



**ICD-10 Coordination and Maintenance Committee Meeting  
March 7-8, 2023  
Diagnosis Agenda**

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

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## ICD-10 TIMELINE

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

March 7-8, 2023	ICD-10 Coordination and Maintenance Committee Meeting.
March 2023	Recordings and slide presentations of the March 7-8, 2023 ICD-10 Coordination and Maintenance Committee Meeting will be posted on the following web pages:  <b>Diagnosis code portion of the recording and related materials–</b> <a href="https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm">https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm</a>  <b>Procedure code portion of the recording and related materials–</b> <a href="https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html">https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html</a>
April 1, 2023	Any new ICD-10 codes will be implemented on April 1, 2023.
<b>April 7, 2023</b>	<b>Deadline for receipt of public comments on proposed new codes and revisions discussed at the March 7-8, 2023 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2023.</b>
April 2023	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 2024 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: <a href="https://www.cms.gov/medicare/medicare-fee-for-service-payment/acuteinpatientpps">https://www.cms.gov/medicare/medicare-fee-for-service-payment/acuteinpatientpps</a>
<b>May 5, 2023</b>	<b>Deadline for receipt of public comments on proposed new codes and revisions discussed at the March 7-8, 2023 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on April 1, 2024.</b>  <b>Deadline for receipt of public comments on proposed new diagnosis codes and revisions discussed at the March 7-8, 2023 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2024.</b>

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**Diagnosis addendum -**

<https://www.cdc.gov/nchs/icd/Comprehensive-Listing-of-ICD-10-CM-Files.htm>

**Procedure addendum -**

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

**June 9, 2023**

**Deadline for requestors: Those members of the public requesting that topics be discussed at the September 12-13, 2023 ICD-10 Coordination and Maintenance Committee Meeting, must have their requests submitted to CMS for procedures and NCHS for diagnoses.**

Requestors should indicate if they are submitting their code request for consideration for an April 1, 2024 implementation date or an October 1, 2024 implementation date.

**July 2023**

Federal Register notice for the September 12-13, 2023 ICD-10 Coordination and Maintenance Committee Meeting will be published. This will include the tentative agenda.

**August 1, 2023**

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, 2023.

This rule can be accessed at:

<https://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/AcuteInpatientPPS/index.html>

**August 2023**

Tentative agenda for the Procedure portion of the September 12, 2023 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 13, 2023 ICD-10 Coordination and Maintenance Committee Meeting will be posted on the NCHS webpage at -

[https://www.cdc.gov/nchs/icd/icd10cm\\_maintenance.htm](https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm)

**September 12-13, 2023**

The September 2023 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.

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- September 2023 Recordings and slide presentations of the September 12-13, 2023 ICD-10 Coordination and Maintenance Committee Meeting will be posted on the following web pages:
- Diagnosis code portion of the recording and related materials–**  
[https://www.cdc.gov/nchs/icd/icd10cm\\_maintenance.htm](https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm)
- Procedure code portion of the recording and related materials–**  
<https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html>
- October 1, 2023 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/Comprehensive-Listing-of-ICD-10-CM-Files.htm>
- Procedure addendum –**  
<https://www.cms.gov/Medicare/Coding/ICD10/>
- October 13, 2023** **Deadline for receipt of public comments on proposed new codes discussed at the September 12-13, 2023 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on April 1, 2024.**
- November 2023 Any new ICD-10 codes that will be implemented on the following April 1 will be announced. Information on any new codes to be implemented April 1, 2024 will be posted on the following websites:
- <https://www.cdc.gov/nchs/icd/Comprehensive-Listing-of-ICD-10-CM-Files.htm>
- <https://www.cms.gov/Medicare/Coding/ICD10/>
- November 15, 2023** **Deadline for receipt of public comments on proposed new codes and revisions discussed at the September 12-13, 2023 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2024.**



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**Contact Information**

Comments on the diagnosis proposals presented at the ICD-10 Coordination and Maintenance Committee meeting should be sent to the following email address: [nchsicd10CM@cdc.gov](mailto:nchsicd10CM@cdc.gov)

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**Continuing Education Credits**

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

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

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

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

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

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

## Anal Fistula

This topic was presented at the March 2022 and September 2022 Coordination and Maintenance meeting. Based on public comments, revisions to the proposal have been made for reconsideration. 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.<sup>1</sup> 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.<sup>2</sup> 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).<sup>3</sup> 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.<sup>2</sup>

The management of patients with anal fistulas varies depending on severity of disease and underlying comorbidities (such as Crohn's disease).<sup>4</sup> 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.<sup>5</sup> 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.<sup>6</sup>

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 recurrence and fecal incontinence.<sup>7</sup>

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. Changes are indicated in **BOLD**.

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**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>
5. Panes J, Reinisch W, Rupniewska E, et al. Burden and outcomes for complex perianal fistulas in Crohn's disease: Systematic review. *World J Gastroenterol*. 2018;24(42):4821-4834. doi:10.3748/wjg.v24.i42.4821
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**

Add	K50	<p>Crohn's disease [regional enteritis]</p> <p>Use additional code to identify any associated fistulas, if applicable: anal fistula (K60.3-) anorectal fistula (K60.5-) rectal fistula (K60.4-)</p>
Add	K51	<p>Ulcerative colitis</p> <p>Use additional code to identify any associated fistulas, if applicable: anal fistula (K60.3-) anorectal fistula (K60.5-) rectal fistula (K60.4-)</p>
<b>Revise</b>	K60	<p>Fissure and fistula of anal and rectal regions</p> <p><b>Excludes1<del>2</del></b>: fissure and fistula of anal and rectal regions with abscess or cellulitis (K61.-)</p> <p>Excludes2: anal sphincter tear (healed) (nontraumatic) (old) (K62.81)</p>
New subcategory	K60.3	Anal fistula
Add		Code first, if applicable: Crohn's disease (K50.-) ulcerative colitis (K51.-)
Add		Excludes1: congenital fistula (Q43.6)

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<b>New code Add</b>	<b>K60.30</b>	<b>Anal fistula, unspecified Anal fistula NOS</b>
New sub-subcategory Add Add	K60.31	Anal fistula, simple Low intersphincteric anal fistula Superficial anal fistula
<b>New code New code Add</b>	<b>K60.311</b>	<b>Anal fistula, simple, initial</b>
	K60.312	Anal fistula, simple, persistent Anal fistula, simple, chronic
New code <b>Add</b>	K60.313	Anal fistula, simple, recurrent <b>Anal fistula simple, occurring following complete healing</b>
New code	K60.319	Anal fistula, simple, unspecified
New sub-subcategory Add Add Add Add	K60.32	Anal fistula, complex Extrasphincteric anal fistula Intersphincteric anal fistula Suprasphincteric anal fistula Transsphincteric anal fistula
Add		Code also, if applicable: perianal abscess (K61.0) rectovaginal fistula (N82.3) stenosis of anus and rectum (K62.4)
<b>New code New code Add</b>	<b>K60.321</b>	<b>Anal fistula, complex, initial</b>
	K60.322	Anal fistula, complex, persistent Anal fistula, complex, chronic
New code <b>Add</b>	K60.323	Anal fistula, complex, recurrent <b>Anal fistula complex, occurring following complete healing</b>
New code	K60.329	Anal fistula, complex, unspecified
New subcategory <b>Add</b>	K60.4	Rectal fistula <b>Code first, if applicable: Crohn's disease (K50.-) ulcerative colitis (K51.-)</b>

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Add		Excludes1: congenital fistula (Q43.6)
<b>New code</b> <b>Add</b>	<b>K60.40</b>	<b>Rectal fistula, unspecified</b> <b>Rectal fistula NOS</b>
New sub-subcategory Add Add	K60.41	Rectal fistula, simple Low intersphincteric rectal fistula Superficial rectal fistula
<b>New code</b> New code Add	<b>K60.411</b> K60.412	<b>Rectal fistula, simple, initial</b> Rectal fistula, simple, persistent Rectal fistula, simple, chronic
New code <b>Add</b>	K60.413	Rectal fistula, simple, recurrent <b>Rectal fistula simple, occurring following complete healing</b>
New code	K60.419	Rectal fistula, simple, unspecified
New sub-subcategory Add Add Add Add	K60.42	Rectal fistula, complex Extrasphincteric rectal fistula Intersphincteric rectal fistula Suprasphincteric rectal fistula Transsphincteric rectal fistula
<b>Add</b>		<b>Code first, if applicable:</b> <b>perianal abscess (K61.0)</b> <b>rectovaginal fistula (N82.3)</b> <b>stenosis of anus and rectum (K62.4)</b>
<b>New code</b> New code Add	<b>K60.421</b> K60.422	<b>Rectal fistula, complex, initial</b> Rectal fistula, complex, persistent Rectal fistula, complex, chronic
New code <b>Add</b>	K60.423	Rectal fistula, complex, recurrent <b>Rectal fistula complex occurring following complete healing</b>
New code	K60.429	Rectal fistula, complex, unspecified

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New subcategory <b>Add</b>	K60.5	Anorectal fistula <b>Code first, if applicable: Crohn's disease (K50.-) ulcerative colitis (K51.-)</b>
Add		Excludes1: congenital fistula (Q43.6)
<b>New code Add</b>	<b>K60.50</b>	<b>Anorectal fistula, unspecified Anorectal fistula NOS</b>
New sub-subcategory Add Add	K60.51	Anorectal fistula, simple Low intersphincteric anorectal fistula Superficial anorectal fistula
<b>New code New code Add</b>	<b>K60.511</b> K60.512	<b>Anorectal fistula, simple, initial</b> Anorectal fistula, simple, persistent Anorectal fistula, simple, chronic
New code <b>Add</b>	K60.513	Anorectal fistula, simple, recurrent <b>Anorectal fistula simple, occurring following complete healing</b>
New code	K60.519	Anorectal fistula, simple, unspecified
New sub-subcategory Add Add Add Add	K60.52	Anorectal fistula, complex Extrasphincteric anorectal fistula Intersphincteric anorectal fistula Suprasphincteric anorectal fistula Transsphincteric anorectal fistula
<b>Add</b>		<b>Code first, if applicable: perianal abscess (K61.0) rectovaginal fistula (N82.3) stenosis of anus and rectum (K62.4)</b>
<b>New code New code Add</b>	<b>K60.521</b> K60.522	<b>Anorectal fistula, complex, initial</b> Anorectal fistula, complex, persistent Anorectal fistula, complex, chronic

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New code <b>Add</b>	K60.523	Anorectal fistula, complex, recurrent <b>Anorectal fistula complex, occurring following complete healing</b>
New code	K60.529	Anorectal fistula, complex, unspecified

**INDEX MODIFICATIONS**

Add	Fistula (cutaneous) L98.8
Revise	- anorectal (infectious) K60.5-
Add	- anus, anal ( <del>recurrent</del> ) (infectious) K60.3-
Add	- rectal (infectious) K60.4-



## **Carcinoid Heart Syndrome (Disease) (Hedinger syndrome)**

The National Center for Health Statistics received a proposal requesting a new ICD-10-CM code for carcinoid heart disease (CaHD) from the Carcinoid Cancer Foundation (CCF).

Carcinoid heart disease is a rare clinical condition in patients with neuroendocrine tumors (NETs) with an estimated population of less than 20,000 individuals in the United States (US).<sup>1-4</sup> CaHD has a unique serotonin-mediated pathology of CaHD versus other valvular disease.<sup>12</sup> Several studies have found a strong correlation between high levels of a serotonin metabolite (i.e., 5-hydroxyindoleacetic acid (5-HIAA)) in patients and the development of CaHD.<sup>4,7,9,10,11,13,14</sup> Serotonin receptors are well represented within cardiac tissues.<sup>15</sup> Once activated, these receptors trigger a fibrotic process which ultimately results in the deposition of pearly, white, carcinoid plaques on valvular cusps, leaflets, papillary muscles, chordae and ventricular walls.<sup>15,16</sup> Key findings of CaHD upon transthoracic echocardiogram (TTE) include thickened, retracted and fixed valve leaflets, valvular regurgitation, and dilated right chambers of the heart.<sup>12</sup> Initially, CaHD predominately affects the right-side of the heart with the tricuspid and pulmonic valves showing early signs of degeneration. However, as the disease progresses, all four valves of the heart can be affected and ultimately heart failure can develop.<sup>17</sup> The irreversible nature of this fibrotic process within the heart makes CaHD pathophysiology unique and insufficiently characterized by the available diagnosis codes currently being used to code patients with CaHD.

Patients with NETs can have “functional” tumors (i.e., tumors that secrete vasoactive substances including serotonin, tachykinins and prostaglandins). The vasoactive substances secreted by functional NETs, especially serotonin, are responsible for the symptoms of CS (e.g., diarrhea, nausea, flushing) as well as the development of CaHD. Carcinoid heart disease typically occurs once a patient’s primary NET has metastasized to the liver. This allows the vasoactive substances to enter the systemic circulation and reach the right side of the heart, which may result in right-sided heart failure, the need for valve replacement surgery and increased risk of mortality.<sup>8</sup>

Data has shown that the majority of CaHD patients are initially asymptomatic.<sup>18</sup> Regular frequent screening is crucial to appropriately identify and manage patients. Several guidelines and expert consensus statements recommend clinicians perform regular TTEs, monitor urine or plasma 5-HIAA and N-terminal pro brain natriuretic peptide (NT-pro-BNP) levels at baseline and at regular 3-12 month intervals.<sup>12,19</sup> However, data suggests that these recommendations are not being followed even at leading NET treatment centers.<sup>20,21</sup> A specific ICD-10-CM code for CaHD will help promote a larger disease awareness and could encourage NET experts to consider CaHD.

There are an estimated 170,000 patients in the US diagnosed with NETs.<sup>5,6</sup> Approximately 19% of patients with NETs (~32,300 patients) have carcinoid syndrome (CS).<sup>2</sup> A systematic literature review which included 26 studies found that majority of studies report between 20-50% of patients with CS will suffer from CaHD.<sup>7</sup> It is estimated that the number of US patients with CaHD is between approximately 6,460 and 16,150.<sup>2,3,6,7</sup>

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It will be beneficial to have a specific ICD-10-CM code to provide coding specificity for CaHD due to the distinct clinical disease characterized by a specific pathophysiologic process in the heart. One study reported a 31% three-year survival rate for patients with CaHD compared to 68% for patients with only CS ( $p=0.0003$ ).<sup>9</sup>

**References**

1. Dasari, Arvind et al. "Trends in the Incidence, Prevalence, and Survival Outcomes in Patients With Neuroendocrine Tumors in the United States." *JAMA oncology* vol. 3,10 (2017): 1335-1342. doi:10.1001/jamaoncol.2017.0589.
2. Halperin, Daniel M et al. "Frequency of carcinoid syndrome at neuroendocrine tumour diagnosis: a population-based study." *The Lancet. Oncology* vol. 18,4 (2017): 525-534. doi:10.1016/S1470-2045(17)30110-9.
3. Macfie, Rebekah et al. "The prevalence, operations, and outcomes of carcinoid heart disease." *American journal of surgery* vol. 224,2 (2022): 665-669. doi:10.1016/j.amjsurg.2022.03.054.
4. Bhattacharyya, Sanjeev et al. "Risk factors for the development and progression of carcinoid heart disease." *The American journal of cardiology* vol. 107,8 (2011): 1221-6. doi:10.1016/j.amjcard.2010.12.025.
5. Dasari A, Shen C, Halperin D, et al. Trends in the Incidence, Prevalence, and Survival Outcomes in Patients With Neuroendocrine Tumors in the United States. *JAMA Oncol.* 2017 3(10):1335-1342.
6. Das S and Dasari A. Epidemiology, incidence, and prevalence of neuroendocrine neoplasms: are there global differences? *Curr Oncol Rep.* 2021 Mar 14;23(4):43.
7. Buchanan-Hughes, Amy et al. "Carcinoid Heart Disease: Prognostic Value of 5-Hydroxyindoleacetic Acid Levels and Impact on Survival: A Systematic Literature Review." *Neuroendocrinology* vol. 111,1-2 (2021): 1-15. doi:10.1159/000506744.
8. Hassan, Saamir A et al. "Carcinoid heart disease." *Heart (British Cardiac Society)* vol. 103,19 (2017): 1488-1495. doi:10.1136/heartjnl-2017-311261.
9. Pellikka, P A et al. "Carcinoid heart disease. Clinical and echocardiographic spectrum in 74 patients." *Circulation* vol. 87,4 (1993): 1188-96. doi:10.1161/01.cir.87.4.1188.
10. Dobson, R et al. "Serial surveillance of carcinoid heart disease: factors associated with echocardiographic progression and mortality." *British journal of cancer* vol. 111,9 (2014): 1703-9. doi:10.1038/bjc.2014.468.
11. Joish, Vijay N et al. "Serotonin levels and 1-year mortality in patients with neuroendocrine tumors: a systematic review and meta-analysis." *Future oncology (London, England)* vol. 15,12 (2019): 1397-1406. doi:10.2217/fon-2018-0960.
12. Davar, Joseph et al. "Diagnosing and Managing Carcinoid Heart Disease in Patients With Neuroendocrine Tumors: An Expert Statement." *Journal of the American College of Cardiology* vol. 69,10 (2017): 1288-1304. doi:10.1016/j.jacc.2016.12.030.
13. Möller, Jacob E et al. "Factors associated with progression of carcinoid heart disease." *The New England journal of medicine* vol. 348,11 (2003): 1005-15. doi:10.1056/NEJMoa021451
14. Lundin, L et al. "Carcinoid heart disease: relationship of circulating vasoactive substances to ultrasound-detectable cardiac abnormalities." *Circulation* vol. 77,2 (1988): 264-9. doi:10.1161/01.cir.77.2.264.
15. Ram, Pradhun et al. "Carcinoid Heart Disease: Review of Current Knowledge." *Texas Heart Institute journal* vol. 46,1 21-27. 1 Feb. 2019, doi:10.14503/THIJ-17-6562
16. Jin, Chengyue et al. "Carcinoid Heart Disease: Pathophysiology, Pathology, Clinical Manifestations, and Management." *Cardiology* vol. 146,1 (2021): 65-73. doi:10.1159/000507847
17. Baron, Emilie et al. "Progression of Carcinoid Heart Disease in the Modern Management Era." *Journal of the American Heart Association* vol. 10,23 (2021): e020475. doi:10.1161/JAHA.120.020475.
18. Bhattacharyya, Sanjeev et al. "Analysis of 150 patients with carcinoid syndrome seen in a single year at one institution in the first decade of the twenty-first century." *The American journal of cardiology* vol. 101,3 (2008): 378-81. doi:10.1016/j.amjcard.2007.08.045.
19. Grozinsky-Glasberg, Simona et al. "European Neuroendocrine Tumor Society (ENETS) 2022 Guidance Paper for Carcinoid Syndrome and Carcinoid Heart Disease." *Journal of neuroendocrinology* vol. 34,7 (2022): e13146. doi:10.1111/jne.13146.
20. Mouser B, Atari O, Howe J et al. Screening for Carcinoid Heart Disease: Future Perspectives. Poster presented at the 71<sup>st</sup> Annual American College of Cardiology Scientific Session and Expo; April 2-4, 2022; Washington, D.C.
21. Duncan K, Law C, Singh S, et al. Carcinoid Heart Disease in Patients Diagnosed with Small Bowel and Lung Neuroendocrine Tumors. Poster presented at The North American Neuroendocrine Tumor Society Annual Meeting; October 27-29, 2022; Washington, D.C.

**TABULAR MODIFICATIONS**

E34 Other endocrine disorders

Excludes1 pseudohypoparathyroidism (E20.1)

E34.0 Carcinoid syndrome

Delete ~~Note: May be used as an additional code to identify functional activity associated with a carcinoid tumor~~

Add Code also the underlying disorder, such as:  
 primary neuroendocrine tumors (C7A.-)  
 secondary neuroendocrine tumors (C7B.-)

New code E34.00 Carcinoid syndrome, unspecified  
 Add Carcinoid disease, unspecified

New code E34.01 Carcinoid heart syndrome  
 Add Carcinoid heart disease  
 Add Hedinger syndrome

New code E34.09 Other carcinoid syndrome  
 Add Carcinoid disease NEC  
 Add Carcinoid syndrome NEC  
 Add Other carcinoid disease

**INDEX MODIFICATIONS**

Revise Argentaffinoma -see also Neoplasm, uncertain behavior, by site  
 - syndrome E34.09

Revise Björck (-Thorson) syndrome (malignant carcinoid) E34.09

Revise Carcinoidosis E34.00  
 Add - heart E34.01

Revise Cassidy (-Scholte) syndrome (malignant carcinoid) E34.09

Revise Flush syndrome E34.09

Add Heart -see condition  
 - carcinoid – see Syndrome, carcinoid, heart

Revise Scholte's syndrome (malignant carcinoid) E34.09

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- Secretion
  - hormone
  - - by
  - Revise - - - carcinoid tumor E34.09
- Syndrome -see also Disease
  - Revise - argentaffin, argintaffinoma E34.09
  - Revise - Björck (-Thorsen) E34.09
  - Revise - carcinoid E34.09
  - Add - - heart E34.01
  - Revise - Cassidy (-Scholte) E34.09
  - Revise - flush E34.09
  - Revise - Hedinger's E34.01
  - Revise - intestinal
    - - carcinoid E34.09
    - Revise - malignant
      - - carcinoid E34.00
      - Revise - metastatic carcinoid E34.00
      - Revise - Scholte's E34.09
      - Revise - Thorson-Björck E34.09
      - Revise Thorson-Björck syndrome E34.09

## **Central Centrifugal Cicatricial Alopecia (CCCA)**

Central Centrifugal Cicatricial Alopecia (CCCA) is a primary cicatricial (scarring) alopecia affecting women of African ancestry that can result in permanent irreversible damage of hair follicles. It occurs predominantly in black women aged 30 to 55 years. The real prevalence of CCCA remains unknown due to lack of population studies; however, its prevalence is estimated to vary from 2.7 to 5.6%.

The etiology of CCCA remains unclear. Previous studies have shown that CCCA does not always relate to hair care practices, as such familial occurrence of CCCA has been found and the genetic susceptibility to the disease is likely to be inherited in an autosomal dominant manner.

CCCA presents with scarring at the vertex or crown of the scalp that tends to spread centrifugally. Symptoms range from itchy, tender scalp; dysesthesias; seborrheic-type scale, follicular papules; and pustules to completely asymptomatic hair breakage at the vertex of the scalp resulting in loss of follicular ostia; but there are patients who are asymptomatic. In addition to the signs and symptoms, CCCA also has a significant negative impact on the quality of life of patients with CCCA.

Currently, there is no specific ICD-10-CM code for CCCA as it is currently coded under (L66.8 Other cicatricial alopecia or L66.9 Cicatricial alopecia, unspecified). Creating a specific ICD-10-CM code for central centrifugal cicatricial alopecia CCCA would be beneficial for the study of this disease and the many people it affects.

The Scarring Alopecia Foundation (SAF) is requesting a new ICD-10-CM diagnosis code for central centrifugal cicatricial alopecia. The American Academy of Dermatology/Association (AAD/A) has reviewed and supports the proposal.

### References

1. Dlova NC, Salkey KS, Callender VD, McMichael AJ. Central centrifugal cicatricial alopecia: new insights and a call for action. *J Invest Dermatol Symp Proc* 2017;18:S54-S56.
2. Filbrandt R, Rufaut N, Jones L, Sinclair R. Primary cicatricial alopecia: diagnosis and treatment. *CMAJ*. 2013 Dec 10;185(18):1579-85. doi: 10.1503/cmaj.111570.
3. Aguh C, McMichael A. Central Centrifugal Cicatricial Alopecia. *JAMA Dermatol*. 2020;156(9):1036. doi:10.1001/jamadermatol.2020.1859
4. Malki L, Sarig O, Romano MT, et al. Variant PADI3 in Central Centrifugal Cicatricial Alopecia. *N Engl J Med*. 2019;380(9):833-841.
5. Dlova NC, Jordaan FH, Sarig O, Sprecher E. Autosomal dominant inheritance of central centrifugal cicatricial alopecia in black South Africans. *J Am Acad Dermatol* 2014;70(4):679-682.e1.
6. Flamm A, Moshiri AS, Roche F, Onyekaba G, Nguyen J, James AJ, Taylor S, Seykora JT. Characterization of the inflammatory features of central centrifugal cicatricial alopecia. *J Cutan Pathol*. 2020 Jun;47(6):530-534
7. Callender VD, Wright DR, Davis EC, Sperling LC. Hair Breakage as a Presenting Sign of Early or Occult Central Centrifugal Cicatricial Alopecia: Clinicopathologic Findings in 9 Patients. *Arch Dermatol*. 2012;148(9):1047-1052.

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8. Maranga A, Roche FC, Alausa M, et al. Quality of life in patients with central centrifugal cicatricial alopecia: a preliminary study. *Br J Dermatol.*2022;187(5):802-804.
9. Valdez R, Ouyang L, Bolen J. Public Health and Rare Diseases: Oxymoron No More. *Prev Chronic Dis.* 2016 Jan 14;13:E05.

**TABULAR MODIFICATIONS**

	L66 Cicatricial alopecia [scarring hair loss]
	L66.8 Other cicatricial alopecia
New code	L66.81 Central centrifugal cicatricial alopecia
Add	CCCA
New code	L66.89 Other cicatricial alopecia

## **Cholestatic Pruritus**

Cholestatic pruritus is the itching of the inside and outside of the skin as caused by liver disease. It is described by patients as intense itching on the outside and inside of the skin and persists even with the various drug treatments, making it almost untreatable. Cholestatic pruritus is a very specific condition, and it would be beneficial to physicians, patients, and caregivers to have its own unique ICD-10-CM code.

Cholestatic pruritus has been defined as the impaired secretion of bile, due to liver diseases. This causes intense itching, loss of sleep, general discomfort, and in intense cases suicidal ideation to make the itching stop. The liver disease can be effectively treated; however patients often feel worse due the drug induced symptom of cholestatic pruritus.

The requested modification to the ICD-10-CM codes related to cholestatic pruritus is timely and relevant to align clinical documentation with the needs of clinical practice, patient and provider education, and epidemiology research. Medical and scientific studies have been rapidly evolving and the public health implications are only now starting to be fully recognized.

The Global Liver Institute propose the following tabular modifications.

### **TABULAR MODIFICATIONS**

L29	Pruritus Excludes1: neurotic excoriation (L98.1) psychogenic pruritus (F45.8)
New code	L29.A Cholestatic pruritus

## Coding of Firearm Injuries

The National Center for Health Statistics received a proposal to change the default in the Alphabetic Index for External Causes. The proposal requests that the default code listed in the Index for External Causes for “gunshot wound” be changed from the current default, “W34, accident by unspecified firearm,” to “X95.9, assault by unspecified firearm.” The change is in the intent category to which firearm injuries would default to assault unless another intent category (accidental, intentional self-harm, legal intervention, terrorism or undetermined) is indicated by the medical documentation.

According to the National Electronic Injury Surveillance System (NEISS),<sup>i</sup> 80% of nonfatal firearm injuries are assaults. For each of the other 22 broad mechanisms of injury (e.g., falls, motor vehicles, cutting/piercing instrument, poisoning, etc.) the vast majority (mean 91%, range 75-100%) are accidents. Default codes in the Alphabetic Index represent “that condition that is most commonly associated with the main term or is the unspecified code for the condition”.

Statewide and national hospital discharge data systems i.e. the Nationwide Emergency Department Sample [NEDS] are vital for firearm injury surveillance but have one major flaw, according to a recent NORC report.<sup>ii</sup> NEDS reports that accidents are the leading type of firearm injury, while injury-focused data systems find assaults are the leading type.<sup>1,iii,iv</sup>

The proposed index change is being requested by: Safe States and by Deborah Azrael, Catherine Barber (Harvard Injury Control Research Center, Harvard T. H. Chan School of Public Health); Matthew Miller (Northeastern University); Eric Goralnick, Erin MacPhaul, Ravali Yenduri, Li Zhou (Mass General Brigham Hospitals); Andrew Bowen, Steve Mooney, Ali Rowhani-Rahbar (Harborview Injury Prevention and Research Center, University of Washington)

<sup>1</sup> CDC National Center for Injury Prevention and Control and U.S. Consumer Product Safety Commission. National Electronic Injury Surveillance System ([NEISS](#)) All Injury Program, 2016. ICPSR [distributor], 2020-08-19.

<sup>1</sup> Roman J & Cook P eds. [Improving Data Infrastructure to Reduce Firearms Violence](#). NORC at the University of Chicago, October 2021

<sup>1</sup> Olufajo OA, Zeineddin A, Nonez H, et al. Trends in firearm injuries among children and teenagers in the United States. *J Surg Res*. 2020 Jan;245:529-536. doi: 10.1016/j.jss.2019.07.056. (Includes data on all age groups.)

<sup>1</sup> WRISS data from Beth Hume, Injury Surveillance Program, Massachusetts Department of Public Health.



## Decreased Blood Glucose Level

Hypoglycemia is a condition in which your blood sugar (glucose) level is lower than the normal range; below 70 mg/dL, with clinically established signs and symptoms. For many years, clinically diagnosed hypoglycemia was classified as mild, moderate, and severe. In 2017, these definitions were adjusted as ‘levels’, creating a classification of hypoglycemia. The glucose levels translate from specific glycemic criteria to natural, whole numbers 1-3. The 3 adopted hypoglycemia levels were published in endocrine and diabetic journals as well as adopted by the American Diabetes Association (ADA). The goal of the ADA was to establish clinically meaningful outcome measures as related to hypoglycemia. Level 1 hypoglycemia is defined as a glucose concentration < 70 mg/dL and should be used as an ‘alert value’ to help individuals avoid more severe hypoglycemia. Level 2 hypoglycemia, defined as a glucose concentration < 54 mg/dL, is the threshold at which neuroglycopenic symptoms begin to occur. Level 3 hypoglycemia is defined as a severe event characterized by altered mental and/or physical functioning.

The level system was created by leading experts from the following clinical groups and non-profit organizations:

- American Association of Clinical Endocrinologists
- American Association of Diabetes Educators
- American Diabetes Association
- Endocrine Society
- Juvenile Diabetes Research Foundation (JDRF) International
- The Leona M. and Harry B. Helmsley Charitable Trust
- Pediatric Endocrine Society
- T1D Exchange

Hypoglycemia largely occurs in diabetes type I and diabetes type II patients. Hypoglycemia can be idiopathic, and it is also identified in non-diabetic patient populations to include: pancreatic carcinoma, benign insulinoma, post bariatric surgery patients and chemotherapy patients.

When the hypoglycemia severity level is utilized and documented, it has impact on patient management, diabetic research, and physician decision to order continuous glucose monitoring systems (CGMs).

Dexcom is requesting new codes to better track the severity of these patients.

### References:

<https://diabetes.org/healthy-living/medication-treatments/blood-glucose-testing-and-control/hypoglycemia>  
<https://diabetesjournals.org/care/article/40/12/1622/36909/Standardizing-Clinically-Meaningful-Outcome>

**TABULAR MODIFICATIONS**

	E16	Other disorders of pancreatic internal secretion
New subcategory Add		E16.A Decreased blood glucose level Excludes1: Hypoglycemia, unspecified (E16.2)
New code Add		E16.A1 Decreased blood glucose level 1 Hypoglycemia level 1
New code Add		E16.A2 Decreased blood glucose level 2 Hypoglycemia level 2
New code Add		E16.A3 Decreased blood glucose level 3 Hypoglycemia level 3
	E08	Diabetes mellitus due to underlying condition
		E08.6 Diabetes mellitus due to underlying condition with other specified complications
Add		E08.64 Diabetes mellitus due to underlying condition with hypoglycemia Use additional code for hypoglycemia level, if applicable (E16.5-)
	E09	Drug or chemical induced diabetes mellitus
		E09.6 Drug or chemical induced diabetes mellitus with other specified complications
Add		E09.64 Drug or chemical induced diabetes mellitus with hypoglycemia Use additional code for hypoglycemia level, if applicable (E16.5-)
	E10	Type 1 diabetes mellitus
		E10.6 Type 1 diabetes mellitus with other specified complications
Add		E10.64 Type 1 diabetes mellitus with hypoglycemia Use additional code for hypoglycemia level, if applicable (E16.5-)

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- E11 Type 2 diabetes mellitus
- E09.6 Type 2 diabetes mellitus with other specified complications
- Add E11.64 Type 2 diabetes mellitus with hypoglycemia  
Use additional code for hypoglycemia level, if applicable (E16.5-)
- E13 Other specified diabetes mellitus
- E13.6 Other specified diabetes mellitus with other specified complications
- Add E13.64 Other specified diabetes mellitus with hypoglycemia  
Use additional code for hypoglycemia level, if applicable (E16.5-)
- E16 Other disorders of pancreatic internal secretion
- Add E16.0 Drug-induced hypoglycemia without coma  
Use additional code for hypoglycemia level, if applicable (E16.5-)
- Add E16.1 Other hypoglycemia  
Use additional code for hypoglycemia level, if applicable (E16.5-)

## Developmental and Epileptic Encephalopathies and Related Disorders

A number of specific proposals to create new codes for particular developmental and epileptic encephalopathies (DEEs) as well as various neurodevelopmental disorders have been received and have been under consideration. A 2022 report by Guerrini et al. described these as follows:

*Developmental and epileptic encephalopathies (DEEs) are a heterogeneous group of disorders characterized by early-onset, often severe epileptic seizures and EEG abnormalities on a background of developmental impairment that tends to worsen as a consequence of epilepsy. DEEs may result from both nongenetic and genetic etiologies. Genetic DEEs have been associated with mutations in many genes involved in different functions including cell migration, proliferation, and organization, neuronal excitability, and synapse transmission and plasticity.<sup>1</sup>*

Guerrini also listed 172 genes that may be involved in various DEEs.<sup>1</sup> Many of these are quite rare, and there have not always been epidemiological data on incidence or prevalence. Many are familial, but many also have a nonfamilial form, based on de novo mutations. There has been some relatively recent work on estimating incidence based on detailed assessment of mutation types and rates. In 2020, López-Rivera provided incidence estimates for over 100 genes with known association to neurodevelopmental disorders, along with over 3100 other genes associated with various disorders.<sup>2</sup> Lemke also in 2020 reviewed certain of these, and noted concordance with epidemiologically based incidence estimates where available, as well as noting how these can be used to provide estimated affected incidence based on U.S. birth rates.<sup>3</sup> While these can provide estimates, it is possible that these could be inaccurate for various reasons. López-Rivera noted that predicted incidence estimates might be overestimates due to accounting for a proportion of variants that may be incompatible with human life.<sup>2</sup> Also, since these are based on de novo mutation rates, it appears possible particularly for familial disorders with variable penetrance, that the familial contribution to incidence and prevalence could make these numbers underestimates.<sup>2,3</sup>

It is being proposed to create a specific code for developmental epileptic encephalopathy, along with codes related to genetic susceptibility to epilepsy and neurodevelopmental disorders. These would be expected to be used along with existing codes to convey the full clinical picture.

### References

1. Guerrini R, Conti V, Mantegazza M, et al. Developmental and epileptic encephalopathies: from genetic heterogeneity to phenotypic continuum. *Physiological Reviews* 103(1):433-513. Pub. Online 2022.  
<https://doi.org/10.1152/physrev.00063.2021>  
<https://journals.physiology.org/doi/full/10.1152/physrev.00063.2021>
2. López-Rivera JA, Pérez-Palma E, Symonds J, et al. A catalogue of new incidence estimates of monogenic neurodevelopmental disorders caused by de novo variants. *Brain*, Volume 143, Issue 4, April 2020, Pages 1099–1105.  
<https://doi.org/10.1093/brain/awaa051>
3. Lemke JR. Predicting incidences of neurodevelopmental disorders. *Brain*. 2020 Apr 1;143(4):1046-1048.  
<https://doi.org/10.1093/brain/awaa079>

## TABULAR MODIFICATIONS

### G93 Other disorders of brain

#### G93.4 Other and unspecified encephalopathy

New code	G93.42	Developmental and epileptic encephalopathy
Add		Early infantile epileptic encephalopathy
Add		Code also, if applicable, associated disorders such as:
Add		developmental disorders of scholastic skills (F81.-)
Add		developmental disorder of speech and language (F80.-)
Add		epilepsy, by specific type (G40.-)
Add		intellectual disabilities (F70-F79)
Add		other neurodevelopmental disorder (F88)
Add		pervasive developmental disorders (F84.-)

### Z15 Genetic susceptibility to disease

New subcategory	Z15.1	Genetic susceptibility to epilepsy and neurodevelopmental disorders
Add		Code also, if applicable, related disorders such as
Add		developmental and epileptic encephalopathy (G93.42)
Add		developmental disorders of scholastic skills (F81.-)
Add		developmental disorder of speech and language (F80.-)
Add		epilepsy, by specific type (G40.-)
Add		intellectual disabilities (F70-F79)
Add		other neurodevelopmental disorder (F88)
Add		pervasive developmental disorders (F84.-)
New code	Z15.10	Genetic susceptibility to epilepsy and neurodevelopmental disorders, unspecified

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New sub-subcategory	Z15.11	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to ion channel genes
New code	Z15.110	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of KCNQ2 gene
Add		Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of the gene for potassium voltage-gated channel, KQT-like subfamily, member 2
New code	Z15.111	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of SCN2A gene
Add		Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of the gene for sodium channel, voltage-gated, type II, alpha subunit
New code	Z15.118	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of other ion channel gene
Add		Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of the SCN8A gene
Add		Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of the KCNQ3 gene
New sub-subcategory	Z15.12	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to glutamate receptor genes

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New code	Z15.120	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of GRIN1 gene
Add		Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of the gene for glutamate receptor, ionotropic, N-methyl d-aspartate 1
New code	Z15.121	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of GRIN2A gene
Add		Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of the gene for glutamate receptor, ionotropic, N-methyl d-aspartate 2A
New code	Z15.122	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of GRIN2B gene
Add		Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of the gene for glutamate receptor, ionotropic, N-methyl d-aspartate 2B
New code	Z15.123	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of GRIN2D gene
Add		Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of the gene for glutamate receptor, ionotropic, N-methyl d-aspartate 2D
New code	Z15.124	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of GRIA1 gene
New code	Z15.125	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of GRIA2 gene

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New code	Z15.126	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of GRIA3 gene
New code	Z15.127	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of GRIA4 gene
New code	Z15.12A	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of GRIK2 gene
New code	Z15.128	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of related to other glutamate receptor gene
New code	Z15.13	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to other receptor genes
New sub-subcategory	Z15.14	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to other transporter and solute carrier genes
New code	Z15.140	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of SLC6A1 gene
Add		Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of the gene for solute carrier family 6 (neurotransmitter transporter, GABA), member 1
New code	Z15.141	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to SLC13A5 gene
New code	Z15.148	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of other transporter and solute carrier genes
New sub-		



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subcategory	Z15.15	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to synapse related genes
New code	Z15.150	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of STXBP1 gene
Add		Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of the gene for syntaxin-binding protein 1
New code	Z15.158	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of other synapse related genes
New code	Z15.16	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to genes associated with transcription and gene expression
Add		Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to FOXP1 gene
New code	Z15.19	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to other genes

## **Discogenic Low Back Pain**

A prior proposal related to this was presented in March 2021 ICD-10 Coordination and Maintenance meeting, titled “Lumbar Degenerative Disc Disease With and Without Pain.” This proposal incorporates changes in response to comments.

Physicians utilize a variety of diagnostic labels which lack granularity with regard to lumbar degenerative disc disease (DDD) associated with either midline axial or sclerotomal, non-radicular/non-sciatic referred leg pain. 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 present in the low back, or may be referred to the lower extremity, or both as a result of lumbar discogenic disease. Symptomatic lumbar discogenic disease represents an increasingly large burden to the health care system.

In addition, absence of pain is generally a sign that the degenerative disc disease is non-noxious. Lumbar disc degeneration also is not a definitive diagnosis as it only represents at most a morphologic sub-grade of disc degeneration by the most widely known T2-based Pfirrmann grading scoring tool available for MRI survey interpretation of the lumbar spine. MRI has provided a paradigm shift in how lumbar spine pathology is managed; the diagnosis of DDD predates MRI and was originally based on X-ray. Treatment expectations have evolved with our understanding of provocative discography and MRI. Restorative/regenerative treatment measures address dark discs, Pfirrmann grades 3-7 out of 8 grades. Lumbar disc degeneration may, however, advance further via atraumatic or traumatic mechanisms from fissuring or bulging to a displaced disc herniation and/or stenosis. Later treatment options for lumbar disc herniation and/or stenosis include surgical decompression to address herniation-induced dermatomal radiculopathy/sciatica and stenosis-induced myelopathy. The degeneration process is a cascade, not a stable, static snapshot but rather a developing, dynamic changing presentation.

Back pain location can be described by region. Expansion of coding for the purposes of this proposal are limited to the lumbar region as MRI Pfirrmann grades are confined to the lumbar spine only and thus major advances in spine care have mostly targeted treatment in the lumbar spine. Sciatica has come to mean dermatomal or radicular leg pain and may be differentiated from nociceptive/referred (sclerotomal)/non-radicular pain by exam. That is to say that radiculopathy is diagnosed clinically by a positive straight leg raise, Lasegue’s sign, crossed Lasegue’s sign, positive bowstring, positive femoral stretch test and motor/sensory/reflex change. Symptomatic lumbar discogenic disease is diagnosed clinically by axial midline back pain, pain with flexion, sitting intolerance, positive provocative with sustained hip flexion and absence of motor/sensory/reflex change and positive discography.

Chronic low back pain (CLBP) or lumbago has 6 sources including: (1) discogenic; (2) facetogenic; (3) neurocompressive including herniation and stenosis; (4) sacro-iliac; (5) vertebrogenic; and (6) psychogenic. The predominant source of CLBP is discogenic low back

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pain (DLBP). DLBP represents 30-50% of CLBP, versus facet joint pain ~31%, sacroiliac joint pain ~18% and other sources ~8%<sup>1,2</sup>.

Discogenic back pain associated with DDD can be multifactorial and difficult to treat. The type of pain present and whether it is primarily LBP or leg pain or both is an important component of the clinical assessment. Treatments for discogenic back pain have ranged from anti-inflammatory medications to invasive procedures including spinal fusion and spinal arthroplasty. There has also been a growing interest in developing strategies that aim to repair the degenerative disc biologically, or to supplement tissue lost to degenerative disc disease<sup>3-7</sup>.

ISASS requests updated ICD-10-CM codes that enable the 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 (axial), the lower extremity only, or both, which will be of benefit for distinguishing, tracking, and improving algorithms and treatments for this common and important clinical issue.

References

1. Laplante BL, Ketchum JM, Saullo TR, DePalma MJ. Multivariable Analysis of the Relationship between Pain Referral Patterns and the Source of Chronic Low Back Pain: Retrospective Review. *Pain Physician* 2012;15:171-178. <https://www.painphysicianjournal.com/current/pdf?article=MTU5OQ%3D%3D&journal=66>
2. Schwarzer AC, Aprill CN, Derby R, et al. The Prevalence and Clinical Features of Internal Disc Disruption in Patients With Chronic Low Back Pain. *Spine* (Sep. 1995) 20(17):1878-1883. <https://doi.org/10.1097/00007632-199509000-00007>
- 3: Fujii K, Yamazaki M, Kang JD, et al. Discogenic Back Pain: Literature Review of Definition, Diagnosis, and Treatment. *JBMR Plus*. 2019 Mar 4;3(5):e10180. <https://doi.org/10.1002/jbm4.10180>. PMID: 31131347; PMCID: PMC6524679.
- 4: Kallewaard JW, Terheggen MA, Groen GJ, et al. 15. Discogenic low back pain. *Pain Pract*. 2010 Nov-Dec;10(6):560-79. <https://doi.org/10.1111/j.1533-2500.2010.00408.x>. Epub 2010 Sep 6. PMID: 20825564.
- 5: Peng BG. Pathophysiology, diagnosis, and treatment of discogenic low back pain. *World J Orthop*. 2013; 4(2):42–52. Published 2013 Apr 18. <https://doi.org/10.5312/wjo.v4.i2.42>.
- 6: Beall DP, Wilson GL, Bishop R, Tally T. VAST Clinical Trial: Safely Supplementing Tissue Lost to Degenerative Disc Disease International Journal of Spine Surgery April 2020, 7033; DOI: <https://doi.org/10.14444/7033>.
- 7: Beall DP, Davis TT, DePalma M, et al. Viable disc tissue allograft supplementation; one- and two-level treatment of degenerated intervertebral discs in patients with chronic discogenic low back pain: one year results of the VAST randomized controlled trial. *Pain Physician*. 2021;24(6):465-477. PMID: 34554689. <http://www.painphysicianjournal.com/linkout?issn=&vol=24&page=465>

## 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 discogenic back pain only

Add

Other intervertebral disc degeneration, lumbar region with axial back pain only

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New code	M51.361	Other intervertebral disc degeneration, lumbar region with lower extremity pain only
Add		Other intervertebral disc degeneration, lumbar region with leg pain only
Add		Other intervertebral disc degeneration, lumbar region with referred sclerotomal pain only
New code	M51.362	Other intervertebral disc degeneration, lumbar region with discogenic back pain and lower extremity pain
Add		Other intervertebral disc degeneration, lumbar region with discogenic back pain and leg pain
Add		Other intervertebral disc degeneration, lumbar region with axial back pain and referred sclerotomal pain
New code	M51.369	Other intervertebral disc degeneration, lumbar region without mention of lumbar back pain or lower extremity pain
Add		Other intervertebral disc degeneration, lumbar region without mention of lumbar back pain or leg pain
Add		Other intervertebral disc degeneration, lumbar region, NOS
	M51.37	Other intervertebral disc degeneration, lumbosacral region
New code	M51.370	Other intervertebral disc degeneration, lumbosacral region with discogenic back pain only
Add		Other intervertebral disc degeneration, lumbosacral region with axial back pain only

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New code	M51.371	Other intervertebral disc degeneration, lumbosacral region with lower extremity pain only
Add		Other intervertebral disc degeneration, lumbosacral region with leg pain only
Add		Other intervertebral disc degeneration, lumbosacral region with referred sclerotomal pain only
New code	M51.372	Other intervertebral disc degeneration, lumbosacral region with discogenic back pain and lower extremity pain
Add		Other intervertebral disc degeneration, lumbosacral region with discogenic back pain and leg pain
Add		Other intervertebral disc degeneration, lumbosacral region with axial back pain and referred sclerotomal pain
New code	M51.379	Other intervertebral disc degeneration, lumbosacral region without mention of lumbar back pain or lower extremity pain
Add		Other intervertebral disc degeneration, lumbosacral region without mention of lumbar back pain or leg pain
Add		Other intervertebral disc degeneration, lumbosacral region, NOS

## Disruption of Gastrointestinal Tract Anastomosis

The National Center of Health Statistics received a proposal from Agency for Healthcare Research and Quality (AHRQ) requesting new ICD-10-CM codes for disruption of gastrointestinal tract anastomosis.

Two types of internal operative wound disruptions in the abdominopelvic region—those involving a gastrointestinal tract anastomosis, a.k.a. anastomotic leak, and abdominal fascial dehiscence—warrant special consideration. Colorectal anastomotic leak occurs in 2-4% of cases.<sup>1,2</sup> Anastomotic leak following esophageal resection ranges from 5 to 40 percent.<sup>3</sup> Such leaks frequently result in sepsis and are associated with markedly increased morbidity and mortality.<sup>4</sup> They are, simply stated, among the most feared technical complications for gastrointestinal surgeons. Abdominal fascial dehiscence is also an important complication, occurring in 1-3% of open elective surgery cases and 5-50% of trauma or emergency general surgery cases.<sup>5</sup> This complication has been associated with increased short-term costs, length of stay, readmissions, and mortality,<sup>6,7</sup> as well as a 69% 10-year cumulative incidence of incisional hernia.<sup>8</sup>

AHRQ requests the creation of new ICD-10-CM codes for coding specificity of “Disruption of internal operation (surgical) wound, not elsewhere classified,” to differentiate disruption of gastrointestinal tract anastomosis, a.k.a. anastomotic leak, and fascial dehiscence from each other and from other types of internal operation (surgical) wound disruption or dehiscence.

### References

- <sup>1</sup> Platell C, Barwood N, Dorfmann G, Makin G. The incidence of anastomotic leaks in patients undergoing colorectal surgery. *Colorectal Dis.* 2007 Jan;9(1):71-9. doi: 10.1111/j.1463-1318.2006.01002.x. PMID: 17181849.
- <sup>2</sup> Suding P, Jensen E, Abramson MA, Itani K, Wilson SE. Definitive Risk Factors for Anastomotic Leaks in Elective Open Colorectal Resection. *Arch Surg.* 2008;143(9):907-912. doi:10.1001/archsurg.143.9.907
- <sup>3</sup> Raymond, Daniel P., J. S. Friedberg, and W. Chen. "Complications of esophageal resection." *UpToDate* (2019).
- <sup>4</sup> Kingham TP, Pachter HL. Colonic anastomotic leak: risk factors, diagnosis, and treatment. *J Am Coll Surg.* 2009 Feb;208(2):269-78. doi: 10.1016/j.jamcollsurg.2008.10.015. Epub 2008 Dec 4. PMID: 19228539.
- <sup>5</sup> Crosen M, Sandhu R. Fascial Dehiscence. In *StatPearls* [Internet], <https://www.ncbi.nlm.nih.gov/books/NBK551644/> (accessed June 10, 2022).
- <sup>6</sup> Rosen AK, Loveland S, Shin M, et al. Examining the impact of the AHRQ Patient Safety Indicators (PSIs) on the Veterans Health Administration: the case of readmissions. *Med Care.* 2013;51(1):37-44.
- <sup>7</sup> Gray DM, 2nd, Hefner JL, Nguyen MC, Eiferman D, Moffatt-Bruce SD. The Link Between Clinically Validated Patient Safety Indicators and Clinical Outcomes. *Am J Med Qual.* 2017;32(6):583-590.
- <sup>8</sup> van't Riet M, de Vos van Steenwijk PJ, Bonjer HJ, Steyerberg EW, Jeekel J. Incisional hernia after repair of wound dehiscence: incidence and risk factors. *Am Surg* 2004;70:281-6.

### TABULAR MODIFICATIONS

J86 Pyothorax  
J86.0 Pyothorax with fistula

Add Code also, if applicable, disruption of internal operation (surgical) wound (T81.32-)

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K31 Other diseases of stomach and duodenum  
K31.6 Fistula of stomach and duodenum

Add Code also, if applicable, disruption of internal operation (surgical)  
wound (T81.32-)

K63 Other diseases of intestine  
K63.2 Fistula of intestine

Add Code also, if applicable, disruption of internal operation (surgical)  
wound (T81.32-)

K82 Other diseases of gallbladder  
K82.3 Fistula of gallbladder

Add Code also, if applicable, disruption of internal operation (surgical)  
wound (T81.32-)

T81 Complications of procedures, not elsewhere classified

The appropriate 7th character is to be added to each code from category  
T81

A - initial encounter  
D - subsequent encounter  
S - sequela

T81.3 Disruption of wound, not elsewhere classified

T81.32 Disruption of internal operation (surgical) wound, not  
elsewhere classified

Delete ~~Deep disruption or dehiscence of operation wound NOS~~

Delete ~~Disruption or dehiscence of closure of internal organ or  
other internal tissue~~

Delete ~~Disruption or dehiscence of closure of muscle or  
muscle flap~~

Delete ~~Disruption or dehiscence of closure of ribs or rib cage~~

Delete ~~Disruption or dehiscence of closure of skull or  
craniotomy~~

Delete ~~Disruption or dehiscence of closure of sternum or  
sternotomy~~

Delete ~~Disruption or dehiscence of closure of tendon or  
ligament~~

Delete ~~Disruption or dehiscence of closure of superficial or  
muscular fascia~~

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New code	T81.320	Disruption or dehiscence of gastrointestinal tract anastomosis, repair, or closure
New code	T81.321	Disruption or dehiscence of closure of internal operation (surgical) wound of abdominal wall muscle or fascia
New code	T81.328	Disruption or dehiscence of closure of other specified internal operation (surgical) wound
Add		Disruption or dehiscence of closure of muscle or muscle flap (other than abdominal wall muscle)
Add		Disruption or dehiscence of closure of ribs or rib cage
Add		Disruption or dehiscence of closure of skull or craniotomy
Add		Disruption or dehiscence of closure of sternum or sternotomy
Add		Disruption or dehiscence of closure of tendon or ligament
Add		Disruption or dehiscence of closure of superficial or muscular fascia (other than abdominal wall fascia)
New code	T81.329	Deep disruption or dehiscence of operation wound, unspecified
Add		Deep disruption or dehiscence of operation wound NOS

T81.4 Infection following a procedure

Add Code also, if applicable, disruption of internal operation (surgical) wound (T81.32-)

T81.8 Other complications of procedures, not elsewhere classified

T81.83 Persistent postprocedural fistula

Add Code also, if applicable, disruption of internal operation (surgical) wound (T81.32-)



## Eating Disorders

This proposal was presented at the September 2022 ICD-10 Coordination and Maintenance Committee meeting. In response to comments received, a revised proposal is presented for reconsideration. Changes are noted in **bold**.

The Fifth Edition of the Diagnostic and Statistical Manual of Mental Disorders (DSM-5), published by the American Psychiatric Association (APA), is a clinical classification with relevant terminology designed to facilitate reliable and consistent diagnosis of mental and behavioral health conditions. The classification subcategorizes many clinical diagnoses by severity, course, and other descriptive feature specifiers.

Most of these specifiers and defining features are reflected in the ICD-10-CM today as unique codes or via inclusion terms (e.g., Bipolar Disorders, Major Depressive Disorder, Substance Use Disorders, and Neurocognitive Disorders).

Kaiser Permanente Medicine requests the following tabular modifications to further the alignment of the two publications by updating the ICD-10-CM to recognize the diagnostic subcategories and other descriptive features of the DSM-5 for feeding and eating disorders (e.g., severity and remission).

The American Psychiatric Association has reviewed and supports this proposal.

### TABULAR MODIFICATIONS

#### F50 Eating disorders

Excludes1: anorexia NOS (R63.0)  
feeding problems of newborn (P92.-)  
polyphagia (R63.2)

Excludes2: feeding difficulties (R63.3)  
feeding disorder in infancy or childhood (F98.2-)

#### F50.0 Anorexia nervosa

Excludes1: loss of appetite (R63.0)  
psychogenic loss of appetite (F50.89)

F50.00 Anorexia nervosa, unspecified

#### **F50.01 Anorexia nervosa, restricting type**

New code

**F50.010 Anorexia nervosa, restricting type, mild**

New code

**F50.011 Anorexia nervosa, restricting type, moderate**

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New code **F50.012 Anorexia nervosa, restricting type, severe**  
New code **F50.013 Anorexia nervosa, restricting type, extreme**  
New code **F50.014 Anorexia nervosa, restricting type, in remission**  
Add **Anorexia nervosa, restricting type, in partial remission**  
Add **Anorexia nervosa, restricting type, in full remission**  
New code **F50.019 Anorexia nervosa, restricting type, unspecified**

**F50.02 Anorexia nervosa, binge eating/purging type**  
Excludes1: bulimia nervosa (F50.2)

New code **F50.020 Anorexia nervosa, binge eating/purging type, mild**  
New code **F50.021 Anorexia nervosa, binge eating/purging type, moderate**  
New code **F50.022 Anorexia nervosa, binge eating/purging type, severe**  
New code **F50.023 Anorexia nervosa, binge eating/purging type, extreme**  
New code **F50.024 Anorexia nervosa, binge eating/purging type, in remission**  
**Anorexia nervosa, restricting type, in partial remission**  
Add **Anorexia nervosa, restricting type, in full remission**  
New code **F50.029 Anorexia nervosa, binge eating/purging type, unspecified**

**F50.2 Bulimia nervosa**

Bulimia NOS

Hyperorexia nervosa

Excludes1: anorexia nervosa, binge eating/purging type (F50.02)

New code **F50.20 Bulimia nervosa, unspecified**  
New code **F50.21 Bulimia nervosa, mild**  
New code **F50.22 Bulimia nervosa, moderate**  
New code **F50.23 Bulimia nervosa, severe**  
New code **F50.24 Bulimia nervosa, extreme**

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<b>New code</b>	<b>F50.25 Bulimia nervosa, in remission</b>
<b>Add</b>	<b>Bulimia nervosa, in partial remission</b>
<b>Add</b>	<b>Bulimia nervosa, in full remission</b>
	<b>F50.8 Other eating disorders</b>
Delete	<del>Excludes2: pica of infancy and childhood (F98.3)</del>
	<b>F50.81 Binge eating disorder</b>
<b>New code</b>	<b>F50.810 Binge eating disorder, mild</b>
<b>New code</b>	<b>F50.811 Binge eating disorder, moderate</b>
<b>New code</b>	<b>F50.812 Binge eating disorder, severe</b>
<b>New code</b>	<b>F50.813 Binge eating disorder, extreme</b>
<b>New code</b>	<b>F50.814 Binge eating disorder, in remission</b>
<b>Add</b>	<b>Binge eating disorder, in partial remission</b>
<b>Add</b>	<b>Binge eating disorder, in full remission</b>
<b>New code</b>	<b>F50.819 Binge eating disorder, unspecified</b>
	<b>F50.82 Avoidant/restrictive food intake disorder</b>
Add	Avoidant/restrictive food intake disorder, in remission
<b>New code</b>	<b>F50.83 Pica in adults</b>
	Pica in adults, in remission
	Excludes1: pica in infancy and childhood (F98.3)
<b>New code</b>	<b>F50.84 Rumination disorder in adults</b>
Add	Rumination disorder in adults, in remission
	Excludes1: Rumination disorder in infancy and childhood (F98.21)
	<b>F50.89 Other specified eating disorder</b>
Delete	<del>Pica in adults</del>
	Psychogenic loss of appetite

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F98.2 Other feeding disorders of infancy and childhood

Excludes2: anorexia nervosa and other eating disorders (F50.-)  
feeding difficulties (R63.3)  
feeding problems of newborn (P92.-)  
pica in infancy or childhood (F98.3)

Revise

F98.21 Rumination disorder of infancy and childhood

Add

Rumination disorder in infancy or childhood, in remission

Add

Excludes1: Rumination disorder in adults (F50.84)

F98.3 Pica of infancy and childhood

Add

Pica in infancy or childhood, in remission

Add

Excludes1: pica in adults (F50.83)

## **Encounter for Sepsis Aftercare**

The National Center of Health Statistics received a proposal requesting a new ICD-10-CM code for post-acute care encounter for sepsis aftercare from University of Colorado/UCHealth.

Sepsis survivors have a high readmission risk due to post-acute complications and sequelae of sepsis after hospital discharge.<sup>1</sup> Around 40% of sepsis survivors discharged to post-acute care are readmitted to the hospital within 90 days. Of those readmitted within 30 days, over 40% do so within seven days post-discharge due to infection, organ dysfunction, or both, that newly develop, reoccur, or worsen.<sup>1,2,3,4</sup> To facilitate rehabilitation and target preventive care after hospital discharge, it is critical that sepsis survivorship is acknowledged in the post-acute and ambulatory medical records.

Post-acute care, and home health care (HHC) in particular, play an essential role in ensuring a continuum of care for sepsis survivors. A recent study, using a dataset of national Medicare claims, revealed that of 165,000 sepsis survivors who transitioned from acute care to HHC, only 7–10% were coded with sepsis-related diagnostic codes in the HHC record.<sup>5</sup> Invisible to post-acute and ambulatory care, many sepsis survivors will not receive the necessary sepsis-focused care to aid recovery and prevent adverse events.

Recognition of sepsis survivorship is essential because the recovery from sepsis is complex and lengthy, lasting months to years.<sup>4,6,7</sup> For example, 62.5% of sepsis survivors experience more than one infection within 12 months post-discharge.<sup>8</sup> Post-sepsis syndrome (PSS) refers to the numerous long-term sequelae of sepsis. PSS, which can include immune dysregulation, cardiovascular and renal problems, cognitive impairment, mental health issues, and physical impairments, affects up to half of all sepsis survivors. Once developed, PSS can increase readmission risk, increase long-term mortality, lead to poor health-related quality of life, and increase health-related costs.<sup>9</sup> A sepsis-specific code for post-acute and ambulatory care is necessary because readmission risk after sepsis is high, independent of illness severity or ICU admission,<sup>10</sup> and the need to rehabilitate neuropsychological and physical impairments are common after sepsis.<sup>11,12,13</sup>

A new code would address post-acute care needs specific to sepsis survivorship and address the limitations of existing codes. Specifically, use of the existing sepsis codes is inappropriate in most post-acute and community-based care situations, as the life-threatening condition of sepsis will have been resolved by hospital discharge. Also, existing alternative codes for aftercare, follow-up, history, and sequelae are not specific to sepsis, missing the opportunity to classify and warn patients, their family members, and clinicians about sepsis-specific risks such as new or recurrent infections and to rehabilitate new impairments. A new ICD-10-CM code will address these limitations by providing coding specificity for the growing population of sepsis survivors who frequently require post-acute and ambulatory care. More specifically, a new code will: 1) increase the awareness among health care providers of the risks of sepsis survivors, 2) improve adherence to best practice recommendations for the management of long-term outcomes of sepsis,<sup>14</sup> 3) optimize care coordination and the delivery of post-acute and ambulatory care services aligned with sepsis survivors' needs, 4) catalyze necessary policy changes to ensure

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improved outcomes of sepsis survivors, and 5) promote epidemiological monitoring to further scientific advances in the management of long-term sequelae of sepsis.<sup>15</sup>

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#### References

1. Shankar-Hari M, Saha R, Wilson J, Prescott HC, Harrison D, Rowan K, Rubenfeld GD, Adhikari NKJ. Rate and risk factors for rehospitalisation in sepsis survivors: systematic review and meta-analysis. *Intensive Care Med.* 2020;46(4):619-636.
2. Gadre SK, Shah M, Mireles-Cabodevila E, Patel B, Duggal A. Epidemiology and Predictors of 30-Day Readmission in Patients With Sepsis. *CHEST.* 2019;155(3):483-490.
3. Prescott HC, Langa KM, Iwashyna TJ. Readmission diagnoses after hospitalization for severe sepsis and other acute medical conditions. *JAMA.* 2015;313(10):1055-1057.
4. Prescott HC, Angus DC. Enhancing Recovery From Sepsis: A Review. *JAMA.* 2018;319(1):62-75.
5. Bowles KH, Murtaugh CM, Jordan L, Barrón Y, Mikkelsen ME, Whitehouse CR, Chase JD, Ryvicker M, Feldman PH. Sepsis Survivors Transitioned to Home Health Care: Characteristics and Early Readmission Risk Factors. *J Am Med Dir Assoc.* 2020;21(1):84-90.
6. Prescott HC, Costa DK. Improving Long-Term Outcomes After Sepsis. *Crit Care Clin.* 2018;34(1):175-188.
7. Gritte RB, Souza-Siqueira T, Curi R, Machado MCC, Soriano FG. Why Septic Patients Remain Sick After Hospital Discharge? *Front Immunol.* 2021;11:605666.
8. Arens C, Bajwa SA, Koch C, Siegler BH, Schneck E, Hecker A, Weiterer S, Lichtenstern C, Weigand MA, Uhle F. Sepsis-induced long-term immune paralysis—results of a descriptive, explorative study. *Crit Care.* 2016;20:93.
9. van der Slikke EC, An AY, Hancock REW, Bouma HR. Exploring the pathophysiology of post-sepsis syndrome to identify therapeutic opportunities. *EBioMedicine.* 2020;61:103044.
10. Riegel B, Huang L, Mikkelsen ME, Kutney-Lee A, Hanlon AL, Murtaugh CM, Bowles KH. Early Post-Intensive Care Syndrome among Older Adult Sepsis Survivors Receiving Home Care. *J Am Geriatr Soc.* 2019;67(3):520-526.
11. Iwashyna TJ, Ely EW, Smith DM, Langa KM. Long-term cognitive impairment and functional disability among survivors of severe sepsis. *JAMA.* 2010;304(16):1787-1794.
12. Odden AJ, Rohde JM, Bonham C, Kuhn L, Malani PN, Chen LM, Flanders SA, Iwashyna TJ. Functional outcomes of general medical patients with severe sepsis. *BMC Infect Dis.* 2013;13:588.
13. Jones LC, Dion C, Efron PA, Price CC. Sepsis and Cognitive Assessment. *J Clin Med.* 2021;10(18):4269.
14. Evans L, Rhodes A, Alhazzani W, Antonelli M, Coopersmith CM, French C, Machado FR, McIntyre L, Ostermann M, Prescott HC, Schorr C, Simpson S, Wiersinga WJ, Alshamsi F, Angus DC, Arabi Y, Azevedo L, Beale R, Beilman G, Belley-Cote E, Burry L, Cecconi M, Centofanti J, Coz Yataco A, De Waele J, Dellinger RP, Doi K, Du B, Estenssoro E, Ferrer R, Gomersall C, Hodgson C, Møller MH, Iwashyna T, Jacob S, Kleinpell R, Klompas M, Koh Y, Kumar A, Kwizera A, Lobo S, Masur H, McGloughlin S, Mehta S, Mehta Y, Mer M, Nunnally M, Oczkowski S, Osborn T, Papathanassoglou E, Perner A, Puskarich M, Roberts J, Schweickert W, Seckel M, Sevransky J, Sprung CL, Welte T, Zimmerman J, Levy M. Surviving sepsis campaign: international guidelines for management of sepsis and septic shock 2021. *Intensive Care Med.* 2021;47(11):1181-1247.
15. Oh S, Mikkelsen ME, O'Connor M, Bowles KH. Why Sepsis Survivors Need an ICD-10 Code for "Sepsis Aftercare". *Chest.* 2022 Nov;162(5):979-981.

### TABULAR MODIFICATION

Z51 Encounter for other aftercare and medical care

Code also condition requiring care

Excludes1: follow-up examination after treatment (Z08-Z09)

New code

Z51.A Encounter for sepsis aftercare

## **Epileptic Seizures Related to External Causes, Intractable**

The National Center for Health Statistics received a proposal requesting new ICD-10-CM codes for epileptic seizures related to external causes, intractable with and without status epilepticus.

Central nervous system toxicity caused by drug exposure is a common reason for presentation to the emergency department. Manifestations include psychoses, various levels of impairment of consciousness, and seizures. No characteristic clinical features differentiate drug-induced seizures from idiopathic epileptic seizures. Generalized seizures with focal features are common, whereas simple partial seizures are rare. Generalized seizures usually present with loss of consciousness, convulsions, tongue biting, and incontinence of urine. There is usually no preceding aura or focal disturbance and no residual neurologic deficit. Severe attacks may lead to status epilepticus, which is drug-induced in 10% of cases.

Drug-induced seizures need to be sorted out the same way as any other form of epileptic seizure. In epileptic patients, differentiation from spontaneous seizures is based on the course of the disorder. Possible seizure-inducing effects of antiepileptic drugs should be considered if seizure frequency increases on introduction of a new drug, is associated with an increase of dose, or occurs despite continued treatment with previous antiepileptic drugs. Suspicion of association with an antiepileptic drug can be verified by discontinuation of the drug followed by improvement of seizures and aggravation of seizures on reintroduction of the drug.

Most drug-induced seizures are self-limiting and do not result in severe sequelae. About 15% may present as status epilepticus, and this has potential for morbidity. The treatment of drug-induced seizures is usually like that of seizures due to other causes, although drug-induced seizures are more difficult to control. Drug-induced seizures are frequently toxic-metabolic in nature, and because they lack a focal brain lesion, they may be less responsive to conventional antiepileptic drugs such as phenytoin.

### **References**

Jain, K. K. Drug-induced seizures. *Neuropharmacology and Neurotherapeutics*, MedLink Neurology. <https://www.medlink.com/articles/drug-induced-seizures>. Published 11/16/1998. Updated 9/11/2021. Accessed 12/21/22.

**TABULAR MODIFICATIONS**

G40 Epilepsy and recurrent seizures

G40.5 Epileptic seizures related to external causes

New  
sub-subcategory

G40.51 Epileptic seizures related to external causes, intractable

Add

G40.511 Epileptic seizures related to external causes,  
intractable, with status epilepticus

Add

G40.519 Epileptic seizures related to external causes,  
intractable, without status epilepticus



## Expansion of Synovitis and Tenosynovitis, unspecified

The American Health Information Management Association (AHIMA) is requesting to expand code M65.9, Synovitis and tenosynovitis, unspecified, to identify specific anatomic sites. This would be consistent with other subcategories in category M65 as well as the World Health Organization (WHO) ICD-10. The WHO version of ICD-10 identifies the anatomic site for unspecified synovitis and tenosynovitis.

### TABULAR MODIFICATIONS

	M65	Synovitis and tenosynovitis
		M65.9 Synovitis and tenosynovitis, unspecified
New code		M65.90 Unspecified synovitis and tenosynovitis, unspecified site
New subcategory		M65.91 Unspecified synovitis and tenosynovitis, shoulder
New code		M65.911 Unspecified synovitis and tenosynovitis, right shoulder
New code		M65.912 Unspecified synovitis and tenosynovitis, left shoulder
New code		M65.919 Unspecified synovitis and tenosynovitis, unspecified shoulder
New subcategory		M65.92 Unspecified synovitis and tenosynovitis, upper arm
New code		M65.921 Unspecified synovitis and tenosynovitis, right upper arm
New code		M65.922 Unspecified synovitis and tenosynovitis, left upper arm
New code		M65.929 Unspecified synovitis and tenosynovitis, unspecified upper arm
New subcategory		M65.93 Unspecified synovitis and tenosynovitis, forearm
New code		M65.931 Unspecified synovitis and tenosynovitis, right forearm
New code		M65.932 Unspecified synovitis and tenosynovitis, left forearm
New code		M65.939 Unspecified synovitis and tenosynovitis, unspecified forearm
New sub-category		M65.94 Unspecified synovitis and tenosynovitis, hand
New code		M65.941 Unspecified synovitis and tenosynovitis, right hand

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New code	M65.942 Unspecified synovitis and tenosynovitis, left hand
New code	M65.949 Unspecified synovitis and tenosynovitis, unspecified hand
New subcategory	M65.95 Unspecified synovitis and tenosynovitis, thigh
New code	M65.951 Unspecified synovitis and tenosynovitis, right thigh
New code	M65.952 Unspecified synovitis and tenosynovitis, left thigh
New code	M65.959 Unspecified synovitis and tenosynovitis, unspecified thigh
New subcategory	M65.96 Unspecified synovitis and tenosynovitis, lower leg
New code	M65.961 Unspecified synovitis and tenosynovitis, right lower leg
New code	M65.962 Unspecified synovitis and tenosynovitis, left lower leg
New code	M65.969 Unspecified synovitis and tenosynovitis, unspecified lower leg
New subcategory	M65.97 Unspecified synovitis and tenosynovitis, ankle and foot
New code	M65.971 Unspecified synovitis and tenosynovitis, right ankle and foot
New code	M65.972 Unspecified synovitis and tenosynovitis, left ankle and foot
New code	M65.979 Unspecified synovitis and tenosynovitis, unspecified ankle and foot
New code	M65.98 Unspecified synovitis and tenosynovitis, other site
New code	M65.99 Unspecified synovitis and tenosynovitis, multiple sites

## Fanconi Anemia

Fanconi anemia (FA) is a rare genetic disorder (estimated incidence of 1:136,000 live births<sup>1</sup>) of defective deoxyribonucleic acid (DNA) repair characterized by congenital malformations, progressive bone marrow failure (aplastic anemia), and increased risk for solid and hematologic malignancies.<sup>2,3</sup> In most cases, FA is an autosomal recessive disease resulting from biallelic mutations in a particular FA gene. To date, at least 23 FA genes have been identified, all of which are involved in the DNA repair pathway.<sup>4</sup> FA-A (i.e., mutations in *FANCA* gene) is the most common (60-70%), followed by FA-C (*FANCC* gene) (14%) and FA-G (*FANCG* gene) (10%).<sup>4</sup> The inability to repair detrimental DNA damage results in chromosomal instability and increased risk of congenital malformations and malignancies<sup>2,5</sup>.

A hallmark of the FA phenotype is severe bone marrow failure (BMF) which develops in 80% of patients during the initial decade of life.<sup>6</sup> The majority (60-70%) of FA patients have at least one major congenital malformation which may vary in severity across any of multiple major organ systems, and which may result in spontaneous abortion or perinatal death.<sup>7,8,9</sup> Additionally, endocrinopathies e.g. glucose/insulin abnormalities, growth hormone insufficiency and hypothyroidism are common.

An accurate diagnosis of FA requires the clinician to first recognize the plethora of possible clinical malformations associated with FA and differentiation of these phenotypic features from other syndromes. To the trained practitioner, most FA patients will have short stature relative to unaffected family members, skin pigmentation abnormalities, hypoplasia of the thenar eminence and microphthalmia. Increased awareness of the complete spectrum of the minor malformations observed in these patients should enable an earlier diagnosis even in the absence of BMF.<sup>7</sup>

Cytopenias and progressive BMF occur early (median age of 7 years) and in almost all patients (90-98%) by age 40.<sup>6,10,11</sup> Severe aplastic anemia, or bone marrow failure (BMF), may lead to death before 10 years of age in the absence of a formal diagnosis.<sup>4</sup> In addition to BMF, over 30% of patients with FA experience hematologic malignancies (most frequently acute myeloid leukemia (AML) or myelodysplastic syndrome (MDS) by the age of 40.<sup>6,12</sup> To date, allogeneic hematopoietic stem cell transplant (HSCT) is the only curative treatment option for the hematologic manifestations of FA.<sup>5</sup> At transplant centers with experience treating FA patients, survival in patients with human leukocyte antigen (HLA) matched donors can exceed 80-90%<sup>12,13</sup>; however, the efficacy of allogeneic transplant is limited by the availability of a suitable donor and risks of mortality and complications, such as infections, graft failure, graft-vs-host disease (GvHD) (acute and chronic) and organ damage.

As a consequence of the rarity of FA, the diagnosis is often delayed or missed in >70% of patients.<sup>14,15,16</sup> In the absence of an appropriate diagnosis code, there is limited opportunity for routine surveillance and effective communication across health care teams.

Establishment of a code for FA will also provide the opportunity to intervene and minimize the impact of learning disabilities, hearing and vision deficits and various endocrinopathies, including growth failure, in children with FA that are likely to be missed. An ICD-10-CM code

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would significantly aid physicians and patients in avoiding potentially serious negative consequences from treatments administered without consideration of the underlying DNA repair defect in FA. For example, FA patients undergoing allogeneic HSCT or cancer treatment must receive modified chemotherapy and/or radiation doses.

Furthermore, a FA-specific ICD-10-CM code may facilitate appropriate FA treatment, while limiting use of inappropriate treatments by physicians and providers who may not otherwise be aware of FA and optimal management standards.

An ICD-10-CM code can also help in identification of patients for clinical trials evaluating potentially valuable treatment options. Finally, an ICD-10-CM will improve the ability to conduct research to facilitate improved understanding of the prevalence and natural history of the disease, genotype-phenotype correlations and enable tailored and long-term monitoring of outcomes from future therapies.

This proposal for a specific code for FA is submitted by John E. Wagner, MD, University of Minnesota, and Magnolia Innovation, with assistance from the medical team at Rocket Pharmaceuticals, Inc.

It is supported by the Fanconi Anemia Research Fund, the lead organization supporting research and education on FA, and the following renowned clinicians and thought leaders in the field: Dr. Jennifer Grandis, American Cancer Society Professor, Otolaryngology, University of California San Francisco; Dr. Rajni Agarwal-Hashmi, Professor of Pediatrics, Stanford University.

#### REFERENCES

1. Che R, Zhang J, Nepal M, Han B, Fei P. Multifaceted Fanconi Anemia Signaling. *Trends Genet.* 2018 Mar; 34(3):171-183.
2. Moreno OM, Paredes AC, Suarez-Obando F, Rojas A. An update on Fanconi anemia: Clinical, cytogenetic and molecular approaches (Review). *Biomed Rep.* 2021;15(3):74.
3. Niraj J, Farkkila A, and D'Andrea A. The Fanconi Anemia Pathway in Cancer. *Annu Rev Cancer Biol* 2019; 3:457-478.
4. Mehta A, and Ebens C. Fanconi Anemia. *GeneReviews.* Seattle (WA): University of Washington, Seattle; 2002-2021.
5. Bhandari J, Thada , Puckett Y. Fanconi Anemia. *Treasure Island (FL): StatPearls Publishing.* 2022
6. Kutler DI, Singh B, Satagopan J, Batish SD, Berwick M, Giampietro PF, Hanenberg H, Auerbach AD. A 20-year perspective on the International Fanconi Anemia Registry (IFAR). *Blood.* 2003 Feb 15;101(4):1249-56.
7. Giampietro PF, Verlander PC, Davis JG, Auerbach AD. Diagnosis of Fanconi anemia in patients without congenital malformations: An International Fanconi Anemia Registry study. *Am J Med Genet* 1997; 68: 58-61.
8. Shimamura A, Alter BP. Pathophysiology and management of inherited bone marrow failure syndromes. *Blood Rev.* 2010 May;24(3):101-22.
9. Wagner JE, Jakub Tolar, Auerbach AD, MacMillan ML. Hematopoietic Cell Transplantation for Fanconi Anemia. Blume KG, Forman SJ, Appelbaum FR, eds. *Thomas' Hematopoietic Cell Transplantation 5th Edition.* Blackwell Publishing Ltd, Oxford, (2015).
10. Ebens CL, DeFor TE, Tryon R, Wagner JE, MacMillan ML. Comparable Outcomes after HLA-Matched Sibling and Alternative Donor Hematopoietic Cell Transplantation for Children with Fanconi Anemia and Severe Aplastic Anemia. *Biol Blood Marrow Transplant.* 2018 Apr;24(4):765-771.
11. Butturini A, Gale RP, Verlander PC, Adler-Brecher B, Gillio AP, Auerbach AD. Hematologic abnormalities in Fanconi anemia: an International Fanconi Anemia Registry study. *Blood.* 1994 Sep 1;84(5):1650-5.
12. Ebens CL, MacMillan ML, Wagner JE. Hematopoietic cell transplantation in Fanconi anemia: current evidence, challenges and recommendations. *Expert Rev Hematol.* 2017; 10:81-97.

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13. Mehta PA, Davies SM, Leemhuis T, Myers K, Kernan NA, Prockop SE, Scaradavou A, O'Reilly RJ, Williams DA, Lehmann L, Guinan E, Margolis D, Baker KS, Lane A, Boulad F. Radiation-free, alternative-donor HCT for Fanconi anemia patients: results from a prospective multi-institutional study. *Blood*. 2017 Apr 20;129(16):2308-2315.
14. Wu ZH. The concept and practice of Fanconi Anemia: from the clinical bedside to the laboratory bench. *Transl Pediatr*. 2013 Jul;2(3):112-9.
15. Giampietro PF, Adler-Brecher B, Verlander PC, Pavlakis SG, Davis JG, Auerbach AD. The need for more accurate and timely diagnosis in Fanconi anemia: a report from the International Fanconi Anemia Registry. *Pediatrics* 1993;91:1116–20.
16. Huck K, Helmut H, Gudowius S, Fenk R, Kalb R, Neveling K, Betz B, Niederacher D, Haas R, Gobel U, Kobbe G, Schilndler D. Delayed diagnosis and complications of Fanconi anaemia at advanced age – a paradigm. *British Journal of Haematology* 2006; 133:188-197.

**TABULAR MODIFICATIONS**

D61 Other aplastic anemias and other bone marrow failure syndromes  
Excludes2: neutropenia (D70.-)

D61.0 Constitutional aplastic anemia

D61.01 Constitutional (pure) red blood cell aplasia

Blackfan-Diamond syndrome

Congenital (pure) red cell aplasia

Familial hypoplastic anemia

Primary (pure) red cell aplasia

Red cell (pure) aplasia of infants

Excludes1: acquired red cell aplasia (D60.9)

New Code

Add

Add

D61.02 Fanconi anemia

Fanconi pancytopenia

Fanconi 's anemia

Delete

D61.09 Other constitutional aplastic anemia

~~Fanconi's Anemia~~

Pancytopenia with malformations

**INDEX MODIFICATIONS**

Add

Add

Fanconi hypoplastic anemia D61.02

Fanconi panmyelopathy D61.02

## Flank Anatomical Specificity

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

This proposal was presented at the March 2021 and September 2022 ICD 10 Coordination and Maintenance meeting. In response to public comments, a revised proposal is being submitted for reconsideration. Changes are noted in **bold**.

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

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

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

### TABULAR MODIFICATIONS

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

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	L02.21 Cutaneous abscess of trunk
Revise	L02.212 Cutaneous abscess of back [any part, except buttock <u>and flank</u> ]
New code	L02.217 Cutaneous abscess of flank
	L02.22 Furuncle of trunk
	Boil of trunk
	Folliculitis of trunk
Revised	L02.222 Furuncle of back [any part, except buttock <u>and flank</u> ]
New code	L02.227 Furuncle of flank
	L02.23 Carbuncle of trunk
Revise	L02.232 Carbuncle of back [any part, except buttock <u>and flank</u> ]
New code	L02.237 Carbuncle of flank
	L03.3 Cellulitis and acute lymphangitis of trunk
	L03.31 Cellulitis of trunk
New code	L03.31A Cellulitis of flank
	L03.32 Acute lymphangitis of trunk
New code	L03.32A Acute lymphangitis of flank
R10	Abdominal and pelvic pain
	Excludes1: renal colic (N23)
Add	Excludes2: costovertebral (angle) tenderness (R39.85)
	dorsalgia (M54.-)
Add	flatulence and related conditions (R14.-)
	R10.1 Pain localized to upper abdomen
Add	<b>Excludes2: pain localized to flank (R10.A-)</b>
Add	pelvic and perineal pain (R10.2-)
	R10.2 Pelvic and perineal pain
Add	Excludes2: pain localized to other parts of lower abdomen(R10.3-)
Add	pain localized to upper abdomen (R10.1-)
New code	R10.20 Pelvic and perineal pain unspecified side
New code	R10.21 Pelvic and perineal pain right side

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New code	R10.22 Pelvic and perineal pain left side
New code	R10.23 Pelvic and perineal pain bilateral
New code	R10.24 Suprapubic pain
	R10.3 Pain localized to other parts of lower abdomen
<b>Add</b>	<b>Excludes2: pain localized to flank (R10.A-)</b>
Add	pelvic and perineal pain (R10.2-)
<b>New subcategory</b>	<b>R10.A Pain localized to flank</b>
<b>Add</b>	<b>Lateral abdomen pain</b>
<b>Add</b>	<b>Lateral flank pain</b>
<b>Add</b>	<b>Latus region pain</b>
Add	Excludes2: pain localized to other parts of lower abdomen (R10.3-)
Add	pain localized to upper abdomen (R10.1-)
New code	R10.A0 Flank pain, unspecified side
New code	R10.A1 Flank pain, right side
New code	R10.A2 Flank pain, left side
New code	R10.A3 Flank pain, bilateral
	R10.8 Other abdominal pain
<b>New subcategory</b>	<b>R10.8A Flank tenderness</b>
New code	R10.8A1 Right flank tenderness
New code	R10.8A2 Left flank tenderness
New code	R10.8A3 Suprapubic tenderness
<b>New code</b>	<b>R10.8A9 Flank tenderness, unspecified</b>
<b>Add</b>	<b>Flank tenderness NOS</b>
New code	R10.85 Abdominal pain of multiple sites
Add	Excludes1: abdominal rigidity NOS (R19.3)
Add	generalized abdominal pain associated with acute abdomen (R10.0)
Add	generalized abdominal pain NOS (R10.84)
Add	localized abdominal pain (R10.1-R10.4-)



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R39.8 Other symptoms and signs involving the genitourinary system

<b>New sub-category</b>	<b>R39.85 Costovertebral (angle) tenderness</b>
<b>Add</b>	<b>Excludes2: abdominal and pelvic pain (R10.-)</b>
<b>New code</b>	<b>R39.851 Costovertebral (angle) tenderness, right side</b>
<b>New code</b>	<b>R39.852 Costovertebral (angle) tenderness, left side</b>
<b>New code</b>	<b>R39.853 Costovertebral (angle) tenderness, bilateral</b>
<b>New code</b>	<b>R39.859 Costovertebral (angle) tenderness, unspecified side</b>

	<b>S30.1 Contusion of abdominal wall and flank</b>
<b>Add</b>	<b>Contusion of abdominal wall</b>
<b>Add</b>	<b>Contusion of latus region</b>
<b>Delete</b>	<del>Contusion of flank</del>
<b>Delete</b>	<del>Contusion of groin</del>

<b>New code</b>	<b>S30.10 Contusion of abdominal wall and flank, unspecified</b>
<b>New code</b>	<b>S30.11 Contusion of abdominal wall and flank</b>
<b>New code</b>	<b>S30.12 Contusion of flank</b>
<b>New code</b>	<b>S30.13 Contusion of groin</b>

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

	S30.81 Abrasion of abdomen, lower back, pelvis, and external genitals
<b>New code</b>	S30.81A Abrasion of flank

	S30.82 Blister (nonthermal) of abdomen, lower back, pelvis, and external genitals
<b>New code</b>	S30.82A Blister (nonthermal) of flank

	S30.84 External constriction of abdomen, lower back, pelvis and external genitals
<b>New code</b>	S30.84A External constriction of flank

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New code	S30.85 Superficial foreign body of abdomen, lower back, pelvis, and external genitals S30.85A Superficial foreign body of flank
New code	S30.86 Insect bite (nonvenomous) of abdomen, lower back, pelvis, and external genitals S30.86A Insect bite (nonvenomous) of flank
New code	S30.87 Other superficial bite of abdomen, lower back, pelvis, and external genitals S30.87A Other superficial bite of flank
New code	S30.9 Unspecified superficial injury of abdomen, lower back, pelvis, and external genitals S30.9A Unspecified superficial injury of flank
	S31.1 Open wound of abdominal wall without penetration into peritoneal cavity
	S31.10 Unspecified open wound of abdominal wall without penetration into peritoneal cavity
New code	S31.106 Unspecified open wound of abdominal wall, right flank without penetration into peritoneal cavity
New code	S31.107 Unspecified open wound of abdominal wall, left flank without penetration into peritoneal cavity
New code	S31.10A Unspecified open wound of abdominal wall, unspecified flank without penetration into peritoneal cavity
Add	Open wound of abdominal wall of flank NOS without penetration into peritoneal cavity

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S31.11 Laceration without foreign body of abdominal wall without penetration into peritoneal cavity

New code S31.116 Laceration without foreign body of abdominal wall, right flank without penetration into peritoneal cavity

New code S31.117 Laceration without foreign body of abdominal wall, left flank without penetration into peritoneal cavity

New code S31.11A Laceration without foreign body of abdominal wall, unspecified flank without penetration into peritoneal cavity

Add Laceration without foreign body of flank NOS without penetration into peritoneal cavity

S31.12 Laceration with foreign body of abdominal wall without penetration into peritoneal cavity

New code S31.126 Laceration with foreign body of abdominal wall, right flank without penetration into peritoneal cavity

New code S31.127 Laceration with foreign body of abdominal wall, left flank without penetration into peritoneal cavity

New code S31.12A Laceration with foreign body of abdominal wall unspecified flank without penetration into peritoneal cavity

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

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

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

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

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

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

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

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

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

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

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

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

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

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New code	S31.157 Open bite of abdominal wall, left flank without penetration into peritoneal cavity
New code	S31.15A Open bite of abdominal wall, unspecified flank without penetration into peritoneal cavity
Add	Open bite of abdominal wall of flank NOS without penetration into peritoneal cavity
S31.6 Open wound of abdominal wall with penetration into peritoneal cavity	
S31.60 Unspecified open wound of abdominal wall with penetration into peritoneal cavity	
New code	S31.606 Unspecified open wound of abdominal wall, right flank with penetration into peritoneal cavity
New code	S31.607 Unspecified open wound of abdominal wall, left flank with penetration into peritoneal cavity
New code	S31.60A Unspecified open wound of abdominal wall, unspecified flank with penetration into peritoneal cavity
Add	Unspecified open wound of abdominal wall of flank NOS, with penetration into peritoneal cavity
S31.61 Laceration without foreign body of abdominal wall with penetration into peritoneal cavity	
New code	S31.616 Laceration without foreign body of abdominal wall, right flank with penetration into peritoneal cavity
New code	S31.617 Laceration without foreign body of abdominal wall, left flank with penetration into peritoneal cavity

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New code	S31.61A Laceration without foreign body of abdominal wall, unspecified flank with penetration into peritoneal cavity
Add	Laceration without foreign body of abdominal wall of flank NOS, with penetration into peritoneal cavity
	S31.62 Laceration with foreign body of abdominal wall with penetration into peritoneal cavity
New code	S31.626 Laceration with foreign body of abdominal wall, right flank with penetration into peritoneal cavity
New code	S31.627 Laceration with foreign body of abdominal wall, left flank with penetration into peritoneal cavity
New code	S31.62A Laceration with foreign body of abdominal wall, unspecified flank with penetration into peritoneal cavity
Add	Laceration with foreign body of abdominal wall, flank NOS, with penetration into peritoneal cavity
	S31.63 Puncture wound without foreign body of abdominal wall with penetration into peritoneal cavity
New code	S31.636 Puncture wound of abdominal wall without foreign body, right flank with penetration into peritoneal cavity
New code	S31.637 Puncture wound of abdominal wall without foreign body, left flank with penetration into peritoneal cavity

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New code	S31.63A Puncture wound of abdominal wall without foreign body, unspecified flank with penetration into peritoneal cavity
Add	Puncture wound of abdominal wall without foreign body, flank NOS, with penetration into peritoneal cavity
	S31.64 Puncture wound with foreign body of abdominal wall with penetration into peritoneal cavity
New code	S31.646 Puncture wound of abdominal wall with foreign body, right flank with penetration into peritoneal cavity
New code	S31.647 Puncture wound of abdominal wall with foreign body, left flank with penetration into peritoneal cavity
New code	S31.64A Puncture wound of abdominal wall with foreign body, unspecified flank with penetration into peritoneal cavity
Add	Puncture wound of abdominal wall with foreign body, flank NOS, with penetration into peritoneal cavity
	S31.65 Open bite of abdominal wall with penetration into peritoneal cavity
New code	S31.656 Open bite of abdominal wall, right flank with penetration into peritoneal cavity
New code	S31.657 Open bite of abdominal wall, left flank with penetration into peritoneal cavity
New code	S31.65A Open bite of abdominal wall, unspecified flank with penetration into peritoneal cavity
Add	Open bite of abdominal wall, flank NOS, with penetration into peritoneal cavity

## **Frontal Fibrosing Alopecia**

Frontal fibrosing alopecia (FFA) is a primary cicatricial (scarring) alopecia first characterized in 1994. FFA is traditionally considered a clinical variant of lichen planopilaris (LPP). LPP historically included three clinically distinct cicatricial alopecias: FFA, classic LPP, and Graham Little Syndrome (GLS). FFA, classic LPP, and GLS were grouped under LPP due to shared histologic features and were considered rare disorders. Increased incidence of FFA has led to advancements in the clinical and pathologic characterization of the disease.

FFA has a clinical presentation unique from GLS and classic LPP. FFA is characterized by irreversible hair loss along the frontotemporal scalp in a band-like distribution and can include loss of the eyebrows, eyelashes, and other body hair. Those with FFA can have associated pruritus and perifollicular erythema/scale but the scalp signs and symptoms in FFA are generally milder than those seen in classic LPP. Patients with FFA may have other unique features including prominent facial veins, facial papules, and frontal vein depression. Both patients with FFA and classic LPP experience a high psychosocial burden of disease including increased depression, impaired self-esteem, and an overall decreased quality of life.

The epidemiology of FFA is not well-established, but recent literature suggests it is increasing in incidence. FFA is most common in Caucasian women following menopause but it also affects men and has an average age of onset between 56 and 66. The cause of irreversible hair loss seen in FFA, classic LPP, and GLS involves inflammation at the hair follicle, leading to the permanent damage of hair stem cells and follicular fibrosis. Recent evidence suggests neural and autoimmune mediated inflammation seen in FFA may be distinct from that seen in other forms of LPP. Additionally, a genome wide association (GWAS) study has identified genetic susceptibility loci unique to FFA.

Currently, there is no specific ICD-10-CM code for FFA. A new ICD-10-CM code for FFA will facilitate improved disease characterization, potentially identify unknown comorbidities and provide quality data for epidemiologic studies at the population level.

The Scarring Alopecia Foundation (SAF) submitted this proposal for consideration. The American Academy of Dermatology/Association (AAD/A) has reviewed and supports the proposal.

### References

1. Kossard S. Postmenopausal frontal fibrosing alopecia. Scarring alopecia in a pattern distribution. *Archives of Dermatology*. 1994;130(6):770-774.
2. Olsen EA, Bergfeld WF, Cotsarelis G, et al. Summary of North American Hair Research Society (NAHRS)-sponsored Workshop on Cicatricial Alopecia, Duke University Medical Center, February 10 and 11, 2001. *Journal of the American Academy of Dermatology*. 2003;48(1):103-110.
3. Mirmirani P, Willey A, Headington JT, Stenn K, McCalmont TH, Price VH. Primary cicatricial alopecia: Histopathologic findings do not distinguish clinical variants. *Journal of the American Academy of Dermatology*. 2005;52(4):637-643.



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4. McSweeney SM, Christou EAA, Dand N, et al. Frontal fibrosing alopecia: a descriptive cross-sectional study of 711 female cases from the U.K. *Br J Dermatol*. Published online July 11, 2020:bjd.19399.
5. Vañó-Galván S, Molina-Ruiz AM, Serrano-Falcón C, et al. Frontal fibrosing alopecia: A multicenter review of 355 patients. *Journal of the American Academy of Dermatology*. 2014;70(4):670-678.
6. Meinhard J, Stroux A, Lünemann L, Vogt A, Blume-Peytavi U. Lichen planopilaris: Epidemiology and prevalence of subtypes - a retrospective analysis in 104 patients: Lichen planopilaris: retrospective study. *JDDG: Journal der Deutschen Dermatologischen Gesellschaft*. 2014;12(3):229-235.
7. Pirmez R, Duque-Estrada B, Barreto T, Quintella DC, Cuzzi T. Successful Treatment of Facial Papules in Frontal Fibrosing Alopecia with Oral Isotretinoin. *Skin Appendage Disord*. 2017;3(2):111-113.
8. Vañó-Galván S, Rodrigues-Barata AR, Urech M, et al. Depression of the frontal veins: A new clinical sign of frontal fibrosing alopecia. *Journal of the American Academy of Dermatology*. 2015;72(6):1087-1088.
9. Alirezaci P, Ahmadpanah M, Rezaejad A, Soltanian A, Sadeghi Bahmani D, Brand S. Compared to Controls, Individuals with Lichen Planopilaris Have More Depression, a Lower Self-Esteem, and a Lower Quality of Life. *Neuropsychobiology*. 2019;78(2):95-103.
10. Chiang YZ, Bundy C, Griffiths CEM, Paus R, Harries MJ. The role of beliefs: lessons from a pilot study on illness perception, psychological distress and quality of life in patients with primary cicatricial alopecia. *Br J Dermatol*. 2015;172(1):130-137. doi:10.1111/bjd.13259
11. Doche I, Wilcox GL, Ericson M, et al. Evidence for neurogenic inflammation in lichen planopilaris and frontal fibrosing alopecia pathogenic mechanism. *Exp Dermatol*. 2020;29(3):282-285.
12. Prasad S, Marks DH, Burns LJ, et al. Patch testing and contact allergen avoidance in patients with lichen planopilaris and/or frontal fibrosing alopecia: A cohort study. *Journal of the American Academy of Dermatology*. 2020;83(2):659-661.
13. Harries M, Hardman J, Chaudhry I, Poblet E, Paus R. Profiling the human hair follicle immune system in lichen planopilaris and frontal fibrosing alopecia: can macrophage polarization differentiate these two conditions microscopically? *Br J Dermatol*. 2020;183(3):537-547.
14. Tziotzios C, Petridis C, Dand N, et al. Genome-wide association study in frontal fibrosing alopecia identifies four susceptibility loci including HLA-B\*07:02. *Nat Commun*. 2019;10(1):1150.

## TABULAR MODIFICATIONS

### L66 Cicatricial alopecia [scarring hair loss]

New Subcategory	L66.1 Lichen planopilaris
Delete	<del>Follicular lichen planus</del>
New code	L66.10 Lichen planopilaris, unspecified
New code	L66.11 Classic lichen planopilaris
Add	Follicular lichen planus
New code	L66.12 Frontal fibrosing alopecia
Add	FFA
New code	L66.19 Other lichen planopilaris
Add	Lassueur Graham-Little Piccardi syndrome

## **Glutamate Receptor, Ionotropic, gene-related Neurodevelopmental Disorders**

The CureGRIN Foundation has proposed new ICD-10-CM codes for nine distinct neurological disorders related to the ionotropic glutamate receptor, each with a specific genetic basis. These include GRIN1-Related Neurodevelopmental Disorder, GRIN2A-Related Neurodevelopmental Disorder, GRIN2B-Related Neurodevelopmental Disorder, GRIN2D-Related Neurodevelopmental Disorder, GRIA1-Related Neurodevelopmental Disorder, GRIA2-Related Neurodevelopmental Disorder, GRIA3-Related Neurodevelopmental Disorder, GRIA4-Related Neurodevelopmental Disorder, and GRIK2-Related Neurodevelopmental Disorder.

These disorders each have a unique phenotype that is related to their membership in a larger family of ionotropic glutamate receptors (iGluRs). iGluRs mediate most excitatory synaptic transmission in the central nervous system. Each iGluR is distinct in its genetics and function in the central nervous system and have been richly studied.<sup>1</sup> The iGluRs play an important role in learning and memory as well as other critical biological functions including regulation of movements and biological rhythms such as breathing. The expression and function of each iGluR is developmentally regulated. Each iGluR has a specific role to play in specific, anatomically distinct central nervous system functions at different times during development. Many are involved in directing the wiring or connections of the central nervous system.<sup>1</sup> All of the aforementioned genes (GRIN1, GRIN2A, GRIN2B, GRIN2D, GRIA1, GRIA2, GRIA3, GRIA4, and GRIK2) are highly expressed in the brain, though there is expression in other tissues.

The specific symptoms, temporal onset of symptoms, and severity of symptoms of each of these disorders are related to the specific alteration of a specific iGluR. The GRIN1, GRIN2A, GRIN2B, and GRIN2D are among the genes that encode the subunits of the NMDA receptor.<sup>1</sup> NMDA receptors are assembled by these subunits in different combinations that are developmentally, functionally, and anatomically regulated. Each of these is associated with a broad spectrum of phenotypes ranging in severity. Symptoms include developmental delay, intellectual disability, autism, speech deficiency, inability to walk, low muscle tone, gastrointestinal issues, feeding difficulties, cortical visual impairments, dystonia, seizures, and paroxysmal sympathetic hyperactivity (PSH).<sup>1-5</sup>

GRIN1-related neurodevelopmental disorder often is associated with developmental delay, intellectual disability, hypotonia, spasticity of the limbs, movement disorders, behavior disorders, cortical visual impairment, feeding difficulties, gastrointestinal abnormalities, microcephaly, and malformations of cortical development.<sup>2</sup> GRIN1 variants are associated with epileptic encephalopathies, with seizure semiology including infantile spasms, tonic and atonic seizures, focal dyscognitive seizures, febrile seizures, hypermotor seizures, generalized tonic-clonic seizures, and status epilepticus.<sup>2</sup> The predicted incidence per 100,000 births is 5.45 for GRIN1.<sup>6,7</sup>

GRIN2A-related neurodevelopmental disorder is may be associated with severe developmental and epileptic encephalopathy (DEE). GRIN2A variants may be associated with epilepsy syndromes such as Landau-Kleffner syndrome.<sup>8,9</sup> Focal seizures are the predominant presentation of GRIN2A-related epilepsy, seen in more than half of affected individuals.<sup>8</sup>

Generalized seizures can occur in up to 10% of cases and comprise atonic, atypical absence, myoclonic, and generalized tonic-clonic seizures. However, some individuals do not have seizures. Speech disorders are often seen in those with GRIN2A variants (over 90%), ranging from mild speech delay to severe speech and language difficulties.<sup>8</sup> About one-third of individuals have normal development and normal intellect.<sup>9</sup> The predicted incidence per 100,000 births is 3.23 for GRIN2A.<sup>6,7</sup>

GRIN2B-related neurodevelopmental disorder is often associated with developmental delay, intellectual disability, hypotonia, spasticity, movement disorders, behavior disorders, cortical visual impairment, feeding difficulties, and microcephaly.<sup>4</sup> The seizure semiology of GRIN2B-related epilepsy includes generalized seizures (mostly tonic or tonic-clonic), focal seizures, and epileptic spasms.<sup>4</sup> Two specific epilepsy syndromes that are evident in some individuals with GRIN2B variants include West syndrome and Lennox-Gastaut syndrome.<sup>4</sup> The prevalence of GRIN2B-related neurodevelopmental disorder among individuals with neurodevelopmental disorders/childhood-onset epilepsy is estimated at 0.2%.<sup>10</sup> The predicted incidence per 100,000 births is 5.91 for GRIN2B.<sup>6,7</sup>

GRIN2D-related neurodevelopmental disorder often has been associated with developmental delay, intellectual disability, hypotonia, spasticity, movement disorders, behavior disorders, cortical visual impairment, feeding difficulties, and microcephaly.<sup>11</sup> It presents with refractory epilepsy in early infancy, and DEE.<sup>11</sup> The predicted incidence per 100,000 births is 4.61 for GRIN2D.<sup>6,7</sup>

GRIA1, GRIA2, GRIA3, GRIA4 encode the subunits of the AMPA receptor.<sup>1</sup> GRIA variants have been associated with intellectual disability and autism spectrum disorder (ASD).<sup>12-21</sup> In addition, some individuals with variants in the GRIA genes may experience behavioral difficulties including features of autism, reduced attention span, anxiety, and hypersensitivity to stimuli. Each GRIA gene disorder is distinct.

GRIA1-related neurodevelopmental disorder is associated with neurodevelopmental delay and speech difficulties.<sup>22</sup> Some patients have moderate to severe cognitive impairment, and some also display anxiety, autism spectrum disorder (ASD), and ADHD phenotypes.<sup>22</sup> Epileptic seizures have also been reported.<sup>22</sup> The predicted incidence per 100,000 births 2.55 for GRIA1.<sup>6,7</sup>

GRIA2-related neurodevelopmental disorder patients display intellectual disability (ID) and neurodevelopmental abnormalities including autism spectrum disorder (ASD), Rett syndrome-like features, and seizures or developmental epileptic encephalopathy (DEE).<sup>14</sup> The predicted incidence per 100,000 births is 2.79 for GRIA2.<sup>6,7</sup>

GRIA3-related neurodevelopmental disorder involves X-linked intellectual disability and is also sometimes referred to as Wu syndrome. For patients with variants in GRIA3, there have been numerous X-linked cases in males, inherited from their mother, and a few de novo cases in females. Findings include epilepsy, mild to severe intellectual disability, bipolar affective disorder, and autistic spectrum disorders.<sup>19</sup> Additional features include hypotonia, asthenic body habitus with poor muscle bulk, and hyporeflexia.<sup>19</sup> Generalized chorea and multifocal myoclonus

have also been observed in patients with GRIA3 variants.<sup>20</sup> The predicted incidence per 100,000 births is 5.45 for 2.48 for GRIA3.<sup>6,7</sup>

GRIA4- related neurodevelopmental disorder results in neurodevelopmental disorder with or without seizure and gait abnormalities, severe developmental delay, limb hypertonia, generalized seizures, retinal hypoplasia, chorioretinal hyperpigmentation, and problems of social behavior.<sup>21,23</sup> The predicted incidence per 100,000 births is 2.08 for GRIA4.<sup>6,7</sup>

GRIK2- related neurodevelopmental disorder results in symptoms such as intellectual disability, developmental delay, seizures, and visual or sensory changes.<sup>24</sup> GRIK2 is a gene that encode the kainate receptor.<sup>1</sup> GRIK2 disorder is distinct from the other iGluR disorders. The predicted incidence per 100,000 births is 1.87 for GRIK2.<sup>6,7</sup>

Accurate and timely diagnosis is required to pursue adequate treatment and precision therapy for affected individuals. Many affected individuals are dependent on the support and care of guardians and caregivers throughout life. Unique ICD-10-CM codes will assist with identifying affected individuals and enabling specific therapies as well as tracking patients and evaluating the impact of each of these disorders on healthcare systems and decision-making.

#### References

1. Hansen KB, Wollmuth LP, Bowie D, et al. Structure, Function, and Pharmacology of Glutamate Receptor Ion Channels. *Pharmacol Rev.* 2021 Oct; 73(4): 298–487. Pub. online 2021. PMID: 34753794  
<https://doi.org/10.1124/pharmrev.120.000131>
2. Platzer K, Lemke JR. GRIN1-Related Neurodevelopmental Disorder. 2019 Jun 20. In: Adam MP, Ardinger HH, Pagon RA, et al., editors. *GeneReviews*® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2021. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK542807>
3. Lemke JR, Lal D, Reinthaler EM, et al. Mutations in GRIN2A cause idiopathic focal epilepsy with rolandic spikes. *Nat Genet.* 2013 Sep;45(9):1067-72. <https://doi.org/10.1038/ng.2728> Epub 2013 Aug 11. PMID: 23933819.
4. Platzer K, Lemke JR. GRIN2B-Related Neurodevelopmental Disorder. 2018 May 31. In: Adam MP, Ardinger HH, Pagon RA, Wallace SE, Bean LJH, Stephens K, Amemiya A, editors. *GeneReviews*® [Internet]. Seattle (WA): University of Washington, Seattle; 1993–2020. PMID: 29851452. <https://www.ncbi.nlm.nih.gov/books/NBK501979/>
5. Xiang Wei W, Kannan V, Xu Y, et al. Heterogeneous clinical and functional features of GRIN2D-related developmental and epileptic encephalopathy. *Brain.* 2019 Oct 1;142(10):3009-3027. <https://doi.org/10.1093/brain/awz232>. PMID: 31504254; PMID: PMC6763743.
6. López-Rivera JA, Pérez-Palma E, Symonds J, Lindy AS, McKnight DA, Leu C, Zuberi S, Brunklaus A, Möller RS, Lal D. A catalogue of new incidence estimates of monogenic neurodevelopmental disorders caused by de novo variants. *Brain.* 2020 Apr 1;143(4):1099-1105. <https://doi.org/10.1093/brain/awaa051>. PMID: 32168371; PMID: PMC7174049.
7. Lemke JR. Predicting incidences of neurodevelopmental disorders. *Brain.* 2020 Apr 1;143(4):1046-1048. <https://doi.org/10.1093/brain/awaa079>. PMID: 32318731.
8. Strehlow V, Heyne HO, Vlaskamp DRM, et al. GRIN2A study group. GRIN2A-related disorders: genotype and functional consequence predict phenotype. *Brain.* 2019 Jan 1;142(1):80-92. <https://doi.org/10.1093/brain/awy304>. PMID: 30544257; PMID: PMC6308310.
9. GRIN2A-related Epilepsy. Epilepsy Foundation. <https://www.epilepsy.com/causes/genetic/grin2a-related-epilepsy>
10. Platzer K, Yuan H, Schütz H, et al. GRIN2B encephalopathy: novel findings on phenotype, variant clustering, functional consequences and treatment aspects. *J Med Genet.* 2017;54:460–70. <https://doi.org/10.1136/jmedgenet-2016-104509>. PMID: PMC5656050. PMID: 28377535
11. Platzer K, Krey I, Lemke JR. GRIN2D-Related Developmental and Epileptic Encephalopathy. 2022 July 28. In: Adam MP, Everman DB, Mirzaa GM, et al., editors. *GeneReviews*® [Internet]. Seattle (WA): University of Washington, Seattle; 1993–2023. <https://www.ncbi.nlm.nih.gov/books/NBK582335/>
12. de Ligt J, Willemsen MH, van Bon BW, et al. Diagnostic exome sequencing in persons with severe intellectual disability. *N Engl J Med.* 2012 Nov 15;367(20):1921-9. <https://doi.org/10.1056/NEJMoal206524>. Epub 2012 Oct 3. PMID: 23033978.
13. Geisheker MR, Heymann G, Wang T, et al. Hotspots of missense mutation identify neurodevelopmental disorder genes and functional domains. *Nat Neurosci.* 2017 Aug; 20(8): 1043–1051. Published online 2017 Jun 19. <https://doi.org/10.1038/nn.4589>. PMID: PMC5539915. PMID: 28628100.

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14. Salpietro V, Dixon CL, Guo H, et al. AMPA receptor GluA2 subunit defects are a cause of neurodevelopmental disorders. *Nat Commun.* 2019 Jul 12;10(1):3094. PMID: 31300657. PMCID: PMC6626132. <https://doi.org/10.1038/s41467-019-10910-w>.
15. Wu Y, Arai AC, Rumbaugh G, et al. Mutations in ionotropic AMPA receptor 3 alter channel properties and are associated with moderate cognitive impairment in humans. *Proc Natl Acad Sci U S A.* 2007 Nov 13;104(46):18163-8. Epub 2007 Nov 7. PMID: 17989220. PMCID: PMC2084314. <https://doi.org/10.1073/pnas.0708699104>
16. Bonnet C, Leheup B, Beri M, et al. Aberrant GRIA3 transcripts with multi-exon duplications in a family with X-linked mental retardation. *Am J Med Genet A.* 2009 Jun;149A(6):1280-9. PMID: 19449417 <https://doi.org/10.1002/ajmg.a.32858>
17. Philippe A, Malan V, Jacquemont ML, et al. Xq25 duplications encompassing GRIA3 and STAG2 genes in two families convey recognizable X-linked intellectual disability with distinctive facial appearance. *Am J Med Genet A.* 2013 Jun;161A(6):1370-5. Epub 2013 May 1. PMID: 23637084. <https://doi.org/10.1002/ajmg.a.35307>
18. Davies B, Brown LA, Cais O, et al. A point mutation in the ion conduction pore of AMPA receptor GRIA3 causes dramatically perturbed sleep patterns as well as intellectual disability. *Hum Mol Genet.* 2017 Oct 15;26(20):3869-3882. PMID: 29016847. PMCID: PMC5639461. <https://doi.org/10.1093/hmg/ddx270>
19. Trivisano M, Santarone ME, Micalizzi A, et al. GRIA3 missense mutation is cause of an x-linked developmental and epileptic encephalopathy. *Seizure.* 2020 Nov;82:1-6. Epub 2020 Sep 22. PMID: 32977175. <https://doi.org/10.1016/j.seizure.2020.08.032>
20. Piard J, Bereau M, XiangWei W, et al. The GRIA3 c.2477G > A Variant Causes an Exaggerated Startle Reflex, Chorea, and Multifocal Myoclonus. *Mov Disord.* 2020 Jul; 35(7): 1224–1232. Published online 2020 May 5. PMCID: PMC9190290. PMID: 32369665. <https://doi.org/10.1002/mds.28058>
21. Martin S, Chamberlin A, Shinde DN, et al. De Novo Variants in GRIA4 Lead to Intellectual Disability with or without Seizures and Gait Abnormalities. *Am J Hum Genet.* 2017 Dec 7;101(6):1013-1020. PMID: 29220673. PMCID: PMC5812909. <https://doi.org/10.1016/j.ajhg.2017.11.004>
22. Ismail V, Zachariassen LG, Godwin A, et al. Identification and functional evaluation of GRIA1 missense and truncation variants in individuals with ID: An emerging neurodevelopmental syndrome. *Am J Hum Genet.* 2022 Jul 7;109(7):1217-1241. Epub 2022 Jun 7. PMID: 35675825 PMCID: PMC9300760. <https://doi.org/10.1016/j.ajhg.2022.05.009>
23. Wang TM, Brown BM, Deng L, et al. A novel NMDA receptor positive allosteric modulator that acts via the transmembrane domain. *Neuropharmacology.* 2017 Jul 15;121:204-218. Epub 2017 Apr 27. PMID: 28457974. <https://doi.org/10.1016/j.neuropharm.2017.04.041>
24. Stolz JR, Foote KM, Veenstra-Knol HV, et al. Clustered mutations in the GRIK2 kainate receptor subunit gene underlie diverse neurodevelopmental disorders. *Am J Hum Genet.* 2021 Sep 2;108(9):1692-1709. Epub 2021 Aug 9. PMID: 34375587 PMCID: PMC8456161. <https://doi.org/10.1016/j.ajhg.2021.07.007>

## TABULAR MODIFICATIONS

### Option #1

Note: this option was created based on a modified approach from the original submission.

F84 Pervasive developmental disorders

F84.8 Other pervasive developmental disorders

New sub-subcategory	F84.83	Glutamate receptor-related neurodevelopmental disorders
Add		Code also, if applicable, related disorders such as
Add		developmental disorders of scholastic skills (F81.-)
Add		developmental disorder of speech and language (F80.-)
Add		epilepsy, by specific type (G40.-)
Add		intellectual disabilities (F70-F79)

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New code	F84.830	GRIN1-related neurodevelopmental disorder
New code	F84.831	GRIN2A-related neurodevelopmental disorder
New code	F84.832	GRIN2B-related neurodevelopmental disorder
New code	F84.833	GRIN2D-related neurodevelopmental disorder
New code	F84.834	GRIA1-related neurodevelopmental disorder
New code	F84.835	GRIA2-related neurodevelopmental disorder
New code	F84.836	GRIA3-related neurodevelopmental disorder
New code	F84.837	GRIA4-related neurodevelopmental disorder
New code	F84.83A	GRIK2-related neurodevelopmental disorder
New code	F84.838	Other glutamate receptor, ionotropic, related neurodevelopmental disorder

**Option #2**

Note: this option was created by NCHS based on a generalized approach for a number of proposals. See also the Developmental Epileptic Encephalopathies topic.

Z15 Genetic susceptibility to disease

New subcategory	Z15.1	Genetic susceptibility to epilepsy and neurodevelopmental disorders
New sub-subcategory	Z15.12	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to glutamate receptor genes
New code	Z15.120	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of GRIN1 gene
Add		Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to

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		pathogenic variant of the gene for glutamate receptor, ionotropic, N-methyl d-aspartate 1
New code	Z15.121	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of GRIN2A gene
Add		Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of the gene for glutamate receptor, ionotropic, N-methyl d-aspartate 2A
New code	Z15.122	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of GRIN2B gene
Add		Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of the gene for glutamate receptor, ionotropic, N-methyl d-aspartate 2B
New code	Z15.123	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of GRIN2D gene
Add		Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of the gene for glutamate receptor, ionotropic, N-methyl d-aspartate 2D
New code	Z15.124	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of GRIA1 gene
New code	Z15.125	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of GRIA2 gene
New code	Z15.126	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of GRIA3 gene

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New code	Z15.127	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of GRIA4 gene
New code	Z15.12A	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of GRIK2 gene
New code	Z15.128	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of related to other glutamate receptor gene



## Gulf War Illness

Gulf War Illness (GWI) is an exposure-induced chronic multisymptom illness affecting personnel who served in the 1990-1991 Persian Gulf conflict and tied to drug/environment exposures associated with that conflict. A solid body of evidence implicates such exposures, including dose-response and gene-environment interaction data. New, unique, and excessive exposure occurred to multiple agents, distributed differently across deployed personnel. This included exposure to organophosphate nerve agents (sarin, cyclosarin), prolonged exposure to multiple pesticides (including organophosphates and carbamates, among others), pyridostigmine bromide (PB) as a nerve agent pretreatment adjunct, anthrax and botulinum toxoid vaccines and multiple vaccines, permethrin-impregnated uniforms, DEET insect repellent, depleted uranium, ciprofloxacin, and other agents. Especially strong evidence ties acetylcholinesterase inhibiting agents including organophosphates (as pesticides and nerve agents) and carbamates (such as PB and carbamate pesticides) to GWI.

Epidemiological studies affirm that deployment to the 1990-1991 conflict and associated exposures are tied to marked increases specifically in an empirically-defined multisymptom profile. Data show no difference in the fraction of individuals reporting symptoms that have persisted for at least six months in 1-2 defined symptom domains, but a marked difference in the fraction reporting symptoms across 3-6 distinct symptom domains in Gulf War-deployed personnel vs. Gulf War-era veterans (who were not deployed to the Gulf War Theatre).

This health condition requires deployment to the Gulf War Theater of Operations anytime between August 1, 1990, and July 31, 1991. It requires chronic symptoms for  $\geq 6$  months, arising during or after Gulf deployment, in  $\geq 3$  of the 6 Kansas criteria questionnaire with symptom domains of fatigue/sleep; neurological/cognitive/mood; pain; gastrointestinal; respiratory; and dermatologic. For a domain to qualify for this condition, symptoms in the domain must be either of at least moderate severity (not mild) and/or there must be multiple symptoms in that domain. Currently, there is no existing ICD-10-CM code which adequately captures this health condition.

The consortium of GWI clinicians, researchers, and patient advocates propose the following tabular modifications to advance epidemiological tracking of this condition and health outcomes in affected veterans.

### TABULAR MODIFICATIONS

	Z77	Other contact with and (suspected) exposures hazardous to health
New Code	Z77.3	Contact with and (suspected) exposure to Persian Gulf theater
Add		Gulf theater exposure NOS
Add		Gulf War illness
Add		Gulf War syndrome
Add		Use additional code to identify associated manifestations

## Injection Drug Use

Despite the devastating toll exacted by injection drug use (IDU) on morbidity and mortality<sup>1-8</sup>, there is currently no existing diagnosis code to identify injection drug use (IDU). The impact of not having a specified code, does impede population-based surveillance, research, and tracking as well as proactive offering of prevention, testing, vaccination, treatment, and harm reduction services to these high-risk patients.

In the absence of ICD-10-CM codes, surveillance efforts require the field to use various combinations of non-specific ICD-10-CM codes to identify IDU with the published literature rife with approaches showing widely varying degrees of sensitivity and specificity<sup>9-12</sup>. The need for ICD-10-CM codes to capture IDU is critical given groundbreaking support from The White House for syringe services programs (SSPs) that “have a proven track record of reducing disease, increasing access to addiction treatment and improving public safety.”<sup>13</sup>.

Moreover, for the first time in its history, the 2022 National Drug Control Strategy includes a focus on evidence-based harm reduction, including SSPs<sup>14</sup>. SSPs are also endorsed by the Centers for Disease Control and Prevention (CDC), Surgeon General of the United States, World Health Organization (WHO), American Medical Association, and American Bar Association<sup>15-19</sup>.

Despite widespread need and support for SSPs, the experience of VHA early adopters of SSPs have identified a clear need for ICD-10-CM codes to enable case finding of people with IDU who would be appropriate for SSPs<sup>20</sup>. Furthermore, not knowing how many persons have IDU also hampers public health from knowing how many SSPs are needed and where they are needed.

Lack of a code has been identified as a major barrier to implementation with patients often being identified too late, downstream, and usually as a result of a complication arising from IDU such as HIV and hepatitis C virus (HCV) infection<sup>3-5</sup>. This is concerning given that the number of acute HCV infections has more than doubled between 2013 to 2020 with IDU the number one risk behavior for these infections.<sup>21</sup>

A unique ICD-10-CM code would identify IDUs to facilitate surveillance, research, treatment, harm reduction and outcomes monitoring among this vulnerable population.

The Veterans Health Administration’s (VHA) Office of Mental Health and Suicide Prevention (OMHSP) has reviewed and supports this proposal developed by Elizabeth Dinges, PharmD Karine Rozenberg, PharmD, and Elizabeth Oliva\*, PhD, VHA National Opioid Overdose Education and Naloxone Distribution Coordinator, VHA OMHSP.

### References

1. Handanagic S, Finlayson T, Burnett JC, et al. HIV Infection and HIV-Associated Behaviors Among Persons Who Inject Drugs - 23 Metropolitan Statistical Areas, United States, 2018. *MMWR Morb Mortal Wkly Rep.* 2021 Oct 22;70(42):1459-1465.
2. O'Donnell J, Tanz LJ, Gladden RM, et al. Trends in and Characteristics of Drug Overdose Deaths Involving Illicitly Manufactured Fentanyl - United States, 2019-2020. *MMWR Morb Mortal Wkly Rep.* 2021 Dec 17;70(50):1740-1746.
3. Zibbell JE, Iqbal K, Patel RC, et al. Increases in hepatitis C virus infection related to injection drug use among persons aged ≤30 years - Kentucky, Tennessee, Virginia, and West Virginia, 2006-2012. *MMWR Morb Mortal Wkly Rep.* 2015 May 8;64(17):453-8.

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4. Harris AM, Iqbal K, Schillie S, et al. Increases in Acute Hepatitis B Virus Infections - Kentucky, Tennessee, and West Virginia, 2006-2013. *MMWR Morb Mortal Wkly Rep.* 2016 Jan 29;65(3):47-50.
5. Peters PJ, Pontones P, Hoover KW, et al. HIV Infection Linked to Injection Use of Oxymorphone in Indiana, 2014-2015. *N Engl J Med.* 2016 Jul 21;375(3):229-39.
6. Dasgupta S, Tie Y, Lemons A, et al. Injection Practices and Sexual Behaviors Among Persons with Diagnosed HIV Infection Who Inject Drugs - United States, 2015-2017. *MMWR Morb Mortal Wkly Rep.* 2019 Aug 2;68(30):653-657.
7. Tookes H, Diaz C, Li H, et al. A Cost Analysis of Hospitalizations for Infections Related to Injection Drug Use at a County Safety-Net Hospital in Miami, Florida. *PLoS One.* 2015 Jun 15;10(6):e0129360.
8. Hall EW, Rosenberg ES, Jones CM, et al. Estimated number of injection-involved drug overdose deaths, United States, 2000 - 2018. *Drug Alcohol Depend.* 2022 May 1;234:109428.
9. Barnes E, Peacock J, Bachmann L. International Classification of Diseases (ICD) Codes Fail to Accurately Identify Injection Drug Use Associated Endocarditis Cases. *J Addict Med.* 2022 Jan-Feb 01;16(1):27-32.
10. Janjua NZ, Islam N, Kuo M, et al. Identifying injection drug use and estimating population size of people who inject drugs using healthcare administrative datasets. *Int J Drug Policy.* 2018 May;55:31-39.
11. Marks LR, Nolan NS, Jiang L, et al. Use of ICD-10 Codes for Identification of Injection Drug Use-Associated Infective Endocarditis Is Nonspecific and Obscures Critical Findings on Impact of Medications for Opioid Use Disorder. *Open Forum Infect Dis.* 2020 Sep 9;7(10):ofaa414.
12. McGrew KM, Carabin H, Garwe T, et al. Validity of ICD-based algorithms to estimate the prevalence of injection drug use among infective endocarditis hospitalizations in the absence of a reference standard. *Drug Alcohol Depend.* 2020 Apr 1;209:107906. doi: 10.1016/j.drugalcdep.2020.107906.
13. The White House. (2021, December 8). White House Releases Model Law to Help States Ensure Access to “Safe, Effective, and Cost-saving” Syringe Services Programs. Retrieved from <https://www.whitehouse.gov/ondcp/briefing-room/2021/12/08/white-house-releases-model-law-to-help-states-ensure-access-to-safe-effective-and-cost-saving-syringe-services-programs/>.
14. Office of National Drug Control Policy. National Drug Control Strategy. 2022. Washington, DC: Executive Office of the President.
15. Centers for Disease Control and Prevention. Syringe Services Programs. <https://www.cdc.gov/ssp/index.html>.
16. World Health Organization. Policy brief: Provision of sterile injecting equipment to reduce HIV transmission. 2004. World Health Organization No. WHO/HIV/2004.03.
17. Adams JM. Making the Case for Syringe Services Programs. *Public Health Reports.* 2020;135(1\_suppl):10S-12S.
18. American Medical Association Policy H-95.958 (Syringe and Needle Exchange Programs), adopted 1995, retained as of 2016. See <https://policysearch.ama-assn.org/policyfinder/detail/syringe?uri=%2FAMADoc%2FHOD.xml-0-5337.xml>.
19. American Bar Association Resolution 106B, House of Delegates 1997 Annual Meeting, (Needle Exchange Programs), adopted August 1997, retained as of August 15, 2017. See [https://www.americanbar.org/content/dam/aba/directories/policy/1997\\_am\\_106b.authcheckdam.pdf](https://www.americanbar.org/content/dam/aba/directories/policy/1997_am_106b.authcheckdam.pdf); <https://www.americanbar.org/content/dam/aba/directories/policy/annual-2017/2017-am-400b.pdf>, p.29.
20. Rife-Pennington T, Dinges E, Ho MQ. Implementing Syringe Services Programs Within the Veterans Health Administration: Facility Experiences and Next Steps. *Journal of the American Pharmacists Association.* 2022.
21. Centers for Disease Control and Prevention. Viral Hepatitis Surveillance Report – United States, 2020. <https://www.cdc.gov/hepatitis/statistics/2020surveillance/index.htm>. Published September 2022. Accessed December 1, 2022.

### TABULAR MODIFICATIONS

Z72 Problems related to lifestyle

Excludes2: problems related to life-management difficulty (Z73.-)  
problems related to socioeconomic and psychosocial  
circumstances (Z55-Z65)

Z72.8 Other problems related to lifestyle

New code	Z72.83 Injection drug use
Add	Code also drug type, if known
Add	Code also any other manifestations, if known

## KCNQ2-Related Epilepsy

KCNQ2-related epilepsy includes two broad phenotypes or different clinical pictures. These include certain self-limited epileptic seizure types, and early-onset epileptic encephalopathy, or developmental epileptic encephalopathy. The gene KCNQ2 is one of the voltage-gated potassium channel genes.<sup>1,2</sup>

The most common self-limited epileptic seizure type involving KCNQ2 is self-limited (familial) neonatal epilepsy (SeLNE), and it has also been known by terms including benign familial neonatal epilepsy, benign neonatal convulsions, benign idiopathic neonatal seizures, and "fifth-day fits" (related to common day of onset). This type may also be caused by another voltage-gated potassium channel gene, KCNQ3. There is also a nonfamilial form due to de novo pathogenic variants in these genes. The familial form is inherited in an autosomal dominant pattern. Seizure onset is within the first week of life, most frequently focal tonic seizures, involving the head, face, and limbs. Focal clonic seizures may also occur. Seizures usually resolve within six weeks to six months but may be persistent. Although development is usually normal, some children have learning difficulties and minor motor delays. For diagnosis, normal brain MRI is required to exclude other causes (as is typical for self-limited epilepsies), and testing for KCNQ2 or KCNQ3 pathogenic variants is necessary.<sup>2</sup>

Self-limited familial neonatal-infantile epilepsy (SeLFNIE) is another autosomal dominant epilepsy that may be caused by pathogenic variants in the KCNQ2 gene, although most cases are due to pathogenic variants in the SCN2A gene. Also previously known as benign familial neonatal-infantile seizures, it generally involves focal tonic seizures with head and eye deviation, or focal clonic seizures may occur. Onset is typically around 11 to 13 weeks but can range from one day through 21 months. The seizures usually resolve by age one to two years.<sup>2</sup>

Self-limited (familial) infantile epilepsy (SeLIE) in most cases is caused by pathogenic variants in the PRRT2 gene, but other genes can be involved, including KCNQ2, KCNQ3, SCN2A, and SCN8A. There is a nonfamilial form due to de novo mutations in PRRT2. The inheritance pattern of familial cases is autosomal dominant with incomplete penetrance. It was formerly called benign familial (and nonfamilial) infantile seizures, and is characterized by focal seizures in an otherwise normal infant, with onset between ages 3 and 20 months, typically about 6 months of age. Focal clonic seizures may progress to bilateral tonic-clonic seizures, or alternate sides.<sup>3</sup>

Benign (childhood) epilepsy with centrotemporal spikes (BECTS) may be nonfamilial or familial with autosomal dominant inheritance and can be associated with pathogenic variants in KCNQ2 and GRIN2A genes. Also known as rolandic epilepsy, and to be termed self-limited epilepsy with centrotemporal spikes, it is the most common type of focal epilepsy affecting children. The most common seizure type is a focal seizure with motor symptoms involving the face. Symptoms correspond to the origin of seizures in the rolandic or perisylvian sensorimotor cortex, which represents the face and oropharynx. These include facial numbness or twitching, guttural vocalizations, hypersalivation, drooling, dysphasia, and speech arrest. Motor activity in the upper extremity is also common. Most seizures occur at night or on awakening. While focal seizures

are the most frequent type, BECTS may present with a secondary generalized tonic-clonic seizure during sleep. Status epilepticus is unusual. BECTS is a benign (self-limited) focal epilepsy of childhood, and has later onset, ranging from 3 to 13 years age, peaking at 7 to 9 years.<sup>4</sup>

Some individuals with de novo missense *KCNQ2* pathogenic variants develop a KCNQ2-related developmental epileptic encephalopathy (KCNQ2-DEE). This presents in the first week of life with neurologic abnormalities (encephalopathy, hypotonia, and lack of visual attentiveness) and severe, treatment-resistant seizures. There are typically tonic seizures, but these may be myoclonic or focal. Brain MRI may show subtle abnormalities in the basal ganglia and thalami. Seizures resolve within a few months to years in most patients, but children with KCNQ2-DEE typically have moderate to severe global neurodevelopmental impairments. Genetic testing for *KCNQ2* pathogenic variants is necessary to confirm the diagnosis of KCNQ2-DEE.<sup>2</sup>

KCNQ2-related epilepsy is one of the most common genetic epilepsy syndromes in childhood. KCNQ2-related epilepsy has a distinctive clinical course characterized by especially early-onset epilepsy, variable degrees of initial drug-refractoriness that later diminishes, and moderate to profound developmental delay that persists even when seizure freedom is achieved. This profile differentiates it from other infantile-onset epilepsies. KCNQ2-related epilepsy has distinctive pathophysiology, manifest in its characteristic clinical course, but also in its relative unresponsiveness to the first line medications most often used in other forms of neonatal-onset epilepsy, and it may have greater responsiveness to certain other drugs which are less-frequently used in the neonatal and early infantile period. There is ongoing work toward specific drug development, including a Phase 3 clinical trial ongoing for KCNQ2-related epilepsy. Creation of specific ICD-10-CM codes for KCNQ2-related epilepsy will enable improved tracking in clinical settings and of epidemiological endpoints, and also has potential to enable broader adoption of recommended treatments, and support for research, and enabling ready access to potential treatments, and thus will benefit people with KCNQ2-related epilepsy.

A request for creation of new codes specific for KCNQ2-related epilepsy was received from the KCNQ2 Cure Alliance, which is dedicated to advancing research and supporting families of children and adults with KCNQ2 developmental and epileptic encephalopathy.

#### References

1. KCNQ2 gene: potassium voltage-gated channel subfamily Q member 2. MedlinePlus. 2013. <https://medlineplus.gov/genetics/gene/kcnq2/>
2. Shellhaas R. Overview of neonatal epilepsy syndromes. UpToDate. Section Eds.: Nordli DR Jr, Garcia-Prats JA. Deputy Ed.: Dashe JF. 2022. <https://www.uptodate.com/contents/overview-of-neonatal-epilepsy-syndromes>
3. Shellhaas R. Overview of infantile epilepsy syndromes. UpToDate. Section Eds.: Nordli DR Jr, Garcia-Prats JA. Deputy Ed.: Dashe JF. 2022. <https://www.uptodate.com/contents/overview-of-infantile-epilepsy-syndromes>
4. Benbadis SR. Benign (self-limited) focal epilepsies of childhood. UpToDate. Section Ed.: Nordli DR Jr. Deputy Ed.: Dashe JF. 2021. <https://www.uptodate.com/contents/benign-self-limited-focal-epilepsies-of-childhood>

## TABULAR MODIFICATIONS

### Option #1

Note: this option was created based on a modified approach from the original submission.

G40 Epilepsy and recurrent seizures

G40.8 Other epilepsy and recurrent seizures

New sub-subcategory	G40.84	KCNQ2-related epilepsy
New code	G40.841	KCNQ2-related epilepsy, not intractable, with status epilepticus
New code	G40.842	KCNQ2-related epilepsy, not intractable, without status epilepticus
Add		KCNQ2-related epilepsy NOS
New code	G40.843	KCNQ2-related epilepsy, intractable, with status epilepticus
New code	G40.844	KCNQ2-related epilepsy, intractable, without status epilepticus

### Option #2

Note: this option was created by NCHS based on a generalized approach for a number of proposals. See also the Developmental Epileptic Encephalopathies topic.

Z15 Genetic susceptibility to disease

New subcategory	Z15.1	Genetic susceptibility to epilepsy and neurodevelopmental disorders
New sub-subcategory	Z15.11	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to ion channel genes

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New code	Z15.110	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of KCNQ2 gene
Add		Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of the gene for potassium voltage-gated channel, KQT-like subfamily, member 2
New code	Z15.118	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of other ion channel gene
Add		Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of the SCN8A gene
Add		Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of the KCNQ3 gene

## Kleefstra Syndrome

Kleefstra syndrome (KS) is a neurodevelopmental disorder caused by a disruption to one copy of the gene *EHMT1* at 9q34.3 (Kleefstra et al., 2006). This disruption can occur due to pathogenic mutations or structural variants (deletions or rearrangements). The *EHMT1* gene is haploinsufficient, and loss of one functional copy will always result in KS.

The disorder is almost always due to a *de novo* genetic event and has only been known to be carried in families due to mosaicism or balanced translocations in one parent (but would hypothetically be inherited in an autosomal dominant manner from a parent with KS). Therefore, the disorder is evenly distributed throughout the population, and affects all regions and ethnic groups equally. The deletion/mutation in *EHMT1* results in a reduction in active EHMT1 protein, triggering the syndromic features.

KS is characterized by a core clinical phenotype of moderate to severe intellectual disability/developmental delay, autism, and childhood hypotonia. A mixture of distinct facial features are usually present. Many KS patients also suffer from epilepsy. Additional risk is present for congenital heart and urogenital defects, behavioral and psychiatric disorders and obesity (Willemsen et al., 2012).

KS patients must be properly evaluated/monitored for these potential comorbidities, and appropriate interventions provided in a timely manner. Importantly, a significant number of KS patients will suffer from severe psychosis/sleep disturbance in adolescence, requiring prompt pharmacologic intervention to stave off a drastic regression in skills (Vermeulen et al., 2017).

Current best estimates for KS prevalence are 1:25-35,000 people (see supporting material), meaning it is expected to affect ~11,000 patients in the US alone, and >250,000 worldwide. This places the disorder in a similar range of prevalence to other rare disorders that have been provided codes, such as SYNGAP1 Encephalopathy and Angelman syndrome. As with these syndromes, the ~11,000 KS patients in the US would greatly benefit from access to the public service that is embodied in an ICD-10 code.

A unique ICD-10-CM code would be a momentous step forward for this rare disease, as coding would aid in: ensuring a standard of care (including emergency interventions, such as for psychosis) research (patient registries, clinical studies), preparation for clinical trials, studying operational and strategic best practices, and refining understanding of prevalence.

This proposal is submitted by Siddharth Srivastava, MD, Tjitske Kleefstra, MD, and IDefine, a patient advocacy organization representing the Kleefstra Syndrome community.

### References:

Kleefstra, T., Brunner, H. G., Amiel, J., Oudakker, A. R., Nillesen, W. M., Magee, A., Geneviè, D., Cormier- Daire, V., van Esch, H., Fryns, J.-P., Hamel, B. C. J., Sistermans, E. A., de Vries, B. B. A., & van Bokhoven, H. (2006). Loss-of-Function Mutations in Euchromatin Histone Methyl Transferase 1 (EHMT1) Cause the 9q34 Subtelomeric Deletion Syndrome. In *The American Journal of Human Genetics* (Vol. 79). www.ajhg.org



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Vermeulen, K., Staal, W. G., Janzing, J. G., van Bokhoven, H., Egger, J. I. M., & Kleefstra, T. (2017). Sleep Disturbance as a Precursor of Severe Regression in Kleefstra Syndrome Suggests a Need for Firm and Rapid Pharmacological Treatment. *Clinical Neuropharmacology*, 40(4), 185–188. <https://doi.org/10.1097/WNF.0000000000000226>

Willemsen, M. H., Vulto-Van Silfhout, A. T., Nillesen, W. M., Wissink-Lindhout, W. M., van Bokhoven, H., Philip, N., Berry-Kravis, E. M., Kini, U., van Ravenswaaij-Arts, C. M. A., Delle Chiaie, B., Innes, A. M. M., Houge, G., Kosonen, T., Cremer, K., Fannemel, M., Stray-Pedersen, A., Reardon, W., Ignatius, J., Lachlan, K., ... Kleefstra, T. (2012). Update on Kleefstra syndrome. *Molecular Syndromology*, 2(3–5), 202–212. <https://doi.org/10.1159/000335648>

### TABULAR MODIFICATIONS

Q87 Other specified congenital malformation syndromes affecting multiple systems

Use additional code(s) to identify all associated manifestations

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

Excludes1: Zellweger syndrome (E71.510)

New code

Q87.86 Kleefstra Syndrome

## Lymphoma in Remission

This proposal was presented at the September 2022 ICD-10 Coordination and Maintenance Committee Meeting of the Lymphoma in Remission proposal. Based on public comments modifications have been made and are in **bold**.

Lymphoma is a cancer of the lymphatic system, which is part of the body's germ-fighting network. The lymphatic system includes the lymph nodes (lymph glands), spleen, thymus gland and bone marrow. Lymphoma can affect all those areas as well as other organs throughout the body.<sup>1</sup>

In general, the goal of treatment is to destroy as many lymphoma cells as possible and to induce a complete remission. Complete remission means that all evidence of disease is eliminated. Patients who go into remission are sometimes cured of their disease. Treatment can also keep non-Hodgkin lymphoma (NHL) in check for many years, even though imaging or other studies show remaining sites of disease.<sup>2</sup>

A new ICD-10-CM code will provide coding specificity for the distinct types of lymphoma in remission. The absence of lymphoma in remission codes will hinder the ability to make meaningful comparisons to assess and evaluate differences in patient care, statistical data, resource consumption (i.e., overall length of stay, additional drugs, etc.), and accurate clinical outcomes of lymphoma cases.

The National Center of Health Statistics received this proposal requesting new ICD-10-CM codes for lymphoma in remission from Alliance Dedicated Cancer Centers (ADCC).

### References

<sup>1</sup> Mayo Clinic Staff. (n.d.). *Lymphoma*. Retrieved from Mayo Clinic:

<https://www.mayoclinic.org/diseasesconditions/lymphoma/symptoms-causes/syc-20352638#:~:text=Lymphoma%20is%20a%20cancer%20of,other%20organs%20throughout%20the%20body.>

<sup>2</sup> *Treatment*. (n.d.). Retrieved from Leukemia & Lymphoma Society: <https://www.lls.org/lymphoma/non-hodgkin-lymphoma/treatment#:~:text=Complete%20remission%20means%20that%20all,show%20remaining%20sites%20of%20disease.>

## TABULAR MODIFICATIONS

C81 Hodgkin lymphoma  
C81.0 Nodular lymphocyte predominant Hodgkin lymphoma

New code C81.0A Nodular lymphocyte predominant Hodgkin lymphoma,  
in remission

**C81.1 Nodular sclerosis Hodgkin lymphoma**  
**Nodular sclerosis classical Hodgkin lymphoma**

New code  
Add  
C81.1A Nodular sclerosis Hodgkin lymphoma in remission  
Nodular sclerosis classical Hodgkin lymphoma in remission

C81.2 Mixed cellularity Hodgkin lymphoma  
Mixed cellularity classical Hodgkin lymphoma

New code  
Add  
C81.2A Mixed cellularity Hodgkin lymphoma, in remission  
Mixed cellularity classical Hodgkin lymphoma, in remission

C81.3 Lymphocyte depleted Hodgkin lymphoma  
Lymphocyte depleted classical Hodgkin lymphoma

New code  
Add  
C81.3A Lymphocyte depleted Hodgkin lymphoma, in remission  
Lymphocyte depleted classical Hodgkin lymphoma, in remission

C81.4 Lymphocyte-rich Hodgkin lymphoma  
Lymphocyte-rich classical Hodgkin lymphoma

New code  
Add  
C81.4A Lymphocyte-rich Hodgkin lymphoma, in remission  
Lymphocyte-rich classical Hodgkin lymphoma, in remission

C81.7 Other Hodgkin lymphoma  
Classical Hodgkin lymphoma NOS  
Other classical Hodgkin lymphoma

New code  
Add  
Add  
C81.7A Other Hodgkin lymphoma, in remission  
Classical Hodgkin lymphoma NOS, in remission  
Other classical Hodgkin lymphoma, in remission

C81.9 Hodgkin lymphoma, unspecified

New code  
C81.9A Hodgkin lymphoma, unspecified, in remission

C82 Follicular lymphoma  
C82.0 Follicular lymphoma grade I

New code  
C82.0A Follicular lymphoma grade I, in remission

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	C82.1	Follicular lymphoma grade II
New code	C82.1A	Follicular lymphoma grade II, in remission
	C82.2	Follicular lymphoma grade III, unspecified
New code	C82.2A	Follicular lymphoma grade III, unspecified, in remission
	C82.3	Follicular lymphoma grade IIIa
New code	C82.3A	Follicular lymphoma grade IIIa, in remission
	C82.4	Follicular lymphoma grade IIIb
New code	C82.4A	Follicular lymphoma grade IIIb, in remission
	C82.5	Diffuse follicle center lymphoma
New code	C82.5A	Diffuse follicle center lymphoma, in remission
	C82.6	Cutaneous follicle center lymphoma
New code	C82.6A	Cutaneous follicle center lymphoma, in remission
	C82.8	Other types of follicular lymphoma
New code	C82.8A	Other types of follicular lymphoma, in remission
	C82.9	Follicular lymphoma, unspecified
New code	C82.9A	Follicular lymphoma, unspecified, in remission
	C83	Non-follicular lymphoma
	C83.0	Small cell B-cell lymphoma
		Lymphoplasmacytic lymphoma
		Nodal marginal zone lymphoma
		Non-leukemic variant of B-CLL
		Splenic marginal zone lymphoma
New code	C83.0A	Small cell B-cell lymphoma, in remission
Add		Lymphoplasmacytic lymphoma, in remission
Add		Nodal marginal zone lymphoma, in remission
Add		Non-leukemic variant of B-CLL, in remission
Add		Splenic marginal zone lymphoma, in remission

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C83.1 Mantle cell lymphoma  
Centrocytic lymphoma

New code  
Add C83.1A Mantle cell lymphoma, in remission  
Centrocytic lymphoma, in remission

C83.3 Diffuse large B-cell lymphoma  
Anaplastic diffuse large B-cell lymphoma  
CD30-positive diffuse large B-cell lymphoma  
Centroblastic diffuse large B-cell lymphoma  
Diffuse large B-cell lymphoma, subtype not specified  
Immunoblastic diffuse large B-cell lymphoma  
Plasmablastic diffuse large B-cell lymphoma  
T-cell rich diffuse large B-cell lymphoma

New code  
Add C83.3A Diffuse large B-cell lymphoma, in remission  
Add Anaplastic diffuse large B-cell lymphoma, in remission  
Add CD30-positive diffuse large B-cell lymphoma, in remission  
Add Centroblastic diffuse large B-cell lymphoma, in remission  
Add Diffuse large B-cell lymphoma, subtype not specified, in remission  
Add Immunoblastic diffuse large B-cell lymphoma, in remission  
Add Plasmablastic diffuse large B-cell lymphoma, in remission  
Add T-cell rich diffuse large B-cell lymphoma, in remission

C83.5 Lymphoblastic (diffuse) lymphoma  
B-precursor lymphoma  
Lymphoblastic B-cell lymphoma  
Lymphoblastic lymphoma NOS  
Lymphoblastic T-cell lymphoma  
T-precursor lymphoma

New code  
Add C83.5A Lymphoblastic (diffuse) lymphoma, in remission  
Add B-precursor lymphoma, in remission  
Add Lymphoblastic B-cell lymphoma, in remission  
Add Lymphoblastic lymphoma NOS, in remission  
Add Lymphoblastic T-cell lymphoma, in remission  
Add T-precursor lymphoma, in remission

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C83.7 Burkitt lymphoma  
Atypical Burkitt lymphoma  
Burkitt-like lymphoma

New code C83.7A Burkitt lymphoma, in remission  
Add Atypical Burkitt lymphoma, in remission  
Add Burkitt-like lymphoma, in remission

C83.8 Other non-follicular lymphoma  
Intravascular large B-cell lymphoma  
Lymphoid granulomatosis  
Primary effusion B-cell lymphoma

New code C83.8A Other non-follicular lymphoma, in remission  
Add Intravascular large B-cell lymphoma, in remission  
Add Lymphoid granulomatosis, in remission  
Add Primary effusion B-cell lymphoma, in remission

C83.9 Non-follicular (diffuse) lymphoma, unspecified

New code C83.9A Non-follicular (diffuse) lymphoma, unspecified, in remission

C84 Mature T/NK-cell lymphomas  
C84.0 Mycosis fungoides

New code C84.0A Mycosis fungoides, in remission

C84.1 Sézary disease

New code C84.1A Sézary disease, in remission

C84.4 Peripheral T-cell lymphoma, not **elsewhere** classified  
Lennert's lymphoma  
Lymphoepithelioid lymphoma  
Mature T-cell lymphoma, not elsewhere classified

New code C84.4A Peripheral T-cell lymphoma, not **elsewhere** classified, in remission  
Add Lennert's lymphoma, in remission  
Add Lymphoepithelioid lymphoma, in remission  
Add Mature T-cell lymphoma, not elsewhere classified, in remission

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C84.6 Anaplastic large cell lymphoma, ALK-positive  
Anaplastic large cell lymphoma, CD30-positive

New code C84.6A Anaplastic large cell lymphoma, ALK-positive, in  
remission  
Add Anaplastic large cell lymphoma, CD30-positive, in  
remission

C84.7 Anaplastic large cell lymphoma, ALK-negative

New code C84.7B Anaplastic large cell lymphoma, ALK-negative, in  
remission

C84.A Cutaneous T-cell lymphoma, unspecified

New code C84.AA Cutaneous T-cell lymphoma, unspecified, in remission

C84.Z Other mature T/NK-cell lymphomas

New code C84.ZA Other mature T/NK-cell lymphomas, in remission

C84.9 Mature T/NK-cell lymphomas, unspecified  
NK/T cell lymphoma NOS

New code C84.9A Mature T/NK-cell lymphomas, unspecified, in  
remission  
Add NK/T cell lymphoma NOS, in remission

C85 Other specified and unspecified types of non-Hodgkin lymphoma

C85.1 Unspecified B-cell lymphoma

New code C85.1A Unspecified B-cell lymphoma, in remission

C85.2 Mediastinal (thymic) large B-cell lymphoma

New code C85.2A Mediastinal (thymic) large B-cell lymphoma, in  
remission

C85.8 Other specified types of non-Hodgkin lymphoma

New code C85.8A Other specified types of non-Hodgkin lymphoma, in  
remission

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C85.9 Non-Hodgkin lymphoma, unspecified  
 Lymphoma NOS  
 Malignant lymphoma NOS  
 Non-Hodgkin lymphoma NOS

New code C85.9A Non-Hodgkin lymphoma, unspecified, in remission  
 Add Lymphoma NOS, in remission  
 Add Malignant lymphoma NOS, in remission  
 Add Non-Hodgkin lymphoma NOS, in remission

C86 Other specified types of T/NK-cell lymphoma  
 C86.0 Extranodal NK/T-cell lymphoma, nasal type

New code C86.00 Extranodal NK/T-cell lymphoma, nasal type not having  
 achieved remission  
 Add Extranodal NK/T-cell lymphoma, nasal type NOS  
 Add Extranodal NK/T-cell lymphoma, nasal type with failed  
 remission

New code C86.01 Extranodal NK/T-cell lymphoma, nasal type, in  
 remission

C86.1 Hepatosplenic T-cell lymphoma  
 Alpha-beta and gamma delta types

New code C86.10 Hepatosplenic T-cell lymphoma not having achieved  
 remission  
 Add Hepatosplenic T-cell lymphoma NOS  
 Add Hepatosplenic T-cell lymphoma with failed remission

New code C86.11 Hepatosplenic T-cell lymphoma, in remission

C86.2 Enteropathy-type (intestinal) T-cell lymphoma  
 Enteropathy associated T-cell lymphoma

New code C86.20 Enteropathy-type (intestinal) T-cell lymphoma not  
 having achieved remission  
 Add Enteropathy associated T-cell lymphoma not having  
 achieved remission  
 Add Enteropathy associated T-cell lymphoma NOS  
 Add Enteropathy associated T-cell lymphoma with failed  
 remission  
 Add Enteropathy-type (intestinal) T-cell lymphoma NOS  
 Add Enteropathy-type (intestinal) T-cell lymphoma with  
 failed remission



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New code	C86.21	Enteropathy-type (intestinal) T-cell lymphoma, in remission
Add		Enteropathy associated T-cell lymphoma, in remission
	C86.3	Subcutaneous panniculitis-like T-cell lymphoma
New code	C86.30	Subcutaneous panniculitis-like T-cell lymphoma not having achieved remission
Add		Subcutaneous panniculitis-like T-cell lymphoma NOS
Add		Subcutaneous panniculitis-like T-cell lymphoma with failed remission
New code	C86.31	Subcutaneous panniculitis-like T-cell lymphoma, in remission
	C86.4	Blastic NK-cell lymphoma Blastic plasmacytoid dendritic cell neoplasm (BPDCN)
New code	C86.40	Blastic NK-cell lymphoma not having achieved remission
Add		Blastic NK-cell lymphoma NOS
Add		Blastic NK-cell lymphoma with failed remission
Add		Blastic plasmacytoid dendritic cell neoplasm (BPDCN) not having achieved remission
Add		Blastic plasmacytoid dendritic cell neoplasm (BPDCN) NOS
Add		Blastic plasmacytoid dendritic cell neoplasm (BPDCN) with failed remission
New code	C86.41	Blastic NK-cell lymphoma, in remission
Add		Blastic plasmacytoid dendritic cell neoplasm (BPDCN), in remission
	C86.5	Angioimmunoblastic T-cell lymphoma Angioimmunoblastic lymphadenopathy with dysproteinemia (AILD)
New code	C86.50	Angioimmunoblastic T-cell lymphoma not having achieved remission
Add		Angioimmunoblastic lymphadenopathy with dysproteinemia (AILD) not having achieved remission
Add		Angioimmunoblastic lymphadenopathy with dysproteinemia (AILD) NOS

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Add		Angioimmunoblastic lymphadenopathy with dysproteinemia (AILD) with failed remission
Add		Angioimmunoblastic T-cell lymphoma NOS
Add		Angioimmunoblastic T-cell lymphoma with failed remission
New code	C86.51	Angioimmunoblastic T-cell lymphoma, in remission
Add		Angioimmunoblastic lymphadenopathy with dysproteinemia (AILD), in remission
	C86.6	Primary cutaneous CD30-positive T-cell proliferations
		Lymphomatoid papulosis
		Primary cutaneous anaplastic large cell lymphoma
		Primary cutaneous CD30-positive large T-cell lymphoma
New code	C86.60	Primary cutaneous CD30-positive T-cell proliferations not having achieved remission
Add		Lymphomatoid papulosis not having achieved remission
Add		Lymphomatoid papulosis NOS
Add		Lymphomatoid papulosis with failed remission
Add		Primary cutaneous anaplastic large cell lymphoma not having achieved remission
Add		Primary cutaneous anaplastic large cell lymphoma NOS
Add		Primary cutaneous anaplastic large cell lymphoma with failed remission
Add		Primary cutaneous CD30-positive large T-cell lymphoma not having achieved remission
Add		Primary cutaneous CD30-positive large T-cell lymphoma NOS
Add		Primary cutaneous CD30-positive large T-cell lymphoma with failed remission
Add		Primary cutaneous CD30-positive T-cell proliferations NOS
Add		Primary cutaneous CD30-positive T-cell proliferations with failed remission
New code	C86.61	Primary cutaneous CD30-positive T-cell proliferations, in remission
Add		Lymphomatoid papulosis, in remission
Add		Primary cutaneous anaplastic large cell lymphoma, in remission
Add		Primary cutaneous CD30-positive large T-cell lymphoma, in remission

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C88 Malignant immunoproliferative diseases and certain other B-cell lymphomas

C88.0 Waldenström macroglobulinemia  
 Lymphoplasmacytic lymphoma with IgM-production  
 Macroglobulinemia (idiopathic) (primary)

- |          |        |  |
|----------|--------|--|
| New code | C88.00 | Waldenström macroglobulinemia not having achieved remission                  |
| Add      |        | Lymphoplasmacytic lymphoma with IgM-production not having achieved remission |
| Add      |        | Lymphoplasmacytic lymphoma with IgM-production, NOS                          |
| Add      |        | Lymphoplasmacytic lymphoma with IgM-production with failed remission         |
| Add      |        | Macroglobulinemia (idiopathic) (primary) not having achieved remission       |
| Add      |        | Macroglobulinemia (idiopathic) (primary) NOS                                 |
| Add      |        | Macroglobulinemia (idiopathic) (primary) with failed remission               |
| Add      |        | Waldenström macroglobulinemia NOS  |
| Add      |        | Waldenström macroglobulinemia with failed remission                          |
| New code | C88.01 | Waldenström macroglobulinemia, in remission                                  |
| Add      |        | Lymphoplasmacytic lymphoma with IgM-production, in remission                 |
| Add      |        | Macroglobulinemia (idiopathic) (primary), in remission                       |

C88.2 Heavy chain disease  
 Franklin disease  
 Gamma heavy chain disease  
 Mu heavy chain disease

- |          |        |   |
|----------|--------|---|
| New code | C88.20 | Heavy chain disease not having achieved remission       |
| Add      |        | Franklin disease not having achieved remission          |
| Add      |        | Franklin disease NOS                                    |
| Add      |        | Franklin disease with failed remission                  |
| Add      |        | Gamma heavy chain disease not having achieved remission |
| Add      |        | Gamma heavy chain disease NOS                           |
| Add      |        | Gamma heavy chain disease with failed remission         |
| Add      |        | Heavy chain disease NOS                                 |
| Add      |        | Heavy chain disease with failed remission               |
| Add      |        | Mu heavy chain disease not having achieved remission    |
| Add      |        | Mu heavy chain disease NOS                              |

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Add		Mu heavy chain disease with failed remission
New code	C88.21	Heavy chain disease, in remission
Add		Franklin disease, in remission
Add		Gamma heavy chain disease, in remission
Add		Mu heavy chain disease, in remission
	C88.3	Immunoproliferative small intestinal disease
		Alpha heavy chain disease
		Mediterranean lymphoma
New code	C88.30	Immunoproliferative small intestinal disease not having achieved remission
<b>Add</b>		<b>Alpha heavy chain disease not having achieved remission</b>
Add		Alpha heavy chain disease NOS
Add		Alpha heavy chain disease with failed remission
Add		Immunoproliferative small intestinal disease NOS
Add		Immunoproliferative small intestinal disease with failed remission
Add		Mediterranean lymphoma not having achieved remission
Add		Mediterranean lymphoma NOS
Add		Mediterranean lymphoma with failed remission
New code	C88.31	Immunoproliferative small intestinal disease, in remission
Add		Alpha heavy chain disease, in remission
Add		Mediterranean lymphoma, in remission
	C88.4	Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue [MALT-lymphoma]
		Lymphoma of bronchial-associated lymphoid tissue [BALT-lymphoma]
		Lymphoma of skin-associated lymphoid tissue [SALT-lymphoma]
New code	C88.40	Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue [MALT-lymphoma] not having achieved remission
Add		Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue [MALT-lymphoma] NOS
Add		Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue [MALT-lymphoma] with failed remission

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Add		Lymphoma of bronchial-associated lymphoid tissue [BALT-lymphoma] not having achieved remission
Add		Lymphoma of bronchial-associated lymphoid tissue [BALT-lymphoma] NOS
Add		Lymphoma of bronchial-associated lymphoid tissue [BALT-lymphoma] with failed remission
Add		Lymphoma of skin-associated lymphoid tissue [SALT-lymphoma] not having achieved remission
Add		Lymphoma of skin-associated lymphoid tissue [SALT-lymphoma] NOS
Add		Lymphoma of skin-associated lymphoid tissue [SALT-lymphoma] with failed remission
New code	C88.41	Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue [MALT-lymphoma], in remission
Add		Lymphoma of bronchial-associated lymphoid tissue [BALT-lymphoma], in remission
Add		Lymphoma of skin-associated lymphoid tissue [SALT-lymphoma], in remission
	C88.8	Other malignant immunoproliferative diseases
New code	C88.80	Other malignant immunoproliferative diseases not having achieved remission
Add		Other malignant immunoproliferative diseases NOS
Add		Other malignant immunoproliferative diseases with failed remission
New code	C88.81	Other malignant immunoproliferative diseases, in remission
	C88.9	Malignant immunoproliferative disease, unspecified
		Immunoproliferative disease NOS
	C88.90	Malignant immunoproliferative disease, unspecified not having achieved remission
Add		Immunoproliferative disease NOS not having achieved remission
Add		Immunoproliferative disease NOS
Add		Immunoproliferative disease NOS with failed remission
Add		Malignant immunoproliferative disease, unspecified NOS
Add		Malignant immunoproliferative disease, unspecified with failed remission

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C88.91 Malignant immunoproliferative disease, unspecified, in  
remission  
Immunoproliferative disease NOS, in remission

## Monogenic Forms of Obesity

Obesity may be caused by mutations in a number of specific genes, particularly certain ones that can lead to disruption of the melanocortin 4 receptor pathway. These include the gene for the melanocortin 4 receptor itself (MC4R), as well as the following genes: leptin (LEP), leptin receptor (LEPR), proopiomelanocortin (POMC), proprotein convertase subtilisin/kexin type 1 (PCSK1), Src homology 2B adaptor signaling protein 1 (SH2B1), and Nuclear Receptor Coactivator 1 (NCOA1). Rhythm Pharmaceuticals, Inc., has proposed the creation of new ICD-10-CM diagnosis codes for describing rare monogenic forms of obesity. Monogenic obesity is different from other forms of obesity, such as those caused by lifestyle. Stratifying the monogenic obesity population, with the creation of new ICD-10-CM diagnosis codes, will assist healthcare providers and researchers in providing and further developing targeted clinical interventions leading to better health outcomes, and bring awareness to these rare diseases.

Obesity is a chronic disease that is defined as having a body mass index (BMI)  $\geq 30.0$  kg/m<sup>2</sup>.<sup>1</sup> Obesity, in general, is the result of a mismatch of consuming more calories beyond what the body expends using energy. Approximately 42.4% of adults<sup>1</sup> and approximately 1 in 5 children in the United States are obese, with incidence rising each year.<sup>2</sup> Obesity can result in a number of immediate and long-term health consequences. Cardiac risks include early signs of vascular dysfunction, subclinical atherosclerosis, and high blood pressure, as well as increased risk of cardiovascular disease (CVD).<sup>3</sup> Obese persons also experience metabolic issues such as type 2 diabetes mellitus (T2DM), hyperinsulinemia, insulin resistance, and prediabetes. Other comorbidities include sleep apnea, nonfatty liver disease, and musculoskeletal problems, cancer, cholestasis and other gallbladder disease, dyslipidemia, hyperuricemia and gout, menstrual abnormalities, musculoskeletal problems, reduction of cerebral blood flow.<sup>4,5</sup> Psychosocial issues include depression, teasing and bullying, social marginalization and discrimination.<sup>5</sup> Additionally, obesity can be associated with disordered eating and in some cases, premature death.<sup>4</sup> Obesity is also associated with functional issues, including disability, low physical fitness, mobility limitations, absenteeism from school or work, disqualification from active military/fire/police services, reduced academic performance, reduced productivity, and unemployment.

It is often assumed that most causes of obesity are related to lifestyle, when in fact, the root of a person's obesity is often due to a myriad of social, psychological, biological and genetic factors, which are complex.<sup>6</sup> Monogenic obesity is a rare and severe early-onset obesity associated with endocrine disorders, mainly due to mutations in genes of the leptin/melanocortin axis involved in food intake regulation. Syndromic obesity is a rare type of obesity associated with additional phenotypes, which may include intellectual impairment, dysmorphic features, and organ-specific developmental abnormalities. Two syndromes frequently linked to obesity are Prader-Willi syndrome (PWS) and Bardet-Biedl syndrome (BBS). Oligogenic obesity is a rare type of obesity characterized by variable severity of obesity, partly dependent on environmental factors and the absence of a specific phenotype. Polygenic obesity is a common clinical presentation of obesity where each susceptibility gene considered individually would only have a slight effect on weight, but when presenting with other susceptibility genes, may be diagnosed as polygenic obesity. Obesity due to genetics is rare; however, recent efforts in increasing genetic testing have been

performed in the United States to better characterize the prevalence of each monogenic obesity disorder.<sup>7,8,9</sup> Various genes play a role in monogenic obesity associated with MC4R pathway. The prevalence of variants associated with MC4R pathway monogenic obesity has been recently evaluated in the Uncover Rare Obesity (URO) testing program in the United States, with estimated prevalence's calculated.<sup>10</sup>

The LEP gene provides instructions for making leptin, which is a hormone involved in the regulation of body weight and the ability to feel fullness. The LEPR gene provides the body with instructions for making the leptin receptor. The leptin receptor is activated by a leptin. Typically, fat cells release leptin, and as fat cells become larger, they produce more leptin. This rise in leptin triggers a series of chemical signals that affect hunger and help produce satiety.<sup>11</sup> The leptin receptor protein is found on the surface of cells in many organs and tissues of the body including the hypothalamus, which controls hunger and thirst, as well as many other functions via release of hormones. Mutations in the LEP and LEPR genes can manifest as deficiency in the effects of leptin.<sup>12,13</sup> The absence of sufficient leptin prevents the receptor from responding to leptin, resulting in severe obesity beginning as early as the first few months of a newborn's life. There is extreme hyperphagia leading to obesity, and also frequent infections, and mild hypothyroidism. Other symptoms include hypogonadotropic hypogonadism, and potentially infertility. Leptin receptor signaling is thought to be involved in regulating the body's response to hormones that control sexual development.<sup>11</sup> It is estimated that there are over 500 individuals with obesity caused by LEP biallelic mutations, and over 8,500 individuals with obesity caused by LEP heterozygote mutations in the United States. For LEPR, it is estimated that there are over 5,800 individuals with obesity caused by LEPR biallelic mutations, and over 115,600 individuals with obesity caused by LEPR heterozygous mutations in the United States.<sup>10</sup>

The POMC gene provides the body with instructions for making proopiomelanocortin, a precursor to peptides are involved in signal pathways that control many important bodily functions.<sup>14</sup> One such peptide, the  $\beta$ -MSH peptide, helps regulate weight by binding to the melanocortin 4 receptor (MC4R). Signaling through this receptor in the brain helps maintain the balance between food intake and energy expenditure, important for maintaining weight. The  $\alpha$ -MSH peptide can also bind to MC4R and help maintain the correct energy balance. Mutations in the POMC gene may lead to POMC deficiency. This may dysregulates the body's energy balance leading to overeating and severe obesity.<sup>14</sup> Variants in POMC can also result in a lack of hypothalamic melanocyte stimulating hormone<sup>15</sup> as well as adrenocorticotrophic hormone deficiency, hypothyroidism, hypogonadism, and hypopigmentation due to loss of POMC-derived melanocortin peptides.<sup>16</sup> It is estimated that there are about 4,800 individuals with obesity caused by POMC biallelic mutations, and over 102,800 individuals with obesity caused by POMC heterozygote mutations in the United States.<sup>10,13</sup>

The PCSK1 gene produces a prohormone which plays a crucial role in signaling, and deficiencies may cause issues with appetite control, eating behavior, and energy balance.<sup>17</sup> Additionally, it activates other hormones involved in metabolism, and variants in the PCSK1 gene have been associated with diabetes and impaired insulin levels.<sup>18</sup> Other conditions caused by variants in PCSK1 are postprandial hypoglycaemia, hypogonadism, hypocortisolism, and malabsorption due to impaired prohormone processing.<sup>19</sup> It is estimated that there are about



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6,400 individuals with obesity caused by PCSK1 biallelic mutations, and 92,200 individuals with obesity caused by PCSK1 heterozygote mutations in the United States.<sup>10</sup>

The NCOA1 gene encodes steroid receptor coactivator-1 (SRC1), a part of the central melanocortin pathway that enhances the anti-obesity effects of leptin.<sup>20-24</sup> Deficiency of SRC1 causes obesity. The Src homology 2B (SH2B1) gene encodes a cytoplasmic adaptor protein involved in leptin and insulin signaling and is a participant in the MC4R pathway. SH2B1 deficiency leads to an autosomal dominant obesity.<sup>25-28</sup> It is estimated that there are over 2,100 individuals with obesity caused by NCOA1 biallelic mutations, and over 111,000 individuals with obesity caused by NCOA1 heterozygote mutations in the United States. For SH2B1, it is estimated that there are approximately 3,200 individuals with obesity caused by SH2B1 biallelic mutations, and over 131,600 individuals with obesity caused by SH2B1 heterozygous mutations in the United States.<sup>10</sup>

The melanocortin-4-receptor gene (MC4R) encodes a key regulator of energy homeostasis, food intake and body weight, working at the level of the hypothalamus.<sup>29-31</sup> MC4R gene mutations have been associated with childhood-onset obesity, hyperphagia and hyperinsulinism.<sup>32</sup> It is estimated that there are over 17,500 individuals with obesity caused by MC4R biallelic mutations, and over 161,000 individuals with obesity caused by MC4R heterozygote mutations in the United States.<sup>10</sup>

#### References

- 1 Hales CM, Carroll MD, Fryar CD, Ogden CL. Prevalence of Obesity and Severe Obesity Among Adults: United States, 2017-2018. *NCHS Data Brief*. 2020 Feb;(360):1-8.
- 2 Centers for Disease Control and Prevention. Overweight and Obesity – Childhood Overweight & Obesity. Accessed on May 23, 2021 and available at <https://www.cdc.gov/obesity/childhood/index.html>.
- 3 Kelly AS, Barlow SE, Rao G, et al. American Heart Association. Atherosclerosis, Hypertension, and Obesity in the Young Committee of the Council on Cardiovascular Disease in the Young, Council on Nutrition, Physical Activity and Metabolism, and Council on Clinical Cardiology. Severe obesity in children and adolescents: identification, associated health risks, and treatment approaches: a scientific statement from the American Heart Association. *Circulation*. 2013 Oct 8;128(15):1689-712.
- 4 Reilly JJ, Methven E, McDowell ZC, et al. Health consequences of obesity. *Arch Dis Child*. 2003 Sep;88:748-752.
- 5 American Academy of Family Physicians. Diagnosis and Management of Obesity. 2013. Accessed on April 30, 2021 and available at [https://www.aafp.org/dam/AAFP/documents/patient\\_care/fitness/obesity-diagnosis-mono.pdf](https://www.aafp.org/dam/AAFP/documents/patient_care/fitness/obesity-diagnosis-mono.pdf).
- 6 van der Valk ES, van den Akker ELT, Savas M, et al. A comprehensive diagnostic approach to detect underlying causes of obesity in adults. *Obes Rev*. 2019 Jun;20:795–804.
- 7 Heymsfield SB, Avena NM, Baier L, Brantley P, Bray GA, Burnett LC, et al. Hyperphagia: current concepts and future directions proceedings of the 2nd international conference on hyperphagia. Obesity (Silver Spring) 2014 Feb;22((S1 Suppl 1)):S1–17.
- 8 Ayers KL, Glicksberg BS, Garfield AS, Longerich S, White JA, Yang P, Du L, Chittenden TW, Gulcher JR, Roy S, Fiedorek F, Gottesdiener K, Cohen S, North KE, Schadt EE, Li SD, Chen R, Van der Ploeg LHT. Melanocortin 4 Receptor Pathway Dysfunction in Obesity: Patient Stratification Aimed at MC4R Agonist Treatment. *J Clin Endocrinol Metab*. 2018 Jul 1;103(7):2601-2612. doi: 10.1210/jc.2018-00258. PMID: 29726959; PMCID: PMC7263790.
- 9 Huvenne H, Dubern B, Clément K, Poitou C. Rare Genetic Forms of Obesity: Clinical Approach and Current Treatments in 2016. *Obes Facts*. 2016;9(3):158-173.
- 10 Bromberg E et al. Frequency of MC4R Pathway Variants in a Large US Cohorts of Patients with Severe Obesity. ObesityWeek, November 2022, San Diego, CA
- 11 Medline Plus. Genetics Home Reference. LEP gene. Accessed on April 30, 2021 and available at <https://medlineplus.gov/genetics/gene/lep/>
- 12 Wabitsch M et al. Severe Early-Onset Obesity Due to Bioinactive Leptin Caused by a p.N103K Mutation in the Leptin Gene. *J Clin Endocrinol Metab*. 2015;100(9):3227-3230.
- 13 Clement K et al. A mutation in the human leptin receptor gene causes obesity and pituitary dysfunction. *Nature*. 1998;392(6674):398-401

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- 14 Medline Plus. Genetics Home Reference. POMC gene. Accessed on April 30, 2021 and available at <https://medlineplus.gov/genetics/gene/pomc/#conditions>
- 15 Kühnen P., Clément K., Wiegand, S., et al. Proopiomelanocortin Deficiency Treated with a Melanocortin-4 Receptor Agonist. *N Engl J Med.* 2016 Jul 21;375(3):240-6.
- 16 Clément, K., van den Akker, E., Argente, J., et al. Efficacy and safety of setmelanotide, an MC4R agonist, in individuals with severe obesity due to LEPR or POMC deficiency: single-arm, open-label, multicentre, phase 3 trials. *Lancet Diabetes Endocrinol* 2020. Published Online October 30, 2020.
- 17 Stijnen P, Ramos-Molina B, O'Rahilly S, et al. PCSK1 Mutations and Human Endocrinopathies: From Obesity to Gastrointestinal Disorders. *Endocr Rev.* 2016 Aug;37(4):347–371.
- 18 Löffler D, Behrendt S, Creemers JWM, et al. Functional and clinical relevance of novel and known *PCSK1* variants for childhood obesity and glucose metabolism. *Mol Metab.* 2016;6(3):295-305. Published 2016 Dec 8. doi:10.1016/j.molmet.2016.12.002
- 19 Martín MG, Lindberg I, Solorzano-Vargas RS, et al. Congenital proprotein convertase 1/3 deficiency causes malabsorptive diarrhea and other endocrinopathies in a pediatric cohort. *Gastroenterology.* 2013;145(1):138-148. doi:10.1053/j.gastro.2013.03.048
- 20 Yang, Y., van der Klaauw, A.A., Zhu, L. *et al.* Steroid receptor coactivator-1 modulates the function of Pomc neurons and energy homeostasis. *Nat Commun.* 2019; 10:1718
- 21 York, B., et al., Steroid receptor coactivator (SRC) family: Masters of systems biology. *Journal of Biological Chemistry.* 2010; 285(50): 38743-38750
- 22 Zhu, L., et al., Steroid receptor coactivator-1 mediates estrogenic actions to prevent body weight gain in female mice. *Endocrinology.* 2013; 154(1): 150-8
- 23 Hill, J., et al., Acute effects of leptin require PI3K signaling in hypothalamic proopiomelanocortin neurons in mice. *Journal of Clinical Investigation.* 2008; 118(5): 1796-1805
- 24 Morton, G., et al., Leptin and the CNS control of glucose metabolism. *Physiological Review.* 2010; 91(2): 389-411
- 25 Ahmed, Z., et al., Adapter protein with a pleckstrin homology (PH) and an Src homology 2 (SH2) domain (APS) and SH2-B enhance insulin-receptor autophosphorylation, extracellular-signal-regulated kinase and phosphoinositide 3-kinase dependent signalling. *Biochemical Journal.* 2003; 371: 371-412
- 26 Duan, C., et al., SH2-B promotes insulin receptor substrate 1 (IRS1)- and IRS2-mediated activation of the phosphatidylinositol 3-kinase pathway in response to leptin. *Journal of Biological Chemistry.* 2004; 279(42):
- 27 Ren D, Li M, Duan C, Rui L. Identification of SH2-B as a key regulator of leptin sensitivity, energy balance, and body weight in mice. *Cell Metab.* 2005; 2(2): 95-104.
- 28 Ren, D., et al., Neuronal SH2B1 is essential for controlling energy and glucose homeostasis. *Journal of Clinical Investigation.* 2007; 117(2): 397-406
- 29 Oswal A, Yeo GS. The leptin melanocortin pathway and the control of body weight: Lessons from human and murine genetics. *Obes Rev.* 2007;8:293–306.
- 30 Tao YX. The melanocortin-4 receptor: Physiology, pharmacology, and pathophysiology. *Endocr Rev.* 2010;31:506–43.
- 31 Farooqi IS, Yeo GS, Keogh JM, et al. Dominant and recessive inheritance of morbid obesity associated with melanocortin 4 receptor deficiency. *J Clin Invest.* 2000;106:271–9.
- 32 Farooqi IS, Keogh JM, Yeo GS, Lank EJ, Cheetham T, O'Rahilly S. Clinical spectrum of obesity and mutations in the melanocortin 4 receptor gene. *N Engl J Med.* 2003;348:1085–95.

## TABULAR MODIFICATIONS

### Option #1

E88 Other and unspecified metabolic disorders

E88.8 Other specified metabolic disorders

New

sub-subcategory

E88.82 Obesity due to disruption of MC4R pathway

Add

Use additional code, if applicable, to identify associated manifestations, such as polyphagia (R63.2)

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Add		Use additional code to identify body mass index (BMI), if known (Z68.-)
New code	E88.820	Obesity due to disruption of MC4R pathway, Leptin (LEP) gene mutation
New code	E88.821	Obesity due to disruption of MC4R pathway, Leptin receptor (LEPR) gene mutation
New code	E88.822	Obesity due to disruption of MC4R pathway, Proopiomelanocortin (POMC) gene mutation
New code	E88.823	Obesity due to disruption of MC4R pathway, Proprotein convertase subtilisin/kexin type 1 (PCSK1) gene mutation
New code	E88.824	Obesity due to disruption of MC4R pathway, Melanocortin 4 Receptor (MC4R) gene mutation
New code	E88.825	Obesity due to disruption of MC4R pathway, Src homology 2B adaptor signaling protein (SH2B1) gene mutation
New code	E88.826	Obesity due to disruption of MC4R pathway, Nuclear receptor coactivator 1 (NCOA1) gene mutation
New code	E88.828	Obesity due to disruption of MC4R pathway, not elsewhere classified

**Option #2**

Z15 Genetic susceptibility to disease

New subcategory

Z15.2 Genetic susceptibility to obesity

Add  
 Add  
 Add  
 Add

Code also, if applicable, any associated manifestations, such as:  
 other obesity (E66.8)  
 polyphagia (R63.2)  
 Use additional code to identify body mass index (BMI), if known (Z68.-)

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New sub-subcategory	Z15.21	Genetic susceptibility to obesity, related to disruption of MC4R pathway
New code	Z15.210	Genetic susceptibility to obesity, related to melanocortin 4 receptor [MC4R] gene mutation
Add		Genetic susceptibility to obesity, related to pathogenic variant of MC4R gene
New code	Z15.211	Genetic susceptibility to obesity, related to leptin [LEP] gene mutation
Add		Genetic susceptibility to obesity, related to pathogenic variant of LEP gene
New code	Z15.212	Genetic susceptibility to obesity, related to leptin receptor [LEPR] gene mutation
Add		Genetic susceptibility to obesity, related to pathogenic variant of LEPR gene
New code	Z15.213	Genetic susceptibility to obesity, related to proopiomelanocortin [POMC] gene mutation
Add		Genetic susceptibility to obesity, related to pathogenic variant of POMC gene
New code	Z15.214	Genetic susceptibility to obesity, related to proprotein convertase subtilisin/kexin type 1 [PCSK1] gene mutation
Add		Genetic susceptibility to obesity, related to pathogenic variant of PCSK1 gene
New code	Z15.215	Genetic susceptibility to obesity, related to src homology 2B adaptor signaling protein [SH2B1] gene mutation
Add		Genetic susceptibility to obesity, related to pathogenic variant of SH2B1 gene
New code	Z15.216	Genetic susceptibility to obesity, related to nuclear receptor coactivator 1 [NCOA1] gene mutation
Add		Genetic susceptibility to obesity, related to pathogenic variant of NCOA1 gene

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New code	Z15.219	Genetic susceptibility to obesity, related to mutation of other gene with disruption of MC4R pathway
New code	Z15.29	Genetic susceptibility to obesity, related to other gene mutation

## Multiple Sclerosis Phenotypes

Multiple sclerosis (MS) is a chronic inflammatory disease of the central nervous system leading to demyelination and neurodegeneration. MS affects an estimated 900,000 people in the United States [1]. Diagnosis is based on a combination of signs and symptoms, radiographic findings, and laboratory findings, which are components of the 2017 McDonald Criteria [2]. The core MS phenotypes are those of relapsing-remitting and progressive disease. The pattern and course of MS is further categorized into several clinical subtypes: clinically isolated syndrome, often representing the first attack of MS, relapsing-remitting multiple sclerosis (RRMS), secondary progressive multiple sclerosis (SPMS), and primary progressive multiple sclerosis (PPMS). The large majority of patients with MS initially follow a relapsing-remitting course, defined by acute exacerbations from which they typically completely or incompletely recover, with periods of relative clinical stability in between [3]. The transition from RRMS to SPMS usually occurs 10 to 20 years after disease onset; SPMS is associated with sustained disability progression and loss of discrete relapse events [4]. Both clinical relapses and new radiographic lesions can occur in the context of progressive MS, and establishing when the transition from relapsing to progressive MS occurs is often difficult to determine prospectively [5]. SPMS can be further characterized as either active (with clinical relapses) or not active (without relapses). PPMS, which represents about 10 percent of adult MS cases at disease onset, is characterized by, often insidious, disease progression from onset, although occasional plateaus, temporary minor improvements, and acute relapses may occur [6]. Currently, there is only one ICD-10-CM code for MS (G35), which does not capture the MS phenotypes.

Several classes of drugs and biologics to treat MS, with varying mechanisms of action and routes of administration, are available for RRMS and active SPMS [1]. However, as of today, only one monoclonal antibody is approved for the treatment of PPMS. The proposed codes follow the recent revamping of U.S. package insert (USPI) indications performed by the U.S. Food and Drug Administration for all approved MS therapeutics and provides a broad separation between PPMS and other phenotypes, a fundamental clinical distinction with therapeutic and prognostic implications.

This phenotype specificity would provide the ability to make inferences at the phenotype level, improve the interpretation of real-world data in the investigation of severe outcomes that might represent safety signals for some treatments approved to treat MS, and improve the design and interpretation of comparative safety and effectiveness studies.

The Center for Drug Evaluation and Research, U.S. Food and Drug Administration, is requesting new ICD-10-CM codes for Multiple Sclerosis (MS) to distinguish between different disease clinical courses, evaluation of disease progression and long-term prognosis of MS in large population-based epidemiological assessments.

### References

1. McGinley, M.P., C.H. Goldschmidt, and A.D. Rae-Grant, *Diagnosis and Treatment of Multiple Sclerosis: A Review*. JAMA, 2021. **325**(8): p. 765-779.
2. Thompson, A.J., et al., *Diagnosis of multiple sclerosis: 2017 revisions of the McDonald criteria*. Lancet Neurol, 2018. **17**(2): p. 162-173.

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3. Katz Sand, I., *Classification, diagnosis, and differential diagnosis of multiple sclerosis*. *Curr Opin Neurol*, 2015. **28**(3): p. 193-205.
4. Kleiter, I., et al., *The transitional phase of multiple sclerosis: Characterization and conceptual framework*. *Mult Scler Relat Disord*, 2020. 44: p. 102242.
5. Ontaneda, D., *Progressive Multiple Sclerosis*. *Continuum (Minneap Minn)*, 2019. **25**(3): p. 736-752.
6. Willis, M.A. and R.J. Fox, *Progressive Multiple Sclerosis*. *Continuum (Minneap Minn)*, 2016. **22**(3): p. 785-98.

**TABULAR MODIFICATIONS**

Delete	G35	Multiple sclerosis
Delete		<del>Disseminated multiple sclerosis</del>
Delete		<del>Generalized multiple sclerosis</del>
Delete		<del>Multiple sclerosis NOS</del>
Delete		<del>Multiple sclerosis of brain stem</del>
Delete		<del>Multiple sclerosis of cord</del>
New code	G35.A	Relapsing-remitting multiple sclerosis
Add		Exclude1: Demyelinating disease of central nervous system, unspecified (G37.9)
Add		Clinically isolated syndromes (G37.9)
New code	G35.B	Primary progressive multiple sclerosis
New	G35.C	Secondary progressive multiple sclerosis
Subcategory		G35.C0 Secondary progressive multiple sclerosis, unspecified
New code		G35.C1 Active secondary progressive multiple sclerosis
New code		G35.C2 Non-active secondary progressive multiple sclerosis
New code	G35.D	Multiple sclerosis, unspecified
Add		Disseminated multiple sclerosis
Add		Generalized multiple sclerosis
Add		Multiple sclerosis NOS
Add		Multiple sclerosis of brain stem
Add		Multiple sclerosis of cord
	G37	Other demyelinating diseases of central nervous system
Add	G37.9	Demyelinating disease of central nervous system, unspecified Clinically isolated syndromes

## Obesity Classes

Obesity is recognized as a highly prevalent chronic disease with complex inflammatory and endocrinological pathophysiology, with serious health and social consequences.<sup>1-3</sup> A previous proposal related to obesity classes was presented at the September 2022 ICD-10 Coordination and Maintenance meeting, and further clinical details are available from that proposal.

Obesity in children and adolescents is determined by age- and gender-specific percentiles. Therefore, a child or adolescent may suffer from obesity at a lower BMI than an adult. For adults, the overweight range is from a BMI of 25.0 to <30. Obesity in adults is subdivided into the following: Class 1: BMI of 30 to < 35; Class 2: BMI of 35 to < 40; Class 3: BMI of 40 or higher (sometimes categorized as “severe” obesity).<sup>4</sup> Obesity in children uses a classification system recognizing BMI  $\geq$ 95th percentile as class I obesity, BMI  $\geq$ 120% of the 95th percentile as class II obesity, and BMI  $\geq$ 140% of the 95th percentile as class III obesity.<sup>5</sup>

Having a high BMI-for-age is associated with clinical risk factors for cardiovascular disease, including high cholesterol and high blood pressure,<sup>6</sup> and other chronic conditions. Obesity has been an ongoing problem in children and adolescents.<sup>6,7</sup> It is recommended that adolescents with class 2 obesity together with a co-morbidity, or with class 3 obesity should be considered for bariatric surgery.<sup>8</sup> In January 2023, the AAP released a Clinical Practice Guideline for the Evaluation and Treatment of Children and Adolescents With Obesity to inform pediatric healthcare providers about the standard of care for youth with overweight and obesity and related comorbidities.<sup>9</sup>

Proposed changes to the ICD-10-CM obesity codes have been received from within CDC, from the Division of Nutrition, Physical Activity, and Obesity, of the National Center for Chronic Disease Prevention and Health Promotion; with further input from additional obesity experts. This proposal is a result of combined input, as well as comments from the previous presentation.

### References

1. Mechanick JI, Garber AJ, Handelsman Y, Garvey WT. American Association of Clinical Endocrinologists' position statement on obesity and obesity medicine. *Endocr Pract* 2012; 18: 642-648.
2. American Medical Association. Recognition of obesity as a disease H-440.842. <https://policysearch.ama-assn.org/policyfinder/detail/obesity?uri=%2FAMADoc%2FHOD.xml-0-3858.xml>.
3. Centers for Disease Control and Prevention. Causes of Obesity. <https://www.cdc.gov/obesity/basics/causes.html>
4. Centers for Disease Control and Prevention. Defining Adult Overweight & Obesity. <https://www.cdc.gov/obesity/basics/adult-defining.html>
5. Skinner AC, Ravanbakht SN, Skelton JA, et al. Prevalence of Obesity and Severe Obesity in US Children, 1999–2016. *Pediatrics* (2018) 141 (3): e20173459. <https://doi.org/10.1542/peds.2017-3459>
6. Skinner AC, Perrin EM, Moss LA, Skelton JA. Cardiometabolic Risks and Severity of Obesity in Children and Young Adults. *N Engl J Med*. 2015 Oct;373(14):1307-17. PMID: 26422721. <https://doi.org/10.1056/NEJMoal502821>
7. Skinner AC, Skelton JA. Prevalence and trends in obesity and severe obesity among children in the United States, 1999-2012. *JAMA Pediatr*. 2014 Jun;168(6):561-6. PMID: 24710576. <https://doi.org/10.1001/jamapediatrics.2014.21>
8. Pratt JSA, Browne A, Browne NT, et al. ASMBS pediatric metabolic and bariatric surgery guidelines, 2018. *Surg Obes Relat Dis*. 2018 Jul;14(7):882-901. Epub 2018 Mar 23. PMID: 30077361 PMID: PMC6097871. <https://doi.org/10.1016/j.soard.2018.03.019>
9. Hampl SE, Hassink SG, Skinner AC, et al. Clinical Practice Guideline for the Evaluation and Treatment of Children and Adolescents With Obesity. *Pediatrics* 2023 Feb 1;151(2):e2022060640. <https://doi.org/10.1542/peds.2022-060640>



**TABULAR MODIFICATIONS**

**Option #1**

	E66	Overweight and obesity
Revise		Use additional code to identify body mass index (BMI), if known, <u>for adults (Z68.1-Z68.45) or pediatrics (Z68.5-)</u>
Delete	<del>Excludes1:</del>	<del> adiposogenital dystrophy (E23.6) lipomatosis NOS (E88.2) lipomatosis dolorosa [Dercum] (E88.2) Prader-Willi syndrome (Q87.11)</del>
Add	Excludes2:	adiposogenital dystrophy (E23.6)
Add		lipomatosis NOS (E88.2)
Add		lipomatosis dolorosa [Dercum] (E88.2)
Add		Prader-Willi syndrome (Q87.11)
	E66.8	Other obesity
New sub-subcategory	E66.81	Obesity class
New code	E66.811	Obesity, class 1
New code	E66.812	Obesity, class 2
New code	E66.813	Obesity, class 3
New code	E66.89	Other obesity not elsewhere classified

**Option #2**

	E66	Overweight and obesity
Revise		Use additional code to identify body mass index (BMI), if known, <u>for adults (Z68.1-Z68.45) or pediatrics (Z68.5-)</u>
Delete	<del>Excludes1:</del>	<del> adiposogenital dystrophy (E23.6) lipomatosis NOS (E88.2) lipomatosis dolorosa [Dercum] (E88.2) Prader-Willi syndrome (Q87.11)</del>
Add	Excludes2:	adiposogenital dystrophy (E23.6)
Add		lipomatosis NOS (E88.2)
Add		lipomatosis dolorosa [Dercum] (E88.2)

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Add		Prader-Willi syndrome (Q87.11)
	E66.8	Other obesity
New sub-subcategory	E66.81	Obesity in children and adolescents
Add		Use additional code to identify body mass index (BMI), pediatric, if known (Z68.5-)
New code	E66.811	Obesity in children and adolescents, class 1
New code	E66.812	Obesity in children and adolescents, class 2
New code	E66.813	Obesity in children and adolescents, class 3
New code	E66.819	Obesity in children and adolescents, unspecified
New sub-subcategory	E66.82	Obesity in adults
Add		Use additional code to identify body mass index (BMI), adult, if known (Z68.1-Z68.45)
New code	E66.821	Obesity in adults, class 1
New code	E66.822	Obesity in adults, class 2
New code	E66.823	Obesity in adults, class 3
New code	E66.829	Obesity in adults, unspecified
New code	E66.89	Other obesity not elsewhere classified

## **Post-exertional malaise/post-exertional symptom exacerbation**

The majority of Long COVID patients experience an exacerbation of some or all their symptoms and a further reduction in functioning following physical or cognitive exertion or emotional, positional, sensory, or other stressors that would have been tolerated before their illness. This symptom is a defining characteristic of myalgic encephalomyelitis/chronic fatigue syndrome (ME/CFS) and is referred to as post-exertional malaise (PEM) or post-exertional symptom exacerbation (PESE). PEM is characterized by exacerbation of some or all a patient's symptoms or occurrence of new symptoms. Symptoms exacerbated can include physical fatigue, cognitive fatigue, exercise intolerance, problems thinking (e.g., slowed information processing speed, memory, concentration), unrefreshing sleep, muscle pain, joint pain, headaches, weakness/instability, light-headedness, flu-like symptoms, sore throat, nausea, orthostatic intolerance or other autonomic dysfunctions, sensory sensitivities, and other symptoms.

Characteristics include pathological loss of stamina and/or functional capacity that is not due to physical deconditioning. An onset that can be immediate or delayed after the exertional stimulus by hours to days. A prolonged, unpredictable time to return to baseline that is not easily relieved by rest or sleep and may last days, weeks, months, or longer. Severity and duration of symptoms that is often out-of-proportion to the type, intensity, frequency, and/or duration of the exertion. Even basic activities of daily living like toileting, bathing, dressing, communicating, and reading can trigger PEM/PESE. In some instances, the specific precipitant cannot be identified. The threshold for a precipitant to trigger PEM/PESE can vary between individuals as well as within the same individual, at different times during their illness.

This symptom has important implications for Long COVID diagnosis, treatment, disability assessment, morbidity tracking, and research using electronic health records and is equally important for ME/CFS and other infection-associated syndromes. For instance:

- In its guidance on fatigue and recommendations for exercise, the American Academy of Physical Medicine and Rehabilitation discusses the importance of identifying PEM and cautions against exercise programs that provoke PEM.
- Numerous Long COVID studies, including those in NIH's RECOVER Initiative and by the CDC, are now using electronic health records to identify important sequelae of an acute SARS CoV-2 infection. PEM/PESE is virtually invisible in these studies.

Currently, the ICD-10-CM does not have a code for the symptom of PEM/PESE as described for Long COVID and myalgic encephalomyelitis/chronic fatigue syndrome (ME/CFS). As a result, this important symptom is not being identified or tracked in electronic health records. A code for this symptom would also facilitate education of physicians, improve communication between a

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patient and clinician and between the PCP and other healthcare providers, and improve the accuracy of Long COVID and ME/CFS research and surveillance using electronic health records.

It's important to note that while PEM is included in a diagnosis of ME/CFS, not all patients with Long COVID and PEM/PESE have been given an ME/CFS diagnosis, either because the patient has not yet met the 6 month diagnostic requirement of ME/CFS in adults or because the patient does not meet the other ME/CFS criteria.

Patient-Led Research Collaborative, with support from #MEAction, Open Medicine Foundation, Solve ME/CFS Initiative, Massachusetts ME/CFS & FM Association, and Pandora Org, is requesting a new ICD-10-CM code to identify patients without the diagnosis of ME/CFS.

## References

1. Davis, H. E.; Assaf, G. S.; McCorkell, L.; et al. Characterizing Long COVID in an International Cohort: 7 Months of Symptoms and Their Impact. *EClinicalMedicine* 2021, 38, 101019. <https://doi.org/10.1016/j.eclinm.2021.101019>
2. Twomey, R.; DeMars, J.; Franklin, K.; et al. Chronic Fatigue and Postexertional Malaise in People Living With Long COVID: An Observational Study. *Physical Therapy* 2022, 102 (4), pzac005. <https://doi.org/10.1093/ptj/pzac005>
3. Kedor, C.; Freitag, H.; Meyer-Arndt, L.; et al. Chronic COVID-19 Syndrome and Chronic Fatigue Syndrome (ME/CFS) Following the First Pandemic Wave in Germany – a First Analysis of a Prospective Observational Study; preprint; *Infectious Diseases (except HIV/AIDS)*, 2021. <https://doi.org/10.1101/2021.02.06.21249256>
4. US Institute of Medicine. Beyond myalgic encephalomyelitis/ chronic fatigue syndrome: redefining an illness. Report guide for clinicians. The National Academies Press. February 2015. <https://www.nap.edu/resource/19012/MECFScliniciansguide.pdf>
5. Bateman, L.; Bested, A. C.; Bonilla, H. F.; et al. Myalgic Encephalomyelitis/Chronic Fatigue Syndrome: Essentials of Diagnosis and Management. *Mayo Clinic Proceedings* 2021, 0 (0). <https://doi.org/10.1016/j.mayocp.2021.07.004>
6. What Are ME/CFS Symptoms? Centers for Disease Control and Prevention. [https://www.cdc.gov/me-cfs/pdfs/interagency/What-are-MECFS-Symptoms\\_508.pdf](https://www.cdc.gov/me-cfs/pdfs/interagency/What-are-MECFS-Symptoms_508.pdf) Accessed June 6, 2022
7. Stussman, B.; Williams, A.; Snow, J.; Gavin, A.; Scott, R.; Nath, A.; Walitt, B. Characterization of Post-Exertional Malaise in Patients with Myalgic Encephalomyelitis/Chronic Fatigue Syndrome. *Front. Neurol.* 2020, 11, 1025. <https://doi.org/10.3389/fneur.2020.01025>
8. Keller BA, Pryor JL, Giloteaux L. Inability of myalgic encephalomyelitis/chronic fatigue syndrome patients to reproduce VO2peak indicates functional impairment. *J Transl Med.* 2014;12(1):104.
9. Herrera, J. E.; Niehaus, W. N.; Whiteson, J.; et al. Multidisciplinary Collaborative Consensus Guidance Statement on the Assessment and Treatment of Fatigue in postacute Sequelae of SARS-CoV -2 Infection ( PASC ) Patients. *PM&R* 2021, 13 (9), 1027–1043. <https://doi.org/10.1002/pmrj.12684>
10. World Physiotherapy response to COVID-19 Briefing paper 9. *World Physiotherapy.* June 2021. <https://world.physio/sites/default/files/2021-06/Briefing-Paper-9-Long-Covid-FINAL-2021.pdf>
11. Podell, R.; Dimmock, M. E.; Comerford, B. B. Documenting Disability in Myalgic Encephalomyelitis/Chronic Fatigue Syndrome (Me/Cfs). *WOR* 2020, 66 (2), 339–352. <https://doi.org/10.3233/WOR-203178>
12. Chu, L.; Fuentes, L. R.; Marshall, O. M.; Mirin, A. A. Environmental Accommodations for University Students Affected by Myalgic Encephalomyelitis/Chronic Fatigue Syndrome (ME/CFS). *WOR* 2020, 66 (2), 315–326. <https://doi.org/10.3233/WOR-203176>
13. Pfaff, E. R.; Girvin, A. T.; Bennett, T. D.; et al. Identifying Who Has Long COVID in the USA: A Machine Learning Approach Using N3C Data. *The Lancet Digital Health* 2022, S2589750022000486. [https://doi.org/10.1016/S2589-7500\(22\)00048-6](https://doi.org/10.1016/S2589-7500(22)00048-6)

**TABULAR MODIFICATIONS**

	R68 Other general symptoms and signs
New code	R68.A Post-exertional malaise
Add	PEM
Add	PESE
Add	Post-exertional symptom exacerbation

## **Primary Central Nervous System Lymphoma**

Primary Central Nervous System Lymphoma (PCNSL) is an aggressive extranodal form of non-Hodgkin's lymphoma which originates directly in the brain, meninges, or spinal cord. There was a specific code for it in ICD-9-CM, and creation of a specific code in ICD-10-CM has been proposed by ONO Pharma USA.

Although under study, the cause of PCNSL is not known. For many years, it was believed that the central nervous system was devoid of lymphatics. In 2015, however, researchers identified lymphatic vessels lining the dural sinuses and draining into the deep cervical lymph nodes. Still, it remains unclear how this may or may not relate to PCNSL. Immune compromise, including inborn or acquired immune disorders as well as immune suppression, eg, post-transplant, is a known risk factor for developing PCNSL.

PCNSL is a rare disorder, comprising about 4-6% of extranodal lymphomas and 1% of all non-Hodgkin's lymphomas. Its incidence has been estimated from about 0.5 to 0.7 per 100,000 in the US. Interestingly, there was an observed spike in incidence in the 1990's, considered to be associated with the HIV epidemic. Overall incidence has decreased since then, but it has continued to increase in older men and women.

There are several cell types which can make up PCNSL. The vast majority of cases, about 90-95%, are diffuse large B-cell lymphoma. The remaining cell types include lymphoblastic, T-cell, and Burkitt lymphoma.

PCNSL is most commonly found in the frontal lobe and basal ganglia of the brain, and occasionally in the spinal cord. Symptoms include focal neurological deficits, behavioral and mental status changes, and indications of increased intracranial pressure such as headaches and papilledema. The diagnosis is generally established by stereotactic biopsy of the lesion. Imaging, including MRI and PET, is used for baseline staging and to detect any disease outside the central nervous system if present.

Treatment of PCNSL is non-surgical, relying on chemotherapy and radiation therapy. Induction chemotherapy typically involves some combination of high-dose methotrexate with other chemotherapeutic agents. The consolidation phase of treatment usually involves radiation, including whole brain radiotherapy, with additional chemotherapy. Although PCNSL is often initially sensitive to chemotherapy and radiotherapy, relapse is unfortunately seen in more than half of patients, usually within five years. The prognosis for PCNSL is dismal if untreated, with average survival of less than two months from diagnosis. With treatment, the five-year survival rate is 30%. It is notable that the prognosis remains poor in comparison to other lymphomas that arise outside the central nervous system.

For data analysis, PCNSL was uniquely identifiable in ICD-9-CM with code 200.50, Primary central nervous system lymphoma, unspecified site, extranodal and solid organ sites. However, ICD-10-CM does not uniquely identify PCNSL, and it has been reported that a variety of codes are being assigned, including C71.9, Malignant neoplasm of brain, unspecified; C72.9,

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Malignant neoplasm of central nervous system, unspecified; C83.39, Diffuse large B-cell lymphoma, extranodal and solid organ sites; and C83.89, Other non-follicular lymphoma, extranodal and solid organ sites. Creating a unique code for PCNSL in ICD-10-CM will help ensure consistent identification of the disorder for accurate tracking and research efforts.

References

- Green K, Hogg JP. Central Nervous System Lymphoma. NIH: National Library of Medicine, StatPearls. June 27, 2022. <https://www.ncbi.nlm.nih.gov/books/NBK545145/>
- Grommes C, DeAngelis LM. Primary CNS lymphoma. *J Clin Oncol* 35:2410-2418. 2017 July 20. <https://doi.org/10.1200/JCO.2017.72.7602>
- Lv C, Wang J, Zhou M, Xu J, Chen B, Wan Y. Primary central nervous system lymphomas in the United States 1975-2017. *Ther Adv Hematol*. 2022 Jan 23. <https://doi.org/10.1177/20406207211066166>
- Menon MP, Nicolae A, Meeker H et al. Primary CNS T-cell lymphomas: A clinical, morphologic, immunophenotypic and molecular analysis. *Am J Surg Pathol*. 2015 December; 39(12): 1719–1729. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4644095/>
- O'Neill BP, Decker PA, Tieu C, Cerhan JR. The changing incidence of primary central nervous system lymphoma is driven primarily by the changing incidence in young and middle-aged men and differs from time trends in systemic diffuse large B-cell non-Hodgkin's lymphoma. *Am J Hematol*. 2013 Dec; 88(12): 997–1000. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4020348/>
- Yang XL, Liu YB. Advances in pathobiology of primary central nervous system lymphoma. *Chin Med J* 2017;130:1973-9. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5555133/>

**TABULAR MODIFICATIONS**

- C83 Non-follicular lymphoma
  - C83.3 Diffuse large B-cell lymphoma
    - C83.39 Diffuse large B-cell lymphoma, extranodal and solid organ sites

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New code	C83.390	Primary central nervous system lymphoma
Add		PCNSL of brain
Add		PCNSL of meninges
Add		PCNSL of spinal cord
Add		PCNSL NOS
Add		Excludes1: Primary central nervous system lymphoma, Burkitt (C83.79)
Add		Primary central nervous system lymphoma, lymphoblastic (C83.59)
Add		Primary central nervous system lymphoma, other (C83.89)
Add		Primary central nervous system lymphoma, T-cell (C84.49)
New code	C83.398	Diffuse large B-cell lymphoma of other extranodal and solid organ sites



## SCN2A-related disorders

The FamilieSCN2A Foundation has requested that a unique ICD-10-CM code or codes be created for SCN2A-related disorders. SCN2A-related disorders are a group of neurodevelopmental disorders characterized by epilepsy, autism spectrum disorder (ASD), and/or intellectual disability (ID).<sup>1-7</sup> Over 1,000 cases of SCN2A-related disorders have been identified to date, with an estimated prevalence of 7.99 in 100,000.<sup>8</sup> The SCN2A-related disorders have three phenotypic categories identified<sup>2</sup>:

1. benign familial neonatal-infantile seizures (SCN2A-BFNIS) (20%)
2. developmental and epileptic encephalopathies (SCN2A-DEE) (60-70%)
3. autism spectrum disorder/intellectual disability (SCN2A-ASD/ID) (16%)

These three SCN2A-related disorders are all caused by variants in the sodium channel, voltage-gated, type II, alpha gene (SCN2A) on chromosome 2.<sup>2</sup> The SCN2A gene encodes the protein for the alpha subunit of the voltage-gated type II sodium channel.<sup>1-6</sup> These channels are expressed in excitatory, glutamatergic neurons of the central nervous system and are involved in action-potential initiation and propagation.<sup>2,5</sup>

Depending on the type and location of the mutation, the result can be either a gain-of-function (GoF), a loss-of-function (LoF), or a mixed non-specific nonspecific abnormal function of the sodium channel. SCN2A mutations resulting in GoF effects in the sodium channel cause increased neuronal excitability, which in turn leads to seizures, while mutations resulting in LoF effects on the channel can cause a decrease in neuronal excitability with downstream effects on cortical circuits, often resulting in autism spectrum disorder and/or developmental delay.<sup>7</sup> The phenotypic categories within SCN2A-related disorders are further described below.

The benign familial neonatal-infantile seizures (SCN2A-BFNIS) phenotype was first reported in 2002 and is caused by a missense GoF mutation in SCN2A, with 82% of mutations inherited in an autosomal dominant manner.<sup>1-3</sup> The SCN2A-BFNIS phenotype is characterized by infantile-onset seizures which start between the first day of life and 23 months of age.<sup>3</sup> Seizures will generally resolve by 2 years of age without long-term neuropsychiatric sequelae.<sup>5</sup>

The second distinct phenotype of SCN2A-related disorders is developmental and epileptic encephalopathies (DEE). First reported in 2009, the SCN2A-DEE phenotype is characterized by developmental delays and epilepsy and accounts for 60-70% of SCN2A-related cases.<sup>1-3</sup> This phenotype is split into two categories based on the age of onset: neonatal and early-infantile, and infantile and childhood.<sup>1,4</sup> Around two-thirds of patients with DEE are considered neonatal and early-infantile, defined as seizure onset prior to 3 months of age.<sup>1</sup> This phenotype is caused by a de novo missense mutation in SCN2A with GoF effects. Infantile and childhood DEE patients usually experience onset after 3 months but prior to 4 years of age and account for 30% of patients with DEE.<sup>1,4</sup> This phenotype usually results from de novo loss-of function (LoF) missense, protein-truncating, and splice site variants in SCN2A.<sup>1</sup> There have also been reports of missense variants with mixed GoF and LoF mutations.<sup>4</sup> Along with seizures, around 75% of SCN2A-DEE patients experience moderate to severe intellectual disability.<sup>1,4</sup> Comorbidities for

SCN2A-DEE phenotypes include ASD, hypotonia, microcephaly, cerebral/cerebellar atrophy, and cortical visual impairment.<sup>1,4</sup> Movement disorders including dystonia, dyskinesia, choreoathetosis, stereotypies, opisthotonos, and oculogyric crisis, with a frequency ranging from 11-84%.<sup>1,4</sup> Childhood survival is also a concern for early-onset SCN2A-DEE patients, with causes of death including sudden unexpected death in epilepsy (SUDEP), autonomic dysfunction, pneumonia, and cardiorespiratory failure.<sup>1</sup>

Finally, the SCN2A-related autism spectrum disorder and intellectual disability phenotype (SCN2A-ASD/ID), described in 2012, is usually caused by a *de novo* truncating LoF mutation in SCN2A.<sup>1,2,4</sup> Sixteen percent of individuals diagnosed with a mutation in the SCN2A gene fall into this cohort, but this is likely significantly lower than the true prevalence because genetic screening for SCN2A variants is more common for patients with epilepsy, than it is for patients with ASD/ID.<sup>1,3</sup> Children with the SCN2A-ASD/ID phenotype develop normally until 6 months of age, after which they begin exhibiting motor, social, and language delay.<sup>1,4</sup> Symptoms of this phenotype include engagement in repetitive activity, reluctance to initiate social interactions, poor eye contact, uncoordinated oral movements, gastrointestinal issues, cortical visual impairment, clumsiness, ataxia, and sleep problems.<sup>1,2,4</sup> Some SCN2A-ASD/ID patients have also presented with schizophrenia.<sup>3</sup>

In summary, the three identified SCN2A-related disorders differ considerably, and yet each will be diagnosed and treated based on their common genetic cause, though the epidemiology of each subcategory will likely be distinct. While most children with *SCN2A* variants fit into one of these three categories, there are some exceptions, including epileptic encephalopathy with choreoathetoid movements, benign infantile seizures with late-onset episodic ataxia, childhood-onset epileptic encephalopathy, and schizophrenia. Further work is required to understand how these disorders relate to the other three distinct categories.

Although many individuals with SCN2A mutations suffer from seizures, this is not present in all cases of SCN2A-related disorders. In addition, intellectual disability is a defining feature of SCN2A-related disorders. Creation of a specific code or codes for SCN2A-related disorders will benefit epidemiological research and support patient well-being, with the ability to track hospital admissions and morbidities.

#### References

1. Wolff, M., Brunklaus, A. & Zuberi, S. M. Phenotypic spectrum and genetics of SCN2A-related disorders, treatment options, and outcomes in epilepsy and beyond. *Epilepsia* 60, (2019).
2. Sanders, S. J. et al. Progress in Understanding and Treating SCN2A-Mediated Disorders. *Trends Neurosci.* 41, 442–456 (2018).
3. Wolff, M. et al. Genetic and phenotypic heterogeneity suggest therapeutic implications in SCN2A-related disorders. *Brain* 140, 1316–1336 (2017).
4. Reynolds, C., King, M. D. & Gorman, K. M. The phenotypic spectrum of SCN2A-related epilepsy. *Eur. J. Paediatr. Neurol.* 24, 117–122 (2020).
5. Howell, K. B. et al. SCN2A encephalopathy: A major cause of epilepsy of infancy with migrating focal seizures. *Neurology* 85, 958–966 (2015).
6. Schwarz, N. et al. Clinical and genetic spectrum of SCN2A-associated episodic ataxia. *Eur. J. Paediatr. Neurol.* 23, 438–447 (2019).

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7. Ben-Shalom, R. et al. Opposing Effects on Na V 1.2 Function Underlie Differences Between SCN2A Variants Observed in Individuals With Autism Spectrum Disorder or Infantile Seizures. *Biol. Psychiatry* 82, 224–232 (2017).
8. López-Rivera, J. A. et al. A catalogue of new incidence estimates of monogenic neurodevelopmental disorders caused by de novo variants. *Brain* (2020) doi:10.1093/brain/awaa051.

TABULAR MODIFICATIONS

**Option #1**

Note: this option was created based on a modified approach from the original submission.

F84 Pervasive developmental disorders

F84.8 Other pervasive developmental disorders

New sub-subcategory	F84.81	SCN2A-related disorders
Add		Code also, if applicable, related disorders such as developmental and epileptic encephalopathy (G93.42)
Add		developmental disorders of scholastic skills (F81.-)
Add		developmental disorder of speech and language (F80.-)
Add		epilepsy, by specific type (G40.-)
Add		intellectual disabilities (F70-F79)
Add		other neurodevelopmental disorder (F88)
New code	F84.811	SCN2A-related disorder with benign familial neonatal-infantile seizures [SCN2A-BFNIS]
New code	F84.812	SCN2A-related disorder with developmental and epileptic encephalopathies [SCN2A-DEE]
New code	F84.813	SCN2A-related disorder with autism spectrum disorder and intellectual disability phenotype [SCN2A-ASD/ID]
New code	F84.819	SCN2A-related disorder, unspecified
New code	F84.89	Other pervasive developmental disorders not elsewhere classified

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**Option #2**

Note: this option was created by NCHS based on a generalized approach for a number of proposals. See also the Developmental Epileptic Encephalopathies topic.

Z15 Genetic susceptibility to disease

New  
subcategory

Z15.1 Genetic susceptibility to epilepsy and neurodevelopmental disorders

New sub-  
subcategory

Z15.11 Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to ion channel genes

New code

Z15.111 Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of SCN2A gene  
Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of the gene for sodium channel, voltage-gated, type II, alpha subunit

Add

## SLC13A5 Citrate Transporter Disorder

TESS Research Foundation (TESS) is proposing that a new specific ICD-10-CM code be created for SLC13A5 Citrate Transporter Disorder (SLC13A5 Deficiency, EIEE25, DEE25, SLC13A5 Epilepsy, OMIM #615905). A unique ICD-10-CM code would also help with tracking and assessing patient morbidity.

Although there are no known cures for SLC13A5 Citrate Transporter Disorder, one treatment specifically for SLC13A5 Citrate Transporter Disorder is currently in development and has received rare pediatric disease and orphan drug designations from the U.S. FDA. This treatment also received orphan drug designation from the European Commission in August of 2021.

SLC13A5 Citrate Transporter Disorder has a unique, consistent, and specific phenotypic presentation. Patients are initially identified by the multiple types of seizures that begin within the first week of life<sup>1,2</sup>. Background electroencephalograms (EEGs) outside the neonatal period are well organized<sup>1,3</sup>. Patients have additional persistent symptoms including teeth hypoplasia, ataxia, poor communication skills, trouble standing or walking, as well as below average growth<sup>1,4</sup>. Importantly, SLC13A5 Citrate Transporter Disorder requires accurate diagnosis in order for patients to receive proper diagnosis and treatment, particularly with the precision therapy currently in development for this disease. Proper diagnosis and care is also important since patients are dependent on caregivers throughout life.

SLC13A5 Citrate Transporter Disorder is caused by mutations in the *SLC13A5* gene that codes for a sodium citrate cotransporter, NaCT. NaCT is responsible for transporting citrate, a key substrate in cellular metabolism, from the extracellular fluid into the cell. NaCT is expressed in multiple tissues with the highest expression in the liver, the brain, and reproductive organs<sup>5</sup>. Pathogenic variants in the *SLC13A5* gene lead to decreased expression and mislocalization of NaCT, as well as impaired citrate transport<sup>6-9</sup>. Consistent with this finding, SLC13A5 Citrate Transporter Disorder patients have elevated citrate levels in the cerebrospinal fluid and plasma<sup>9</sup>.

SLC13A5 Citrate Transporter Disorder is a recently identified, rare disease with an estimated prevalence of 1,900 patients in the United States and Europe<sup>10</sup>. Creation of a unique code for SLC13A5 Citrate Transporter Disorder will enable progress towards patient identification, treatment, and research. It will also help to improve patient access to new treatments and to enable measuring outcomes in clinical trials, and thus to develop a better understanding of this disease.

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References

1. Yang, Q.-Z., Spelbrink, E. M., Nye, K. L., Hsu, E. R. & Porter, B. E. Epilepsy and EEG Phenotype of SLC13A5 Citrate Transporter Disorder. *Child Neurol. Open* 7, (2020).
2. Matricardi, S. et al. Neonatal developmental and epileptic encephalopathy due to autosomal recessive variants in SLC13A5 gene. *Epilepsia* 61, 2474–2485 (2020).
3. Thevenon, J. et al. Mutations in SLC13A5 Cause Autosomal-Recessive Epileptic Encephalopathy with Seizure Onset in the First Days of Life. *Am. J. Hum. Genet.* 95, 113–120 (2014).
4. Hardies, K. et al. Recessive mutations in SLC13A5 result in a loss of citrate transport and cause neonatal epilepsy, developmental delay and teeth hypoplasia. *Brain J. Neurol.* 138, 3238–3250 (2015).
5. Bergeron, M. J., Clémenton, B., Hediger, M. A. & Markovich, D. SLC13 family of Na<sup>+</sup>-coupled di- and tri-carboxylate/sulfate transporters. *Mol. Aspects Med.* 34, 299–312 (2013).
6. Klotz, J., Porter, B. E., Colas, C., Schlessinger, A. & Pajor, A. M. Mutations in the Na<sup>+</sup>/Citrate Cotransporter NaCT (SLC13A5) in Pediatric Patients with Epilepsy and Developmental Delay. *Mol. Med.* 22, 310–321 (2016).
7. Henke, C. et al. Disruption of the sodium-dependent citrate transporter SLC13A5 in mice causes alterations in brain citrate levels and neuronal network excitability in the hippocampus. *Neurobiol. Dis.* 143, 105018 (2020).
8. Selch, Stefan, Anja Chafai, Heinrich Sticht, Andreas L. Birkenfeld, Martin F. Fromm, and Jörg König. 2018. “Analysis of Naturally Occurring Mutations in the Human Uptake Transporter NaCT Important for Bone and Brain Development and Energy Metabolism.” *Scientific Reports* 8 (July): 11330. <https://doi.org/10.1038/s41598-018-29547-8>.
9. Bainbridge, M. N. et al. Analyses of SLC13A5-epilepsy patients reveal perturbations of TCA cycle. *Mol. Genet. Metab.* 121, 314–319 (2017).
10. Taysha Gene Therapies Announces New Data on Multiple Preclinical Programs and Upcoming R&D Day | Taysha Gene Therapies. <https://ir.tayshagtx.com/news-releases/news-release-details/taysha-gene-therapies-announces-new-data-multiple-preclinical/>.

**TABULAR MODIFICATIONS**

**Option #1**

Note: this option was created based on a modified approach from the original submission.

E74 Other disorders of carbohydrate metabolism

E74.8 Other specified disorders of carbohydrate metabolism

New

sub-subcategory

E74.82 Disorders of citrate metabolism

New code

E74.820 SLC13A5 Citrate Transporter Disorder

New code

E74.829 Other disorders of citrate metabolism

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**Option #2**

Note: this option was created by NCHS based on a generalized approach for a number of proposals. See also the Developmental Epileptic Encephalopathies topic.

Z15 Genetic susceptibility to disease

New  
subcategory

Z15.1 Genetic susceptibility to epilepsy and neurodevelopmental disorders

New  
sub-subcategory

Z15.14 Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to other transporter and solute carrier genes

New code

Z15.141 Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to SLC13A5 gene

## SLC6A1-Related Disorders

The SLC6A1 Connect patient advocacy organization has requested that a specific ICD-10-CM code be created for SLC6A1-related disorder.

Solute carrier family 6 member 1 disorders (SLC6A1-related disorders) have emerged as a common cause of developmental and epileptic encephalopathies<sup>1</sup> and epilepsy with myoclonic- atonic seizures (EMAS)<sup>2</sup> since initial descriptions in 2014<sup>3</sup> and 2015.<sup>4</sup> In the years since the disorder was first described, the rate of diagnosis has increased, with the prevalence of SLC6A1 disorders now estimated to be 1 in 38,000 births, and currently 333 cases are reported in published literature,<sup>1</sup> with 128 patients in the registry maintained by patient-advocacy organization.<sup>5</sup>

The SLC6A1 gene encodes the voltage dependent gamma-aminobutyric acid (GABA) transporter (GAT-1) which is responsible for GABA reuptake from synapses and is predominantly expressed in the nerve terminal of GABAergic interneurons in the brain.<sup>6</sup> SLC6A1-related disorders are caused by mutations in the SLC6A1 gene, which result in a loss of function for GAT-1.<sup>7</sup> There are more than 20 known variants identified to date<sup>8</sup> and the vast majority of these occur de novo and affect highly conserved nucleotide sequences.<sup>4</sup> Some variants impair GAT-1 localization to the cell membrane, others change the hydrophobicity of amino acid residues which de-stabilize GAT-1 protein structure, resulting in mis-folding and aggregation in the endoplasmic reticulum.<sup>8</sup> These impairments in GAT-1 protein folding and trafficking both ultimately lead to reduced GAT-1 function,<sup>9</sup> and diminished clearance of GABA from the synapse,<sup>8</sup> however the GAT-1 transporter is not involved in GABA metabolism. There is no clear correlation between mutational variants and clinical symptoms, nor symptom severity in SLC6A1-related disorders.<sup>7</sup>

The symptoms of SLC6A1-related disorder present in infancy or childhood, and include global developmental delay, sleep issues,<sup>10</sup> severe speech impairment,<sup>11</sup> stereotypies,<sup>3</sup> deficits in fine and gross motor skills, ataxia,<sup>7</sup> intellectual disability, autism and autistic traits, aggression,<sup>7</sup> and seizures.<sup>11</sup> Onset and type of epilepsy is also quite variable with atonic, absence and myoclonic seizures most commonly reported.<sup>11</sup> Disorders related to SLC6A1 variants are often associated with epilepsy with myoclonic atonic seizures (EMAS)<sup>7</sup> although individuals with EMAS often have multiple seizure semiologies.<sup>7</sup> It is important to note that although seizures are common in individuals with SLC6A1 variants, there are patients who do not have epilepsy.<sup>11</sup> Similarly, some, but not all, patients experience significant intellectual disability.<sup>7,11</sup> Further, symptom severity is variable among cases, with a range of seizure semiology and frequency, response to anti-seizure medication, variable language abilities, and variable levels of gross and fine motor abilities.<sup>7,12</sup> Furthermore, stalled neurodevelopment and/or regression in motor and cognitive function has been observed in children with SLC6A1-related disorders with the onset of seizures,<sup>11,13</sup> but some patients experience intellectual disability before seizure onset.<sup>11</sup> This variability in presentation belies the use of ICD-10-CM codes based on symptomatology for SLC6A1-related disorders.



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Ultimately, a unique ICD-10-CM code for SLC6A1-related disorder will expedite understanding of its epidemiology and enable better tracking and understanding of the broad spectrum of phenotypes, which will facilitate patient care for SLC6A1-related disorder.

References

1. López-Rivera JA, Pérez-Palma E, Symonds J, Lindy AS, McKnight DA, Leu C, et al. A catalogue of new incidence estimates of monogenic neurodevelopmental disorders caused by de novo variants. *Brain*. 2020;
2. Routier L, Verry F, Barcia G, Chemaly N, Desguerre I, Colleaux L, et al. Exome sequencing findings in 27 patients with myoclonic-atonic epilepsy: Is there a major genetic factor? *Clin Genet*. 2019 Sep;96(3):254–60.
3. Dikow N, Maas B, Karch S, Granzow M, Janssen JWG, Jauch A, et al. 3p25.3 microdeletion of GABA transporters SLC6A1 and SLC6A11 results in intellectual disability, epilepsy and stereotypic behavior. *Am J Med Genet A*. 2014 Dec;164(12):3061–8.
4. Carvill GL, McMahon JM, Schneider A, Zemel M, Myers CT, Saykally J, et al. Mutations in the GABA transporter SLC6A1 cause epilepsy with myoclonic-atonic seizures. *Am J Hum Genet*. 2015 May;96(5):808–15.
5. SLC6A1 Patient Registries | SLC6A1 Connect [Internet]. [cited 2021 May 28]. Available from: <https://slc6a1connect.org/registry/>
6. Scimemi A. Structure, function, and plasticity of GABA transporters. *Front Cell Neurosci*. 2014;8:161.
7. Johannesen KM, Gardella E, Linnankivi T, Courage C, de Saint Martin A, Lehesjoki A-E, et al. Defining the phenotypic spectrum of SLC6A1 mutations. *Epilepsia*. 2018 Feb;59(2):389–402.
8. Wang J, Poliquin S, Mermer F, Eissman J, Delpire E, Wang J, et al. Endoplasmic reticulum retention and degradation of a mutation in SLC6A1 associated with epilepsy and autism. *Mol Brain*. 2020 May 12;13(1):76.
9. Mermer F, Poliquin S, Rigsby K, Rastogi A, Shen W, Romero-Morales A, et al. Common molecular mechanisms of SLC6A1 variant-mediated neurodevelopmental disorders in astrocytes and neurons. *Brain J Neurol*. 2021 May 24;
10. Kahen A, Kavus H, Geltzeiler A, Kentros C, Taylor C, Brooks E, et al. Neurodevelopmental phenotypes associated with pathogenic variants in SLC6A1. *J Med Genet*. 2021 May 18;
11. Goodspeed K, Pérez-Palma E, Iqbal S, Cooper D, Scimemi A, Johannesen KM, et al. Current knowledge of SLC6A1-related neurodevelopmental disorders. *Brain Commun*. 2020;2(2):fcaa170.
12. Epi4K Consortium, Epilepsy Phenome/Genome Project. De novo mutations in epileptic encephalopathies. *Nature*. 2013 Sep;501(7466):217–21.
13. Islam MP, Herman GE, de los Reyes EC. Language Regression in an Atypical SLC6A1 Mutation. *Semin Pediatr Neurol*. 2018 Jul;26:25–7.

## TABULAR MODIFICATIONS

### Option #1

Note: this option was created based on a modified approach from the original submission. Also note that a concurrent proposal could create a code F84.81, SCN2A-related disorders.

F84 Pervasive developmental disorders

F84.8 Other pervasive developmental disorders

New code F84.82 SLC6A1-related disorder

New code F84.89 Other pervasive developmental disorders not elsewhere classified

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**Option #2**

Note: this option was created by NCHS based on a generalized approach for a number of proposals. See also the Developmental Epileptic Encephalopathies topic.

Z15 Genetic susceptibility to disease

New  
subcategory

Z15.1 Genetic susceptibility to epilepsy and neurodevelopmental disorders

New sub-  
subcategory

Z15.14 Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to other transporter and solute carrier genes

New code

Z15.140 Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of SLC6A1 gene  
Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of the gene for solute carrier family 6 (neurotransmitter transporter, GABA), member 1

Add

## Social Determinants of Health

This proposal was presented at the September 2022, ICD-10 Coordination and Maintenance Committee meeting and is being represented with modifications from the comments.

Health Insurance Coverage, Healthy People 2030 includes several objectives that relate to improving the proportion of people with some form of health or dental insurance or reducing the proportion of people under 65 who are uninsured. Earlier last year, the Centers for Medicare and Medicaid Services (CMS) unveiled an initiative to reduce the uninsured rate among children and increase Medicaid enrollment for parents and pregnant people.

Lack of insurance affects access to care and preventative services as well as an increase in mortality. The current code Z59.7, Insufficient social insurance and welfare support, this code is not specific enough. Social insurance is a broad-based term and welfare support can include different types of assistance programs outside of insurance coverage.

The Gravity Project community determined that a specific code to identify insufficient health insurance coverage is needed to properly identify this situation.

### TABULAR MODIFICATIONS

	Z56	Problems related to employment and unemployment Excludes2:occupational exposure to risk factors (Z57.-) problems related to housing and economic circumstances (Z59.-)
Add	Z59.7	Insufficient social insurance and welfare support Insufficient social and welfare insurance
New code	Z59.71	Insufficient health insurance coverage
Add		Inadequate social insurance
Add		Insufficient social insurance
Add		No health insurance coverage
New code	Z59.72	Insufficient welfare support
Add		Inadequate welfare support

## **STXBP1-related disorders**

The STXBP1 Foundation, a nonprofit patient advocacy organization, has requested that an ICD-10-CM code be created for STXBP1-related disorders.

Mutations in the syntaxin-binding protein 1 (STXBP1) gene, located on chromosome 9q34.11, cause a complex neurodevelopmental disease, known as STXBP1-related disorder or STXBP1-encephalopathy.<sup>1</sup> STXBP1 is a protein that is highly expressed in the brain and plays a critical role in the release of neurotransmitters.<sup>2,3</sup> Specifically, each of the three domains of the STXBP1 protein separately contribute to binding the soluble N-ethylmaleimide-sensitive factor attachment protein receptor (SNARE) complex, promoting synaptic vesicle priming, fusion, and neurotransmitter release into the synaptic cleft.<sup>2</sup> Defects in neurotransmitter release deleteriously affect how neurons communicate with one another and maintain proper synapses; the number and strength of these neuronal connections are the foundations of learning and memory in the brain.<sup>3</sup> Thus, disruption in this system through mutation in STXBP1 leads to the observed symptoms of the disorder.

There is a wide spectrum of phenotypic presentation from each type of mutation in the STXBP1 gene, including missense, nonsense, frameshift, deletion, and splice site mutations.<sup>2,4</sup> Neither the type of mutation, nor the location of the mutation, predict which symptoms will be present in patients. This lack of a clear link between genotype and phenotype results from studies of patients with STXBP1-related disorders and in disease models, suggests that haploinsufficiency is the primary disease mechanism underlying STXBP1-related disorders.<sup>4,5</sup> STXBP1-related disorders are characterized by epilepsy, intellectual disability, motor disturbances, and autism, but not all of these symptoms are shared by all patients.<sup>1,2,4</sup> Seizures typically develop in the first year of life, although onset has been reported in older children.<sup>1,4</sup> The range of symptoms can vary in severity (with some experiencing mild cognitive impairments, or no seizures), but in a vast majority of patients, intellectual disability is considered to be severe to profound, with global delays.<sup>1,4</sup>

In the decade since STXBP1-related disorders were first described in 2008,<sup>6</sup> another 282 patients have been reported in the literature.<sup>4</sup> A recent analysis by Dr. Dennis Lal at Cleveland Clinic predicted an updated estimate of incidence for STXBP1-related disorders to approximately 1 in 30,000 births,<sup>7</sup> suggesting that many more cases will be diagnosed in upcoming years. The new estimate of incidence includes both mild and severe forms of STXBP1-related disorder, and exceeds previous reports by 3 to 4-fold, likely due to prior underdiagnosis of the milder cases.<sup>1,4,7</sup> Many symptoms are shared with other neurodevelopmental syndromes, which are often misdiagnosed without sufficient genomic testing.<sup>7,8</sup>

Additionally, this updated incidence rate concurs with previous findings recognizing STXBP1 mutation as the 5th most common genetic source of epileptic encephalopathies and other neurodevelopmental diseases.<sup>9</sup>

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References

1. Stamberger H, Nikanorova M, Willemsen MH, Accorsi P, Angriman M, Baier H, et al. STXBP1 encephalopathy: A neurodevelopmental disorder including epilepsy. *Neurology*. 2016 Mar 8;86(10):954–62.
2. Suri M, Evers JMG, Laskowski RA, O'Brien S, Baker K, Clayton-Smith J, et al. Protein structure and phenotypic analysis of pathogenic and population missense variants in STXBP1. *Mol Genet Genomic Med*. 2017 Sep;5(5):495–507.
3. Verhage M. Synaptic Assembly of the Brain in the Absence of Neurotransmitter Secretion. *Science*. 2000 Feb 4;287(5454):864–9.
4. Abramov D, Guiberson NGL, Burré J. STXBP1 encephalopathies: Clinical spectrum, disease mechanisms, and therapeutic strategies. *J Neurochem*. 2021 Apr;157(2):165–78.
5. Kovačević J, Maroteaux G, Schut D, Loos M, Dubey M, Pitsch J, et al. Protein instability, haploinsufficiency, and cortical hyper-excitability underlie STXBP1 encephalopathy. *Brain*. 2018 May 1;141(5):1350–74.
6. Saitsu H, Kato M, Mizuguchi T, Hamada K, Osaka H, Tohyama J, et al. De novo mutations in the gene encoding STXBP1 (MUNC18-1) cause early infantile epileptic encephalopathy. *Nat Genet*. 2008 Jun;40(6):782–8.
7. López-Rivera JA, Pérez-Palma E, Symonds J, Lindy AS, McKnight DA, Leu C, et al. A catalogue of new incidence estimates of monogenic neurodevelopmental disorders caused by de novo variants. *Brain*. 2020;
8. Deciphering Developmental Disorders Study. Prevalence and architecture of de novo mutations in developmental disorders. *Nature*. 2017 Feb;542(7642):433–8.
9. Symonds JD, McTague A. Epilepsy and developmental disorders: Next generation sequencing in the clinic. *Eur J Paediatr Neurol*. 2020 Jan;24:15–23.

**TABULAR MODIFICATIONS**

**Option #1**

Note: this option was created based on a modified approach from the original submission.

G40 Epilepsy and recurrent seizures

G40.4 Other generalized epilepsy and epileptic syndromes

New code	G40.43	Syntaxin-binding protein 1-related disorders
Add		STXBP1-related disorders

**Option #2**

Note: this option was created by NCHS based on a generalized approach for a number of proposals. See also the Developmental Epileptic Encephalopathies topic.

Z15 Genetic susceptibility to disease

New subcategory	Z15.1	Genetic susceptibility to epilepsy and neurodevelopmental disorders
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New sub-subcategory	Z15.15	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to synapse related genes
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New code	Z15.150	Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of STXBP1 gene
Add		Genetic susceptibility to epilepsy and neurodevelopmental disorders, related to pathogenic variant of the gene for syntaxin-binding protein 1

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Add	D65	Disseminated intravascular coagulation [defibrination syndrome] COVID-19- associated diffuse or disseminated intravascular coagulopathy
Add		Code first, if applicable, associated condition
Add	D68	Other coagulation defects D68.8 Other specified coagulation defects COVID-19-associated coagulopathy
Add		Code first, if applicable, associated condition
Add	D68.69	Other thrombophilia COVID-19- associated hypercoagulability Hypercoagulable states NEC Secondary hypercoagulable state NOS
Add		Code first, if applicable, associated condition
Delete	G31	Other degenerative diseases of nervous system, not elsewhere classified <del>Use additional For codes G31.0-G31.83, G31.85-G31.9, use additional code, if applicable, to identify:</del>
Add		Use additional code, if applicable, for codes G31.0-G31.83, G31.85-G31.9, to identify:
Revise	G93	Other disorders of brain G93.3 Postviral and related fatigue syndromes Excludes1: chronic fatigue syndrome NOS (R53.82) neurasthenia (F48.8)
Revise	Q85	Phakomatoses, not elsewhere classified Q85.8 Other phakomatoses, not elsewhere classified Q85.81 PTEN <u>hamartoma</u> tumor syndrome PHTS
Delete		<del>PTEN hamartoma tumor syndrome</del> PTEN related Cowden syndrome

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R53 Malaise and fatigue  
R53.8 Other malaise and fatigue  
R53.82 Chronic fatigue, unspecified

Revise Excludes1: chronic fatigue syndrome (G93.32)  
myalgic encephalomyelitis (G93.32)  
other post infection and related fatigue  
syndromes (G93.39)

U07 Emergency use of U07  
U07.1 COVID-19

Add Use additional code, if applicable, for associated conditions such  
as:

Add COVID-19 associated coagulopathy (D68.8)  
Add disseminated intravascular coagulation (D65)  
Add hypercoagulable states (D68.69)  
Add thrombophilia (D68.69)

Delete Excludes2: coronavirus as the cause of diseases classified  
elsewhere (B97.2-)  
~~coronavirus infection, unspecified (B34.2)~~



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	Aneurysm (anastomotic) (artery) (cirroid) (diffuse) (false) (fusiform) (multiple) (saccular) I72.9
	- brain I67.1
Revise	- - mycotic <del>I33.0</del> <u>I67.1</u>
Add	- - - with endocarditis - see also Endocarditis
	Complication(s) (from) (of)
	- joint prosthesis, internal T84.9
	- - mechanical
Add	- - - displacement T84.02
	Disease, diseased -see also Syndrome
Add	-Hunt's
Add	- - disease or syndrome (herpetic geniculate ganglionitis) B02.21
Add	- - - dyssynergia cerebellaris myoclonica G11.19
Add	- - neuralgia B02.21
	Defect, defective Q89.9
	- coagulation (factor) -see also Deficiency, factor D68.9
	- - with
Add	- - - COVID-19-associated coagulopathy D68.8
Revise	Ramsay-Hunt disease or syndrome -see also Hunt's, disease B02.21
Add	Recrudescence
Add	- deficit
Add	- - cerebral infarction (see also Sequelae, infarction, cerebral) I69.398
Add	- - stroke (see also Sequelae, infarction, cerebral) I69.398
Add	- sequelae
Add	- - cerebral infarction (see also Sequelae, infarction, cerebral) I69.398
Add	- - stroke (see also Sequelae, infarction, cerebral) I69.398
	Syndrome -see also Disease
Add	-Hunt's
Add	- - disease or syndrome (herpetic geniculate ganglionitis) B02.21
Add	- - - dyssynergia cerebellaris myoclonica G11.19
Add	- - neuralgia B02.21
Revise	- Schwachman's <del>-see Syndrome, Schwachman's</del> D61.02

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	A04	Other bacterial intestinal infections
Add	A04.7	Enterocolitis due to Clostridium difficile Clostridioides difficile Foodborne intoxication by Clostridium difficile Pseudomembraneous colitis
	D05	Carcinoma in situ of breast
Add		Excludes2: Malignant neoplasm of breast (C50.-)
	E87	Other disorders of fluid, electrolyte and acid-base balance
	E87.0	Hyperosmolality and hypernatremia
Add		Excludes2: Diabetes with hyperosmolality (E08, E09, E11, E13 with ending .00 or .01)
	G04	Encephalitis, myelitis and encephalomyelitis
Revise		Excludes2: acute transverse myelitis ( <del>G37.3</del> ) ( <u>G37.3</u> )
	G92	Toxic encephalopathy
	G92.0	Immune effector cell-associated neurotoxicity syndrome
Delete		<del>Code also associated signs and symptoms, such as seizures and cerebral edema</del>
Revise		<u>Code also, if applicable, associated sign and symptoms, such as:</u> cerebral edema (G93.6) unspecified convulsions (R56.9)
	G93.4	Other and unspecified encephalopathy
Delete		<del>Excludes1: alcoholic encephalopathy (G31.2) encephalopathy in diseases classified elsewhere (G94) hypertensive encephalopathy (I67.4)</del>
Add		Excludes2: alcoholic encephalopathy (G31.2) encephalopathy in diseases classified elsewhere (G94) hypertensive encephalopathy (I67.4)

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H26	Other cataract	
	H26.2 Complicated cataract	
	H26.21 Cataract with neovascularization	
Revise		Code also, <u>if applicable</u> , associated condition, such as: chronic iridocyclitis (H20.1-)
I16	Hypertensive crisis	
	I16.1 Hypertensive emergency	
Add		Use additional code, if applicable, to identify specific organ dysfunction, such as:
Add		acute kidney injury (N17.-)
Add		acute myocardial infarction (I21.-)
Add		acute pulmonary edema (left and/or right ventricular failure) (J81.0, I50.-)
Add		aortic dissection (I71.0-)
Add		cerebral hemorrhage (I60.-, I61.-, I62.-)
Add		cerebral infarction (I63.-)
Add		eclampsia (O15.-)
Add		hypertensive encephalopathy (I67.4)
Add		seizure (R56.9)
K56	Paralytic ileus and intestinal obstruction without hernia	
	K56.4 Other impaction of intestine	
	K56.41 Fecal impaction	
Delete		Excludes1: constipation (K59.0-) <del>incomplete defecation (R15.0)</del>
Add		Excludes2: incomplete defecation (R15.0)
K59	Other functional intestinal disorders	
	K59.0 Constipation	
Delete		Excludes1: fecal impaction (K56.41) <del>incomplete defecation (R15.0)</del>
Add		Excludes2: incomplete defecation (R15.0)
P28	Other respiratory conditions originating in the perinatal period	
	P28.5 Respiratory failure of newborn	
Delete		<del>Excludes1: respiratory arrest of newborn (P28.81)</del>

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Delete		<del>respiratory distress of newborn (P22.0)</del>
Add	Excludes2:	respiratory arrest of newborn (P28.81)
Add		respiratory distress syndrome of newborn (P22.0)
R41		Other symptoms and signs involving cognitive functions and awareness
Revise	Excludes1:	dissociative [conversion] disorders (F44.-) delirium or acute confusional state with dementia F05 <del>mild cognitive impairment, so stated</del> <u>mild cognitive impairment of uncertain or unknown etiology (G31.84)</u>
S06.0		Concussion
S06.3		Focal traumatic brain injury
Delete	Excludes1:	<del>any condition classifiable to S06.4-S06.6</del>
S22		Fracture of rib(s), sternum and thoracic spine
Revise		Code first, <u>if applicable</u> , any associated <u>condition such as:</u> injury of intrathoracic organ (S27.-) spinal cord injury (S24.0-, S24.1-)
Add		traumatic hemothorax (S27.1-)
Add		traumatic pneumothorax (S27.0)
Add		traumatic hemopneumothorax (S27.2)
V92		Drowning and submersion due to accident on board watercraft, without accident to watercraft
V92.0		Drowning and submersion due to fall off watercraft
V92.07		Drowning and submersion due to fall off water-skis
Revise	Excludes1:	drowning and submersion due to falling off burning water-skis (V90.27) drowning and submersion due to falling off crushed water-skis (V90.37) hit by boat while water-skiing NOS ( <del>V94.X</del> ) <u>(V94.-)</u>

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- Add Bigorexia F45.22
- Blister (nonthermal)
- Revise - thermal -see Burn, by site, second degree, ~~by site~~
- Revise ~~Cesarean delivery, previous, affecting management of pregnancy O34.219~~
- Add - affecting management of pregnancy O34.219
- Revise - - classical (vertical) scar O34.212
- Revise - - isthmocele (non-pregnant state) N85.A
- Revise - - - maternal care for O34.22
- Revise - - low transverse scar O34.211
- Revise - - mid-transverse T incision O34.218
- Revise - - scar
- Revise - - - defect (non-pregnant state) N85.A
- Revise - - - - maternal care for O34.22
- Revise - - - specified type NEC O34.218
- Add - isthmocele (non-pregnant state) N85.A
- Add - scar
- Add - - defect (non-pregnant state) N85.A
- Revise Clostridium (C.) ~~perfringens, as cause of disease classified elsewhere B96.7 - see~~  
also Infection, Clostridium
- Add - perfringens, as cause of disease classified elsewhere B96.7
- Revise Colitis (acute) (catarrhal) (chronic) (noninfective) (hemorrhagic) (-see also  
Enteritis) K52.9
- Add - Clostridioides difficile
- Add - - not specified as recurrent A04.72
- Add - - recurrent A04.71
- toxic NEC K52.1
- Add - - due to Clostridioides difficile
- Add - - - not specified as recurrent A04.72
- Add - - - recurrent A04.71
- Complication(s) (from) (of)
- prosthetic device or implant T85.9
- Revise - - vascular -see Complications, cardiovascular device, graft or implant
- Compression
- Revise - eustachian tube -see Obstruction, eustachian tube, ~~cartilaginous~~ cartilagenous

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- Revised Degeneration, degenerative Oct 2024  
- disc disease -see Degeneration, intervertebral disc NOS, by site
- Diarrhea, diarrheal (disease) (infantile) (inflammatory) R19.7  
-- due to  
Add -- Clostridioides difficile  
Add --- not specified as recurrent A04.72  
Add --- recurrent A04.71
- infectious NOS A09  
Add - Clostridioides difficile  
Add -- not specified as recurrent A04.72  
Add -- recurrent A04.71
- toxic NEC K52.1  
Add -- due to Clostridioides difficile  
Add --- not specified as recurrent A04.72  
Add --- recurrent A04.71
- Revised Disorder (of) -see also Disease  
- myelodysplastic (~~-see also Syndrome, myelodysplasia~~ myelodysplastic) C94.6
- Revised Dwarfism -see also ~~Stature, short~~ Short, stature E34.328  
Revised - congenital -see also ~~Stature, short~~ Short, stature E34.328  
Revised - infantile -see also ~~Stature, short~~ Short, stature E34.328  
Revised - Laron-type -see also ~~Stature, short~~ Short, stature E34.321
- Dystrophy, dystrophia 2024  
- Leyden-Möbius -see also Dystrophy, muscular, limb-girdle, by type G71.039  
Delete ~~— meaning Limb-girdle muscular dystrophy, other specified type, see by type~~
- Endometriosis N80.9  
- intestine N80.50  
Revised - \_small N80.569  
Revised - - \_deep (multifocal) N80.562  
Revised - - \_superficial N80.561
- Entanglement  
- umbilical cord(s) O69.82  
Revised - - around neck (~~with compression~~) O69.81  
- - - with compression O69.1  
- - - without compression O69.81
- Add - - other, with compression O69.2  
Add - - other, without compression O69.82

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- Enterocolitis -see also Enteritis K52.9
- Add - Clostridioides difficile
  - Add - - not specified as recurrent A04.72
  - Add - - recurrent A04.71
- 
- necrotizing K55.30
  - Add - - due to Clostridioides difficile
  - Add - - - - not specified as recurrent A04.72
  - Add - - - - recurrent A04.71
- 
- Hypertension, hypertensive (accelerated) (benign) (essential) (idiopathic) (malignant) (systemic) I10
- complicating
  - - childbirth (labor) O16.4
  - Revise - - - pre-existing ~~O10.92~~ O10.02
  - - - - essential O10.02
- 
- - pregnancy O16.-
  - Revise - - - pre-existing ~~O10.91~~ O10.01-
- 
- Revise Inability to swallow—see Aphagia
- 
- Infection, infected, infective (opportunistic) B99.9
- Add - Clostridioides
  - Add - - difficile
  - Add - - - as cause of disease classified elsewhere B96.89
  - Add - - - foodborne (disease)
  - Add - - - - not specified as recurrent A04.72
  - Add - - - - recurrent A04.71
  - Add - - - necrotizing enterocolitis
  - Add - - - - not specified as recurrent A04.72
  - Add - - - - recurrent A04.71
  - Add - - - sepsis A41.4
- 
- Add Injury -see also specified injury type T14.90  
thalamic, thalamus - see Injury, intracranial, intracerebral hemorrhage, traumatic
- 
- Intoxication
- foodborne A05.9
  - - due to
  - Add - - - Clostridioides
  - Add - - - - difficile
  - Add - - - - - not specified as recurrent A04.72
  - Add - - - - - recurrent A04.71

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- - - Clostridium
- - - - botulinum A05.1
- Add - - - - difficile
- Add - - - - - not specified as recurrent A04.72
- Add - - - - - recurrent A04.71
  
- Add Isthmocele (non-pregnant state) N85.A
- Add - affecting management of current pregnancy – see Cesarean delivery, previous, affecting management of pregnancy
  
- Leak, leakage
- device, implant or graft -see also Complications, by site and type, mechanical
- Revise - - arterial graft NEC -see Complication, vascular, graft, mechanical, leakage T82.838
  
- Megacolon (acquired) (functional) (not Hirschsprung's disease) (in) K59.39
- toxic NEC K59.31
- Add - - due to - Clostridioides difficile
- Add - - - not specified as recurrent A04.72
- Add - - - recurrent A04.71(1142)
  
- Nutrition deficient or insufficient -see also Malnutrition E63.9
- Revise - sequelae -see ~~Sequelae~~ Sequelae, nutritional deficiency
  
- Osteoporosis (female) (male) M81.0
- age-related M81.0
- - with current pathologic fracture M80.00
- Revise - - - rib(s) -see ~~Osteoporosis, specified site~~ NEC M80.0A
  
- Revise Pneumonitis (acute) (primary) (-see also Pneumonia) J98.4
- Add - noninfectious J98.4
- Add - specified NEC J98.4



**EXTERNAL CAUSE OF INJURIES INDEX MODIFICATION PROPOSED ADDENDA**  
**All proposed effective October 1, 2024**

- Accident (to) X58
- Revise - motor scooter -see Accident, transport, ~~motorecyclist~~ motorcycle
- Revise - motorcycle NOS -see Accident, transport, ~~motorecyclist~~ motorcycle
- transport (involving injury to) V99
- Revise - - e-bicycle -see Accident, transport, electric (assisted) ~~bicylist~~ bicyclist
- Revise - - e-bike -see Accident, transport, electric (assisted) ~~bicylist~~ bicyclist
- Revise - - ~~motorecyclist~~ motorcycle V29.99

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<sup>i</sup> CDC National Center for Injury Prevention and Control and U.S. Consumer Product Safety Commission. National Electronic Injury Surveillance System ([NEISS](#)) All Injury Program, 2016. ICPSR [distributor], 2020-08-19.

<sup>ii</sup> Roman J & Cook P eds. [Improving Data Infrastructure to Reduce Firearms Violence](#). NORC at the University of Chicago, October 2021

<sup>iii</sup> Olufajo OA, Zeineddin A, Nonez H, et al. Trends in firearm injuries among children and teenagers in the United States. *J Surg Res.* 2020 Jan;245:529-536. doi: 10.1016/j.jss.2019.07.056. (Includes data on all age groups.)

<sup>iv</sup> WRISS data from Beth Hume, Injury Surveillance Program, Massachusetts Department of Public Health.