ICD-10 Coordination and Maintenance Committee Meeting
March 8-9, 2022
Diagnosis Agenda

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

Diagnosis Topics:
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ICD-10 TIMELINE

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

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 **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	New ICD-10 codes to capture new diseases and technology will be implemented on April 1, 2022.
April 8, 2022	**Deadline for receipt of public comments on proposed new 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

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
May 9, 2022	**Deadline for receipt of public comments on proposed new codes and revisions discussed at the March 8-9, 2022, ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2023**.
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 expected to be published in the Federal Register as mandated by Public Law 99-509. This rule will also include links to all the final codes to be implemented on October 1, 2022. This rule can be accessed at: https://www.cms.gov/medicare/medicare-fee-for-service-payment/acuteinpatientpps
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

August 12, 2022

On-line registration opens for the September 13-14, 2022, ICD-10 Coordination and Maintenance Committee Meeting at: https://www.eventbrite.com/

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 is anticipated to be fully virtual by zoom and dial-in. Those who wish to attend must participate via Zoom Webinar or by dialing in.

September 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

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, 2022, 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, 2022, ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2023.**

Contact Information
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Continuing Education Credits

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

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

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

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

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

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

Anal Fistula

An anal fistula is an inflammatory tract or connection between the surface of the anal canal and, most frequently, the perianal skin or perineum, typically evolving from an anal abscess.[1] The disease has significant implications for a patient's quality of life, as symptoms range from pain and discharge to fecal incontinence.

Anal fistulas are typically classified using the Parks classification system, which considers the external sphincter as a central point of reference to describe five distinct types of fistulas: superficial, intersphincteric, transsphincteric, suprasphincteric, and extrasphincteric.[2] The classification system describes the anatomic location of the fistula and facilitates the identification of a treatment pathway. The system is also useful in describing the complexity of the condition and related treatment protocols.

While clinical definitions of complex anal fistula can vary, clinicians are aligned on a consistent definition of simple fistula. According to several clinical guidelines, an anal fistula is considered to be "simple" when the tract is intersphincteric or low intersphincteric (crossing <30% of the external anal sphincter).[3] In addition, simple fistulas have a single external and internal opening, are associated with no pain or fluctuation to suggest presence of perianal abscess and have no evidence of a rectovaginal fistula or anorectal stricture.[2]

The management of patients with anal fistulas varies depending on severity of disease and underlying comorbidities (such as Crohn's disease).[4] Treatment and management of simple fistulas are relatively straightforward compared with complex anal fistulas. Complex anal fistulas can be much more challenging to manage, resulting in high disease burden, diminished health-related quality of life, and increased healthcare resource use and costs.[5] Treatments vary by location and fistula type, and include fistulotomies, endoanal advancement flap or ligation of the intersphincteric fistula tract (LIFT), proctectomies, and fecal diversions.[6]

A common complication of anal fistula surgery is recurrence of fistulas after surgery, which represents a challenging problem as these fistulas are usually associated with higher risk of re-recurrence and fecal incontinence.[7]

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

Takeda Pharmaceuticals America, Incorporated is proposing the following tabular modifications to enable better tracking of complex fistulas, facilitating greater understanding of anal fistula epidemiology, and improving treatment paradigms. The American Gastroenterological Association (AGA) has reviewed and supports this proposal.

References:

1. Champagne, Bradley J. UpToDate, 2019, http://www.uptodate.com/contents/operative-management-of- anorectal-fistulas
2. American Gastroenterological Association Clinical Practice Committee. American Gastroenterological Association medical position statement: perianal Crohn's disease. *Gastroenterology*. 2003;125(5):1503-1507. doi:10.1016/j.gastro.2003.08.024
3. Vogel JD, Johnson EK, Morris AM, et al. Clinical Practice Guideline for the Management of Anorectal Abscess, Fistula-in-Ano, and Rectovaginal Fistula. *Dis Colon Rectum*. 2016;59(12):1117-1133. doi:10.1097/DCR.0000000000000733
4. Vogel, Jon D. UpToDate, 2020, https://www.uptodate.com/contents/anorectal-fistula-clinical-manifestations- diagnosis-and-management-principles
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6. Williams G, Williams A, Tozer P, et al. The treatment of anal fistula: second ACPGBI Position Statement - 2018. *Colorectal Dis*. 2018;20 Suppl 3:5-31. doi:10.1111/codi.14054
7. Emile SH. Recurrent anal fistulas: When, why, and how to manage?. *World J Clin Cases*. 2020;8(9):1586-1591.doi:10.12998/wjcc.v8.i9.1586

TABULAR MODIFICATIONS

	K60	Fissure and fistula of anal and rectal regions	
New subcategory	K60.3	Anal fistula	
New sub-subcategory	K60.31	Anal fistula, simple	
New code		K60.311	Anal fistula, simple, persistent
New code		K60.312	Anal fistula, simple, recurrent
New code		K60.319	Anal fistula, simple, unspecified
New sub-subcategory	K60.32	Anal fistula, complex	
New code		K60.321	Anal fistula, complex, persistent
New code		K60.322	Anal fistula, complex, recurrent
New code		K60.329	Anal fistula, complex, unspecified
New code	K60.39	Other anal fistula	
Add		Anal fistula NOS	

Appendicitis with Generalized Peritonitis with or without Perforation

When appendicitis leads to a frank perforation or rupture, that will usually cause severe peritonitis, which is commonly generalized peritonitis, although it can sometimes become walled off and localized. However, there can also be appendicitis with microperforations, which can lead to some degree of peritonitis, but milder. It is possible for appendicitis to present with generalized peritonitis, even without a frank perforation or rupture of the appendix.

It is being proposed to create codes for acute appendicitis with generalized peritonitis, with perforation and without perforation, and unspecified as to perforation. This proposal is based on internal discussions within CDC and CMS.

TABULAR MODIFICATIONS

K35 Acute appendicitis

K35.2 Acute appendicitis with generalized peritonitis

Delete ~~Appendicitis (acute) with generalized (diffuse) peritonitis following rupture or perforation of appendix~~

K35.20 Acute appendicitis with generalized peritonitis, without abscess

Delete ~~(Acute) appendicitis with generalized peritonitis NOS~~

New code K35.200 Acute appendicitis with generalized peritonitis, without perforation or abscess

Add (Acute) appendicitis with generalized peritonitis without rupture or perforation of appendix, NOS

New code K35.201 Acute appendicitis with generalized peritonitis, with perforation, without abscess

Add Appendicitis (acute) with generalized (diffuse) peritonitis following rupture or perforation of appendix, NOS

New code K35.209 Acute appendicitis with generalized peritonitis, without abscess, unspecified as to perforation

Add (Acute) appendicitis with generalized peritonitis NOS

	K35.21	Acute appendicitis with generalized peritonitis, with abscess	
New code	K35.210	Acute appendicitis with generalized peritonitis, without perforation, with abscess	
Add		(Acute) appendicitis with generalized peritonitis without rupture or perforation of appendix, with abscess	
New code	K35.211	Acute appendicitis with generalized peritonitis, with perforation and abscess	
Add		Appendicitis (acute) with generalized (diffuse) peritonitis following rupture or perforation of appendix, with abscess	
New code	K35.219	Acute appendicitis with generalized peritonitis, with abscess, unspecified as to perforation	
Add		(Acute) appendicitis with generalized peritonitis and abscess NOS	

Bardet-Biedl Syndrome and Laurence-Moon Syndrome

Bardet-Biedl Syndrome (BBS) is a rare genetic disorder of obesity, affecting approximately 1 in 140,000-160,000 people in North America and Europe,[1] and estimated to affect approximately 3000 people in the U.S. and Canada alone.[2] Patients with BBS can experience endocrine abnormalities, such as hypogonadism; visual impairment; cognitive disabilities; polydactyly; renal dysfunction; genital, renal, and dental anomalies; speech; and developmental delays.[3]

BBS has been studied since the early 1900's.[4] It is caused by genetic variants located on the MC4R pathway that affects signals between the brain and body. The underlying cause, regardless of gene type, is malfunction of the primary cilia, a key component of cellular communication. There are at least 21 genes associated with BBS, including the following: BBS1, BBS2, ARL6 (BBS3), BBS4, BBS5, MKKS (BBS6), BBS7, TTC8 (BBS8), BBS9, BBS10, TRIM32 (BBS11), BBS12, MKS1 (BBS13), CEP290 (BBS14), WDPCP (BBS15), SDCCAG8 (BBS16), LZTFL1 (BBS17), BBIP1 (BBS18), IFT27 (BBS19), IFT72 (BBS20), and C8ORF37 (BBS21).[5] Genetic testing to confirm BBS has evolved from targeted sequencing of common genetic variants, including the common BBS1 p.M390R, BBS2 p.Y24X, BBS2 p.R275X, and BBS10 c.91fsX5 mutations to next-generation sequencing gene panels containing all known BBS genes.[4]

While there is no clear link between the different mutations identified and disease severity, patients with mutations in the BBS1 gene seem to have milder ophthalmologic involvement, whereas patients with mutations in the BBS2, BBS3 and BBS4 genes experience classic deterioration of their vision, which in some cases leads to blindness due to retinitis pigmentosa. Patients with mutations in the BBS10 gene generally have significantly increased tendency toward obesity and insulin resistance.[6] Despite the large number of genes identified as being associated with BBS, genetic mutations have not been identified in an estimated 20-30 percent of individuals with BBS.[6]

Genetic testing has also been developed to identify BBS and has become the diagnostic tool of choice.**Error! Bookmark not defined.** However, a diagnosis of BBS does not necessarily indicate any particular symptom or severity of symptoms.

Because BBS can potentially impact multiple body systems, the current treatment modality is to manage the BBS patient's presenting symptoms. Because obesity is a common manifestation, controlling the patient's weight is important as doing so can have a positive impact on other body systems as well. More recently, therapies that target the genetic mutation are being developed and may provide relief to BBS patients in the future.**Error! Bookmark not defined.**

To properly diagnose BBS, providers rely on the identification of four of the primary characteristics (listed below), or if the person has three primary characteristics and at least two secondary characteristics. Note that most people with BBS do not have all of the characteristics listed below.[1,3,4] The primary characteristics of BBS (and approximate percent affected) are: visual impairment caused by retinal abnormalities (90-100); obesity, typically apparent by age one (72-92); polydactyly (extra fingers or toes) (63-81); hypogonadism (59-98); renal

anomalies (kidney malformations and/or malfunctions) (20-53); and learning disabilities (50-61). Secondary characteristics include: developmental delays; speech disorders / delay; dental anomalies (small teeth, small lower jaw, short teeth); behavioral problems; neurological problems; hypertension; lack of a sense of smell (anosmia); flat, wide feet; no arches; thyroid problems; strabismus (with "lazy eye"); short stature relative to parents' height; toe and finger variations including short digits (brachydactyly); curved digits (clinodactyly), especially the outer fingers or toes; mild webbing (syndactyly) especially between the 2nd and 3rd toes.

Laurence-Moon syndrome (LMS) is a rare autosomal recessive condition defined by visual degeneration compounded with pituitary dysfunction. For some time LMS and BBS were grouped together and termed Laurence-Moon-Bardet-Biedl syndrome, because of similarities in these conditions. There is also similarity to Oliver-McFarlane syndrome (OMS). All three conditions are characterized by progressive blindness, obesity, and learning disabilities.[7]

The pituitary gland serves to regulate the major chemicals that drive body processes, ranging from growth and metabolism to reproduction potential. LMS is characterized by childhood neurological problems including loss of control over movement and loss of peripheral nerve function, which can result in a stiffness-contraction of the limbs. Intellectual disabilities may be associated. [7]

LNMS is most commonly associated with mutations in the PNPLA6 gene, with an autosomal recessive inheritance. The PNPLA6 gene is responsible for the production of proteins that drive the breakdown of cell membranes. The PNPLA6 protein is an enzyme that is thought to drive the growth of nerve and non-nerve cells as they grow and mature. This gene is notably associated not only with LNMS but also Oliver-McFarlane syndrome, and a number of other identified syndromes as well (Boucher-Neuhauser syndrome, Gordon-Holmes syndrome, and spastic paraplegia type 39). LNMS is estimated to have a prevalence of 1 in 100,000 in North America.[7]

Creation of a new ICD-10-CM diagnosis code for Bardet-Biedl syndrome (BBS) has been proposed by Rhythm Pharmaceuticals, Inc. A new code is needed to bring awareness to the BBS population, as well as to identify, diagnose and track patients and the clinical interventions used to treat and manage patients, and the outcomes of treatments. Given the way that LNMS and BBS have been grouped in the past, a separate code for LNMS is also being proposed.

References

1. M'hamdi O, Ouertani I, Chaabouni-Bouhamed H: Update on the Genetics of Bardet-Biedl Syndrome. *Mol Syndromol* 2014;5:51-56. https://doi.org/10.1159/000357054
2. Bardet Biedl Syndrome Foundation. What is BBS? Accessed at https://www.bardetbiedl.org/what-is-bbs.
3. Han C, Reyes-Capo DP, Liu CY, et al. Comprehensive Endocrine-Metabolic Evaluation of Patients with Alström Syndrome Compared with BMI-Matched Controls. *J Clin Endocrinol Metab*. 2018 Jul 1;103:2707-2719. https://doi.org/10.1210/jc.2018-00496
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6. National Organization for Rare Disorders (NORD). Rare Disease Database. Bardet-Biedl Syndrome. Accessed at https://rarediseases.org/rare-diseases/bardet-biedl-syndrome/

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TABULAR MODIFICATIONS

Q87 Other specified congenital malformation syndromes affecting multiple systems

Q87.8 Other specified congenital malformation syndromes, not elsewhere classified

New code Q87.83 Bardet-Biedl syndrome

Add Code also associated conditions, such as:
Add obesity (E66.8)
Add polydactyly (Q69.-)
Add retinal dystrophy (H35.5-)

New code Q87.84 Laurence-Moon syndrome

Q87.89 Other specified congenital malformation syndromes, not elsewhere classified

Delete ~~Laurence-Moon (-Bardet) Biedl syndrome~~

INDEX MODIFICATIONS

Syndrome …
Add - Oliver-McFarlane Q87.89

Crohn's Disease

Crohn's disease is an idiopathic inflammatory bowel disease that most commonly involves the ileum and colon but also involves the esophagus in approximately 10% of cases. In pediatric patients, the rate of esophageal involvement may be twice as high.[1] Approximately 5 to 15 percent have involvement of the mouth or gastroduodenal area.[2]

Currently, Crohn's disease can be classified by ICD-10-CM with sites involving colon, duodenum, ileum, jejunum, large intestine and small intestine. However, Crohn's disease can affect the entire gastrointestinal tract from the mouth to the anus.

The Regulatory Committee of the Association of Clinical Documentation Integrity Specialists (ACDIS) is proposing the following tabular modifications to accurately capture the site specificity of this condition. The American Gastroenterologist Association (AGA) has reviewed and supports this proposal.

References:
[1] https://www.sciencedirect.com/topics/nursing-and-health-professions/crohn-disease
[2] https://www.uptodate.com/contents/clinical-manifestations-diagnosis-and-prognosis-of-crohn-disease-in-adults

TABULAR MODIFICATIONS

	K50	Crohn's disease [regional enteritis]	
New subcategory	K50.2	Crohn's disease of oropharynx	
New code	K50.20	Crohn's disease of oropharynx without complications	
New sub-subcategory	K50.21	Crohn's disease of oropharynx with complications	
New code	K50.211	Crohn's disease of oropharynx with rectal bleeding	
New code	K50.212	Crohn's disease of oropharynx with intestinal obstruction	
New code	K50.213	Crohn's disease of oropharynx with fistula	
New code	K50.214	Crohn's disease of oropharynx with abscess	
New code	K50.218	Crohn's disease of oropharynx with other complications	
New code	K50.219	Crohn's disease of oropharynx with unspecified complications	

New subcategory	K50.3	Crohn's disease of esophagus		
New code		K50.30	Crohn's disease of esophagus without complications	
New sub-subcategory		K50.31	Crohn's disease of esophagus with complications	
New code			K50.311	Crohn's disease of esophagus with rectal bleeding
New code			K50.312	Crohn's disease of esophagus with intestinal obstruction
New code			K50.313	Crohn's disease of esophagus with fistula
New code			K50.314	Crohn's disease of esophagus with abscess
New code			K50.318	Crohn's disease of esophagus with other complications
New code			K50.319	Crohn's disease of esophagus with unspecified complications
New subcategory	K50.4	Crohn's disease of stomach		
New code		K50.40	Crohn's disease of stomach without complications	
New sub-subcategory		K50.41	Crohn's disease of stomach with complications	
New code			K50.411	Crohn's disease of stomach with rectal bleeding
New code			K50.412	Crohn's disease of stomach with intestinal obstruction
New code			K50.413	Crohn's disease of stomach with fistula
New code			K50.414	Crohn's disease of stomach with abscess
New code			K50.418	Crohn's disease of stomach with other complications
New code			K50.419	Crohn's disease of stomach with unspecified complications

Coma Due to Underlying Condition

Subsequently after the recent coding guideline changes which limits Glasgow coma scale codes to traumatic brain injury (TBI), National Center for Health Statistics received a proposal for the creation of a new ICD-10-CM code for "Coma NEC."

This is a representation from the September 2021 C&M meeting**;** edits are based on public comments and are **bolded**.

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 the coma; for example, coma secondary to spontaneous brain hemorrhage. Unspecified coma does not seem because it is known(specified) to be a non-TBI coma.

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 **R40.2A** **Coma due to underlying condition**
Add **Secondary coma**

Add **Code first underlying condition**

Add **Excludes1: Coma associated with traumatic brain injury (R40.21 – R40.244)**

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.

A proposal was presented at the September 2021 ICD10 Coordination and Maintenance meeting. In response to public comments, a revised proposal is being submitted for reconsideration. Changes are noted in **bold.**

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) in the tabular, but will remain indexed.

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.

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) to provide more clinical granularity.[1,2]

This proposal is supported by the American Academy of Pediatrics.

References

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TABULAR MODIFICATIONS

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

Q75.0 Craniosynostosis

Delete ~~Acrocephaly~~
Delete ~~Imperfect fusion of skull~~
Delete ~~Oxycephaly~~
Delete ~~Trigonocephaly~~

New code Q75.01 Sagittal craniosynostosis
Add **Non-deformational dolichocephaly**
Add **Non-deformational scaphocephaly**

New subcategory Q75.02 Coronal craniosynostosis
Add Anterior plagiocephaly
New code Q75.021 Coronal craniosynostosis unilateral
Add **Non-deformational anterior plagiocephaly**
New code Q75.022 Coronal craniosynostosis bilateral
Add **Non-deformational brachycephaly**
New code Q75.029 Coronal craniosynostosis unspecified

New code Q75.03 Metopic craniosynostosis
Add Trigonocephaly

New subcategory	Q75.04	Lambdoid craniosynostosis
Add		**Non-deformational 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**	**Multi-suture craniosynostosis**
New code		**Q75.051 Cloverleaf skull**
Add		**Kleeblattschaedel skull**
New code		**Q75.052 Pansynostosis**
New code		**Q75.058 Other multi-suture craniosynostosis**
New subcategory	**Q75.08**	**Other single-suture craniosynostosis**
New code		**Q75.081 Other single-suture craniosynostosis, unilateral**
New code		**Q75.082 Other single-suture craniosynostosis, bilateral**
New code		**Q75.089 Other single-suture craniosynostosis, unspecified**
New subcategory	**Q75.09**	**Craniosynostosis unspecified**
Add		**Craniosynostosis NOS**
New code		**Q75.091 Craniosynostosis unspecified, unilateral**
New code		**Q75.092 Craniosynostosis unspecified, bilateral**
New code		**Q75.099 Craniosynostosis unspecified**
Add		**Imperfect fusion of skull**

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.

The National Center for Health Statistics (NCHS) received a request to create ICD-10-CM codes for desmoid tumors. This is a representation from the September 2021 ICD10 Coordination and Maintenance (C&M) meeting**;** edits are based on public comments and are **bolded**.

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 β-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 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.

References

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3. Trovato, M. J., Schwartz , R. A., & Lambert , P. C. (2019). Home / For Patients and Families / Rare Disease Information / Desmoid Tumor. Retrieved May 29, 2019, from NORD National Organization for Rare Disorders: https://rarediseases.org/rare-diseases/desmoid-tumor/
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

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 upper extremity and shoulder girdle**
New code		**D48.116**	**Desmoid tumor of lower extremity and pelvic girdle**
Add			**Desmoid tumor of buttock**
New code		**D48.117**	**Desmoid tumor of back**
New code		D48.118	Desmoid tumor of other site
New code		D48.119	Desmoid tumor of unspecified site
New code	D48.19	Other specified neoplasm of uncertain behavior of connective and other soft tissue	

Encounter for Follow-Up Examination after Completed Treatment for Malignant Neoplasm

This topic was presented at the September 2021 ICD10 Coordination and Maintenance meeting and based on comments received during the public comment period it is being represented for consideration. Changes are noted in **bold**. 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 (AAD/A) 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.

Based on the current coding guidelines, Section I.C.21.8 for Factors influencing health status and contact with health services *follow-up codes are used to explain continuing surveillance following completed treatment of a disease, condition, or injury. They imply that the condition has been fully treated and no longer exists. They should not be confused with aftercare codes, or injury codes with a 7th character for subsequent encounter, that explain ongoing care of a healing condition or its sequelae. Follow-up codes may be used in conjunction with history codes to provide the full picture of the healed condition and its treatment. The follow-up code is sequenced first, followed by the history code.*

However, should a condition be found to have recurred on the follow-up visit, then an active malignant neoplasm diagnosis code should be used in place of the follow-up code because the patient now defaults back into active treatment of the malignant condition.

The AAD/A believes that not having a full range of codes to capture follow-up encounters of completed treatment modalities hamper the ability for healthcare providers and statisticians to aggregate patient outcomes and efficacy of malignant treatment modalities.

The encounter for follow-up examination after treatment for malignant neoplasm can be reported as long as there is continued surveillance of the patient. Duration on surveillance for individual malignancies should be determined by specialty clinical guidelines. For example, dermatology, patients are followed and surveilled after treatment of malignant lesions for the rest of their life.

The American Academy of Dermatology/Association is requesting 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

Add **Excludes1: Malignant neoplasm (C00 - C96)**

New code Z08.0 Encounter for follow-up examination after surgery for malignant neoplasm

Add **Medical surveillance following completed treatment, surgery only**

New code Z08.1 Encounter for follow-up examination after radiotherapy for malignant neoplasm

Add **Medical surveillance following completed treatment, radiotherapy only**

New code Z08.2 Encounter for follow-up examination after chemotherapy for malignant neoplasm

Add **Medical surveillance following completed treatment, chemotherapy only**

New code **Z08.7 Encounter for follow-up examination after combined treatment for malignant neoplasm**

Add **Medical surveillance following completed treatment of malignant neoplasm, multiple therapies**

Add **Use additional code to identify personal history of medical treatment (Z92.-)**

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

Add **Use additional code to identify personal history of medical treatment (Z92.-)**

New code Z08.9 Encounter for follow-up examination after unspecified treatment for malignant neoplasm

Encounter for Observation for Suspected Newborn Problem

The American Academy of Pediatrics (AAP) respectfully submits the following code proposal for observation and evaluation of a newborn who presents with a physiological monitoring device concern, however, is found to not have a medical issue.

This request is in response to comments received and mirrors the proposal Z03.83, Encounter for observation for suspected conditions related to home physiologic monitoring device ruled out which was presented and supported at the September 2021 ICD10 Coordination and Maintenance (C&M) meeting.

Almost all of the babies diagnosed with apnea while inpatient 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.

TABULAR MODIFICATIONS

Z05 Encounter for observation and evaluation of newborn for suspected diseases and conditions ruled out

This category is to be used for newborns, within the neonatal period (the first 28 days of life), who are suspected of having an abnormal condition, but without signs or symptoms, and which, after examination and observation, is ruled out.

New subcategory Z05.8 Observation and evaluation of newborn for other specified suspected condition ruled out

New code Z05.81 Observation and evaluation of newborn for suspected condition related to home physiologic monitoring device ruled out

Add Encounter for observation for apnea alarm without findings

Add		Encounter for observation for bradycardia alarm without findings
Add		Encounter for observation for malfunction of home cardiorespiratory monitor
Add		Encounter for observation for non-specific findings home physiologic monitoring device
Add		Encounter for observation for pulse oximeter alarm without findings
Add		Excludes1: neonatal bradycardia (P29.12)
Add		other newborn apnea (P28.4-)
Add		primary asleep apnea of newborn (P28.3-)
New code	Z05.89	Observation and evaluation of newborn for other specified suspected condition ruled out

Extraocular Muscle Entrapment

The National Center for Health Statistics (NCHS) received a proposal requesting the creation of ICD-10-CM codes for extraocular muscle entrapment for coding specificity and research. This is a re-presentation from the September 2021 ICD10 Coordination and Maintenance (C&M) meeting**;** edits are based on public comments and are **bolded**.

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.[1]

An article published by AO Surgery Reference states, "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".[2]

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

References
1. Grant, M. P., Mahoney, N. R., & Merali, F. I. (2015, 06 19). Orbital Floor Fracture with Atypical Extraocular Muscle Entrapment Pattern and Intraoperative Asystole in an Adult. Retrieved from Craniomaxillofac Trauma Reconstruction: https://journals.sagepub.com/doi/10.1055/s-0035-1556052

2. Cornelius, C.-P., Gellrich, N., Hillerup, S., Kusumoto, K., & Schubert, W. (n.d.). Emergency treatment. Retrieved from Surgery Reference: https://surgeryreference.aofoundation.org/cmf/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 **eye**
New code		H50.622	Inferior oblique muscle entrapment, left **eye**
New code		H50.629	Inferior oblique muscle entrapment, unspecified **eye**
New sub-subcategory	H50.63	Inferior rectus muscle entrapment	
New code		H50.631	Inferior rectus muscle entrapment, right **eye**
New code		H50.632	Inferior rectus muscle entrapment, left **eye**

New code		H50.639	Inferior rectus muscle entrapment, unspecified **eye**
New sub-subcategory	H50.64	Lateral rectus muscle entrapment	
New code		H50.641	Lateral rectus muscle entrapment, right **eye**
New code		H50.642	Lateral rectus muscle entrapment, left **eye**
New code		H50.649	Lateral rectus muscle entrapment, unspecified **eye**
New sub-subcategory	H50.65	Medial rectus muscle entrapment	
New code		H50.651	Medial rectus muscle entrapment, right **eye**
New code		H50.652	Medial rectus muscle entrapment, left **eye**
New code		H50.659	Medial rectus muscle entrapment, unspecified **eye**
New sub-subcategory	H50.66	Superior oblique muscle entrapment	
New code		H50.661	Superior oblique muscle entrapment, right **eye**
New code		H50.662	Superior oblique muscle entrapment, left **eye**
New code		H50.669	Superior oblique muscle entrapment, unspecified **eye**
New sub-subcategory	H50.67	Superior rectus muscle entrapment	
New code		H50.671	Superior rectus muscle entrapment, right **eye**
New code		H50.672	Superior rectus muscle entrapment, left **eye**
New code		H50.679	Superior rectus muscle entrapment, unspecified **eye**
New sub-subcategory	**H50.68**	**Extraocular muscle entrapment**	
New code		**H50.681**	**Extraocular muscle entrapment unspecified, right eye**
New code		**H50.682**	**Extraocular muscle entrapment unspecified, left eye**
New code		**H50.689**	**Unspecified extraocular muscle entrapment, unspecified eye**

Foreign Body Sensation

The National Center for Health Statistics (NCHS) received a request to create ICD-10-CM codes for foreign body sensation of the throat. This is a re-presentation from the September 2021 ICD10 Coordination and Maintenance (C&M) meeting; edits are based on public comments and are **bolded.**

Foreign body sensation is the persistent feeling of a lump in the throat or that something is stuck in the throat; patients often describe the sensation as throat fullness. It is usually not painful but described as annoying, it is a common condition.[1]

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 American Gastroenterological Association (AGA) and American Academy of Ophthalmology (AAO) 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

H57 Other disorders of eye and adnexa
H57.8 Other specified disorders of eye and adnexa

New subcategory	**H57.8A**	**Foreign body sensation eye (ocular)**	
New code		**H57.8A1**	**Foreign body sensation, right eye**
New code		**H57.8A2**	**Foreign body sensation, left eye**
New code		**H57.8A3**	**Foreign body sensation, bilateral eyes**
New code		**H57.8A9**	**Foreign body sensation, unspecified eye**

R09 Other symptoms and signs involving the circulatory and respiratory system

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

Add	Abnormal chest percussion
	Bruit (arterial)
Delete	~~Abnormal chest percussion~~
Add	Chest tympany
Add	Choking sensation
Delete	~~Feeling of foreign body in throat~~
	Friction sounds in chest
Delete	~~Chest tympany~~
Delete	~~Choking sensation~~
	Rales
	Weak pulse

New subcategory R09.A Foreign body sensation of the circulatory and respiratory system

New code R09.A0 Foreign body sensation, unspecified

New code R09.A1 Foreign body sensation, nose

New code R09.A2 Foreign body sensation, throat

Add Foreign body sensation, globus

New code R09.A9 Foreign body sensation, other site

Gadolinium Toxicity

The National Center for Health Statistics NCHS) has received a proposal requesting a unique code with a higher level of specificity for gadolinium toxicity. This will allow for accuracy in reporting gadolinium induced disease and used to track such disease and treatment.

Gadolinium is a heavy metal with paramagnetic properties in the middle of the lanthanides in the F block of the periodic table of elements. Gadolinium has many uses in electronics, medical imaging (as a contrast agent for MRI), phosphor in polymer matrix in x-ray detector, microwave applications, fabrication of various optical components, computer memory, refrigeration and in alloys making magnets and data storage discs.

Gadolinium toxicity has the potential to cause disease in humans, and even in small amounts may be associated with significant morbidity and mortality[1]. Gadolinium toxicity can affect many body systems,[2] including the musculoskeletal, brain, skin, renal and neurologic systems.

This toxicity can manifest itself in various symptoms and effects on the body including but not limited to central nervous system symptoms, including impairment of cognition, memory, impairment of sight, painful tinnitus, and pseudoangioedema. Additional manifestations can include impairment of voice and pharyngeal swallowing mechanisms, cardiac arrythmias, changes in blood pressure, and impaired function of the gastrointestinal tract and urinary system. Symptoms can be mild in some patients, while others develop severe life-threatening illness similar to cytokine storm response [3].

Additional rationale as to why a new code is being requested includes: to ensure gadolinium toxic patients are recognized, diagnosed properly, treated appropriately[4] and timely in order to prevent progressive disease and damages in the human body that is caused by gadolinium toxicity.

References:

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TABULAR MODIFICATIONS

T56 Toxic effect of metals
Includes: toxic effects of fumes and vapors of metals
toxic effects of metals from all sources, except medicinal substances
Use additional code to identify any retained metal foreign body, if applicable (Z18.0-, T18.1-)
Excludes1: arsenic and its compounds (T57.0)
manganese and its compounds (T57.2)

The appropriate 7th character is to be added to each code from category T56
A initial encounter
D subsequent encounter
S sequela

T56.8 Toxic effects of other metals

New subcategory	T56.82 Toxic effect of gadolinium	
New code	T56.821	Toxic effect of gadolinium, accidental (unintentional) Toxic effect of gadolinium NOS
New code	T56.822	Toxic effect of gadolinium, intentional self-harm
New code	T56.823	Toxic effect of gadolinium, assault
New code	T56.824	Toxic effect of gadolinium, undetermined

Immunoglobulin G4-Related Disease

Immunoglobulin G4-Related Disease (IgG4-RD) is a recently characterized, rare, multifocal disease of unknown origin. It is considered a chronic, relapsing-remitting, immune-mediated fibroinflammatory disorder that if not diagnosed and left untreated can lead to impaired organ function.[1] IgG4-RD is not associated with pain and may be asymptomatic, which can lead to intensified disease progression before it is identified by specialty physicians.

Over the past two decades, research has helped identify patients with this disease and distinctly classify them to alleviate confounding features that resemble other immune-mediated or malignancy-related conditions with similar initial characteristics. Despite these classification efforts, the complicated clinical diagnosis and nature of disease manifestation in multiple organs (e.g., lung, kidney, pancreas) present barriers to accurate classification, diagnosis, and treatment of the condition. This challenge may lead to an underappreciation of the disease by many physicians and indirectly contributes to unfavorable patient outcomes as a result. In general, IgG4-RD tends to possess the following characteristics in the majority of cases:[2]

- Tumefactive lesions (causing swelling)
- Dense lymphoplasmacytic infiltrate
- IgG4-positive plasma cells present in large numbers in tissues
- Storiform fibrosis (distinctive histopathological feature)
- Elevated serum IgG4 concentrations

Underappreciation, coupled with mis- or under-diagnosis, has hindered accurate estimates of the incidence and prevalence of IgG4-RD.[3] There have been no studies to assess the prevalence of IgG4-RD in the US, but studies performed in other countries lead to current estimates of its prevalence in the US at around 30,000.[4,5] Although data are not robust, there is a tendency for higher incidence in older male adults (61%) with an average age of onset of 58.8 years,[3,4] but women and children can also manifest hallmark IgG4-RD, with some data demonstrating particular localizations in those populations.[3]

As many as 85% of patients with IgG4-RD have active disease at the time of diagnosis. Pancreatic involvement is present in about 20-25% of IgG4-RD cases, making it the most common manifestation. In rare pediatric cases, periorbital IgG4-RD is more common and has been seen in as many as 44% of studied cases in that population.[6]

While there are no diagnostic biomarkers for IgG4-RD, there are themes to show how clinical presentation may occur and what markers are most predictive of accurate diagnosis. For example, serum IgG4 levels predict disease in most cases,[2] but can be elevated independent of the disease, making it helpful but not definitive.

A global steering committee organized by The American College of Rheumatology (ACR) and The European Alliance of Associations for Rheumatology (EULAR) have identified, weighted, and tested potential classification criteria for IgG4-RD, beginning in 2011 and most recently in 2019.[7] This effort is a consensus-driven source for IgG4-RD identification and diagnostic work

up procedures. Despite the ACR and EULAR guidelines, as well established clinical nomenclature[8], diagnosis and treatment of IgG4-RD remains low.

Although time to diagnosis can often be months to years due to difficulty with early detection, time to treatment after a clinical diagnosis can be shorter. Clinical management of IgG4-RD is varied, and detection can be delayed. The root causes for this are driven by the multi-factorial nature of the disease. The diagnosis of IgG4-RD is made by many specialists including neurologists, gastroenterologists, pulmonologists, and rheumatologists who recognize various characteristics of the disease. patients who are not able to receive the proper diagnosis and treatment face the loss of function of critical organs like the pancreas, kidney, and liver.

Patients with IgG4-RD are generally responsive to treatment with glucocorticoids. Earlier diagnosis has vast potential to alleviate burden of this disease, to the benefit of both patients and the healthcare system. Clinical testing has also shown utility of rituximab for treatment, which maybe allow less glucocorticoid use (and avoid potential for side effects from glucocorticoids).[9,10] Limited clinical studies are underway to test other compounds.

A specific code for IgG4-RD has been requested by John H. Stone, MD, MPH; Division of Rheumatology, Allergy and Immunology; Massachusetts General Hospital; Boston, MA. A new ICD-10-CM code for IgG4-RD would enhance the epidemiological capabilities and evidence generation for the nature of the disease, and aid in unifying healthcare provider specialties on the same disease and shorten the time-to-diagnosis and treatment, thereby improving patient outcomes. It will lead to improved access to care, tracking, and disease management efforts. To reiterate, IgG4-RD is particularly amenable to treatment, so the potential to alleviate burden from both a patient and health care system perspective can be realized with a distinction that IgG4-RD is unique and can be affirmatively diagnosed as a stand-alone disease.

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TABULAR MODIFICATIONS

D89 Other disorders involving the immune mechanism, not elsewhere classified

D89.8 Other specified disorders involving the immune mechanism, not elsewhere classified

New code	D89.84	IgG4-related disease
Add		Immunoglobulin G4-related disease
Add		Excludes2: chronic pancreatitis (K86.1)

Impairing Emotional Outbursts

A significant number of children and adolescents present to outpatient departments, emergency rooms, and inpatient units and/or are suspended from school because they respond to relatively ordinary frustrations and disappointments with volcanic anger or distress. Such impairing emotional outbursts occur in the context of a number of different mental disorders (e.g., attention-deficit/hyperactivity disorder, autism spectrum disorder, oppositional defiant disorder, generalized anxiety disorder, posttraumatic stress disorder, mood and psychotic disorders) and are often the reason for families seeking treatment.

For example, in a study of 107 outpatients (ages 7-17) with anxiety disorders, 55.1% had rage episodes in the prior week, with 7.5% having daily rages (Johnco et al., 2015). In another study of 462 patients (ages 3-25) from the Interactive Autism Network, 24% of patients ages 3-11 and 28% of patients ages 12-25 had severe tantrums that lead to crisis (Vasa et al., 2020). Impairing emotional outbursts can also occur independent of other conditions, as is often the case in young children. For example, in a community sample of preschoolers obtained from the waiting rooms of 5 large pediatric practices, pathological tantrums were found in 8.6% of the sample and a suburban community sample of 6 year-olds found that 11.0% had "severe tantrums," lasting at least 15 minutes at least three times a week (Carlson et al., 2016;Wakschlag et al., 2012).

To facilitate appropriate care for such children and adolescents, it is necessary to identify them reliably and communicate the nature of their problems with caregivers and other professionals. The availability of a symptom code for impairing emotional outbursts will facilitate the identification of outbursts as a focus for care, alongside the other conditions for which the child is being treated. Finally, through the collection of research and medical records data over time, the inclusion of such a symptom code will allow for improvements in the understanding, assessment, and treatment of impairing emotional outbursts in youth.

Because of its clinical significance, the American Psychiatric Association is planning to add impairing emotional outbursts to the 5th edition of the *Diagnostic and Statistical Manual of Mental Disorders* as one of the Other Conditions That May Be a Focus of Attention. Impairing Emotional Outbursts will be defined as follows: "*Displays of anger or distress manifested verbally (e.g., verbal rages, uncontrolled crying) and/or behaviorally (e.g., physical aggression toward people, property, or self) that lead to significant functional impairment."*

In ICD-10-CM, there is currently no symptom code for noting such outbursts. The American Psychiatric Association is proposing a new code to capture this behavior.

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TABULAR MODIFICATIONS

R45 Symptoms and signs involving emotional state

R45.8 Other symptoms and signs involving emotional state

New code R45.8A Impairing emotional outbursts

Inappropriate Sinus Tachycardia

Inappropriate sinus tachycardia (IST) is defined as a sinus heart rate >100 bpm at rest (with a mean 24-hour heart rate >90 bpm not due to primary causes) and is associated with distressing symptoms of palpitations.

The prevalence of IST was estimated in a middle-aged population of people with and without hypertension. Using a definition of a resting heart rate >100 bpm and an average heart rate of >90 bpm on 24-hour Holter monitoring, the IST prevalence was 1.2% (7 of 604 patients), including both symptomatic and asymptomatic patients.[2]

The mechanisms leading to IST are not completely understood, but there are several underlying diseases that can result in this syndrome, including increased sinus node automaticity, beta-adrenergic hypersensitivity, decreased parasympathetic activity, and impaired neurohumoral modulation. β-Adrenergic receptor antibodies can sensitize β- adrenergic receptors in some patients, while other patients might have increased sympathetic activity and sensitivity, with or without inherent impaired sinus node automaticity.

The National Center for Health Statistics (NCHS) received a request to create an ICD-10-CM code for inappropriate sinus tachycardia for coding specificity to accurately track cases, allowing for etiology related research, patient segmentation, and therapeutic selection.

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TABULAR MODIFICATIONS

I47 Paroxysmal tachycardia

I47.1 Supraventricular tachycardia

Delete ~~Atrial (paroxysmal) tachycardia~~
Delete ~~Atrioventricular [AV] (paroxysmal) tachycardia~~
Delete ~~Atrioventricular re-entrant (nodal) tachycardia [AVNRT] [AVRT]~~
Delete ~~Junctional (paroxysmal) tachycardia~~
Delete ~~Nodal (paroxysmal) tachycardia~~

New code	I47.10	Supraventricular tachycardia, unspecified
New code	I47.11	Inappropriate sinus tachycardia
New code	I47.19	Other supraventricular tachycardia
Add		Atrial (paroxysmal) tachycardia
Add		Atrioventricular [AV] (paroxysmal) tachycardia
Add		Atrioventricular re-entrant (nodal) tachycardia [AVNRT] [AVRT]
Add		Junctional (paroxysmal) tachycardia
Add		Nodal (paroxysmal) tachycardia

Insulin Resistant Syndrome

The National Center for Health Statistics (NCHS) received a request to create ICD-10-CM codes for Type A and Type B insulin resistance-syndrome for coding specificity. This is a re-presentation from the September 2021 ICD10 Coordination and Maintenance (C&M) meeting**;** edits are based on public comments and are **bolded**.

The National Institute of Health (NIH) 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 **and B** 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.[1]

The Office of Genomics Precision Public Health and American College of Medical Genetics and Genomics have reviewed and support the proposal.

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TABULAR MODIFICATIONS

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

E88.8 Other specified metabolic disorders

Revise E88.81 Metabolic syndrome **<u>and other insulin resistance</u>**

Delete ~~Dysmetabolic syndrome X~~

Use additional codes for associated manifestations, such as:
obesity (E66.-)

New code	**E88.810**	**Metabolic syndrome**
Add		**Dysmetabolic syndrome X**
New code	**E88.811**	**Insulin resistance syndrome, Type A**
New code	**E88.818**	**Other insulin resistance**
Add		**Insulin resistance Type B**
New code	**E88.819**	**Insulin resistance, unspecified**

Intestinal Failure-Associated Liver Disease

Intestinal failure is the inability to consume and absorb sufficient nutrition and fluid in order to maintain nutritional autonomy (Pironi et al. 2015). This serious disease is highly complex and require multidisciplinary management teams. Some patients develop complications and require intestinal transplantation. Upwards of 25-60% of these patients may develop liver disease, which may be reversible, or irreversible. If irreversible, death results in the absence of intestinal or intestine/liver transplantation. The disease intestinal failure-associated liver disease (IFALD) has been misclassified by clinicians as a form of non-alcoholic fatty liver disease (NAFLD), including non-alcoholic steatohepatitis (NASH), when in fact, the pathophysiology, biology, histology and prognosis are very distinct. IFALD is not NAFLD/NASH and is in fact, a different disease entity. (Buchman, 2017) IFALD is the leading indication for intestinal transplantation, (Buchman, 2021) yet it does not currently have a disease code.

IFALD, which occurs in patients dependent on parental nutrition support, is characterized by hepatic steatosis, cholestasis, fibrosis and rapid progression of liver disease through to hepatic failure and death in the absence of intestine-liver transplant. IFALD carries a relatively poor prognosis, with a 15–34% death rate within 1–4 years (Chan et al., 1999; Cavicchi et al., 2000). When IFALD presents with symptoms of liver disease in children, mortality is even higher (23–40%) (Willis et al., 2010; Pichler et al., 2012). The prevalence of IFALD in patients dependent upon PN increases as the number of years receiving PN increases, with up to 72% of patients presenting with IFALD after 6 years of chronic dependence on PN (Cavicchi et al., 2000).

In considering IFALD as a disease entity, it is important to note that IFALD is a distinct disease from nonalcoholic fatty liver disease (NAFLD) and nonalcoholic steatohepatitis (NASH), as salient medical characteristics, etiology, pathophysiology, epidemiology, and progression differ substantially between IFALD and these diseases (Buchman et al., 2017).

Histologically, both IFALD and NAFLD/NASH patients exhibit hepatic steatosis; however, patients with IFALD often exhibit an uncommon but characteristic subtype of steatosis, namely, a combination of both macro- and micro-vesicular steatosis (Cavicchi et al., 2000) and a low rate of steatohepatitis. A majority of IFALD patients exhibit microvesicular steatosis, in which small intracytoplasmic fat vacuoles (liposomes) accumulate and are diffusely dispersed throughout the cytoplasm in hepatic cells (Buchman et al., 2017). In contrast, microvesicular steatosis is not a common histological finding of NAFLD/NASH, which are commonly associated with macrovesicular steatosis. IFALD and NAFLD/NASH are described more fully as separate diseases in the published literature (Buchman et al., 2017).

Alan Buchman, MD, Professor of Clinical Surgery and Medical Director, Intestinal Rehabilitation and Transplant Center, University of Illinois at Chicago, is proposing the following tabular modifications for better tracking of these patients. The American Gastroenterologist Association (AGA) has reviewed and supports this proposal.

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TABULAR MODIFICATIONS

K76 Other diseases of liver

K76.8 Other specified diseases of liver

New code K76.83 Intestinal failure-associated liver disease

Lafora Body Disease

Lafora Body Disease, or Lafora progressive myoclonus epilepsy, is a neurodegenerative condition caused by a glycogen metabolism disorder that results in the accumulation of abnormal glycogen aggregates called Lafora Bodies in the brain, heart, and liver. This accumulation of Lafora Bodies causes the progressive degeneration of the nervous system resulting in generalized tonic-clonic seizures, occipital seizures, myoclonic seizures, tonic, absence and atonic seizures. In addition to the progression of seizures, patients with Lafora Disease also experience a rapid cognitive decline and gross and fine motor regression. Other symptoms of Lafora Disease include behavioral changes such as depression and confusion, and physical symptoms such as "ataxia (difficulty controlling muscles), difficulty walking, difficulty eating, dysarthria (difficulty speaking), and childhood dementia." [1] Children are born with the Lafora bodies but typically do not begin showing symptoms until late childhood or adolescence. There is no treatment for Lafora, and therapy is primarily palliative and focused on reducing seizures. Life expectancy extends 10 years after the onset of symptoms, which is generally around the mid-twenties. [1]

Lafora Disease is typically caused by a mutation in the EPM2A or EMP2B (NHLRC1) gene, both of which are responsible for Laforin and Malin regulation. [1] The EMP2A gene belongs to a family of genes that provide instructions for making a protein called Laforin, which plays a vital role in maintaining neurons in the brain.[2] Laforin has various functions within the cell, and it must interact with other proteins to carry out these functions, such as Malin which is produced by the EMP2B gene. The EMP2B (NHLRC1) gene belongs to a family of genes that provide "instructions for making a protein called Malin.[6] Although this protein is active in cells throughout the body, it appears to play a critical role in the survival of nerve cells (neurons) in the brain." [2] Malin helps break down unwanted proteins within cells and helps to tag damaged and excess proteins which aids in the degradation of these proteins. Malin targets several proteins such as Laforin which is produced by the EPM2A gene. Both Laforin and Malin control the way cells store glycogen, which is a form of sugar; and a mutation of either EMP2A or EMP2B results in a toxic buildup of unbranched, long chain glycogen molecules that accumulate in cells in the form of polyglucason bodies or Lafora bodies. The EMP2A and EMP2B genes are inherited, with Lafora body disease having autosomal recessive inheritance.[1-6]

Lafora disease has a prevalence of about four cases per one million persons.[4] It has been found that there is a "higher incidence of the disease in children of Middle Eastern, Southern European (Spain, France and Italy), South Asian (India and Pakistan) and North African descent. The disease appears to affect males and females equally." [1] It is estimated that the number is higher due to mis-and undiagnosed cases in undeveloped countries.[4]

The Lafora Cure Initiative (LECI) has four independent platforms working towards future treatments in Antisense Oligonucleotides, Antibody Enzyme Fusion, Small Molecules, and Gene Therapy. Through LECI, the Fundación Jiménez Díaz Hospital in Madrid, Spain has also established the Lafora Disease Registry which holds all patient data for future clinical trials.

There is currently no specific ICD-10-CM code for Lafora Disease. While it is now recognized to involve a disorder of carbohydrate metabolism, it has been classified with the generalized

idiopathic epilepsies (although it is no longer idiopathic). Chelsea's Hope Lafora Children Research's Fund has requested the creation of a specific ICD-10-CM code for Lafora Disease. A specific and unique code will be beneficial in more accurately capturing and tracking prevalence, enhancing research efforts, and aiding in proper diagnosis, treatment, and services for patients. Related neurodegenerative, childhood dementia, neurological and cognitive issues should also be coded.

It is proposed to create codes for Lafora progressive myoclonus epilepsy, and an inclusion term for Lafora body disease, as a new expansion at G40, Epilepsy and recurrent seizures. Clinically, Lafora progressive myoclonus epilepsy may be associated with seizures that can be intractable and can involve status epilepticus.

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TABULAR MODIFICATIONS

	G40	Epilepsy and recurrent seizures	
New subcategory		G40.C	Lafora progressive myoclonus epilepsy
Add			Lafora body disease
Add			Code also, if applicable, associated conditions such as dementia (F02.8-)
New sub-subcategory		G40.C0	Lafora progressive myoclonus epilepsy, not intractable
New code			G40.C01 Lafora progressive myoclonus epilepsy, not intractable, with status epilepticus

New code		G40.C09	Lafora progressive myoclonus epilepsy, not intractable, without status epilepticus
New sub-subcategory	G40.C1	Lafora progressive myoclonus epilepsy, intractable	
New code		G40.C11	Lafora progressive myoclonus epilepsy, intractable, with status epilepticus
New code		G40.C19	Lafora progressive myoclonus epilepsy, intractable, without status epilepticus

Leukodystrophies

Leukodystrophies, or more specifically inherited leukodystrophies, are a group of diseases affecting the white matter of the brain, that cause significant morbidities and death in 1 of 3 patients by age 8 years.[1] The Global Leukodystrophy Alliance (GLIA), an NIH-funded consortium composed of clinicians, scientists, patients, and patient advocacy groups, is requesting new ICD-10-CM codes for 31 separate and genetically distinct leukodystrophy diseases. NCHS has received letters of support from various professional and patient groups.

Leukodystrophies may present at any age from preterm infants and neonates to late adulthood and have been reported across all ethnicities and regions of the world. Thirty years ago only a few leukodystrophies were recognized as distinct disease entities, but in the past 10 years over 400 genetically unique leukodystrophies have been reported.[2] Even though most leukodystrophies are individually rare, as a group leukodystrophies affect close to 1 in 4,000 live births.[3,4] Further, consensus work in the community has defined a group of leukodystrophies with unique genetic causes and well-studied, distinct clinical and pathophysiological features.

Currently there are only specific ICD-10-CM codes for six of the primary leukodystrophies (X-linked Adrenoleukodystrophy, ALD- E71.52x; Metachromatic leukodystrophy, MLD- E75.25; Krabbe disease- E75.23, Refsum's disease- G60.1; Zellweger syndrome- E71.510; and E71.511 Neonatal Adrenoleukodystrophy). Otherwise, many leukodystrophies are indexed under a single ICD-10-CM code E75.29, Other sphingolipidosis.

Prior to ICD-10-CM, there were not specific ICD codes for ALD, MLD, or Krabbe. The advent of specific ICD-10-CM codes for ALD, Krabbe, and MLD enabled clinical trials, newborn screening, and studies of racial disparities.[6,7]

The leukodystrophies proposed for unique ICD-10-CM codes all have unique genetic causes; distinct clinical courses and morbidities; and have different treatments-either currently available or in clinical trials. For example, VWM has a sputtering clinical course[8] and has a clinical trial with the α-agonist guanabenz.[9] In contrast, Canavan's disease has an early rapid progression[10] and potential treatment with antisense oligonucleotides.[11]

Creation of specific codes for this heterogenous, complex group of disorders known as leukodystrophies is critical for patient care, clinical trials, and research. The diversity in causes should be reflected by a diversity of codes to best represent these disorders. The importance of and difference between these leukodystrophy disorders can be seen in the codes already created for ICD-11. In ICD-11, Leukodystrophy has its own ICD-11 category, there are five new leukodystrophy codes, and there are also fourteen new leukodystrophy indices.

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TABULAR MODIFICATIONS

D81 Combined Immunodeficiencies
D81.8 Other combined immunodeficiencies

New sub-subcategory — D81.82 Heritable interferonopathies
New code — D81.820 Aicardi-Goutières syndrome
New code — D81.828 Other heritable interferonopathies
New code — D81.829 Heritable interferonopathies, unspecified

E71 Disorders of branched-chain amino-acid metabolism and fatty-acid metabolism
E71.3 Disorders of fatty-acid metabolism

New sub-subcategory — E71.39 Other disorders of fatty-acid metabolism

New code		E71.390 Sjögren-Larsson syndrome
New code		E71.398 Other disorders of fatty-acid metabolism
New code		E71.399 Other disorders of fatty-acid metabolism, unspecified
	E71.5	Peroxisomal Disorders
New sub-subcategory		E71.53 Other Group 2 peroxisomal disorders
New code		E71.530 Peroxisomal D-bifunctional enzyme deficiency
New code		E71.531 Sterol carrier protein deficiency
New code		E71.532 Peroxisomal acyl-CoA oxidase deficiency
E72		Other disorders of amino-acid metabolism
	E72.8	Other specified disorders of amino-acid metabolism
New sub-subcategory		E72.82 Disorders of organic acid metabolism
New code		E72.820 Canavan disease
E74		Other disorders of carbohydrate metabolism
	E74.0	Glycogen storage disease
New sub-subcategory		E74.09 Other glycogen storage disease
		Andersen disease
Delete		~~Hers disease~~
Delete		~~Tauri disease~~
		Glycogen storage disease, types 0, IV, VI-XI
Add		Hers disease
		Liver phosphorylase deficiency
		Muscle phosphofructokinase deficiency
Add		Tauri disease
New code		E74.091 Polyglucosan body disease
Add		PGBD
New code		E74.098 Other glycogen storage disease

E74.8 Other specified disorders of carbohydrate metabolism

New sub-subcategory — E74.82 Disorders of sialic acid metabolism

Add — Excludes1: Sialidosis [mucolipidosis I] (E77.1)

New code — E74.820 Salla disease

New code — E74.821 Infantile sialic acid storage disease

New code — E74.822 Intermediate severe sialic acid storage disease

E75 Disorders of sphingolipid metabolism and other lipid storage disorders

E75.2 Other sphingolipidosis

New code — E75.27 Pelizaeus-Merzbacher disease

Add — PMD

E77 Disorders of glycoprotein metabolism

New sub-subcategory — E77.1 Defects in glycoprotein degradation

Aspartylglucosaminuria

Delete — ~~Fucosidosis~~

Mannosidosis

Sialidosis [mucolipidosis I]

New code — E77.10 Fucosidosis

New code — E77.18 Other defects in glycoprotein degradation

New code — E77.19 Unspecified defects in glycoprotein degradation

E78 Disorders of lipoprotein metabolism and other lipidemias

E78.7 Disorders of bile acid and cholesterol metabolism

New code — E78.73 Cerebrotendinous xanthomatosis

Add — CTX

E88 Other and unspecified metabolic disorders

E88.4 Mitochondrial metabolism disorders

New sub-subcategory — E88.43 Disorders of mitochondrial tRNA synthetases

New code — E88.431 Leukoencephalopathy with brainstem - spinal cord involvement - lactate elevation

Add — LBSL

New code		E88.432 Hypomyelination with brainstem - spinal cord involvement - leg spasticity
Add		HBSL
New code		E88.433 Leukoencephalopathy with thalamus - brainstem involvement - high lactate
Add		LTBL
New code		E88.438 Other disorders of mitochondrial tRNA synthetases
New code		E88.439 Other disorders of mitochondrial tRNA synthetases, unspecified
	G11	Hereditary ataxia
New code		G11.5 Hypomyelination - hypogonadotropic hypogonadism - hypodontia
Add		4H syndrome
Add		Pol III-related leukodystrophy
New code		G11.6 Leukodystrophy with vanishing white matter disease
	G23	Other degenerative diseases of basal ganglia
New code		G23.3 Hypomyelination with atrophy of the basal ganglia and cerebellum
Add		H-ABC
	G31	Other degenerative diseases of nervous system, not elsewhere classified
		G31.8 Other specified degenerative diseases of nervous system
New sub-subcategory		G31.89 Other specified degenerative diseases of nervous system
New code		G31.890 Alexander Disease
New code		G31.891 Pelizaeus-Merzbacher-like disease
Add		PMLD
New code		G31.898 Other specified degenerative diseases of the nervous system
New code		G31.899 Other specified degenerative diseases of the nervous system, unspecified

G90 Disorders of autonomic nervous system

New code G90.A LMNB1-related autosomal dominant leukodystrophy

G93 Other Disorders of the Brain

G93.4 Other and unspecified encephalopathy

New sub-subcategory G93.42 Leukoencephalopathy, non-infectious causes

New code G93.420 Chloride ion channel 2 (CIC-2) related leukoencephalopathy with intramyelinic oedema

New code G93.421 Megaloencephalic leukoencephalopathy with subcortical cysts

Add MLC

New code G93.422 Leukoencephalopathy with calcifications and cysts

New code G93.423 Adult-onset leukodystrophy with axonal spheroids

New code G93.424 RNASE T2-deficient cystic leukoencephalopathy

New code G93.428 Other leukoencephalopathy, non-infectious causes

New code G93.429 Other leukoencephalopathy, non-infectious causes unspecified

Q07 Other congenital malformations of nervous system

New code Q07.1 SOX-10 associated peripheral demyelinating neuropathy-central demyelinating leukodystrophy-Waardenburg syndrome-Hirschsprung disease

Add PCWH

Q12 Congenital lens malformations

New sub-subcategory Q12.0 Congenital cataract

New code Q12.01 Hypomyelination with congenital cataract

Add HCC

New code		Q12.08 Other congenital cataract
New code		Q12.09 Congenital cataract unspecified
	Q87	Other specified congenital malformation syndromes affecting multiple systems
New sub-subcategory		Q87.0 Other specified congenital malformation syndromes predominantly affecting facial appearance
New code		Q87.01 Oculo-dento-digital dysplasia
New code		Q87.08 Other specified congenital malformation syndromes predominantly affecting facial appearance
	Q93	Monosomies and deletions from the autosomes, not elsewhere classified
		Q93.8 Other deletions from the autosomes
New code		Q93.83 Deletions of the long arm of chromosome 18

Lumbar Degenerative Disc Disease With and Without Pain

The International Society for the Advancement of Spine Surgery (ISASS) is proposing the creation of new ICD-10-CM diagnosis codes for describing pain associated with lumbar and lumbosacral degenerative disc disease. The presence or absence of pain associated with degenerative disc disease in the low back is an important factor in clinical decision making in regard to selecting the appropriate treatment. Pain may be present in the low back, or may occur in the leg, or both. Absence of pain is generally a sign that the degenerative disc disease is non-noxious.

Low back pain or lumbago has 6 sources including discogenic, facetogenic, neurocompressive, sacroiliac, vertebrogenic, and psychogenic. The association between lumbar degenerative disc disease (LDDD) and low back pain (LBP) has been well established.[1,2]

Discogenic back pain associated with degenerative disc disease can be multifactorial and difficult to treat. The type of pain present and whether it is primarily low back pain or sciatica pain is an important component of the clinical assessment.

Treatments for discogenic back pain have ranged from anti-inflammatory medications to invasive procedures including spine fusion and spinal arthroplasty. There has also been a growing interest in developing strategies that aim to repair or regenerate the degenerated disc biologically, or to supplement tissue lost to degenerative disc disease.[3,4]

New ICD-10-CM codes that enable identification of pain present with lumbar and lumbosacral degenerative disc disease and enable the pain to be characterized as involving either the lumbar region only, the leg only, or both the back and leg will be of benefit for characterizing, tracking and improving treatments for this common and important clinical issue.

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TABULAR MODIFICATIONS

M51 Thoracic, thoracolumbar, and lumbosacral intervertebral disc disorders

M51.3 Other thoracic, thoracolumbar and lumbosacral intervertebral disc degeneration

M51.36 Other intervertebral disc degeneration, lumbar region

New code M51.360 Other intervertebral disc degeneration, lumbar region with lumbar back pain only

New code M51.361 Other intervertebral disc degeneration, lumbar region with leg pain only

New code M51.362 Other intervertebral disc degeneration, lumbar region with lumbar back pain and leg pain

New code M51.369 Other intervertebral disc degeneration, lumbar region without mention of lumbar back pain or leg pain

M51.37 Other intervertebral disc degeneration, lumbosacral region

New code M51.370 Other intervertebral disc degeneration, lumbosacral region with lumbar back pain

New code M51.371 Other intervertebral disc degeneration, lumbosacral region with leg pain only

New code M51.372 Other intervertebral disc degeneration, lumbosacral region with lumbar back pain and leg pain

New code M51.379 Other intervertebral disc degeneration, lumbosacral region without mention of lumbar back pain or leg pain

Metabolic Acidemia in Newborn

The National Center for Health Statistics (NCHS) received a request to revise P19.9 Metabolic acidemia, unspecified to include "in newborn". Currently "in newborn" is included in the category P19 Metabolic acidemia in newborn, but not in the P19.9 Metabolic acidemia, unspecified. Changes in the index are also proposed.

TABULAR MODIFICATIONS

P19 Metabolic acidemia in newborn
Includes: metabolic acidemia in newborn
P19.0 Metabolic acidemia in newborn first noted before onset of labor
P19.1 Metabolic acidemia in newborn first noted during labor
P19.2 Metabolic acidemia noted at birth
Revise P19.9 Metabolic acidemia in newborn, unspecified

INDEX MODIFICATIONS

Acidemia E87.2-
Revise - metabolic - see also Acidosis, metabolic ~~(newborn)~~ ~~P19.9~~
Add - - newborn P19.9
Revise - - - first noted before onset of labor P19.0
Revise - - - first noted during labor P19.1
Revise - - - noted at birth P19.2

Non-Traumatic Peritoneal Hemorrhage

Retroperitoneal hemorrhage is a particularly important site of occult or concealed hemorrhage.[1] 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.[1] 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%.[2] 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.[3]

The National Center for Health Statistics (NCHS) received a request to create ICD-10-CM codes for nontraumatic retroperitoneal hemorrhage and retroperitoneal fibrosis. This is a representation from the September 2021 ICD10 Coordination and Maintenance (C&M) meeting**;** edits are based on public comments and are **bolded**.

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.

The Agency for Healthcare Research and Quality (AHRQ) is requesting the creation of ICD-10-CM codes for non-traumatic peritoneal hemorrhage and retroperitoneal fibrosis for coding specificity and to improve quality of care for patients.

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TABULAR MODIFICATIONS

K66 Other disorders of peritoneum

K66.1 Hemoperitoneum
Add **Peritoneal hematoma**
Add **Peritoneal hemorrhage**

Add Excludes2: retroperitoneal hematoma (K68.3)
Add retroperitoneal hemorrhage (K68.**3**)

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 **Retroperitoneal hemorrhage**

INDEX MODIFICATIONS

Hematoma (traumatic) (skin surface intact) -see also Contusion
Revise - retroperitoneal (nontraumatic) ~~K66.1~~ K68.3

Revise - retroperitoneal ~~R58~~ **K68.3**

Syndrome -see also Disease
Revise - retroperitoneal fibrosis ~~N13.5~~ K68.2

Parkinson's Disease with OFF Episodes

This topic was presented at the September 2021 ICD10 Coordination and Maintenance (C&M) meeting. Based on feedback received during the public comment period, the proposal has been revised for consideration. Changes are noted in **bold**.

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. [1] 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.[2]

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.[3,4] Currently, no cure or disease-modifying therapies exist and treatment relies mainly upon levodopa to relieve motor and nonmotor symptoms.[5] 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.[2,6,7] Each patient's experience with PD is unique with some patients experiencing dyskinesias, OFF episodes, or both.[7-9]

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.[10,11] 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.[11-13] 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.[14-15] 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.[15]

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.[15,16] Response fluctuations may also present as nonmotor symptoms.[15,16] Non-motor symptoms that have been reported to occur during fluctuations include apathy, anxiety, irritability, mood changes, cognitive changes, fatigue, pain, and drenching sweats.[15,16]

Fluctuations may have a significant impact on patients.[17] 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.[18] 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.[18]

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.

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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TABULAR MODIFICATIONS

G20 Parkinson's disease
Hemiparkinsonism
Idiopathic Parkinsonism or Parkinson's disease
Paralysis agitans

Delete ~~Parkinsonism or Parkinson's disease NOS~~
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 **mention of** fluctuations
Add **Parkinson's disease NOS**
Add Parkinson's disease without dyskinesia, without **mention of** 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 **mention of** fluctuations
Add Parkinson's disease with dyskinesia, without **mention of** 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
Add **Excludes1: Parkinson's disease with dyskinesia (G20.B-)**
Add **Parkinson's disease without dyskinesia (G20.A-)**
Add **secondary parkinsonism (G21-)**

Problems Related to Upbringing

The American Academy of Pediatrics (AAP) has previously presented a proposal on Problems Related to Upbringing at the September 2019, March 2020 and September 2021 ICD10 Coordination and Maintenance (C&M) meetings. 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. Changes are noted in **bold**.

In addition, at the September 2021 C&M meeting AAP requested 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").

Today there are a greater variety of family dynamics that are more extended then 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.

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. It is the intent of the Academy that this revised proposal will better capture these expanded "family" dynamics and conflicts that can complicate a medical encounter.

TABULAR MODIFICATIONS

Z02 Encounter for administrative examination

Z02.8 Encounter for other administrative examinations

New code **Z02.84 Encounter for child welfare exam**

Add **Encounter for child welfare screening exam**

Add Excludes2: encounter for examination and observation for alleged child physical abuse (Z04.72)
encounter for examination and observation for alleged child rape (Z04.42)

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

Add **Child in welfare guardianship**

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

Z62.22 Institutional upbringing

Add **Child living in orphanage**

Add **Child living in group home**

Add **Code also, if applicable, child in welfare custody (Z62.21)**

New code Z62.23 **Child in custody of non-parental relative**

Add Child in care of non-parental family member

Add Child in custody of grandparent

Add Child in kinship care

Add **Guardianship by non-parental relative**

Add **Code also, if applicable, child in welfare custody (Z62.21)**

New code **Z62.24 Child in custody of non-relative guardian**

Add **Code also, if applicable, child in welfare custody (Z62.21)**

Z62.8 Other specified problems related to upbringing

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-)**
	Z62.82	Parent-child conflict
		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
New subcategory	Z62.83	Non-parental relative or guardian-child conflict
New code		Z62.831 Non-parental relative-child conflict
Add		Grandparent-child conflict
Add		Kinship-care child conflict
Add		Non-parental relative legal guardian-child conflict
Add		Other relative-child conflict
Add		**Excludes1: Group home staff-child conflict (Z62.833)**
New code		**Z62.832 Non-relative guardian-child conflict**
Add		**Excludes1: Group home staff-child conflict (Z62.833)**
New code		Z62.833 Group home staff-child conflict
	Z62.89	Other specified problems related to upbringing
New code		Z62.892 Runaway [from current living environment]
Add		Child leaving living situation without permission

Resistant Hypertension

Resistant hypertension (RH) is a condition where the blood pressure (BP) of a patient with hypertension remains above goal in spite of the concurrent use of at least three antihypertensive medications of different pharmacologic classes, commonly including a long-acting calcium channel blocker, a blocker of the renin-angiotensin system (angiotensin converting enzyme inhibitor or angiotensin receptor blocker) and a diuretic.[1] The definition of RH stipulates that all pharmacologic agents should be administered at maximum or maximally tolerated doses and at the appropriate dosing interval. RH also includes patients who achieve their BP target levels on four or more antihypertensive medications, a condition termed "controlled RH". Thus, the designation of RH refers to patients with both uncontrolled and controlled hypertension depending on the number of antihypertensive agents administered.[1] True RH is defined as RH in which the causes of pseudo-resistance, as discussed below, have been excluded; when any of the causes of pseudo-resistance cannot be ruled out, the term "apparent treatment resistant hypertension (aTRH)" is applied.

Pseudo-resistance can result in a misdiagnosis of RH, due to error in BP measurement, the "white coat effect," or medication non-adherence. Inaccurate BP measurement may result from improper preparation of the patient, non-ideal environmental conditions, incorrect cuff size and improper measurement technique. To minimize BP variability, diagnostic BP recordings should include an average of ≥2 readings obtained on ≥2 separate occasions. Therefore, before diagnosis of RH accurate BP measurement is imperative. Similarly, out-of-office BP monitoring including home BP monitoring (HBPM) requires use of correct technique. The "white coat effect" is defined as having treated office BP above goal, but out-of-office BP by ambulatory blood pressure monitoring (ABPM) (or HBPM) at goal, in a patient taking ≥3 antihypertensive agents. The risk of cardiovascular disease (CVD) complications in patients with the white coat effect is similar to the risk in patients with controlled hypertension. Accurate BP measurement sharply reduces but does not totally eliminate the white coat effect. Out-of-office BP monitoring is required to make the diagnosis of true RH. Medication non-adherence is highly prevalent in patients with aTRH. This is in part due to the large pill burden, dosing complexity, expense and high frequency of adverse reactions that may occur with multi-drug antihypertensive regimens. Exclusion of non-adherence includes frank and nonjudgmental clinician-patient discussion and monitoring of the most recent prescription drug refills, pill counts, and, if available, biochemical assay of drugs or metabolites in urine or plasma.

RH, as defined above, identifies patients who are at significantly higher risk for target organ damage, morbid CVD events, end-stage kidney disease and death compared with hypertensive patients without treatment resistance.[1,2] In addition, patients with RH are much more likely to have medication adverse effects or a secondary cause for their hypertension compared with patients with hypertension without drug resistance.[1] Patients with RH require special expertise for careful assessment and may benefit from special diagnostic and/or therapeutic approaches to control their BP.

In reporting the prevalence of RH, the term 'apparent' treatment resistant hypertension (aTRH) has been employed when one or more of the following data elements is (are) missing: medication

dose, level of adherence, or out-of-office BP. Among treated adults with hypertension, aTRH occurs in approximately 12% -15% of population-based and 15%-18% of clinic-based reports.[1,3] In a recent study of RH prevalence using the BP cutoff of <140/90 mm Hg compared to the more recently adopted cutoff of <130/80 mm Hg to define BP control, 17.7 and 19.7% of patients, respectively, were estimated to have aTRH.[4]

RH is not a new diagnostic term and has been recognized since the 1980s. It was highlighted as a distinct clinical entity in the Seventh Report of the Joint National Committee on the Prevention, Detection, Evaluation and Treatment of High Blood Pressure (JNC-7) in 2003.[5] The first American Heart Association (AHA) Scientific Statement on Resistant Hypertension, published in 2008, provided a precise definition of RH (as given above).[6] The 2008 Scientific Statement codified the various causes of pseudo-resistance and reviewed lifestyle factors and secondary causes resulting in drug resistance. The document recommended the most appropriate modification steps in pharmacologic treatment and indications for hypertension specialist referral if BP control could not be achieved. The updated 2018 AHA Scientific Statement[1] definition of RH differed from that in the 2008 document in 3 important ways: (1) BP should be measured and the BP threshold for diagnosis and treatment goals should be in accord with current clinical practice guidelines[7]; (2) patients with the white-coat effect should not be classified as having RH; and (3) the diagnosis of RH requires exclusion of antihypertensive medication non-adherence. These criteria distinguish pseudo-resistance, which can be managed with accurate BP monitoring and improved medication adherence, from true RH which warrants further evaluation and adjustments of therapy.

An essentially identical definition of RH was published by the 2018 European Society of Cardiology/European Society of Hypertension and the 2020 International Society of Hypertension (ISH) clinical practice guidelines.[8,9] The ISH document further recommended that, because of its complexity, RH should be managed in a hypertension specialty center.[9]

RH is a complex disorder necessitating special clinical expertise and devotion of additional time and effort to validate the diagnosis, assess the extent of target organ damage, determine potential specific causes and contributing factors and recommend changes in the treatment regimen that will enhance the chance of success in lowering BP to goal.[1] Explicit specialized knowledge is requisite to conduct ABPM and accurately interpret its results (to exclude the white coat effect or detect masked uncontrolled hypertension) and to train patients to perform accurate HBPM for monitoring the response to pharmacologic dose-titration. Detection and reversal of suboptimal medication adherence also requires special expertise, time and sensitivity to the barriers to optimal adherence. Secondary hypertension is a frequent cause of RH demanding knowledge of the various drug classes and disorders that can cause treatment resistance (e.g., sleep disorders, renal parenchymal and vascular disease, endocrine disorders such as primary aldosteronism, Cushing's syndrome, pheochromocytoma and other endocrine disorders). If a specific cause of RH cannot be identified during a systematic search, treatment usually entails a combination of lifestyle modification (diet, sodium and potassium intake, exercise and limitation of alcohol consumption) and drug titration steps, initiated on the basis of the most likely pathophysiologic mechanisms of the RH in each individual patient.[1]

The 2018 AHA Scientific Statement[1] presented a new evidence-based template for the therapeutic sequence in RH. Regardless of the particular antihypertensive agents employed, intensive BP lowering is superior to standard treatment in terms of CVD outcomes in RH.[10] Regarding the prognosis of RH using the new 2018 AHA guideline BP goal of <130/80 mm Hg, a large RH cohort study from Korea recently demonstrated that the risk for major adverse CVD events and adverse kidney outcomes is similar under the 2018 AHA as compared with earlier definitions of RH with no significant difference for predicting major adverse CVD events.[11,12]

To summarize, based on the complexity of accurately diagnosing and effectively treating RH, and requirements for expertise in conducting out-of-office BP monitoring (i.e. ABPM and HBPM), documentation of target organ damage, identification of secondary causes of and contributing factors to resistance, institution of rigorous lifestyle modification and intricate pharmacological management, the American Heart Association Hypertension Council requests consideration of a new ICD-10 code for RH separate from the existing codes for hypertension. The American Heart Association Hypertension Council *Ad Hoc* Writing Committee includes Robert M. Carey, MD, MACP, Chair; George L. Bakris, MD; Jan N. Basile, MD; John Flack, MD; Daiichi Shimbo, MD; Sandra J. Taler, MD; and Lillie Noe, Council Manager.

A specific code for RH will enable specific identification of patients with RH within the general hypertensive population; help to overcome existing suboptimal antihypertensive therapy; enable proper evaluation of the contributing factors/causes of true RH, including lifestyle factors and secondary causes that may be reversible with specific treatment; and enable optimal BP control, thus improving cardiovascular and renal morbidity and mortality. In addition, clear identification and categorization of persons with RH with subsequent optimal treatment will help to alleviate certain existing racial health disparities in BP management.

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TABULAR MODIFICATIONS

New Category	I1A	Other hypertension	
New code		I1A.0	Resistant hypertension
Add			Apparent treatment resistant hypertension
Add			True resistant hypertension
Add			Code first specific type of existing hypertension, if known, such as:
Add			essential hypertension (I10)
Add			secondary hypertension (I15.-)

Sickle-Cell Dactylitis and Vaso-Occlusive Crisis

Vaso-occlusive crisis is the most frequent reason for inpatient care of children with sickle cell anemia. Acute episodes of pain, also commonly referred to as sickle cell pain crises, or vaso-occlusive crises (VOCs), are not only the primary presenting morbidity associated with sickle cell disease, (SCD) but also the cause of hospitalization in approximately 95% of cases." (Darbarai 2020).

This diagnosis of sickle cell vasoocclusive crisis can result in a prolonged hospital stay with intractable pain, and lead to development of a chronic pain syndrome. Difficulties in pain control may potentiate the risk of pneumonia, acute chest syndrome, and even acute respiratory failure. The frequency and severity of vaso-occlusive crisis may be predictive of developing other life-threatening complications of sickle cell disease including splenic sequestration crisis, pulmonary hypertension, and stroke.

This proposal, submitted by the Regulatory Committee of the Association of Clinical Documentation Integrity Specialists (ACDIS) propose changes to ICD-10-CM to enhance the specificity of reporting for sickle cell disease. It is being requested that (1) unique codes be created for sickle cell disease with dactylitis for various types of sickle cell disease and (2) making the term vaso-occlusive crisis a non-essential modifier in the Index and Tabular entries for sickle cell disease.

Dactylitis is a severe inflammation of the fingers and toes commonly seen in infants with sickle cell anemia. In the pre-verbal child, it may be the only clinical indication of vaso-occlusive pain crisis. Early recognition of dactylitis and care for the underlying condition helps prevent later complications of sickle cell disease.

Dactylitis is not currently able to be specified within the ICD-10-CM code set. ICD-10-CM is clear in promoting additional specificity in clinical coding when such specificity is possible; the potential for further specificity exists in the reporting of manifestations of sickle cell disease. As early dactylitis has been suggested as a factor suggesting more severe disease, accurate reporting of the nature and frequency of dactylitis as a manifestation of acute sickle cell vaso-occlusive crisis may guide clinicians to early intervention to prevent long-term complications, morbidity, and mortality from sickle cell disease.

ACDIS proposes that "vaso-occlusive" be made a non-essential modifier for sickle cell disease with pain. It is of the opinion of ACDIS, that this request is supported by review of hospitalization documentation that demonstrates that the documentation of 'pain' in SCD patients is commonly not further described as 'vaso-occlusive' pain. Clinically, pain in a patient treated for sickle cell disease is vaso-occlusive. As reported by Darabi et. al., "Acute episodes of pain, also commonly referred to as sickle cell pain crises, or vaso-occlusive crises (VOCs), are not only the primary presenting morbidity associated with SCD.

The enhanced specificity promoted by these ICD-10-CM codes will enable improved tracking of patient severity of illness and promote a more accurate assessment of the need and intensity of care, in patients with sickle cell disease.

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TABULAR MODIFICATIONS

D57 Sickle-cell disorders
Use additional code for any associated fever (R50.81)
Excludes1: other hemoglobinopathies (D58.-)

D57.0 Hb-SS disease with crisis
Sickle-cell disease with crisis
Revise Hb-SS disease with (vaso-occlusive) pain

D57.00 Hb-SS disease with crisis, unspecified
Hb-SS disease with (painful) crisis NOS
Revise Hb-SS disease with (vaso-occlusive) pain NOS

New code D57.04 Hb-SS disease with dactylitis

D57.09 Hb-SS disease with crisis with other specified complication

Revise ~~Use additional code~~ Code also, if applicable to identify complications, such as:
cholelithiasis (K80.-)
priapism (N48.32)

D57.2 Sickle-cell/Hb-C disease
Hb-SC disease
Hb-S/Hb-C disease

D57.21 Sickle-cell/Hb-C disease with crisis

New code
D57.214 Sickle-cell/Hb-C disease with dactylitis

D57.219 Sickle-cell/Hb-C disease with crisis, unspecified
Sickle-cell/Hb-C disease with crisis NOS

Revise
Sickle-cell/Hb-C disease with <u>(vaso-occlusive)</u> pain NOS

D57.4 Sickle-cell thalassemia
Sickle-cell beta thalassemia
Thalassemia Hb-S disease

D57.41 Sickle-cell thalassemia, unspecified, with crisis
Sickle-cell thalassemia with (painful) crisis NOS

Revise
Sickle-cell thalassemia with <u>(vaso-occlusive)</u> pain NOS

New code
D57.414 Sickle-cell thalassemia, unspecified, with dactylitis

D57.418 Sickle-cell thalassemia, unspecified, with crisis with other specified complication

Revise
~~Use additional code~~ <u>Code also, if applicable</u> to identify complications, such as:
cholelithiasis (K80.-)
priapism (N48.32)

D57.419 Sickle-cell thalassemia, unspecified, with crisis
Sickle-cell thalassemia with (painful) crisis NOS

Revise
Sickle-cell thalassemia with <u>(vaso-occlusive)</u> pain NOS

D57.43 Sickle-cell thalassemia beta zero with crisis
HbS-beta zero with crisis
Sickle-cell beta zero with crisis

New code
D57.434 Sickle-cell thalassemia beta zero with dactylitis

D57.438 Sickle-cell thalassemia beta zero with crisis with other specified complication
HbS-beta zero with other specified complication
Sickle-cell beta zero with other specified complication

Revise

~~Use additional code~~ Code also, if applicable to identify complications, such as:
cholelithiasis (K80.-)
priapism (N48.32)

D57.439 Sickle-cell thalassemia beta zero with crisis, unspecified
HbS-beta zero with other specified complication
Sickle-cell beta zero with crisis unspecified
Sickle-cell thalassemia beta zero with (painful) crisis NOS

Revise

Sickle-cell thalassemia beta zero with (vaso-occlusive) pain NOS

D57.45 Sickle-cell thalassemia beta plus with crisis
HbS-beta plus with crisis
Sickle-cell beta plus with crisis

New code

D57.454 Sickle-cell thalassemia beta plus with dactylitis

D57.458 Sickle-cell thalassemia beta plus with crisis with other specified complication
HbS-beta plus with crisis with other specified complication
Sickle-cell beta plus with crisis with other specified complication

Revise

~~Use additional code~~ Code also, if applicable to identify complications, such as:
cholelithiasis (K80.-)
priapism (N48.32)

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

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

Revise — Sickle-cell thalassemia beta plus with (vaso-occlusive) pain NOS

D57.8 Other sickle-cell disorders
Hb-SD disease
Hb-SE disease

D57.81 Other sickle-cell disorders with crisis

New code — D57.814 Other sickle-cell disorders with dactylitis

D57.818 Other sickle-cell disorders with crisis with other specified complication

Revise — ~~Use additional code~~ Code also, if applicable to identify complications, such as:
cholelithiasis (K80.-)
priapism (N48.32)

D57.819 Other sickle-cell disorders with crisis, unspecified
Other sickle-cell disorders with crisis NOS

Revise — Other sickle-cell disorders with (vaso-occlusive) pain NOS

INDEX MODFICATIONS

Disease
- sickle-cell D57.1
-- with
--- crisis(painful) D57.00

Add ---- with dactylitis D57.04
Add --- dactylitis D57.04
Revise --- ~~vasoocclusive~~ pain (vaso-occlusive) D57.00
Add -- priaprism D57.09
-- Hb-C D57.20
--- with
---- crisis D57.219
Add ----- with dactylitis D57.214
Add ---- dactylitis D57.214

Revise	---- ~~vasoocclusive~~ pain (vaso-occlusive) D57.219
Add	-- priaprism D57.218
	-- Hb-SD D57.80
	--- with
	---- crisis D57.819
Add	----- with dactylitis D57.814
Add	---- dactylitis D57.814
Revise	---- ~~vasoocclusive~~ pain (vaso-occlusive) D57.819
	-- Hb-SE D57.80
	--- with
	---- crisis D57.819
Add	----- with dactylitis D57.814
Add	---- dactylitis D57.814
Revise	---- ~~vasoocclusive~~ pain (vaso-occlusive) D57.819
	-- specified NEC D57.80
	--- with
	---- crisis D57.819
Add	----- with dactylitis D57.814
Add	---- dactylitis D57.814
Revise	---- ~~vasoocclusive~~ pain (vaso-occlusive) D57.819
	-- thalassemia D57.40
	--- with
	---- acute chest syndrome D57.411
Add	----- with dactylitis D57.414
	----- with specified complication NEC D57.418
Add	---- dactylitis D57.414
Revise	---- ~~vasoocclusive~~ pain (vaso-occlusive) D57.419
	--- beta plus D57.44
	---- with
	----- acute chest syndrome D57.451
Add	------with dactylitis D57.454
Add	-----dactylitis D57.454
Revise	-----~~vasoocclusive~~ pain (vaso-occlusive) D57.459
	--- beta zero D57.42
	---- with
	----- acute chest syndrome D57.431
Add	------ with dactylitis D57.434
Add	---- dactylitis D57.434
Revise	----- ~~vasoocclusive~~ pain (vaso-occlusive) D57.439

Social Determinants of Health

This proposal was originally submitted by the Gravity Project (GP) and presented at the March 2021 and September 2021 ICD10 Coordination and Maintenance meetings. Parts of the proposal were previously approved and will be implemented on October 1, 2022. Subsequently, we have received additional code requests from the GP and AHIMA that have been incorporated in the proposal.

The Gravity Project requests a revision of the original request for Z59.87 Material Hardship and requests that it be titled Z59.87 Material hardship, due to limited financial resources to clarify economics as the driver. Furthermore, a request for a new term under Z58 to cover basic necessities unavailable in the environment, Z58.81 Material hardship, inadequate physical environment.

The Gravity Project's continues in collaboration with the US Department of Veteran Affairs the need to clarify for a distinct code separate from Z91.82 Personal history of military deployment. The risks of deployment and service are distinct and need to be independently identified.

The Gravity Project worked with international experts on social connection over the course of 2021. Through that work, they identified missing concepts across the types of lack of social connection - social isolation, loneliness, and lack of social support. Social isolation is adequately covered in the taxonomy. However, to allow for consistent documentation of "loneliness" and "lack of social support" they are requesting for a "loneliness" inclusion term under R45.89 Other symptoms and signs involving emotional stateand "lack of emotional support" under Z60.8 Other problems related to social environment.

The Gravity Project requests specific codes for intimate partner violence (IPV) (confirmed and suspected.) At present, to represent IPV across the age spectrum, one would have to code adult and child abuse codes across the abuse subtypes (neglect, physical, psychological, sexual exploitation, etc.)

The Gravity Project is requesting two areas of Elder Abuse terms. All terms align with the Center for Disease Control and Prevention 2016 publication "Elder Abuse Surveillance: Uniform Definitions and Recommended Core Data Elements." Elder Abuse is defined as "An intentional act or failure to act by a caregiver or another person in a relationship involving an expectation of trust that causes or creates a risk of harm to an older adult and can be in the form of physical abuse, psychological abuse, sexual abuse, financial abuse, and neglect by someone in a caregiving role. (The Gravity Project, 2021)"

The previously proposed Y07.2 Acquaintance or friend, perpetrator of maltreatment and neglect presented at the September 2021 meeting is requested to be placed at the Y07.5 Non-family member, perpetrator of maltreatment and neglect. The is also a need for financial abuse terms (confirmed and suspected.) The Gravity Project requests missing critical perpetrator

codes to align with the CDC core data elements and the literature of elder abuse.

TABULAR MODIFICATIONS

R45 Symptoms and signs involving emotional state
R45.8 Other symptoms and signs involving emotional state
R45.89 Other symptoms and signs involving emotional state

Add — Flat affect
Add — Loneliness

T74 Adult and child abuse, neglect, and other maltreatment, confirmed
Use additional code, if applicable, to identify any associated current injury
The appropriate 7th character is to be added to each code from category T74
A initial encounter
D subsequent encounter
S sequela

T74.1 Physical abuse, confirmed
New code — T74.13 Intimate partner physical abuse, confirmed

T74.2 Sexual abuse, confirmed
New code — T74.23 Intimate partner sexual abuse, confirmed

T74.3 Psychological abuse, confirmed
New code — T74.33 Intimate partner psychological abuse, confirmed

New subcategory — T74.A Financial abuse, confirmed
New code — T74.A1 Adult financial abuse, confirmed

T76 Adult and child abuse, neglect, and other maltreatment, suspected
Use additional code, if applicable, to identify any associated current injury

The appropriate 7th character is to be added to each code from category T76
A initial encounter
D subsequent encounter
S sequela

T76.1 Physical abuse, suspected
New code — T76.13 Intimate partner physical abuse, suspected

T76.2 Sexual abuse, suspected
New code T76.23 Intimate partner sexual abuse, suspected

T76.3 Psychological abuse, suspected
New code T76.33 Intimate partner psychological abuse, suspected

New subcategory T76.A Financial abuse, suspected
New code T76.A1 Adult financial abuse, suspected

Z58 Problems related to physical environment
New subcategory Z58.8 Other problems related to physical environment
New code Z58.81 Material hardship, inadequate physical environment
Add Unable to obtain electricity, due to inadequate physical environment
Add Unable to obtain internet service, due to inadequate physical environment

New code Z58.89 Other problems related to physical environment

Z59 Problems related to housing and economic circumstances
Z59.8 Other problems related to housing and economic circumstances

New code Z59.87 Material hardship, due to limited financial resources, NEC
Add Material deprivation
Add Unable to obtain adequate childcare due to limited financial resources
Add Unable to obtain adequate clothing due to limited financial resources
Add Unable to obtain adequate utilities due to limited financial resources
Add Unable to obtain basic needs, due to limited financial resources

Z60 Problems related to social environment
Z60.8 Other problems related to social environment
Add Inadequate social support
Add Lack of emotional support

Z65 Problems related to other psychosocial circumstances
Z65.8 Other specified problems related to psychosocial circumstances
Add At risk for loneliness

Z91 Personal risk factors, not elsewhere classified
Z91.1 Patient's noncompliance with medical treatment and regimen
New sub subcategory Z91.14 Patient's other noncompliance with medication regimen
Patient's underdosing of medication NOS
New code Z91.141 Patient’s other noncompliance with medication regimen due to financial hardship
New code Z91.148 Patient’s other noncompliance with medication regimen for other reason
New sub subcategory Z91.15 Patient's noncompliance with renal dialysis
New code Z91.151 Patient's noncompliance with renal dialysis due to financial hardship
New code Z91.158 Patient's noncompliance with renal dialysis for other reason

Z91.4 Personal history of psychological trauma, not elsewhere classified
Z91.41 Personal history of adult abuse
New code Z91.413 Personal history of adult financial abuse

Z91.A Caregiver’s noncompliance with patient’s medical treatment and regimen
New sub subcategory Z91.A4 Caregiver's other noncompliance with patient’s medication regimen
Caregiver’s underdosing with patient’s medication NOS
New code Z91.A41 Caregiver's other noncompliance with patient’s medication regimen due to financial hardship
New code Z91.A48 Caregiver's other noncompliance with patient’s medication regimen for other reason

New sub subcategory Z91.A5 Caregiver's noncompliance with patient’s renal dialysis
New code Z91.A51 Caregiver's noncompliance with patient’s renal dialysis due to financial hardship
New code Z91.A58 Caregiver's noncompliance with patient’s renal dialysis for other reason

New sub subcategory	Z91.A9	Caregiver's noncompliance with patient's other medical treatment and regimen	
Add		Nonadherence to medical treatment	
New code		Z91.A91	Caregiver's noncompliance with patient's other medical treatment and regimen due to financial hardship
New code		Z91.A98	Caregiver's noncompliance with patient's other medical treatment and regimen for other reason
	Z91.8	Other specified personal risk factors, not elsewhere classified	
	Z81.82	Personal history of military deployment Individual (civilian or military) with past history of military war, peacekeeping and humanitarian deployment (current or past conflict) Returned from military deployment	
Add		Excludes2: personal history of military service (Z91.85)	
New code	Z91.85	Personal history of military service	
Add		Excludes2:	personal history of military deployment (Z91.82)
Add			served in the armed forces
Add			veteran
	Z91.89	Other specified personal risk factors, not elsewhere classified	
Add		Increased risk for social isolation	

Shwachman-Diamond Syndrome

Shwachman-Diamond syndrome (SDS) is a genetic multi-system disorder characterized by bone marrow failure, exocrine pancreatic dysfunction, and predisposition to myeloid malignancies.[1] The estimated incidence of SDS is 1:76,000.[2] With a reduced life expectancy of about 40 years (estimated), there are expected to be several thousand patients in the US alone. It is anticipated that substantially more individuals will be identified as genetic and biochemical testing becomes more widely adopted.

The diagnosis of SDS is established with the classic clinical findings of exocrine pancreatic insufficiency and bone marrow failure and/or identification of biallelic pathogenic variants in *DNAJC21*, *EFL1*, or *SBDS,* or a heterozygous pathogenic variant in *SRP54* by molecular genetic testing.[3,4] Biallelic mutations in *SBDS* account for over 90% of cases of SDS,[5] while the other genes, *EFL1*,[6] *DNAJC21*,[7] and *SRP54*[8] account only for a small percentage of cases. For close to 10% of cases, no genetic cause has yet been identified. All the genes associated with SDS are involved in the ubiquitous pathway of ribosome biogenesis and function, and thus are necessary for the ribosomal subunits to assemble into translationally competent ribosomes and enable adequate levels of translation.[3,4]

The major cause of mortality from SDS are hematological complications, such as severe bone marrow failure, myelodysplastic syndrome (MDS) and acute myeloid leukemia (AML), which are estimated to occur in about 30% of patients with biallelic *SBDS* mutations by age 30.[3,4,9,10] This risk has not been confirmed in SDS patients without biallelic *SBDS* mutations, emphasizing the need to distinctly demark *SBDS* cases from other gene variants resulting in the SDS phenotype.

Clinically, the distinction of the various genetic causes of SDS is important as different underlying genetic causes may give rise to different natural histories, symptoms, and phenotypes and directly impact patient anticipatory counseling and clinical monitoring. Additionally, patients in the different categories will require different management and treatment strategies for optimal outcomes including lifesaving interventions, especially with precision medicine approaches and gene targeting therapies currently in development.

Creation of a new code would enable more readily tracking morbidity rates, hospital admissions, and treatment outcomes for SDS, and will be of benefit for clinical and policy research efforts. The Shwachman-Diamond Syndrome Alliance, a nonprofit patient advocacy organization, formally requests that an ICD-10-CM code be created for Shwachman-Diamond Syndrome (SDS).

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TABULAR MODIFICATIONS

D61 Other aplastic anemias and other bone marrow failure syndromes

D61.0 Constitutional aplastic anemia

New code	D61.02	Shwachman-Diamond Syndrome
Add		Code also, if applicable, associated conditions such as:
Add		acute myeloblastic leukemia (C92.0-)
Add		exocrine pancreatic insufficiency (K86.81)
Add		myelodysplastic syndrome (D46.-)
Add		Use additional code, if applicable, for genetic susceptibility to other malignant neoplasm (Z15.09)

Wasting Disease (Syndrome) Due to Underlying Condition

The National Center for Health Statistics (NCHS) received a request to create an ICD-10-CM code for wasting disease (syndrome) due to underlying condition. This will improve coding specificity and aid in capturing severity of illness for morbidity of the underlying conditions and help with improved treatments for these conditions.

Wasting disease (syndrome) is an involuntary, on-going loss of more than 10% of body weight with reduction in muscle mass, with or without loss of fat due to underlying condition. The manifestations of the disease occur in multiple conditions as an indicator of end-stage progression and complicate those concurrent conditions.

Wasting disease (syndrome) is a metabolic-catabolic syndrome that is a severe complication of a chronic, primary disease. It has a constellation of signs and symptoms and is a manifestation signaling the later end-stage or morbidity of an underlying condition and is typically irreversible.

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TABULAR MODIFICATIONS

E88 Other and unspecified metabolic disorders

E88.8 Other specified metabolic disorders

New code E88.A Wasting disease (syndrome) due to underlying condition

Add Code first underlying condition

TABULAR MODIFICATIONS PROPOSED ADDENDA

All proposed effective October 1, 2023

Diseases of liver (K70-K77)

Excludes2: hemochromatosis (E83.11-)
Reye's syndrome (G93.7)
viral hepatitis (B15-B19)
Revise Wilson's disease (E83.01)

D64 Other anemias
D64.2 Secondary sideroblastic anemia due to drugs and toxins

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

D70 Neutropenia

Use additional code for any associated:
fever (R50.81)
Delete ~~mucositis (J34.81, K12.3-, K92.81, N76.81)~~

Add Code also, if applicable, mucositis (J34.81, K12.3-, K92.81, N76.81)

D77 Other disorders of blood and blood-forming organs in diseases classified elsewhere

Code first underlying disease, such as:
Revise congenital early syphilis (A50.0-)

E03 Other hypothyroidism
E03.2 Hypothyroidism due to medicaments and other exogenous substances

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

E09 Drug or chemical induced diabetes mellitus

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

E35 Disorders of endocrine glands in diseases classified elsewhere

Revise Code first underlying disease, such as: late congenital syphilis of thymus gland [Dubois disease] (A50.5-)

E87 Other disorders of fluid, electrolyte and acid-base balance
Excludes1: diabetes insipidus (E23.2)
Add metabolic acidemia in newborn, unspecified (P19.9)

F02 Dementia in other diseases classified elsewhere
Code first the underlying physiological condition, such as:
Revise hepatolenticular degeneration (E83.01)

F05 Delirium due to known physiological condition
Revise Code first the underlying physiological condition such as:
Dementia (F03.9-)

F11 Opioid related disorders
F11.1 Opioid abuse
F11.18 Opioid abuse with other opioid-induced disorder
F11.188 Opioid abuse with other opioid-induced disorder
Add Opioid-associated amnestic syndrome with opioid abuse

F11.2 Opioid dependence
F11.28 Opioid dependence with other opioid-induced disorder
F11.288 Opioid dependence with other opioid-induced disorder
Add Opioid-associated amnestic syndrome with opioid dependence

F11.9 Opioid use, unspecified
F11.98 Opioid use, unspecified with other specified opioid-induced disorder
F11.988 Opioid use, unspecified with other opioid-induced disorder
Add Opioid-associated amnestic syndrome without use disorder

F33 Major depressive disorder, recurrent
F33.8 Other recurrent depressive disorders
Recurrent brief depressive episodes
Seasonal affective disorder

Code first the underlying physiological condition, such as:
Revise hepatolenticular degeneration (E83.01)

F50 Eating disorders
Revise Excludes2: feeding difficulties (R63.3-)

F98 Other behavioral and emotional disorders with onset usually occurring in childhood and adolescence
F98.2 Other feeding disorders of infancy and childhood

Excludes2: anorexia nervosa and other eating disorders (F50.-)
Revise feeding difficulties (R63.3-)

G04 Encephalitis, myelitis and encephalomyelitis

Delete ~~Excludes1: acute transverse myelitis (G37.3-)~~

Add Excludes2: acute transverse myelitis (G37.3-)

G05 Encephalitis, myelitis and encephalomyelitis in diseases classified elsewhere
G05.3 Encephalitis and encephalomyelitis in diseases classified elsewhere

Add Code also, if applicable, underlying disease

G73 Disorders of myoneural junction and muscle in diseases classified elsewhere
G73.7 Myopathy in diseases classified elsewhere

Code first underlying disease, such as:
Revise glycogen storage disease (E74.0-)

G92 Toxic encephalopathy
G92.8 Other toxic encephalopathy
Revise Code first poisoning due to drug or toxin, if applicable, (T36-T65 with fifth or sixth character 1-4 ~~or 6~~)

G92.9 Unspecified toxic encephalopathy
Revise Code first poisoning due to drug or toxin, if applicable, (T36-T65 with fifth or sixth character 1-4 ~~or 6~~)

G96 Other disorders of central nervous system
G96.0 Cerebrospinal fluid leak
G96.08 Other cranial cerebrospinal fluid leak
Postoperative cranial cerebrospinal fluid leak
Traumatic cranial cerebrospinal fluid leak
Code also if applicable:
Revise head injury ~~(S00.- to S09.-)~~ (S00 – S09)

G96.09 Other spinal cerebrospinal fluid leak

Code also if applicable:
Revise head injury ~~(S00.- to S09.-)~~ (S00 – S09)

G99 Other disorders of nervous system in diseases classified elsewhere
G99.8 Other specified disorders of nervous system in diseases classified elsewhere

Code first underlying disorder, such as:
Revise avitaminosis (E56.~~9~~ -)

H35 Other retinal disorders
H35.3 Degeneration of macula and posterior pole
H35.38 Toxic maculopathy
Revise Code first poisoning due to drug or toxin, if applicable (T36-T65 with fifth or sixth character 1-4 ~~or 6~~)

H54 Blindness and low vision Note: For definition of visual impairment categories see table below
H54.5 Low vision, one eye
Visual impairment categories 1 or 2 in one eye [normal vision in other eye]
H54.51 Low vision, right eye, normal vision left eye
Revise H54.511 Low vision, right eye, category 1~~,2~~
H54.511A Low vision right eye category 1, normal vision left eye
New subcategory H54.512 Low vision, right eye, category 2
H54.512A Low vision right eye category 2, normal vision left eye

H62 Disorders of external ear in diseases classified elsewhere
H62.4 Otitis externa in other diseases classified elsewhere
Code first underlying disease, such as:

Revise impetigo (L01.0-)

H91 Other and unspecified hearing loss
H91.0 Ototoxic hearing loss
Revise Code first poisoning due to drug or toxin, if applicable (T36-T65 with fifth or sixth character 1-4 ~~or 6~~)

I26 Pulmonary embolism
Add Excludes1: cor pulmonale without embolism (I27.81)

I27 Other pulmonary heart diseases
I27.8 Other specified pulmonary heart diseases
I27.81 Cor pulmonale (chronic)
Add Code also, if applicable, Right heart failure (I50.81-)

I42 Cardiomyopathy
I42.7 Cardiomyopathy due to drug and external agent
Revise Code first poisoning due to drug or toxin, if applicable (T36-T65 with fifth or sixth character 1-4 ~~or 6~~)

I60 Nontraumatic subarachnoid hemorrhage
Add Use additional code, if known, to indicate National Institutes of Health Stroke Scale (NIHSS) score (R29.7-)

I61 Nontraumatic intracerebral hemorrhage
Add Use additional code, if known, to indicate National Institutes of Health Stroke Scale (NIHSS) score (R29.7-)

I62 Other and unspecified nontraumatic intracranial hemorrhage
Add Use additional code, if known, to indicate National Institutes of Health Stroke Scale (NIHSS) score (R29.7-)

I63 Cerebral infarction
Add Excludes2: chronic, without residual deficits (sequelae) Z86.73

I67 Other cerebrovascular diseases
Add Excludes1: Occlusion and stenosis of cerebral artery causing cerebral infarction (I63.3-I63.5-)
Add Occlusion and stenosis of precerebral artery causing cerebral infarction (I63.2-)

I67.4 Hypertensive encephalopathy

Add
Code also, if applicable, associated hypertensive conditions such as:
essential (primary) hypertension (I10)
hypertensive heart disease (I11)
hypertensive chronic kidney disease (I12)
hypertensive heart and chronic kidney disease (I13)

I87 Other disorders of veins

I87.2 Venous insufficiency (chronic) (peripheral)

Add
Use additional, if applicable, code to specify site and severity of ulcer (L97.-)

J31 Chronic rhinitis, nasopharyngitis and pharyngitis

Delete
~~Use additional code to identify:~~
~~exposure to environmental tobacco smoke (Z77.22)~~
~~exposure to tobacco smoke in the perinatal period (P96.81)~~
~~history of tobacco dependence (Z87.891)~~
~~occupational exposure to environmental tobacco smoke (Z57.31)~~
~~tobacco dependence (F17.-)~~
~~tobacco use (Z72.0)~~

J32 Chronic sinusitis

Delete
~~Use additional code to identify:~~
~~exposure to environmental tobacco smoke (Z77.22)~~
~~exposure to tobacco smoke in the perinatal period (P96.81)~~
~~history of tobacco dependence (Z87.891)~~
~~occupational exposure to environmental tobacco smoke (Z57.31)~~
~~tobacco dependence (F17.-)~~
~~tobacco use (Z72.0)~~

J33 Nasal polyp

Delete
~~Use additional code to identify:~~
~~exposure to environmental tobacco smoke (Z77.22)~~
~~exposure to tobacco smoke in the perinatal period (P96.81)~~
~~history of tobacco dependence (Z87.891)~~
~~occupational exposure to environmental tobacco smoke (Z57.31)~~
~~tobacco dependence (F17.-)~~
~~tobacco use (Z72.0)~~

J35 Chronic diseases of tonsils and adenoids

Delete
~~Use additional code to identify:~~
~~exposure to environmental tobacco smoke (Z77.22)~~
~~exposure to tobacco smoke in the perinatal period (P96.81)~~
~~history of tobacco dependence (Z87.891)~~

~~occupational exposure to environmental tobacco smoke (Z57.31)~~
~~tobacco dependence (F17.-)~~
~~tobacco use (Z72.0)~~

J38 Diseases of vocal cords and larynx, not elsewhere classified

Delete ~~Use additional code to identify:~~
~~exposure to environmental tobacco smoke (Z77.22)~~
~~exposure to tobacco smoke in the perinatal period (P96.81)~~
~~history of tobacco dependence (Z87.891)~~
~~occupational exposure to environmental tobacco smoke (Z57.31)~~
~~tobacco dependence (F17.-)~~
~~tobacco use (Z72.0)~~

J43 Emphysema

Delete ~~Use additional code to identify:~~
~~exposure to environmental tobacco smoke (Z77.22)~~
~~exposure to tobacco smoke in the perinatal period (P96.81)~~
~~history of tobacco dependence (Z87.891)~~
~~occupational exposure to environmental tobacco smoke (Z57.31)~~
~~tobacco dependence (F17.-)~~
~~tobacco use (Z72.0)~~

J44 Other chronic obstructive pulmonary disease

Delete ~~Use additional code to identify:~~
~~exposure to environmental tobacco smoke (Z77.22)~~
~~exposure to tobacco smoke in the perinatal period (P96.81)~~
~~history of tobacco dependence (Z87.891)~~
~~occupational exposure to environmental tobacco smoke (Z57.31)~~
~~tobacco dependence (F17.-)~~
~~tobacco use (Z72.0)~~

J84 Other interstitial pulmonary diseases
J84.1 Other interstitial pulmonary diseases with fibrosis
J84.17 Other interstitial pulmonary diseases with fibrosis in diseases classified elsewhere

J84.170 Interstitial lung disease with progressive fibrotic phenotype in diseases classified elsewhere
Code first underlying disease, such as:
Revise sarcoidosis (D86.-)

J93 Pneumothorax and air leak
J93.1 Other spontaneous pneumothorax
J93.12 Secondary spontaneous pneumothorax
Code first underlying condition, such as:
Revise eosinophilic pneumonia (J82.81 -J82.82)
Revise Marfan's syndrome (Q87.4 -)

J99 Respiratory disorders in diseases classified elsewhere
Code first underlying disease, such as:
Revise ankylosing spondylitis (M45.-)
Revise congenital syphilis (A50.5-)
Revise early congenital syphilis (A50.0-, A51.2)

K22 Other diseases of esophagus
K22.1 Ulcer of esophagus
Revise Code first poisoning due to drug or toxin, if applicable (T36-T65 with fifth or sixth character 1-4 ~~or 6~~)

Code first underlying disease, such as:
Revise ankylosing spondylitis (M45.-)
Revise congenital syphilis (A50.5-)
Revise early congenital syphilis (A50.0-, A51.2)

K55 Vascular disorders of intestine
Add Excludes2: angioectasia (angiodysplasia) duodenum (K31.81-)

K56 Paralytic ileus and intestinal obstruction without hernia
K56.6 Other and unspecified intestinal obstruction
K56.69 Other intestinal obstruction
Delete ~~Excludes1~~: ~~intestinal condition code to condition obstruction due to specified~~

K71 Toxic liver disease
Revise Code first poisoning due to drug or toxin, if applicable (T36-T65 with fifth or sixth character 1-4 ~~or 6~~)

L24 Irritant contact dermatitis
L24.A Irritant contact dermatitis due to friction or contact with body fluids

Revise
L24.A9 Irritant contact dermatitis due to friction or contact with other specified body fluids
Irritant contact dermatitis related to endotracheal tube

Revise
Irritant contact dermatitis related to ~~W~~wound fluid~~s,~~ exudate

L53 Other erythematous conditions
L53.0 Toxic erythema

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

Metabolic disorders (E70-E88)

Excludes1: androgen insensitivity syndrome (E34.5-)
congenital adrenal hyperplasia (E25.0)
hemolytic anemias attributable to enzyme disorders (D55.-)

Revise
Marfan~~'s~~ syndrome (Q87.4-)

M24 Other specific joint derangements
M24.1 Other articular cartilage disorders

Excludes2: chondrocalcinosis (M11.1, M11.2-)

Revise
metastatic calcification (E83.59)

M34 Systemic sclerosis [scleroderma]
M34.2 Systemic sclerosis induced by drug and chemical

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

M36 Systemic disorders of connective tissue in diseases classified elsewhere
M36.8 Systemic disorders of connective tissue in other diseases classified elsewhere
Code first underlying disease, such as:

Revise
alkaptonuria (E70.29)

Revise
ochronosis (E70.29)

M41 Scoliosis

Revise
Excludes2: postprocedural scoliosis ~~(M96.-)~~ (M96.89)

Add
postradiation scoliosis (M96.5)

M41.1 Juvenile and adolescent idiopathic scoliosis
Revise M41.12 Adolescent idiopathic scoliosis

M83 Adult osteomalacia
Excludes1: infantile and juvenile osteomalacia (E55.0)
renal osteodystrophy (N25.0)
rickets (active) (E55.0)
rickets (active) sequelae (E64.3)
Revise vitamin D-resistant osteomalacia (E83.31)
Revise vitamin D-resistant rickets (active) (E83.31)

M89 Other disorders of bone
M89.7 Major osseous defect
Code first underlying disease, if known, such as:
Revise osteolysis (M89.5-)

M90 Osteopathies in diseases classified elsewhere
M90.8 Osteopathy in diseases classified elsewhere
Code first underlying disease, such as:
Revise vitamin-D-resistant rickets (E83.31)

M97 Periprosthetic fracture around internal prosthetic joint
Add Code first, if known, the specific type and cause of fracture, such as traumatic or pathological

N14 Drug- and heavy-metal-induced tubulo-interstitial and tubular conditions
Revise Code first poisoning due to drug or toxin, if applicable (T36-T65 with fifth or sixth character 1-4 ~~or 6~~)

N16 Renal tubulo-interstitial disorders in diseases classified elsewhere
Code first underlying disease, such as:
Revise Wilson's disease (E83.01)

N20 Calculus of kidney and ureter
Revise Excludes1: nephrocalcinosis (E83.59)

N29 Other disorders of kidney and ureter in diseases classified elsewhere
Code first underlying disease, such as:
Revise nephrocalcinosis (E83.59)

O04 Complications following (induced) termination of pregnancy
Delete ~~Excludes1: encounter for elective termination of pregnancy, uncomplicated (Z33.2)~~
~~failed attempted termination of pregnancy (O07.)~~

Add — Excludes2: encounter for elective termination of pregnancy, uncomplicated (Z33.2)
failed attempted termination of pregnancy (O07.-)

O75 Other complications of labor and delivery, not elsewhere classified
O75.8 Other specified complications of labor and delivery
O75.82 Onset (spontaneous) of labor after 37 completed weeks of gestation but before 39 completed weeks gestation, with delivery by (planned) cesarean section

Code first to specify reason for planned cesarean section such as:
Revise — previous cesarean delivery (O34.21-)

O87 Venous complications and hemorrhoids in the puerperium
O87.0 Superficial thrombophlebitis in the puerperium

Add — Use additional code, if applicable, to identify the superficial vein thrombosis, such as thrombosis of superficial vessels of lower extremities (I80.0-)

P00 Newborn affected by maternal conditions that may be unrelated to present pregnancy
P00.8 Newborn affected by other maternal conditions
P00.89 Newborn affected by other maternal conditions

Revise — Excludes-2: newborn affected by positive maternal group B streptococcus (GBS)colonization (P00.82)

P04 Newborn affected by noxious substances transmitted via placenta or breast milk
P04.1 Newborn affected by other maternal medication
Revise — Code first, if applicable, withdrawal symptoms from maternal use of drugs of addiction~~, if applicable~~ (P96.1)
Add — withdrawal symptoms from therapeutic use of drugs in newborn (P96.2)

P09 Abnormal findings on neonatal screening
P09.3 Abnormal findings on neonatal screening for congenital hematologic disorders

Revise — Abnormal findings for ~~hemoglobinothies~~ hemoglobinopathy screening

P58 Neonatal jaundice due to other excessive hemolysis
P58.4 Neonatal jaundice due to drugs or toxins transmitted from mother or given to newborn
Revise — Code first poisoning due to drug or toxin, if applicable (T36-T65 with fifth or sixth character 1-4 ~~or 6~~)

P92 Feeding problems of newborn
Revise — Excludes2: feeding problems in child over 28 days old (R63.3-)

Q84 Other congenital malformations of integument
Q84.1 Congenital morphological disturbances of hair, not elsewhere classified
Excludes1: Menkes' kinky hair syndrome (E83.09)

Q87 Other specified congenital malformation syndromes affecting multiple systems
Revise — Q87.4 Marfan~~'s~~ syndrome
Revise — Q87.40 Marfan~~'s~~ syndrome, unspecified
Revise — Q87.41 Marfan~~'s~~ syndrome with cardiovascular manifestations
Revise — Q87.410 Marfan~~'s~~ syndrome with aortic dilation
Revise — Q87.418 Marfan~~'s~~ syndrome with other cardiovascular manifestations
Revise — Q87.42 Marfan~~'s~~ syndrome with ocular manifestations
Revise — Q87.43 Marfan~~'s~~ syndrome with skeletal manifestation

R78 Findings of drugs and other substances, not normally found in blood

Delete — ~~Excludes1: mental or behavioral disorders due to psychoactive substance use (F10-F19)~~

Add — Excludes2: mental or behavioral disorders due to psychoactive substance use (F10-F19)

S22 Fracture of rib(s), sternum and thoracic spine

Revise — Code ~~first~~ also, if applicable, any associated:
injury of intrathoracic organ (S27.-)
spinal cord injury (S24.0-, S24.1-)

S42 Fracture of shoulder and upper arm
Add — Excludes2: periprosthetic fracture around internal prosthetic shoulder joint (M97.3)

S52 Fracture of forearm
Excludes2: fracture at wrist and hand level (S62.-)
Add periprosthetic fracture around internal prosthetic elbow joint (M97.4)

S72 Fracture of femur
S72.0 Fracture of head and neck of femur
Excludes2: physeal fracture of upper end of femur (S79.0-)
Add physeal fracture of lower end of femur (S79.1-)

S82 Fracture of lower leg, including ankle
Excludes2: fracture of foot, except ankle (S92.-)
Add periprosthetic fracture around internal prosthetic ankle joint (M97.2)
periprosthetic fracture around internal prosthetic implant of knee joint (M97.1-)

T80 Complications following infusion, transfusion and therapeutic injection
Excludes2: any encounters with medical care for postprocedural conditions in which no complications are present, such as:
Revise poisoning and toxic effects of drugs and chemicals (T36-T65 with fifth or sixth character 1-4 ~~or 6~~)

T81 Complications of procedures, not elsewhere classified
Excludes2: complications following immunization (T88.0-T88.1)
Revise poisoning and toxic effects of drugs and chemicals (T36-T65 with fifth or sixth character 1-4 ~~or 6~~)

T81.1 Postprocedural shock
Shock during or resulting from a procedure, not elsewhere classified
Excludes1: anaphylactic shock NOS (T78.2)
Delete ~~septic shock (R65.21)~~

T81.8 Other complications of procedures, not elsewhere classified
T81.83 Persistent postprocedural fistula
Add Code also, if applicable, site of fistula such as:
Add anal fistula (K60.3)
Add anorectal fistula (K60.5)
Add bladder fistula (N32.2)
Add other female intestinal-genital tract fistulae (N82.4)

T88 Other complications of surgical and medical care, not elsewhere classified

Excludes2: complication following infusion, transfusion and therapeutic injection (T80.-)

Revise poisoning and toxic effects of drugs and chemicals (T36-T65 with fifth or sixth character 1-4 ~~or 6~~)

INDEX MODIFICATION PROPOSED ADDENDA

All proposed effective October 1, 2023

Abuse
- - opioid F11.10
- - - with
Add - - - - opioid-associated amnestic syndrome F11.118

Accident
Delete ~~- cerebral I63.9~~
Revise - cerebrovascular ~~(embolic)~~ (ischemic) ~~(thrombotic)~~ I63.9
Add - - chronic (old) (remote) (imaging) (without sequelae) Z86.73
Add - - - with residual defects - see Sequelae, disease, cerebrovascular
Add - - embolic I63.-
Add - - thrombotic I63.-

Revise Acrochondrohyperplasia -see Syndrome, Marfan~~'s~~

Additional -see also Accessory
Revise - chromosome(s) (see also Trisomy) Q99.8
Add - - marker – see Extra, marker chromosomes

Aneurysm (anastomotic) (artery) (cirsoid) (diffuse) (false) (fusiform) (multiple) (saccular) I72.9
Revise - sinus of Valsalva ~~Q25.49~~ Q25.43

Revise Angiospasm (peripheral) (traumatic) (vessel) (see also vasospasm) I73.9

Revise Arachnodactyly -see Syndrome, Marfan~~'s~~

Arteriosclerosis, arteriosclerotic (diffuse) (obliterans) (of) (senile) (with calcification) I70.90
- brain I67.2
Add - - with infarction— see Occlusion, artery, brain or cerebral, with infarction
- central nervous system I67.2
Add - - with infarction— see Occlusion, artery, cerebral or precerebral, with infarction
- cerebral I67.2
Add - - with infarction— see Occlusion, artery, brain or cerebral, with infarction
- cerebrovascular I67.2
Add - - with infarction— see Occlusion, artery, brain or cerebral, with infarction

Revise - heart (disease) -see Arteriosclerosis, coronary (artery)~~,~~

- vertebral (artery) I67.2
Add - - with infarction— see Occlusion, artery, vertebral, with infarction

	Asphyxia, asphyxiation (by) R09.01
Revise	- mucus -see also Foreign body, respiratory tract, causing, ~~asphyxia~~ asphyxiation
	Checking (of)
	- wound Z48.0-
Add	- - postoperative – see also Aftercare
	Cor
Revise	- pulmonale ~~(chronic)~~ I27.81
	- - acute I26.09
Add	- - - without pulmonary embolism I27.81
Add	- - chronic I27.81
Add	- - - with chronic pulmonary embolism I27.82
	Counseling (for) Z71.9
	- medical (for) Z71.9
Revise	- - person living alone (see also Consultation, specified reason NEC) Z60.2
	Dependence (on) (syndrome) F19.20
	- - opioid F11.20
	- - - with
Add	- - - - opioid-associated amnestic syndrome F11.228
	Dermatitis (eczematous) L30.9
	- due to
Add	- - exudate (wound fluids) L24.A9
	- contact (occupational) L25.9
	- - irritant L24.9
	- - - due to
	- - - - body fluids L24.A0
Add	- - - - - feces L24.A2
Add	- - - - - urine L24.A2
Add	- - - - - wound exudate L24.A9
Add	- - - - exudate L24.A9
Add	- - - - friction L24.A0
	- due to
Add	- - exudate -see Dermatitis, contact, irritant
	Diabetes, diabetic (mellitus) (sugar) E11.9
	- due to
Add	- - pancreatectomy - see Diabetes, specified type NEC

Difficult, difficulty (in)
- feeding R63.30
Add - - elderly R63.39
Add - - infant NOS R63.39

Disease, diseased -see also Syndrome
Revise - arterial (see also Disease, artery) I77.9
Revise - - occlusive -see also Occlusion, by site
Revise - artery (see also Disease, arterial) I77.9
- lung J98.4
- - interstitial J84.9
Add - - - drug-induced – see Disorder, lung, interstitial, drug-induced
- - - of childhood, specified NEC J84.848
Add - - - - drug-induced – see Disorder, lung, interstitial, drug-induced
- peripheral
- - vascular NOS I73.9
Add - - - in diabetes mellitus -see Diabetes, by type, with peripheral angiopathy

Revise Dolichostenomelia -see Syndrome, Marfan's

Duodenitis (nonspecific) (peptic) K29.80
Add - erosive - see Ulcer, duodenum

Duplication, duplex -see also Accessory
Revise - chromosome NEC – see also Trisomy

Elevated, elevation
Revise - troponin ~~R77.8~~ R79.89
Add - - due to
Add - - - myocardial infarction – see Infarction, myocardial
Add - - - myocardial injury – see Injury, myocardial

Embolism (multiple) (paradoxical) I74.9
- vein (acute) I82.90
Add - - calf muscular I82.46-
Add - - - chronic I82.56-
Add - - peroneal I82.45-
Add - - - chronic I82.55-

Encephalitis (chronic) (hemorrhagic) (idiopathic) (nonepidemic) (spurious) (subacute) G04.90
- in (due to)
Revise - - systemic lupus erythematosus M32.19 [G05.3]
Revise - lupus erythematosus, systemic M32.19 [G05.3]

Encounter (with health service) (for) Z76.89
Add - postoperative – see also Aftercare

Enlargement, enlarged -see also Hypertrophy
- prostate N40.0
- - with lower urinary tract symptoms (LUTS) N40.1
Add - - - nodular N40.3
Add - - nodular N40.2
Add - - - with lower urinary tract symptoms (LUTS) N40.3
- - without lower urinary tract symtpoms (LUTS) N40.0
Add - - - nodular N40.2

Epilepsy, epileptic, epilepsia (attack) (cerebral) (convulsion) (fit) (seizure) G40.909
Add - Partial – see Epilepsy, localization-related, symptomatic, with simple partial seizures

Erosion - come back to
Add - cameron – see Ulcer, stomach

Esophagitis (acute) (alkaline) (chemical) (chronic) (infectional) (necrotic) (peptic) (postoperative) (without bleeding) K20.90
- reflux K21.00
Add - - with bleeding K21.01

Exudate
Add - causing irritant dermatitis L24.A9
Revise - wound fluids causing irritant dermatitis L24.A9

Failure, failed
- hepatic K72.90
Add - - end stage K72.10
Add - - - with coma K72.11

Fistula (cutaneous) L98.8
Revise - postoperative, persistent (see also Fistula, by site, if known) T81.83
Delete ~~- - specified site -see Fistula, by site~~

Granuloma L92.9
Revise - Hodgkin C81.9-

Add Hemimegalencephaly Q04.5

Hygroma (congenital) (cystic) D18.1
Add - subdural – see Leak, cerebrospinal fluid

Hyperplasia, hyperplastic
Revise - prostate (adenofibromatous) ~~(nodular)~~ N40.0
- - with lower urinary tract symptoms (LUTS) N40.1
Add - - - nodular N40.3
Add - - nodular N40.2
Add - - - with lower urinary tract symptoms (LUTS) N40.3
- - without lower urinary tract symtpoms (LUTS) N40.0
Add - - - nodular N40.2

Infarct, infarction
Revise - cerebral (acute) ~~(chronic)~~ -see also Occlusion, artery cerebral or precerebral, with infarction I63.9-
Add - - chronic (old) (remote) (imaging) (without sequelae) Z86.73
Add - - - with residual defects - see Sequelae, disease, cerebrovascular
- myocardium, myocardial (acute) (with stated duration of 4 weeks or less) I21.9
Revise - - Q wave (see also, Infarct, myocardium, ST elevation, by site) I21.3
Revise - - transmural (see also, Infarct, myocardium, ST elevation, by site) I21.3

Keratosis
Add - lichenoid L82.0

Add Lactate, elevated – see Acidosis, lactic

Lesion(s) (nontraumatic)
Add - cameron – see Ulcer, stomach

Add Lichenoid, keratosis – see Keratosis, lichenoid

Loose -see also condition
- body
- - joint M24.00
Add - - - temporomandibular M24.08

Lymphoma (of) (malignant) C85.90
Revise - Hodgkin C81.9-

Revise Marfan~~'s~~ syndrome -see Syndrome, Marfan~~'s~~

Microangiopathy (peripheral) I73.9
- thrombotic M31.10
Revise - - hematopoietic stem cell transplantation-associated [HSCT-TMA] ~~M31.10~~ M31.11

Add Monoparesis – see Monoplegia

	Myelopathy (spinal cord) G95.9
	- in (due to)
Add	- - disease classified elsewhere G99.2
Add	Non-accidental trauma – see Abuse, physical
	Osteoporosis (female) (male) M81.0
	- age-related M81.0
	- - with current pathologic fracture M80.00
Add	- - - femur M80.05
Add	- - - hip M80.05
Revise	- - - ilium ~~M80.05~~ M80.0A
Revise	- - - ischium ~~M80.05~~ M80.0A
Revise	- - - pelvis ~~M80.05~~ M80.0A
Add	- - - pubis ramus – see Osteoporosis, pelvis
	- disuse M81.8
	- - with current pathological fracture M80.80
Add	- - - femur M80.85
Add	- - - hip M80.85
Revise	- - - ilium ~~M80.85-~~ M80.8A
Revise	- - - ischium ~~M80.85-~~ M80.8A
Revise	- - - pelvis ~~M80.85-~~ M80.8A
Add	- - - pubis ramus – see Osteoporosis, pelvis
	- postmenopausal M81.0
	- - with pathological fracture M80.00
Add	- - - femur M80.05
Add	- - - hip M80.05
Revise	- - - ilium ~~M80.05-~~ M80.0A
Revise	- - - ischium ~~M80.05-~~ M80.0A
Revise	- - - pelvis ~~M80.05-~~ M80.0A
Add	- - - pubis ramus – see Osteoporosis, pelvis
	- specified type NEC M81.8
	- - with pathological fracture M80.80
Add	- - - femur M80.85
Add	- - - hip M80.85
Revise	- - - ilium ~~M80.85-~~ M80.8A
Revise	- - - ischium ~~M80.85-~~ M80.8A
Revise	- - - pelvis ~~M80.85-~~ M80.8A
Add	- - - pubis ramus M80.8A

Revise	Postoperative (postprocedural) -see also Complication, postoperative
Add	- visit – see also Aftercare
Add	- wound check – see also Aftercare
	Problem (with) (related to)
Revise	- feeding (elderly) (infant) NOS R63.39
Add	Rhinosinusitis – see Sinusitis
	Schizophrenia, schizophrenic F20.9
Revise	- childhood type ~~F84.5~~ F20.9
Revise	Sciatica (infective) M54.3-
	Scoliosis (acquired) (postural) M41.9
Add	- postprocedural scoliosis M96.89
	Sepsis (generalized) (unspecified organism) A41.9
Revise	- escherichia coli (E. coli) A41.51
Revise	- gram-negative (organism) A41.50
Add	- other gram-negative A41.59
Add	- pseudomonas (pseudomonas aeruginosa) A41.52
Add	- serratia A41.53
	Spider
Revise	- fingers -see Syndrome, Marfan~~'s~~
Revise	- toes -see Syndrome, Marfan~~'s~~
	Spondyloarthritis
Revise	- axial -see also ~~Spondlyitis~~ Spondylitis, ankylosing
	Spondylosis M47.9
	- specified NEC M47.899
Revise	- - facet joint ~~-see also Spondylosis~~ M47.819
	Stasis
	- ulcer -see Varix, leg, with, ulcer
Revise	- - without varicose veins (see also Ulcer, by site) I87.2
Revise	Stroke (apoplectic) (brain) ~~(embolic)~~ (ischemic) (paralytic) ~~(thrombotic)~~ I63.9
Add	- cerebrovascular (ischemic) I63.9
Add	- - chronic (old) (remote) (imaging) (without sequelae) Z86.73
Add	- - - with residual defects - see Sequelae, disease, cerebrovascular
Add	- - embolic I63.-
Add	- - thrombolic I63.-

	Syndrome -see also Disease
Revise	- schizophrenic, of childhood NEC ~~F84.5~~ F20.9
Add	- Snyder-Robinson Q87.89
	Therapy
	- drug, long-term (current) (prophylactic)
Add	- - oral antidiabetic Z79.84
Add	- - oral hypoglycemic Z79.84
	Thrombosis, thrombotic (bland) (multiple) (progressive) (silent) (vessel) I82.90
Add	- - calf muscular I82.46-
Add	- - - chronic I82.56-
Add	- - peroneal I82.45-
Add	- - - chronic I82.55-
	Trauma, traumatism -see also Injury
Add	- non-accidental – see Abuse, physical
	Ulcer, ulcerated, ulcerating, ulceration, ulcerative
Add	- cameron – see Ulcer, stomach
	- duodenum, duodenal (eroded) (peptic) K26.9
Revise	- - chronic (erosive) K26.7
	- stasis (venous) -see Varix, leg, with, ulcer
Revise	- - without varicose veins (see also Ulcer, by site) I87.-
	Use (of)
	- opioid F11.90
	- - with
Add	- - - opioid-associated amnestic syndrome F11.988
Revise	Vasospasm (vasoconstriction) (see angiospasm) I73.9
	Wound check Z48.0-
Add	- postoperative – see also Aftercare

External Cause of Morbidity Index Addenda

	Accident
Delete	~~- animal-rider -see Accident, transport, animal-rider~~
	- animal-drawn vehicle -see Accident, transport, animal-drawn vehicle occupant
Add	- animal-rider -see Accident, transport, animal-rider

Place of occurrence Y92.9
Add - bar Y92.59
Add - tavern Y92.59

Prolonged
Revise - sitting in transport vehicle -see ~~Travel, by type of vehicle~~ Sitting