Medicine, published an extensive evidence review of “myalgic encephalomyelitis/chronic fatigue syndrome” (ME/CFS) and recommended new diagnostic criteria. The CDC, disease experts, and many clinical guidance and medical education providers have already adopted the IOM criteria along with the term “myalgic encephalomyelitis/chronic fatigue syndrome.” Postviral Fatigue Syndrome proposals were presented at the September 2011 and September 2018 C&M meetings; the comments did not support the proposals. An updated proposal is included. This new proposal was submitted by The International Association for Chronic Fatigue Syndrome/Myalgic Encephalomyelitis, #MEAction, Open Medicine Foundation, Solve M.E., Massachusetts ME/CFS & FM Association, the Minnesota ME/CFS Alliance, and Pandora Org.

Currently, the term does not exist in the ICD-10-CM and the ICD-10-CM code most often used is the one for “chronic fatigue syndrome,” which is the same code as the symptom of “chronic fatigue”, (R53.82). As a result, it is difficult to accurately track ME/CFS separate from the symptom of chronic fatigue. This could also have secondary effects on healthcare resource planning, fiscal support for clinical care, use of medical records in future research, provisioning of workplace/school accommodations, and determination of disability benefits.

In its 2015 report, the Institute of Medicine concluded that ME/CFS is “*a serious, chronic, complex, and multisystem disease,*” characterized by the hallmark symptom of post-exertional malaise (PEM) in which even small amounts of cognitive and physical exertion can exacerbate symptoms that can last for days, weeks or sometimes months. ME/CFS is debilitating with an estimated 25% of patients homebound or bedbound and as many as 75% unable to work or go to school. ME/CFS is known to often follow a viral infection

The rationale for including ME/CFS as a synonym to ME is that the ME and ME/CFS criteria, including the IOM criteria, all require the hallmark symptoms of post-exertional malaise, unrefreshing sleep, profound fatigue, and significant impairment in function. These criteria also include other symptoms such as cognitive impairment, orthostatic intolerance, pain, and sensory sensitivity. The US ME/CFS Clinician Coalition recommends the IOM criteria for diagnosis in the US, but also acknowledges the similarities across these definitions and notes that some experts use the 2003 Canadian Consensus Criteria (labeled as ME/CFS) and the 2011 ME International Consensus Criteria (labeled as ME) to validate an IOM-criteria clinical diagnosis of ME/CFS.

The IOM report established new diagnostic criteria that include PEM and substantial impairment in functioning, profound fatigue, unrefreshing sleep, and either cognitive issues or orthostatic intolerance. Other common but non-required symptoms include joint and muscle pain, headaches, and sensitivity to noise and light. Published peer-reviewed studies have demonstrated neurological, immunological, autonomic, and energy metabolism impairment associated with these symptoms.

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In addition to the new clinical diagnostic criteria, the IOM also recommended a new name, “Systemic exertion intolerance disease (SEID).” Federal agencies including CDC and NIH, disease experts, and medical education and clinical guidance providers have adopted the term “ME/CFS,” instead of SEID, along with the new clinical diagnostic criteria recommended by the IOM in the 2015 report.

The IOM noted that not all persons previously diagnosed with CFS using the Fukuda CFS definition, used in the US prior to adoption of the IOM criteria, would meet the new IOM criteria for ME/CFS. The IOM did not intend the term ME/CFS to be a replacement for the term chronic fatigue syndrome or an amalgamation of all ME and CFS diagnoses.

Consideration was given as a potential alternative is to have coders separately apply the two codes for the terms, myalgic encephalomyelitis and chronic fatigue syndrome when the doctor diagnoses ME/CFS. However, this does not capture the name of the disease as specified by the doctor or in clinical guidance and medical education. Additionally, this currently could not be done due to existing instructional (Excludes1) notes.

The code title for ICD-10-CM code, G93.3, is currently, postviral fatigue syndrome (PVFS) and ME is an inclusion term. Because the G93.3 title specifies postviral illness, some doctors have declined to diagnose ME when viral illness is not proven. However, according to practice and established criteria, ME can be triggered by both viral and non-viral precipitants, including non-viral infections and non-infectious causes. This is also true for the definitions that use the ME/CFS label, including the IOM criteria.

It is also being recommended the code title G93.3, Postviral fatigue syndrome, be revised to Postviral and related fatigue syndromes to include other precipitants and still maintain the code title’s original wording. By its name, the term postviral fatigue syndrome is intended only for post-viral illness. But as noted above, the terms ME and ME/CFS include both viral and nonviral precipitants.

The ME and ME/CFS definitions further specify that symptoms should persist for 6 months while PVFS is used even if patients have not been sick for six months.

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  - Medscape 2019 ME/CFS CME - [https://www.medscape.org/viewarticle/907632?src=acdmprt\\_cdc](https://www.medscape.org/viewarticle/907632?src=acdmprt_cdc)
  - Kaiser Permanente. ME/CFS medical information. <https://healthy.kaiserpermanente.org/northern-california/health-wellness/health-encyclopedia/he.myalgic-encephalomyelitis-chronic-fatigue-syndrome.hw32907>  
Content provided by Healthwise which provides content to a number of other sites as well
  - Cleveland Clinic - <https://my.clevelandclinic.org/health/diseases/17720-myalgicencephalomyelitischronic-fatigue-syndrome-mecfs>

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11. Institute of Medicine 2015 report. Page 5
12. Institute of Medicine 2015 report. Page 8, 77
13. See the evidence review in the Institute of Medicine report. Also see Komaroff AL. Advances in understanding the pathophysiology of chronic fatigue syndrome. *JAMA*. 2019;322(6):499.

### TABULAR MODIFICATIONS

	A85 Other viral encephalitis, not elsewhere classified
Revise	Excludes1: <del>Benign</del> Myalgic encephalomyelitis (G93.32)
	G04 Encephalitis, myelitis and encephalomyelitis
Revise	Excludes2: <del>Benign</del> Myalgic encephalomyelitis (G93.32)
	G93 Other disorders of brain
Revise	G93.3 Postviral <u>and related</u> fatigue syndromes
Delete	<del>Benign myalgic encephalomyelitis</del>
	Excludes1: chronic fatigue, unspecified (R53.82)
Add	neurasthenia (F48.8)
New code	G93.31 Postviral fatigue syndrome
New code	G93.32 Myalgic encephalomyelitis/chronic fatigue syndrome
Add	Chronic fatigue syndrome
Add	ME/CFS

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Add	Myalgic encephalomyelitis
New code	G93.39 Other post infection and related fatigue syndromes
R53	Malaise and fatigue
	R53.8 Other malaise and fatigue
	R53.82 Chronic fatigue, unspecified
Delete	<del>Chronic fatigue syndrome NOS</del>
Add	Excludes1: chronic fatigue syndrome (G93.32)
Add	myalgic encephalomyelitis (G93.32)
Add	post infection and related fatigue syndromes (G93.39)
Revise	postviral fatigue syndrome (G93.31)

**INDEX MODIFICATIONS**

Revise	Akureyri's disease G93.3 .39
	Disease, diseased - see also Syndrome
Revise	- Iceland <del>G93.3</del> <u>G93.39</u>
	Encephalomyelitis G04.90 - see also Encephalitis
Delete	- <del>benign myalgia</del> <del>G93.3</del>
Revise	- myalgic, (benign) <del>G93.3</del> <u>G93.32</u>
Revise	Neuromyasthenia (epidemic) (postinfectious) <del>G93.3</del> <u>G93.39</u>
	Syndrome - see also Disease
Add	- postbacterial fatigue G93.39
Add	- postinfectious fatigue G93.39
Revise	- postviral NEC <del>G93.3</del> <u>G93.31</u>
Revise	- - fatigue <del>G93.3</del> <u>G93.31</u>
Add	Systemic exertion intolerance disease [SEID] G93.32

## Problems Related to Upbringing

The American Academy of Pediatrics (AAP) has previously presented a proposal on Problems Related to Upbringing at the September 2019 and March 2020 Coordination and Maintenance meeting. In response to comments received, the Academy is submitting a revised proposal for consideration to better identify problems related to upbringing and to better clarify the specific caregiver (or situation) the child is involved.

Today there are a greater variety of family dynamics that are more extended than the traditional nuclear family. A child may be living with a step-parent or non-parental guardian, such as a grandparent, almost as often as living with a biological or adopted parent. Children living with non-parental caregivers often present similar situations that may contribute to the child's wellbeing and need to seek medical attention.

It is the intent of the Academy this revised proposal will better capture these expanded "family" dynamics and conflicts that can complicate a medical encounter. The current ICD-10-CM codes identifying problems related to upbringing and parent-child conflict do not cover some of these other family situations. These types of circumstances often present unique situations that frequently contribute to the child being brought to seek medical attention.

The American Academy of Pediatrics (AAP) requests that the code set at Z62, Problems related to upbringing, be expanded to represent "family" dynamics and conflicts that can complicate an encounter.

**New to this proposal, the Academy is also requesting an expansion at Z02.8, Encounter for other administrative examinations, in order to show when a child is brought to medical attention by a welfare or law enforcement agency for examination unrelated to alleged physical or sexual abuse, but prior to placement outside of parental care (e.g., "medical clearance").**

### TABULAR MODIFICATIONS

Z02 Encounter for administrative examination

#### **Z02.8 Encounter for other administrative examinations**

**New code  
Add**

**Z02.84 Encounter for child welfare screening exam  
Excludes2: encounter for examination and observation  
for alleged child physical abuse (Z04.72)  
encounter for examination and observation  
for alleged child rape (Z04.42)**

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Z62 Problems related to upbringing  
Includes: current and past negative life events in childhood current and past problems of a child related to upbringing

Excludes2: maltreatment syndrome (T74.-)  
problems related to housing and economic circumstances (Z59.-)

Z62.2 Upbringing away from parents

Excludes1: problems with boarding school (Z59.3)

Z62.21 Child in welfare custody

Delete ~~Child in care of non-parental family member~~  
Child in foster care

Delete ~~Excludes2: problem for parent due to child in welfare custody (Z63.5)~~

Z62.22 Institutional upbringing

Child living in orphanage or group home

New code

Z62.23 Child in custody of non-parental guardian

Add Child in care of non-parental family member

Add Child in custody of grandparent

Add Child in kinship care

Z62.8 Other specified problems related to upbringing

Z62.82 **Parent-child conflict**

Add Code also, if applicable:

Add absence of family member (Z63.3-)

Add disappearance and death of family member (Z63.4)

Add disruption of family by separation and divorce  
(Z63.5)

Add other specified problems related to primary support  
group (Z63.8)

Add other stressful life events affecting family and  
household (Z63.7-)

Z62.820 Parent-biological child conflict

Parent-child problem NOS

Z62.821 Parent-adopted child conflict

Z62.822 Parent-foster child conflict

New code Z62.823 Parent-step child conflict



## **Progressive collapsing foot deformity: flexible and rigid**

The American Podiatric Medical Association (APMA) and American Society of Podiatric Surgeons (ASPS), request modifications to the ICD-10-CM for reporting of progressive collapsing foot deformity. Currently there is no ICD-10-CM code to represent progressive collapsing foot deformity.

Traditionally, when a patient experiences progressive flattening of the arch of the foot, it has been referred to as adult acquired flatfoot deformity. In some cases, this pathology may be referred to as pes planus. Since the establishment of these terms, the medical community has enjoyed the introduction of new imaging modalities, new research, more clinical experience, and new literature related this pathology, all of which, considered together, makes it clear that the terms “adult acquired flatfoot deformity” and “pes planus” do not adequately represent the different forms of the pathology they are intended to represent<sup>5</sup>. Furthermore, clinicians now know there are many different forms of this pathology and these now-antiquated terms do not provide adequate opportunity to identify pathology to its greatest specificity.

The term “progressive collapsing foot deformity” has been widely accepted, evidenced by the fact it already appears in 16 National Library of Medicine PubMed.gov<sup>4</sup> search results at the time of the writing of this request in May 2021, a startling number given the term was just introduced in October 2020. At the same time the consensus group advocated for the use of “progressive collapsing foot deformity”, it also proposed a new classification system which has been widely accepted. Differentiating between flexible and rigid progressive collapsing foot deformity is an important component of accurate documentation, epidemiology, and treatment planning, to name a few reasons.

The APMA and ASPS are requesting the following new ICD-10-CM codes.

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## **TABULAR MODIFICATIONS**

- M21 Other acquired deformities of limbs  
Excludes1: acquired absence of limb (Z89.-)

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congenital absence of limbs (Q71-Q73)  
congenital deformities and malformations of limbs (Q65-Q66,  
Q68-Q74)

Excludes2: acquired deformities of fingers or toes (M20.-)  
coxa plana (M91.2)

M21.4 Flat foot [pes planus] (acquired)

Excludes1: congenital pes planus (Q66.5-)

Add progressive collapsing foot deformity, flexible  
(M21.63-)

Add progressive collapsing foot deformity, rigid (M21.64-)

M21.40 Flat foot [pes planus] (acquired), unspecified foot

M21.41 Flat foot [pes planus] (acquired), right foot

M21.42 Flat foot [pes planus] (acquired), left foot

M21.6 Other acquired deformities of foot

Excludes2: deformities of toe (acquired) (M20.1-M20.6-)

New  
subcategory

M21.63 Progressive collapsing foot deformity, flexible

Add Excludes 1: congenital pes planus (Q66.5-)  
flat foot [pes planus] (acquired) (M21.4-)

Add M21.631 Progressive collapsing foot deformity, flexible,  
right foot

New code M21.632 Progressive collapsing foot deformity, flexible,  
left foot

New code M21.639 Progressive collapsing foot deformity, flexible,  
unspecified foot

New  
subcategory

M21.64 Progressive collapsing foot deformity, rigid

Add Excludes 1: congenital pes planus (Q66.5-)  
flat foot [pes planus] (acquired) (M21.4-)

Add M21.641 Progressive collapsing foot deformity, rigid,  
right foot

New code M21.642 Progressive collapsing foot deformity, rigid, left  
foot

New code M21.649 Progressive collapsing foot deformity, rigid,  
unspecified foot

## **PTEN Hamartoma Tumor Syndrome (PHTS)**

PHTS is a rare syndrome caused by germline heterozygous loss-of-function mutation in the Phosphatase and Tensin Homolog (PTEN) tumor suppressor gene. Most PTEN mutations are inherited in a family for generations, following an autosomal dominant pattern, but 10-45% of cases are due to new (de novo) mutations (Mester & Eng, 2012). Estimating the prevalence of PHTS is complex due to the varied presentations and diagnoses patients can have and because some features (e.g. benign breast lesions) also commonly occur in the general population. However, it is estimated that about 1 in 200,000 individuals could have PHTS (corresponding to around 2000 individuals in the US population, and around 47,000 individuals worldwide). PHTS has a range of clinical manifestations including benign hamartomas, macrocephaly, neurocognitive deficits (including autism spectrum disorder and cognitive impairment), and an increased risk of malignancy (particularly breast, thyroid, and endometrial cancer) (Ngeow & Eng, 2015). Clinical symptoms and signs of PHTS vary in incidence and severity. The most common clinical features are macrocephaly (>90% of patients) and increased lifetime cancer risk (>85% of patients).

Before the identification of the PTEN gene and routine genetic testing of patients with rare congenital conditions, several syndromes were described based on clinical features. These included Cowden syndrome (CS), Bannayan-Riley-Ruvalcaba syndrome (BRRS) and Proteus-like syndrome (PLS). PTEN germline mutations have been identified in only a proportion of patients diagnosed with CS (25-85%) or BRRS (60%), and mutations in other (often related) genes have been identified in some of these cases (Yehia et al, 2019).

The diverse manifestations of PHTS and marked differences in severity mean that patients present for the first time and are treated by a range of clinicians, including pediatric and adult neurologists, vascular surgeons, interventional radiologists, dermatologists and oncologists. Current treatment for many manifestations of PHTS is supportive, but guidelines recommend a comprehensive program of surveillance and early intervention for cancer in all patients with PHTS (Daly et al, 2020; Tischkowitz et al, 2020). Failure to recognize that a patient has PHTS may result in a missed opportunity to initiate surveillance and potentially, prophylactic surgery for cancer, which is the mainstay of cancer prevention in these patients.

The requestors are proposing the creation of a new ICD-10-CM code to specifically identify patients with PTEN Hamartoma Tumor Syndrome (PHTS). Currently, there is no unique ICD-10-CM code to identify these patients and no existing ICD-10-CM code is specific enough to capture the multi-systemic effects of this rare genetic disorder.

A unique ICD-10-CM code will also improve clinical care by making it possible to track outcomes from clinical interventions, and by facilitating the development of standard care protocols to improve consistency between different specialists. Finally, an ICD-10-CM code will improve understanding of the severity and risk of the different manifestations of PHTS, and the impact of different interventions on patient well-being and survival.

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This proposal is based on a request from PTEN Research and is supported by CDC’s Office of Genomics and Precision Public Health, and Division of Cancer Prevention and Control, and American College of Medical Genetics Genomics (ACMG).

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**TABULAR MODIFICATIONS**

	Q85	Phakomatoses, not elsewhere classified
		Q85.8 Other phakomatoses, not elsewhere classified
Delete		<del>Peutz-Jeghers Syndrome</del>
Delete		<del>Sturge-Weber(-Dimitri) syndrome</del>
Delete		<del>von Hippel-Lindau syndrome</del>
New code	Q85.81	PTEN hamartoma tumor syndrome
Add		PHTS
Add		PTEN related Cowden syndrome
Add		Code also, if applicable, genetic susceptibility to malignant neoplasm Z15.0-
New code	Q85.82	Other Cowden syndrome
New code	Q85.89	Other PTEN hamartoma tumor syndrome
Add		Peutz-Jeghers Syndrome
Add		Sturge-Weber(-Dimitri) syndrome

Note: von Hippel-Lindau syndrome will be addressed in a separate proposal.

## **Rib fracture due to cardiopulmonary resuscitation**

For rib fracture due to cardiopulmonary resuscitation (CPR), ICD-10-CM/PCS Coding Clinic, First Quarter ICD-10 2021, pages: 5-6, Effective with discharges: March 10, 2021, instructs coders to assign code M96.89 “Other intraoperative and postprocedural complications and disorders of the musculoskeletal system”, along with external cause of injury code Y84.8, “Other medical procedures as the cause of abnormal reaction of the patient, or of later complication, without mention of misadventure at the time of the procedure”.

This Coding Clinic guidance is consistent with previous guidance to avoid assigning traumatic injury codes for injuries that occur during, or as a result of, a medical intervention (even when those injuries are a result of mechanical trauma). However, it has previously been common practice for coders to assign traumatic injury codes to all fractures that occur in the course of medical care.

Rib fractures resulting from the performance of CPR are a known risk, especially among elderly individuals and those with osteoporosis. Reports on conventional CPR in adults suggest an incidence of rib fractures ranging from 13 to 97%, and of sternal fractures from 1 to 43%. In fact, rib fractures may be an unavoidable consequence of providing sufficient external pressure to support perfusion of the brain and other vital organs, and detection of these fractures may be incomplete.

AHRQ’s PSI 06, “Iatrogenic Pneumothorax Rate”, excludes diagnoses that could reasonably be expected to involve entering into the pleural space. Cases involving rib fracture due to performance of CPR should be excluded from PSI 06, because iatrogenic pneumothorax would be an expected outcome in this clinical setting. This exclusion has been historically accomplished using S codes, but this approach is no longer feasible in light of the recent Coding Clinic guidance.

Many other conditions are also assigned to code M96.89 through the Diagnosis Index; for example:

- Complication(s) (from) (of)
  - intraoperative (intraprocedural)
    - specified NEC
    - musculoskeletal structure M96.89
- Unstable
  - joint - see Instability, joint
    - secondary to removal of joint prosthesis M96.89

AHRQ is requesting new codes to specifically identify thoracic fractures due to performance of CPR or chest compressions.

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**TABULAR MODIFICATIONS**

	M96	Intraoperative and postprocedural complications and disorders of musculoskeletal system, not elsewhere classified
New subcategory		M96.A Fracture of ribs, sternum and thorax associated with compression of the chest and cardiopulmonary resuscitation
New code	M96.A1	Fracture of sternum associated with chest compression and cardiopulmonary resuscitation
New code	M96.A2	Fracture of one rib associated with chest compression and cardiopulmonary resuscitation
New code	M96.A3	Multiple fractures of ribs associated with chest compression and cardiopulmonary resuscitation
New code	M96.A4	Flail chest associated with chest compression and cardiopulmonary resuscitation
New code	M96.A9	Other fracture associated with chest compression and cardiopulmonary resuscitation

## 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 and March 2021 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 statural growth. In puberty sex steroid hormones facilitate the pubertal growth spurt.<sup>1</sup>

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.<sup>2</sup> Of those, approximately 5%, or 1:1,000 children have short stature due to endocrine disorders.<sup>3</sup>

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.<sup>4</sup> Growth hormone binds to GH receptors (GHR), mainly on cells in the liver, although most tissues contain GHRs.<sup>5</sup> 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.<sup>4</sup>

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.<sup>6</sup>

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.<sup>7</sup> Its prevalence is estimated to be between 1:4,000 to 1:10,000.<sup>4</sup> 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

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secretion.<sup>8</sup> Some studies suggest imaging the hypothalamic-pituitary region via MRI may be more helpful in diagnosing growth hormone deficiency than laboratory assays.<sup>9</sup> GHD is treated with recombinant human GH (rhGH), also known as somatotropin.<sup>7</sup>

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).<sup>8,10,11</sup> 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.<sup>11</sup> 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.<sup>3</sup> 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.<sup>8</sup>

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

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.<sup>8</sup> 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

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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**

Add	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) short stature due to endocrine disorder (E34.3-)
New subcategory Delete Delete	E34	Other endocrine disorders Excludes1: pseudohypoparathyroidism (E20.1) E34.3 Short stature due to endocrine disorder <del>Constitutional short stature</del> <del>Laron type short stature</del>
Add		Excludes1: short stature (child) (R62.52)
New Code		E34.30 Short stature due to endocrine disorder, unspecified
New code Add		E34.31 Constitutional short stature Constitutional delay of growth, puberty, or maturation
New subcategory New code		<b>E34.32 Genetic causes of short stature</b>
Add		<b>E34.321 Primary insulin-like growth factor-1 (IGF-1) deficiency</b> <b>Acid-labile subunit gene (<i>IGFALS</i>) defect</b>
Add		<b>Growth hormone gene 1 (<i>GH1</i>) defect with growth hormone neutralizing antibodies</b>
Add		<b>Growth hormone insensitivity syndrome (GHIS)</b>
Add		<b>Insulin-like growth factor 1 gene (<i>IGF1</i>) defect</b>
Add Add		<b>Laron type short stature</b> <b>Severe primary insulin-like growth factor-1 deficiency (SPIGFD)</b>
Add		<b>Signal transducer and activator of transcription 5B gene (<i>STAT5b</i>) defect</b>
New code		<b>E34.322 Insulin-like growth factor-1 (IGF-1) resistance</b>

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<b>Add</b>	<b>Genetic syndrome with resistance to insulin-like growth factor-1</b>
<b>Add</b>	<b>Insulin-like growth factor-1 receptor (<i>IGF-1R</i>) defect</b>
<b>Add</b>	<b>Post-insulin-like growth factor-1 receptor signaling defect</b>
<b>New code</b>	<b>E34.328 Other genetic causes of short stature</b>
<b>New code</b>	<b>E34.329 Unspecified genetic causes of short stature</b>
<b>New code</b>	<b>E34.39 Other short stature due to endocrine disorder</b>

## Social Determinants of Health

This proposal was originally submitted by the Gravity Project and presented at the March 2021 ICD10 Coordination and Maintenance meeting. Parts of the proposal were previously approved and will be implemented on October 1, 2021. Subsequently, we have received additional code requests and a proposal from the American Academy of Pediatrics (AAP). New codes and revisions have been made (**noted in bold**) and resubmitted for reconsideration.

Over the last decades growing literature has clarified and further identified the social determinants of health 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.

The domain of transportation security represents both health risks and management complexities as health care systems consider transportation barriers. The Gravity Project proposes an ICD-10-CM code for transportation insecurity.

Health insurance insecurity is at an all-time high. Health care insurance is becoming cost prohibitive to all financial status of persons. Health care for all is not just important to achieve, but imperative. Lack of health insurance significantly impacts people seeking care.

Financial Insecurity and Material Hardship- Currently ICD-10-CM contains terminology for low income and poverty. However, the health risks driven by limited financial resources are not limited to low income or impoverished individuals.

Financial Insecurity (“A subjective evaluation of one's current financial situation that includes perceived inadequacy of financial resources and financial concerns or worries, including expectations regarding one's future economic situation.”). The Material Hardship (“unable to obtain basic needs”) can be considered broad terms that identify all economic driven social risk. A clear outcome of

Gravity’s work was the understanding that there is a need for a general concept for financial insecurity and material hardship as the health risks and management needs of each are clear in the peer-reviewed literature and excluded from the individual domains such as food insecurity, housing instability, or transportation insecurity. Furthermore, it is critical to define risk beyond low income and poverty thresholds. These two concepts also streamline the granular recommendations of the UHC/AMA submission.

Socioeconomic Risk Counseling- the need for a specific counseling code to represent the effort of assessing and patient centered goal setting required to address socioeconomic risks.

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The American Academy of Pediatrics (AAP) is proposing codes to identify when noncompliance is due to the primary caregiver and not the patient. Under the current code category Z91.1, Patient's personal history of noncompliance with medical treatment and regimen, non-compliance is currently assumed to be due to the patient's action or lack thereof. Unfortunately, the code also appears to place the responsibility of non-compliance on those who may not have any direct control, because someone else cares for them, including children, the elderly and the disabled. A unique set of codes is needed to better track these circumstances.

The existing code Z91.82, Personal history of military deployment, does not adequately reflect veteran service. The proposed revision will capture the data element of service.

### TABULAR MODIFICATIONS

Z59 Problems related to housing and economic circumstances

Z59.8 Other problems related to housing and economic circumstances

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	<b>Z59.83 Health insurance insecurity</b>
Add	<b>High deductible health care</b>
Add	<b>Lack of health insurance</b>
Add	<b>Medical cost burden</b>
Add	<b>Excludes2: caregiver's intentional underdosing of patient's medication regimen due to financial hardship (Z91.A20) patient's intentional underdosing of medication regimen due to financial hardship (Z91.120)</b>

New code	Z59.86 Financial insecurity
Add	Bankruptcy
Add	Burdensome debt
Add	Economic strain
Add	Financial strain
Add	Money problems
Add	Running out of money
Add	Unable to make ends meet

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**Add** **Excludes2: extreme poverty (Z59.5)**  
**Add** **low income (Z59.6)**  
**Add** **material hardship, not elsewhere classified**  
**(Z59.87)**

New code Z59.87 Material hardship  
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: extreme poverty (Z59.5)**  
**Add financial insecurity, not elsewhere classified**  
**(Z59.86)**  
**Add low income (Z59.6)**

Z91 Personal risk factors, not elsewhere classified  
Excludes2: contact with and (suspected) exposures hazardous to health  
(Z77.-)  
exposure to pollution and other problems related to physical  
environment (Z77.1-)  
female genital mutilation status (N90.81-)  
**Delete ~~personal history of physical injury and trauma (Z87.81,~~**  
**~~Z87.82-)~~**  
occupational exposure to risk factors (Z57.-)  
**Add personal history of physical injury and trauma (Z87.81,**  
**Z87.82-)**

**Add Z91.1 Patient's noncompliance with medical treatment and regimen**  
**Excludes 2: caregiver noncompliance with patient's medical**  
**treatment and regimen (Z91.A-)**

New sub Z91.11 Patient's noncompliance with dietary regimen  
subcategory  
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  
**Add Inability to comply with dietary regimen**  
**Add Code also, if applicable, lack of adequate food**  
**(Z59.41)**

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New code	Z91.119 Patient's noncompliance with dietary regimen due to unspecified reason
New sub category	Z91.19 Patient's noncompliance with other medical treatment and regimen
Add	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
New subcategory	<b>Z91.A Caregiver's noncompliance with patient's medical treatment and regimen</b>
New code	<b>Z91.A1 Caregiver's noncompliance with patient's dietary regimen</b>
Add	<b>Caregiver's inability to comply with patient's dietary regimen</b>
New code	<b>Z91.A10 Caregiver's noncompliance with patient's dietary regimen due to financial hardship</b>
New code	<b>Z91.A18 Caregiver's noncompliance with patient's dietary regimen for other reason</b>
Add	<b>Code also, if applicable, lack of adequate food (Z59.41)</b>
New sub category	<b>Z91.A2 Caregiver's intentional underdosing of patient's medication regimen</b>
Add	<b>Code first underdosing of medication (T36-T50) with fifth or sixth character 6</b>
Add	<b>Excludes1: adverse effect of prescribed drug taken as directed-code to adverse effect poisoning (overdose) -code to poisoning</b>
New code	<b>Z91.A20 Caregiver's intentional underdosing of patient's medication regimen due to financial hardship</b>

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New code	<b>Z91.A28 Caregiver's intentional underdosing of medication regimen for other reason</b>
New code	<b>Z91.A3 Caregiver's unintentional underdosing of patient's medication regimen</b>
Add	<b>Code first underdosing of medication (T36-T50) with fifth or sixth character 6</b>
Add	<b>Excludes1: adverse effect of prescribed drug taken as directed-code to adverse effect poisoning (overdose)-code to poisoning</b>
New code	<b>Z91.A4 Caregiver's other noncompliance patient's with medication regimen</b>
Add	<b>Caregiver's underdosing of patient's medication NOS</b>
New code	<b>Z91.A5 Caregiver's noncompliance with patient's renal dialysis</b>
New code	<b>Z91.A9 Caregiver's noncompliance with patient's other medical treatment and regimen</b>
Add	<b>Nonadherence to medical treatment</b>
	Z91.8 Other specified personal risk factors, not elsewhere classified
Revise	<b>Z91.82 Personal history of military <u>service and deployment</u></b>
Revise	Individual (civilian or military) with past history of military war, peacekeeping and humanitarian deployment <b>(<del>current or past conflict</del>)</b> Returned from military deployment

## **Transfusion-Associated Dyspnea (TAD)**

Transfusion-Associated Dyspnea (TAD) is defined by CDC's National Hemovigilance Network as 'Acute respiratory distress occurring within 24 hours of cessation of transfusion and allergic reaction, TACO, and TRALI are not applicable' <sup>(1)</sup>. Similarly, according to the 2019 SHOT report <sup>(2)</sup>, TAD is 'a pulmonary complication post transfusion that cannot be classified as TACO or TRALI, nor can it be ascribed to patient's pre-existing diseases.' In a study of transfusion-related adverse events by CDC's National Healthcare Safety Network (NHSN) using reporting facilities <sup>(3)</sup>, transfusion-related pulmonary complications (TACO, TRALI, and TAD) accounted for 35% of serious reactions and 65% of fatalities. Of all reported reactions (N=23,083), 2% (N=456) had TAD, with 74.3% satisfying case definition and 22% being serious reactions with one fatality. <sup>(3)</sup> The literature <sup>(1-5)</sup> suggests that TAD may be serious transfusion related pulmonary complication and needs population-based national monitoring, including understanding potential risk factors in elderly and other populations, and further characterization to better understand pathophysiology of cases reported under TAD. TAD occurred with RBCs, platelets, and plasma transfusions, and has also been reported as an adverse event of COVID-19 Convalescent Plasma. <sup>(2, 3, 4, 6)</sup>

This is a request for a new ICD-10-CM code for Transfusion-Associated Dyspnea (TAD) that distinguishes this transfusion-related adverse event from Transfusion associated circulatory overload (TACO) (E87.71) and Transfusion-related acute lung injury (TRALI) (J95.84) based on the established definition by CDC's National Healthcare Safety Network <sup>(1)</sup>. Currently, there is no specific coding for TAD. Therefore, it has been proposed to introduce a new condition-specific code to result in the following coding:

In summary, the introduction of a condition-specific code for TAD will improve coding accuracy, increase provider awareness of this potentially serious transfusion related adverse event, allow better understanding of its pathophysiology, enhance monitoring of its occurrence and understanding of risk factors, and therefore will help in development and evaluation of appropriate prevention and treatment strategies.

This proposal was submitted by the Office of Biostatistics and Epidemiology, Center for Biologics Evaluation and Research, U. S. Food & Drug Administration.

This proposal is supported by Centers of Disease Control and Prevention, Division of Developmental and Intellectual Disabilities, National Center for Emerging and Zoonotic Infectious Diseases, Division of Healthcare Quality Promotion CDC/DDID/NCEZID/DHQ.

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### TABULAR MODIFICATIONS

- J95 Intraoperative and postprocedural complications and disorders of respiratory system, not elsewhere classified
- Excludes2: aspiration pneumonia (J69.-)  
emphysema (subcutaneous) resulting from a procedure (T81.82)  
hypostatic pneumonia (J18.2)  
pulmonary manifestations due to radiation (J70.0-J70.1).
- J95.8 Other intraoperative and postprocedural complications and disorders of respiratory system, not elsewhere classified

New code                      J95.87      Transfusion-associated dyspnea (TAD)

## **Underimmunization for COVID-19**

During the current time of the COVID-19 pandemic, immunizations have provided protection for many people, but there is interest in being able to track people who are not immunized or only partially immunized. At the current time, this is a significant modifiable risk factor for morbidity and mortality, and of interest for clinical reasons, as well as of value for public health.

NCHS is proposing creation of codes for unvaccinated for COVID-19, and for partially vaccinated for COVID-19.

**Early implementation is proposed on April 1, 2022, and comments are requested by October 15, 2021.**

### **TABULAR MODIFICATIONS**

	Z28	Immunization not carried out and underimmunization status
	Z28.3	Underimmunization status
New sub-subcategory	Z28.31	Underimmunization for COVID-19 status
New code	Z28.310	Unvaccinated for COVID-19
New code	Z28.311	Partially vaccinated for COVID-19
New code	Z28.39	Other underimmunization status

## **von Hippel-Lindau Disease**

von Hippel-Lindau (VHL) disease is a rare genetic disorder caused by mutation, deletion or hypermethylation in the *VHL* gene, a tumor-suppressor gene located on chromosome 3p25-26.<sup>1</sup> VHL disease can be inherited in an autosomal dominant manner; however, *VHL* mutations can also arise *de novo*. Published research estimates that approximately 80% of VHL-associated renal cell carcinoma (RCC) cases are linked to family history of VHL disease, and approximately 20% of cases are linked to *de novo* mutations in the *VHL* gene.<sup>2,3</sup>

Patients affected by VHL disease are at increased risk of developing various benign and malignant tumors and/or cysts in multiple organs of the body throughout their life.<sup>4,5,6</sup> The manifestations of VHL disease include: <sup>4,5,6</sup> renal cysts, clear cell RCC (ccRCC), pheochromocytomas, retinal hemangioblastomas, central nervous system (CNS) (spinal and cerebellar) hemangioblastomas (HB), liver hemangiomas, pancreatic cysts, pancreatic microcystic serous adenomas, pancreatic neuroendocrine tumors, epididymal and broad ligament cystadenomas, and endolymphatic sac tumors.

The incidence of VHL disease in the US is thought to be about one in 36,000 births.<sup>7,8</sup> The prevalence of VHL disease in the US ranges from approximately one in every 30,000 people to one in every 50,000 people, with up to 10,000 people estimated to be living with the disease.<sup>7,8</sup> VHL disease has a high penetrance rate (approximately 90% by the age of 65 years), leading to early-onset of the disease.<sup>6,9</sup> The majority of patients (90%) with VHL disease develop clinical symptoms before the age of 65,<sup>10</sup> with symptom onset most commonly occurring between the ages of 18 to 30.<sup>9</sup>

RCC is the most frequent visceral manifestation and most frequent type of malignant tumor in VHL disease.<sup>6</sup> In the US, the reported frequency of RCC in VHL disease is estimated at 42-57.5%.<sup>11,12</sup> Relative to sporadic RCC which has a mean age of onset of 63 years, the mean age of onset for VHL-associated RCC is 39 years (range: 31 years to 62 years).<sup>5,12,13</sup> Similarly, patients with VHL-associated RCC can be asymptomatic for years before presenting varying symptoms.

VHL disease is associated with a high humanistic burden, as it has a negative impact on a patients' physical and psychosocial well-being, affecting their overall quality of life.<sup>14,15,16,17</sup> In addition to poor health and physical impairment, patients with VHL disease tend to experience emotional distress associated with the wide number of possible manifestations,<sup>15,17</sup> the variable age of onset (from early childhood into adulthood),<sup>15,17</sup> and the 'watch and wait' approach of active surveillance, which is restricted to periodic ultrasounds or scans to check for growth of the existing tumors or occurrence of new tumors.<sup>4</sup>

Moreover, anxiety associated with prolonged screening and observation in patients with VHL disease can negatively impact a patient's adherence to the surveillance program.<sup>18</sup> VHL disease has a negative psychosocial impact not only on VHL patients themselves, but also on their caregivers and families, highlighting the impact of the disease on society.<sup>15</sup> Caregivers of VHL

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patients report considerable physical and psychosocial challenges, such as: little reprieve from caregiving responsibilities, difficulties balancing the needs of VHL-affected and other unaffected family members, fears about the future, and guilt regarding the onset of tumors in their children.<sup>16</sup>

Early detection and treatment of VHL disease is important to patient care.<sup>8</sup> The VHL Alliance (VHLA) is the preeminent resource and clearinghouse for those affected by von Hippel-Lindau disease, including patients, caregivers, researchers, and the medical community. The VHLA Guidelines recommend specific testing of patients who are at risk of VHL disease who do not yet have symptoms, and patients who do not have VHL disease symptoms in a particular area.<sup>19</sup> Furthermore, VHL Alliance Guidelines recommend active surveillance to prevent severe VHL complications in patients with VHL-associated RCC, or patients with RCC and at risk of VHL disease, except those who have had a DNA test and do not carry the altered *VHL* gene.

The National Comprehensive Cancer Network (NCCN) guidelines for Kidney Cancer (Version 2.2021) includes guidance on Diagnosis, Screening and Surveillance for VHL-Disease.<sup>20</sup> Genetic testing is recommended in individuals with a close blood relative (first-degree or second-degree) with a known *VHL* pathogenic/likely pathogenic variant. Furthermore, risk assessment and counseling are recommended in individuals with RCC who present syndrome features or clinical manifestations of VHL disease.<sup>21</sup> If positive for *VHL* pathogenic or likely pathogenic variant, the guidelines recommend radiological assessment of kidneys, pancreas, and adrenals every two years starting at age 15. In patients without known familial pathogenic/likely pathogenic variant, the recommendation is to consider testing with kidney cancer multi-gene panel or clinically directed single-gene testing.<sup>20</sup>

Regarding treatment for malignancies associated with VHL, on August 13, 2021, the Food and Drug Administration approved belzutifan, which is a hypoxia-inducible factor inhibitor, for adult patients with VHL disease who require therapy for associated renal cell carcinoma (RCC), central nervous system (CNS) hemangioblastomas, or pancreatic neuroendocrine tumors (pNET), not requiring immediate surgery.<sup>22</sup>

Currently, ICD-10-CM classifies von Hippel-Lindau (VHL) disease under Other Phakomatoses, along with a number of other syndromes. These conditions represent a group of largely rare, neurocutaneous disorders characterized by the involvement of structures that arise from the embryonic ectoderm, such as central nervous system, skin, eyes and other organs.<sup>23</sup> However, this grouping does not represent distinct genetic causes currently recognized as underlying various different disorders, nor does it differentiate for the range of different phenotypic features among these conditions.

A specific ICD-10-CM code for VHL disease will help to differentiate it from the other syndromes. Also, with a treatment being newly available, an ICD-10-CM code will aid in identifying patients and tracking, which is anticipated to improve disease reporting and identification of patients in routine clinical practice, and also to enhance patient experience and

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clinical outcomes, and adherence to practice guidelines. In addition, the ICD-10-CM code specific for VHL disease will facilitate epidemiologic and health services research, with potential to improve understanding of disease natural history, disease burden, care and management patterns, and unmet needs for this rare disease and its many manifestations. More information in these areas will help raise awareness for this disease, potentially contributing to development of new diagnostic and treatment options in future. It is anticipated this will work to enable access to more timely and appropriate care, including surgery, imaging, and novel therapeutics. In summary, given the early onset and high burden of VHL disease, an improved understanding of the disease and its management is needed to help clinicians, researchers, policymakers and others provide the best possible care for these patients.

This proposal is based on a request for a new code for VHL disease, received from Eric Jonasch, MD, Professor of Medicine at the MD Anderson Cancer Center, University of Texas. The VHL Alliance reviewed the original proposal and extended its support for the request.

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### TABULAR MODIFICATIONS

Note that a different proposal is anticipated to also affect expansion of Q85.8.

	Q85	Phakomatoses, not elsewhere classified
	Q85.8	Other phakomatoses, not elsewhere classified
Delete		<del>Peutz-Jeghers Syndrome</del>
Delete		<del>Sturge-Weber(-Dimitri) syndrome</del>
Delete		<del>von Hippel-Lindau syndrome</del>
New code	Q85.83	von Hippel-Lindau syndrome
New code	Q85.89	Other phakomatoses, not elsewhere classified
Add		Peutz-Jeghers Syndrome
Add		Sturge-Weber(-Dimitri) syndrome

## WHIM Syndrome

WHIM syndrome is a rare autosomal-dominant inherited combined primary immunodeficiency syndrome caused by pathogenic gain-of-function variants in the chemokine type 4 receptor gene (CXCR4).<sup>1</sup> This proposal for a specific ICD-10-CM code for WHIM syndrome is based on a request by X4 Pharmaceuticals, Inc. It is supported by the American Academy of Allergy, Asthma, and Immunology (AAAAI), the Clinical Immunology Society (CIS) and the Jeffrey Modell Foundation (JMF).

The acronym “WHIM” stands for four main clinical manifestations of the syndrome: Warts, due to human papillomavirus (HPV) infections, variable Hypogammaglobinemia, Infections, and Myelokathexis, the abnormal retention of mature neutrophils in the bone marrow.<sup>2</sup> Based on the current International Union of Immunological Societies (IUIS) criteria, WHIM syndrome is listed among innate immune defects linked to HPV infection.<sup>3</sup> In addition to severe neutropenia, many WHIM patients have additional immune defects including lymphopenia with variable severity of T and B cells dysfunction; therefore, WHIM Syndrome can be considered to be a variant of combined immunodeficiency.<sup>4</sup> The Food and Drug Administration (FDA) has recognized WHIM syndrome as a “severely debilitating or life-threatening hematologic disorder.”<sup>5</sup>

WHIM may manifest in early childhood with incomplete penetrance and is characteristically marked by frequent or severe bacterial infections before the appearance of warts; later in life, life-threatening complications become more common. Recurrent pneumonia, bronchiectasis and its complications, hearing loss from recurrent otitis media, disfiguring and disabling warts and HPV-associated malignancies --notably surgically-treated squamous cell carcinoma and precancerous genital lesions-- are WHIM’s most serious complications.<sup>6,7</sup> Case studies suggest that WHIM patients are unable to mount a sufficient immune response to the HPV vaccine, leading to the need for focused monitoring for HPV.<sup>8</sup>

Clinical presentation of the disease is quite heterogeneous as most patients will not present with all the symptoms associated with the syndrome. WHIM is often managed by specialists who focus on specific symptoms or complications, rather than addressing the underlying *pathomechanism* resulting from disease-causing gain-of-function CXCR4 variants. The result of all of these factors is that diagnosis of WHIM may be delayed or missed and, consequently, underdiagnosed.<sup>5</sup>

The prognosis for WHIM depends in part on early recognition of the disorder and subsequent aggressive therapeutic intervention for any suspected infection. Current clinical management of WHIM includes treatment with G-CSF, IVIG, prophylactic antibiotics, and aggressive surveillance for and surgical extirpation of dysplastic skin and mucosal HPV related lesions.<sup>9</sup> Patients with extensive bronchiectasis, if diagnosed with WHIM, could be referred for close management by a pulmonary specialist, because many existing specialized interventions may serve to benefit these patients.<sup>6</sup>

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A specific code for WHIM syndrome is proposed at D81.83, in subcategory D81.8, Other combined immunodeficiencies. Having a specific ICD-10-CM code for WHIM Syndrome is anticipated to enable improved tracking, and following the multisystemic effects of the condition, along with early recognition of the patients who are asymptomatic in early childhood but may progress to severe disease with age. It will assist with implementing options for familial genetic counseling, along with coordinated patient monitoring and heightened HPV-associated cancer surveillance, as well as monitoring for common clinical conditions, such as neutropenia and hypogammaglobulinemia that can be treated if symptomatic. It will also enable targeted treatment protocols including current and new pharmacologic agents, and lead to a better understanding of the natural history of the disease. It is also anticipated that the promise of a new ICD-10-CM code will lead to enhanced disease awareness, more frequent diagnosis, and preventive interventions and aggressive medical management to avoid the numerous, serious complications of this syndrome, hopefully improving quality of life for these patients.

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**TABULAR MODIFICATIONS**

Note that a different proposal is anticipated to also affect expansion of D81.8.

D81 Combined immunodeficiencies

D81.8 Other combined immunodeficiencies

New code	D81.83	WHIM Syndrome
Add		Immunodeficiency with neutropenia and HPV susceptibility

**ICD-10-CM TABULAR OF DISEASES - PROPOSED ADDENDA**  
**All proposed effective October 1, 2022**

Revise	C61	Malignant neoplasm of prostate Use additional code, <u>if appropriate</u> , to identify: hormone sensitivity status (Z19.1-Z19.2) rising PSA following treatment for malignant neoplasm of prostate (R97.21)
Revise	C84	Mature T/NK-cell lymphomas Peripheral T-cell lymphomas Excludes1: personal history of non-Hodgkin lymphoma (Z85.72) C84.0 Mycosis fungoides Excludes1: peripheral T-cell lymphoma, not <u>elsewhere</u> classified (C84.4-)
Revise	C84.4	Peripheral T-cell lymphoma, not <u>elsewhere</u> classified Mature T-cell lymphoma, not elsewhere classified
Revise	C84.40	Peripheral T-cell lymphoma, not <u>elsewhere</u> classified, unspecified site
Revise	C84.41	Peripheral T-cell lymphoma, not <u>elsewhere</u> classified, lymph nodes of head, face, and neck
Revise	C84.42	Peripheral T-cell lymphoma, not <u>elsewhere</u> classified, intrathoracic lymph nodes
Revise	C84.43	Peripheral T-cell lymphoma, not <u>elsewhere</u> classified, intra-abdominal lymph nodes
Revise	C84.44	Peripheral T-cell lymphoma, not <u>elsewhere</u> classified, lymph nodes of axilla and upper limb
Revise	C84.45	Peripheral T-cell lymphoma, not <u>elsewhere</u> classified, lymph nodes of inguinal region and lower limb
Revise	C84.46	Peripheral T-cell lymphoma, not <u>elsewhere</u> classified, intrapelvic lymph nodes
Revise	C84.47	Peripheral T-cell lymphoma, not <u>elsewhere</u> classified, spleen
Revise	C84.48	Peripheral T-cell lymphoma, not <u>elsewhere</u> classified, lymph nodes of multiple sites
Revise	C84.49	Peripheral T-cell lymphoma, not <u>elsewhere</u> classified, extranodal and solid organ sites

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- C94 Other leukemias of specified cell type  
Revise C94.6 Myelodysplastic and myeloproliferative disease,  
not elsewhere classified  
Revise Myelodysplastic disease, not elsewhere classified  
Add Myelodysplastic/myeloproliferative neoplasm, unclassifiable

**Disorders of muscles (M60-M63)**

- Delete Excludes1: dermatopolymyositis (M33.-)  
~~muscular dystrophies and myopathies (G71-G72)~~  
Add Excludes2: muscular dystrophies and myopathies (G71-G72)

- F07 Personality and behavioral disorders due to known physiological condition  
Code first the underlying physiological condition  
F07.0 Personality change due to known physiological condition  
Delete ~~Code first underlying physiological condition~~

- F31 Bipolar disorder  
Includes: bipolar I disorder  
bipolar type I disorder  
manic-depressive illness  
manic-depressive psychosis  
manic-depressive reaction  
Add seasonal bipolar disorder

- F33 Major depressive disorder, recurrent  
Includes: recurrent episodes of depressive reaction  
recurrent episodes of endogenous depression  
recurrent episodes of major depression  
recurrent episodes of psychogenic depression  
Add recurrent episodes of seasonal affective disorder  
recurrent episodes of reactive depression  
recurrent episodes of seasonal depressive disorder  
recurrent episodes of vital depression

- F84 Pervasive developmental disorders  
Revise ~~Use additional code~~ Code also to identify any associated  
medical condition and intellectual disabilities.

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- G96 Other disorders of central nervous system  
G96.0 Cerebrospinal fluid leak  
G96.00 Cerebrospinal fluid leak, unspecified  
Code also if applicable:  
head injury (S00.- to S09.-)
- Revise
- I69 Sequelae of cerebrovascular disease  
I69.0 Sequelae of nontraumatic subarachnoid hemorrhage  
I69.09 Other sequelae of nontraumatic subarachnoid hemorrhage  
I69.091 Dysphagia following nontraumatic subarachnoid hemorrhage  
Use additional code to identify the type of dysphagia, if known (~~R13.1~~) (R13.11-R13.19)
- Revise
- I69.1 Sequelae of nontraumatic intracerebral hemorrhage  
I69.19 Other sequelae of nontraumatic intracerebral hemorrhage  
I69.191 Dysphagia following nontraumatic intracerebral hemorrhage  
Use additional code to identify the type of dysphagia, if known (~~R13.1~~) (R13.11-R13.19)
- Revise
- I69.2 Sequelae of other nontraumatic intracranial hemorrhage  
I69.29 Other sequelae of other nontraumatic intracranial hemorrhage  
I69.291 Dysphagia following other nontraumatic intracranial hemorrhage  
Use additional code to identify the type of dysphagia, if known (~~R13.1~~) (R13.11-R13.19)
- Revise
- I69.8 Sequelae of other cerebrovascular diseases  
I69.89 Other sequelae of other cerebrovascular disease  
I69.891 Dysphagia following other cerebrovascular disease  
Use additional code to identify the type of dysphagia, if known (~~R13.1~~) (R13.11-R13.19)
- Revise
- I69.9 Sequelae of unspecified cerebrovascular diseases  
I69.99 Other sequelae of unspecified cerebrovascular disease  
I69.991 Dysphagia following unspecified cerebrovascular disease  
Use additional code to identify the type of dysphagia, if known (~~R13.1~~) (R13.11-R13.19)
- Revise

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- I70 Atherosclerosis  
I70.3 Atherosclerosis of unspecified type of bypass graft(s)  
of the extremities  
I70.32 Atherosclerosis of unspecified type of bypass graft(s) of  
the extremities with rest pain  
Includes: any condition classifiable to I70.31-  
chronic limb-threatening ischemia of unspecified  
type of bypass graft(s) of the extremities  
with rest pain, ~~right leg~~
- Revise
- I77 Other disorders of arteries and arterioles  
I77.8 Other specified disorders of arteries and arterioles  
I77.81 Aortic ectasia  
Ectasis aorta  
Excludes1: aortic aneurysm and dissection (I71.0- - I71.9)
- Revise
- J44 Other chronic obstructive pulmonary disease  
Includes: asthma with chronic obstructive pulmonary disease  
chronic bronchitis with ~~airways~~ airway obstruction
- Revise
- J45 Asthma  
Excludes1: detergent asthma (J69.8)  
~~eosinophilic asthma (J82)~~  
Excludes2: asthma with chronic obstructive pulmonary  
disease (J44.9)  
eosinophilic asthma (J82.83)
- Delete
- Add
- J91 Pleural effusion in conditions classified elsewhere  
J91.0 Malignant pleural effusion  
Code first underlying neoplasm (C00-D49)
- Revise
- K35 Acute appendicitis  
K35.3 Acute appendicitis with localized peritonitis  
K35.32 Acute appendicitis with perforation, localized peritonitis,  
and gangrene, without abscess  
K35.33 Acute appendicitis with perforation, localized peritonitis,  
and gangrene, with abscess
- Revise
- Revise
- K77 Liver disorders in diseases classified elsewhere  
Code first underlying disease, such as:  
amyloidosis (E85.-)  
congenital syphilis (A50.0, A50.5)

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		congenital toxoplasmosis (P37.1)
Revise		infectious mononucleosis with liver disease (B27.0-B27.9 with <u>fifth character 9</u> )
	K80	Cholelithiasis
		K80.4 Calculus of bile duct with cholecystitis
		Any condition listed in K80.5 with cholecystitis (with cholangitis)
Revise		<del>Codes</del> <u>Code</u> also fistula of bile duct (K83.3)
	M50	Cervical disc disorders
Delete		<del>Note: code to the most superior level of disorder</del>
	M67	Other disorders of synovium and tendon
		Excludes1: palmar fascial fibromatosis [Dupuytren] (M72.0)
		tendinitis NOS (M77.9-)
Revise		xanthomatosis localized to tendons ( <del>E78.2</del> ) ( <u>E75.2</u> )
	O10	Pre-existing hypertension complicating pregnancy, childbirth and the puerperium
		O10.0 Pre-existing essential hypertension complicating pregnancy, childbirth and the puerperium
Revise		O10.01 Pre-existing essential hypertension complicating pregnancy,
		pregnancy,
	P29	Cardiovascular disorders originating in the perinatal period
Delete		<del>Excludes1: congenital malformations of the circulatory system (Q20-Q28)</del>
Add		Excludes2: congenital malformations of the circulatory system (Q20-Q28)
	S23	Dislocation and sprain of joints and ligaments of thorax
Revise		Code also any associated; open wound
	U09	Post COVID-19 condition U09.9 Post COVID-19 condition, unspecified
Delete		<del>Note: This code enables establishment of a link with COVID-19.</del>
		<del>This code is not to be used in cases that are still presenting with active COVID-19. However, an exception is made in cases of re-infection with COVID-19, occurring with a condition related to prior COVID-19.</del>
Add	Note:	This code enables establishment of a link with COVID-19.
Add		This code is not to be used in cases that are still presenting with active COVID-19. However, an exception is made in

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cases of re-infection with COVID-19, occurring with a  
condition related to prior COVID-19.

Add	W08	Fall from other furniture Fall from stool
Add	X15	Contact with hot household appliances
Add	X15.3	Contact with hot saucepan or skillet
Add		Contact with hot cooking pan
		Contact with hot cooking pot
Delete	Z87	Personal history of other diseases and conditions
	Z87.7	Personal history of (corrected) congenital malformations
		<del>Excludes1: congenital malformations that have been partially corrected or repair but which still require medical treatment—code to condition</del>
Add		Excludes2: <u>congenital malformations that have been partially corrected or repair but which still require medical treatment - code to condition</u>

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**All proposed to be effective October 1, 2022**

- Abscess (connective tissue) (embolic) (fistulous) (infective) (metastatic)  
(multiple) (pernicious) (pyogenic)(septic) L02.91
- Revise - gingival ~~-see Periodontitis, localized~~  
Add - - aggressive K05.20  
Add - - - generalized K05.229  
Add - - - - moderate K05.222  
Add - - - -severe K05.223  
Add - - - - slight K05.221  
Add - - - localized K05.219  
Add - - - - moderate K05.212  
Add - - - - severe K05.213  
Add - - - - slight K05.211
- Revise - gum ~~-see Periodontitis, localized~~  
Add - - aggressive K05.20  
Add - - - generalized K05.229  
Add - - - - moderate K05.222  
Add - - - - severe K05.223  
Add - - - - slight K05.221  
Add - - - localized K05.219  
Add - - - - moderate K05.212  
Add - - - - severe K05.213  
Add - - - slight K05.211
- Revise - peritoneum, peritoneal (perforated) (ruptured) K65.1(2060)  
- - postoperative ~~T81.49~~ T81.43
- Revise Alcohol, alcoholic, alcohol-induced  
- delirium (acute) (tremens) (withdrawal) ~~F10.231~~F10.921  
Add - - dependence (acute) (tremens) (withdrawal) F10.231
- Revise Aneurysm (anastomotic) (artery) (cirroid) (diffuse) (false)  
(fusiform) (multiple)(saccular) I72.9  
- brain I67.1  
- - arteriosclerotic I67.1  
- - - ruptured -see Hemorrhage, intracranial, subarachnoid  
Revise - - - - ruptured -see Aneurysm, arteriovenous, brain, ruptured I60.8\_  
Revise - - - ruptured -see Aneurysm, arteriovenous, brain, ruptured I60.8\_
- Revise Antritis J32.0  
- stomach (see also Gastritis) ~~K29.60~~ K29.50  
Revise - - with bleeding ~~K29.61~~ K29.51

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- Appendicitis (pneumococcal) (retrocecal) K37  
- with  
-- gangrene K35.89  
Add --- with localized peritonitis K35.31  
Revise - acute (catarrhal) (fulminating) (~~gangrenous~~) (obstructive)  
(retrocecal) (suppurative) K35.80  
-- specified NEC K35.890  
--- with gangrene K35.891  
Add --- with localized peritonitis K35.31  
Revise - gangrenous -see Appendicitis, with, gangrene acute
- Breakdown  
Revise -- joint prosthesis -see Complications---, joint prosthesis, internal, mechanical,  
by site
- Revise Bullet wound ~~see also Wound, open~~ see also Puncture, open
- Calcification  
- adrenal (capsule) (gland) E27.49  
Revise -- tuberculous ~~E35 [B90.8]~~ B90.8 [E35]
- Cardiomyopathy (familial) (idiopathic) I42.9  
- ischemic I25.5  
Add -- non-ischemic I42.8  
Add - other cardiomyopathies NEC I42.8
- Revise Cervicitis (acute) (chronic) (nonvenereal) (senile) (atrophic)  
(subacute) (with ulceration) N72
- Collapse R55  
- vertebra M48.50-  
-- in (due to)  
Delete --- ~~metastasis~~ see Collapse, vertebra, in, specified disease NEC  
Add -- - neoplasm (metastasis) M84.50
- Complication(s) (from) (of)  
- coronary artery (bypass) graft T82.9  
-- atherosclerosis -see Arteriosclerosis, coronary (artery),  
Revise -- embolism ~~T82.818~~ T82.817  
Revise -- fibrosis ~~T82.828~~ T82.827  
Revise -- hemorrhage ~~T82.838~~ T82.837  
Revise -- pain ~~T82.848~~ T82.847  
Revise -- stenosis ~~T82.858~~ T82.857  
Revise -- thrombosis ~~T82.868~~ T82.867



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- Revise Death (cause unknown) (of) (unexplained) (unspecified cause) R99  
- cardiac (sudden) (with successful resuscitation) - ~~code to underlying disease~~  
see Arrest, cardiac
- Revise Deficiency, deficient  
- nutrition, nutritional E63.9 – see also Nutritional deficiency
- Deformity, limb, clawhand  
Deformity Q89.9  
- limb (acquired) M21.90  
- - clawfoot M21.53-  
- - clawhand M21.51-  
Add - - - congenital Q68.1
- Revise Degeneration, degenerative  
- disc disease -see Degeneration, intervertebral disc-~~NEC~~, by site
- Revise Derangement  
- ankle (internal) -see Derangement, joint, articular cartilage, ankle  
Revise - elbow (internal) -see Derangement, joint, articular cartilage, elbow  
Revise - hip (joint) (internal) (old) -see Derangement, joint, articular cartilage, hip
- Revise Dermatormucosomyositis – see also Dermatomyositis, M33.10
- Dermatomyositis (acute) (chronic)  
- juvenile M33.00  
Add - - amyopathic M33.03  
- specified NEC M33.10  
Add - - amyopathic M33.13
- Dermatopolymyositis M33.90  
- with  
Add - - amyopathic M33.03  
Add - - without myopathy M33.03  
- specified NEC M33.10  
Add - - amyopathic M33.13  
Add - - without myopathy M33.13
- Revise Diabetes, diabetic (mellitus) (sugar) E11.9  
- inadequately controlled –~~code to~~ – see Diabetes, by type, with hyperglycemia  
Revise - out of control - ~~code to~~ – see Diabetes, by type, with hyperglycemia  
Revise - poorly controlled –~~code to~~ – see Diabetes, by type, with hyperglycemia  
Add - retina, hemorrhage E13.39  
- type 2 E11.9

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- - with
- - - coma due to
- Revise - - - - ketoacidosis E11.1-
  
- Disease, diseased -see also Syndrome
- liver (chronic) (organic) K76.9
- Revise - - end stage ~~K72.90~~ K72.1-
- Revise - myelodysplastic, ~~not classified~~ C94.6 -see also Syndrome, myelodysplastic
- Add - myelodysplastic/myeloproliferative neoplasm, unclassifiable C94.6
- Revise - myeloproliferative, ~~not classified~~ ~~C94.6~~ D47.1
- Add - - not classified C94.6
- Add - - specified NEC C94.6
- Add - - unclassifiable C94.6
  
- Disorder
- Revise - adrenogenital E25.9 – see also Adrenogenital syndrome
  
- Embolism (multiple) (paradoxical) I74.9
- vein (acute) I82.90
- Add - - calf, muscle I82.46-
- Add - - - chronic I82.56-
- Add - - peroneal I82.45-
- Add - - - chronic I82.55-
- Add - - gastrocnemial I82.46-
- Add - - - chronic I82.56-
- Add - - soleal I82.46-
- Add - - - chronic I82.56-
  
- Granulomatosis L92.9
- Revise - with polyangiitis M31.3-
  
- Hemorrhage, hemorrhagic (concealed) R58
- retina, retinal (vessels) H35.6-
- Revise - - diabetic -see ~~Diabetes, retinal, hemorrhage~~ Disorder, retina, microaneurysm, diabetic
  
- Infarct, infarction
- myocardium, myocardial (acute) (with stated duration of 4 weeks or less) I21.9
- - subsequent (recurrent) (reinfarction) I22.9
- Revise - - - transmural ~~I21.3~~ I22.9
  
- Injury -see also specified injury type T14.90
- nerve NEC T14.8
- Revise - - lumbar spinal -see Injury, ~~nerve,~~ spinal, lumbar
- Revise - - sacral spinal -see Injury, ~~nerve,~~ spinal, sacral

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- Insufficiency, insufficient  
Revise - nourishment T73.0 – see also Deficiency, nutrient element
- Leak, leakage  
Revise - device, implant or graft -see also Complications, by site and type, mechanical  
- - arterial graft NEC -see Complication, ~~cardiovascular device,~~  
~~mechanical~~, vascular, graft, mechanical, leakage T82.833
- Lymphoma (of) (malignant) C85.90  
Revise - peripheral T-cell, ~~not classified~~ NEC C84.4-
- Malnutrition E46  
- degree  
- - first E44.1  
- - - intermediate form E42  
- - - - with  
Revise - - - - kwashiorkor (~~and marasmus~~) E42
- Main en griffe (acquired) -see also Deformity, limb, clawhand  
Revise - congenital ~~Q74.0~~ Q68.1
- Migraine (idiopathic) G43.909  
- without refractory migraine G43.909  
- - with status migrainosus G43.901  
Revise - - without status migrainosus ~~G43.919~~ G43.909
- Menopause, menopausal (asymptomatic) (state) Z78.0  
Add - postirradiation (postprocedural)  
Add - - asymptomatic E89.40  
Add - - symptomatic E89.41
- Neuritis (rheumatoid) M79.2  
- cranial nerve  
Revise - - fifth or trigeminal ~~G51.0~~ G50.-
- Nutrition deficient or insufficient E63.9 -see also Malnutrition ~~E46~~  
Add - sequelae – see Sequelae, nutritional deficiency  
Add - specified NEC E63.8
- Osteoarthritis M19.90  
Revise - ankle M19.07\_  
Add - - post-traumatic M19.17-  
Add - - primary M19.07-  
Add - - secondary M19.27-

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Revise - elbow M19.02\_  
 Add - - post-traumatic M19.12-  
 Add - - primary M19.02-  
 Add - - secondary M19.22-  
 Revise - foot joint M19.07\_  
 Add - - post-traumatic M19.17-  
 Add - - primary M19.07-  
 Add - - secondary M19.27-  
 Revise - hand joint M19.04\_  
       - - first carpometacarpal joint M18.9  
 Add - - - post-traumatic – see Osteoarthritis, post-traumatic NEC, hand  
 joint, first carpometacarpal joint  
 Add - - - primary – see Osteoarthritis, primary, hand joint, first  
 carpometacarpal joint  
 Add - - - secondary – see Osteoarthritis, secondary, hand joint, first  
 carpometacarpal joint  
 Add - - post-traumatic M19.14-  
 Add - - primary M19.04-  
 Add - - secondary M19.24-  
 Revise - hip ~~M16.1~~ M16.9  
       - - bilateral M16.0  
 Add - - - due to hip dysplasia M16.2  
 Add - - - post-traumatic M16.4  
 Add - - - secondary M16.6  
       - - due to hip dysplasia (unilateral) M16.3-  
       - - - bilateral M16.2  
 Add - - post-traumatic – see Osteoarthritis, post-traumatic, hip  
 Add - - primary M16.1-  
 Add - - - bilateral M16.0  
 Add - - secondary - see Osteoarthritis, secondary, hip  
 Add - - unilateral M16.1-  
 Add - - - due to hip dysplasia M16.3-  
 Add - - - post-traumatic M16.5-  
 Add - - - primary M16.1-  
 Add - - - secondary NEC M16.7  
 Revise - knee ~~M17.1~~ M17.9  
       - - bilateral M17.0  
 Add - - - post-traumatic M17.2  
 Add - - - secondary M17.4  
 Add - - post-traumatic – see Osteoarthritis, post-traumatic, knee  
 Add - - primary M17.1-  
 Add - - - bilateral M17.0  
 Add - - secondary - see Osteoarthritis, secondary, knee  
 Add - - unilateral M17.1-  
 Add - - - post-traumatic M17.3-  
 Add - - - primary M17.1-

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- Add
  - - - secondary NEC M17.5
  - post-traumatic NEC M19.92
- Revise
  - - hip M16.5<sub>2</sub>
- Revise
  - - knee M17.3<sub>2</sub>
  - primary M19.91
- Revise
  - - hip M16.1<sub>2</sub>
- Revise
  - - knee M17.1<sub>2</sub>
  - secondary M19.93
- Revise
  - - hip M16.7<sub>2</sub>
- Revise
  - - knee M17.5<sub>2</sub>
- Revise
  - shoulder M19.01<sub>2</sub>
- Add
  - - post-traumatic M19.11-
- Add
  - - primary M19.01-
- Add
  - - secondary M19.21-
- Revise
  - wrist M19.03<sub>2</sub>
- Add
  - - first carpometacarpal joint – see Osteoarthritis, hand joint,  
first carpometacarpal joint
- Add
  - - post-traumatic M19.13-
- Add
  - - primary M19.03-
- Add
  - - secondary M19.23-
  
- Revise
  - Osteochondromatosis ~~D48.0~~ D16.9
  
  - Osteoporosis (female) (male) M81.0
    - age-related M81.0
    - - with current pathologic fracture M80.00
- Add
  - - - rib(s) – see Osteoporosis, specified site NEC
- Add
  - - - specified site NEC M80.0A
  
- Add
  - Periodontitis (chronic) (complex) (compound) (local) (simplex) K05.30
    - aggressive K05.20
    - - generalized K05.229
    - - - moderate K05.222
    - - - severe K05.223
    - - - slight K05.221
    - - localized K05.219
    - - - moderate K05.212
    - - - severe K05.213
    - - - slight K05.211
  
- Revise
  - Perityphlitis (see also Cecitis) K37
  
- Add
  - Poikilodermatomyositis M33.10
    - amyopathic M33.13
    - without myopathy M33.13

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- Presence (of)
- Revise - cardioverter-~~defibrillator~~ defibrillator (ICD) Z95.810
- Pseudohermaphroditism Q56.3
- Revise - female Q56.2 - see also Disorder, adrenogenital
- Delete —~~unspecified E25.9~~
- Revise - male Q56.1 - see also Disorder, adrenogenital
- Delete —~~unspecified E25.9~~
- Schizophrenia, schizophrenic F20.9
- Revise - undifferentiated (type) F20.3
- - chronic ~~F20.5~~ F20.9
- Scoliosis (acquired) (postural) M41.9
- Revise - degenerative ~~M41.8~~ M41.5
- Scotoma (arcuate) (Bjerrum) (central) (ring) -see also Defect, visual field, localized, scotoma
- Revise - scintillating ~~H53.19~~ H53.12
- Subluxation - see also Dislocation
- Revise - symphysis (pubis) - see also Dislocation, symphysis pubis
- Supervision (of)
- Revise - high-risk pregnancy -see Pregnancy, ~~complicated by,~~ supervision of, high, risk high-risk
- Add Transgender F64.9
- Revise Typhlitis -see ~~Appendicitis~~ Cecitis
- Withdrawal state -see also Dependence, drug by type, with withdrawal - alcohol
- Revise - - abuse – see Abuse, alcohol, with, withdrawal
- Revise - - dependence – see Dependence, alcohol, with, withdrawal
- Revise - - use – see Use, alcohol, with, withdrawal
- - with perceptual disturbances F10.232
- Add - - - due to alcohol abuse F10.132
- Add - - - due to alcohol use F10.932
- - without perceptual disturbances F10.239
- Add - - - due to alcohol abuse F10.139
- Add - - - due to alcohol use F10.939

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**ICD-10-CM EXTERNAL CAUSE OF MORBIDITY INDEX PROPOSED ADDENDA**  
**All proposed to be effective October 1, 2022**

Accident (to) X58  
- transport (involving injury to) V99  
- - motorcyclist V29.9  
- - - driver  
- - - - collision (with)  
Add - - - - - pedestrian  
Add - - - - - traffic V20.4  
Add - - - - - nontraffic V20.0  
- - - passenger  
- - - - collision (with)  
Add - - - - - pedestrian  
Add - - - - - traffic V20.5  
Add - - - - - nontraffic V20.1  
- - pedestrian  
- - - conveyance (occupant) V09.9  
Revise - - - - ~~skate board~~ skateboard V00.138

Place of occurrence Y92.9  
Revise - highway (~~interstate~~) ~~Y92.411~~ Y92.410  
Add - - interstate Y92.411  
Add - interstate Y92.411  
Revise - lake ~~Y92.828~~ Y92.838  
Add - - wilderness Y92.828

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**ICD-10-CM TABLE OF DRUGS AND CHEMICALS PROPOSED ADDENDA**  
**All proposed to be effective October 1, 2022**

Revise	Chloroethane - <i>see Ethyl<sub>2</sub> chloride</i>						
Revise	Chlorethyl - <i>see Ethyl<sub>2</sub> chloride</i>						
Revise	Cuprous sulfate - <i>see also Copper<sub>2</sub> sulfate</i>						
Revise	DMSO - <i>see Dimethyl<sub>2</sub> sulfoxide</i>						
Revise	Epilim - <i>see Sodium<sub>2</sub> valproate</i>						
	Fumes (from)						
Revise	- pesticides - <i>see Pesticides-Pesticide</i>						
Revise	Gas NEC						
	- utility (for cooking, heating, or lighting) (piped)NEC	T59.891	T59.892	T59.893	T59.894	--	--
	- - incomplete combustion of - <i>see Carbon, monoxide, fuel, <del>utility</del>-utility</i>						
Revise	Iodine (antiseptic, external) (tincture) NEC	T49.0X1	T49.0X2	T49.0X3	T59.0X4	T49.0X5	T49.0X6
	- 125 - <i>see also Radiation sickness, and</i>	T50.8X1	T50.8X2	T50.8X3	T50.8X4	T50.8X5	T50.8X6
Revise	<i>Exposure to <del>radioactive</del> radioactive isotopes</i>						
	- 131 - <i>see also Radiation sickness, and</i>	T50.8X1	T50.8X2	T50.8X3	T50.8X4	T50.8X5	T50.8X6
	<i>Exposure to <del>radioactive</del> radioactive</i>						
	Isotopes						
Revise	Moth balls – <i>see Pesticides-Pesticide</i>	T60.2X1	T60.2X2	T60.2X3	T60.2X4	--	--
	Spray (aerosol)						
Revise	- pesticides – <i>see Pesticides Pesticide</i>						