



October 2020 Annual ICD-10 Update Part 1: ICD-10-CM

| Total 2020 Codes | 2021 Deletions | 2021 Additions | Total 2021 Codes | Code Description Revisions for 2021 |
|------------------|----------------|----------------|------------------|-------------------------------------|
| 72,198 | 58 | 490 | 72,616 | 47 |

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Code Changes

- Sub-Category D84.8 Other Immunodeficiencies was expanded
 - D84.81 Immunodeficiency due to conditions classified elsewhere (*Manifestation Code- cannot be PDX*)
 - D84.821 Immunodeficiency due to drugs
 - D84.822 Immunodeficiency due to external causes
 - D84.89 Other immunodeficiencies

CC Status

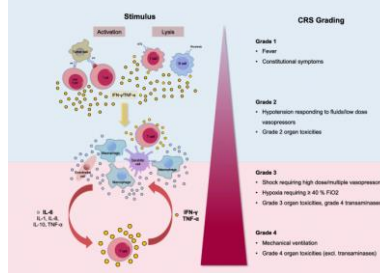
D84.81 Immunodeficiency due to conditions classified elsewhere [X]
 Code first underlying condition, such as:
 chromosomal abnormalities (Q90-Q99)
 diabetes mellitus (E08-E13)
 malignant neoplasms (C00-C96)

[X] certain disorders involving the immune mechanism (D80-D83, D84.0, D84.1, D84.9)
 human immunodeficiency virus (HIV) disease (B20)

D84.821 Immunodeficiency due to drugs [X]
 Immunodeficiency due to (current or past) medication
 Use additional code for adverse effect if applicable, to identify adverse effect of drug (T36-T50 with fifth or six character 5)
 Use additional code, if applicable, for associated long term (current) drug therapy drug or medication such as: long term (current) drug therapy systemic steroids (Z79.52) other long term (current) drug therapy (Z79.899)

D84.822 Immunodeficiency due to external causes [X]
 Code also, if applicable, radiological procedure and radiotherapy (Y84.2)
 Use additional code for external cause such as:
 exposure to ionizing radiation (W88)

- New Codes under Sub-Category D89.8 Other Specified Disorders Involving the Immune Mechanism NEC have been created for Cytokine Release Syndrome
 - D89.831 Cytokine release syndrome, grade 1
 - D89.832 Cytokine release syndrome, grade 2
 - D89.833 Cytokine release syndrome, grade 3 **CC**
 - D89.834 Cytokine release syndrome, grade 4 **CC**
 - D89.835 Cytokine release syndrome, grade 5 **CC**
 - D89.839 Cytokine release syndrome, grade unspecified
- *These codes are unacceptable as PDX on inpatient encounters



<https://jtc.biomedcentral.com/articles/10.1186/s40425-018-0343-9>

- In code block F10-F19 Mental and Behavioral Disorders due to Psychoactive Substance Use, new codes were added for selected substance use and abuse with withdrawal.
- Substances for abuse with withdrawal added were: Alcohol, Opioids, Cannabis, Sedatives/Hypnotics, Cocaine, Stimulants, and Other Psychoactive Substances.
- Substances for use with withdrawal that are new are: Alcohol and Cocaine
- All new codes are CCs except for Cannabis use with withdrawal F12.13

| Code | Description |
|--------|--|
| F10130 | Alcohol abuse with withdrawal, uncomplicated |
| F10131 | Alcohol abuse with withdrawal delirium |
| F10132 | Alcohol abuse with withdrawal with perceptual disturbance |
| F10139 | Alcohol abuse with withdrawal, unspecified |
| F10930 | Alcohol use, unspecified with withdrawal, uncomplicated |
| F10931 | Alcohol use, unspecified with withdrawal delirium |
| F10932 | Alcohol use, unspecified with withdrawal with perceptual disturbance |
| F10939 | Alcohol use, unspecified with withdrawal, unspecified |
| F1113 | Opioid abuse with withdrawal |
| F1213 | Cannabis abuse with withdrawal |
| F13130 | Sedative, hypnotic or anxiolytic abuse with withdrawal, uncomplicated |
| F13131 | Sedative, hypnotic or anxiolytic abuse with withdrawal delirium |
| F13132 | Sedative, hypnotic or anxiolytic abuse with withdrawal with perceptual disturbance |
| F13139 | Sedative, hypnotic or anxiolytic abuse with withdrawal, unspecified |
| F1413 | Cocaine abuse, unspecified with withdrawal |
| F1493 | Cocaine use, unspecified with withdrawal |
| F1513 | Other stimulant abuse with withdrawal |
| F19130 | Other psychoactive substance abuse with withdrawal, uncomplicated |
| F19131 | Other psychoactive substance abuse with withdrawal delirium |
| F19132 | Other psychoactive substance abuse with withdrawal with perceptual disturbance |
| F19139 | Other psychoactive substance abuse with withdrawal, unspecified |

- Clinically, it was originally thought that a withdrawal syndrome only developed in individuals with a diagnosis of substance dependence; however, substance withdrawal can occur in clinical situations involving individuals who use substances regularly and then suddenly stop using them, but who do not have a diagnosis of substance dependence.
- Such situations include:
 1. Individuals taking prescribed medication daily exactly as directed who are physiologically addicted to the substance but who do not have the behavioral elements required for a diagnosis of substance dependence
 2. Individuals who abuse substances regularly (which qualifies for a diagnosis of substance abuse) but lack the loss of control required for a diagnosis of substance dependence.

- New codes for Intracranial Hypotension have been added
 - G96.810 Intracranial hypotension, unspecified
 - G96.811 Intracranial hypotension, spontaneous
 - G96.819 Other intracranial hypotension
 - G97.83 Intracranial hypotension following lumbar cerebrospinal fluid shunting **CC**
 - G97.84 Intracranial hypotension following other procedure **CC**

- Sub-Category G96.0 CSF Leak has been expanded.
 - G96.00 Cerebrospinal fluid leak, unspecified
 - G96.01 Cranial cerebrospinal fluid leak, spontaneous
 - G96.02 Spinal cerebrospinal fluid leak, spontaneous
 - G96.08 Other cranial cerebrospinal fluid leak
 - G96.09 Other spinal cerebrospinal fluid leak

CC Status

G96.0 Cerebrospinal fluid leak
 ▶ Code also if applicable: ◀
 ▶ Intraaxial hypotension G96.81 ◀
DEFINITION cerebrospinal fluid leak from spinal puncture (G97.8)
 AHA, 2018, Q2, 13
 DEF: Cerebrospinal fluid discharging from the nose or the ear caused by a fracture of the frontal bone with tearing of dura mater and arachnoid. It is characterized by watery drainage usually from only one side of the nose or one ear, headache, vision changes, and hearing loss.

G96.00 Cerebrospinal fluid leak, unspecified
 Code also if applicable:
 Head injury (S80- to S89-.)

G96.01 Cranial cerebrospinal fluid leak, spontaneous
 Otorrhea due to spontaneous cerebrospinal fluid leak
 Rhinorrhea due to spontaneous cerebrospinal fluid leak
 Spontaneous cerebrospinal fluid leak from skull base

G96.02 Spinal cerebrospinal fluid leak, spontaneous
 Spontaneous cerebrospinal fluid leak from spine

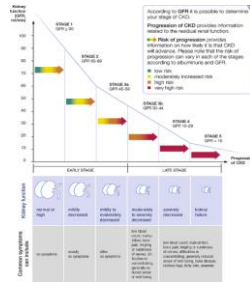
G96.08 Other cranial cerebrospinal fluid leak
 Postoperative cranial cerebrospinal fluid leak
 Traumatic cranial cerebrospinal fluid leak
 Code also if applicable:
 head injury (S80- to S89-.)

G96.09 Other spinal cerebrospinal fluid leak
 Other spinal CSF leak
 Postoperative spinal cerebrospinal fluid leak
 Traumatic spinal cerebrospinal fluid leak
 Code also if applicable:
 head injury (S80- to S89-.)

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- New Combination Codes for Esophagitis with and without Bleeding have been created
 - K20.80 Other esophagitis without bleeding
 - K20.81 Other esophagitis with bleeding **MCC**
 - K20.90 Esophagitis, unspecified without bleeding
 - K20.91 Esophagitis, unspecified with bleeding **MCC**
 - K21.00 Gastro-esophageal reflux disease with esophagitis, without bleeding
 - K21.01 Gastro-esophageal reflux disease with esophagitis, with bleeding **MCC**

- Sub-Category N18.3 CKD Stage 3 has been expanded
 - N18.30 Chronic kidney disease, stage 3 unspecified
 - N18.31 Chronic kidney disease, stage 3a
 - N18.32 Chronic kidney disease, stage 3b



<https://www.nephrocare.com/patients-home/kidney-kidney-disease/my-disease-stage.html>

- Sub-Category O34.2 Maternal Care due to Uterine Scar from Previous Surgery has been expanded
 - O34.218 Maternal care for other type scar from previous cesarean delivery
 - The American Hospital Association's (AHA) Editorial Advisory Board (EAB) for coding clinic has received inquiries on how to code a mid-transverse T incision.
 - O34.22 Maternal care for cesarean scar defect (isthmocele)

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- Sub-Category R74.0 Nonspecific Elevation of Levels of Transaminase and Lactic Acid Dehydrogenase (LDH) has been expanded
 - R74.01 Elevation of levels of liver transaminase levels
 - R74.02 Elevation of levels of lactic acid dehydrogenase [LDH]

- Category S20, Superficial injury of thorax, expanded and codes created to identify the middle and bilateral walls of the front thorax
 - Contusion (S20.2-)
 - Unspecified superficial injury (S20.30-)
 - Abrasion (S20.31-)
 - Blister (S20.32-)
 - External constriction (S20.34-)
 - Superficial foreign body (S20.35-)
 - Insect bite (S20.36-)
 - Other superficial bite (S20.37-)
- Results in 54 new codes

- The anterior thorax is one of the most common locations of traumatic injury. Blunt, high energy injuries as seen with vehicle collisions are responsible for upward of 25% of trauma related deaths.
- Unlike penetrating thorax trauma which may be to one (or both sides) of the anterior or posterior thorax, blunt trauma is usually to the mid-chest region.
- Anatomically, the sternum and the underlying heart are in the center of the chest as opposed to one side or the other.

- Compared to motorcycle riders, e-scooter riders stand rather than sit, travel at lower speeds (15-20 MPH versus >30 MPH), and may operate their vehicles in a variety of spaces (sidewalks, bike lanes, streets versus in-street only).
- Additionally, at under 50 pounds, e-scooters are considered “ultralight” roadway devices and are not regulated as motorcycles by most transportation departments.



YES



NO

- Sub-Category Z03.82- Encounter for Observation for Suspected Foreign Body Ruled Out has been added
 - Z03.821 Encounter for observation for suspected ingested foreign body ruled out
 - Z03.822 Encounter for observation for suspected aspirated (inhaled) foreign body ruled out
 - Z03.823 Encounter for observation for suspected inserted (injected) foreign body ruled out

- Codes in subcategory T86.84, Complications of corneal transplant, have been expanded to identify laterality (right eye, left eye, bilateral and unspecified eye)
 - Results in 20 new codes.

CC Status

- Under Category D57 Sickle Cell Disorders
 - New codes have been added for cerebral vascular involvement and for other specified complications
 - New codes have been added to further specify the Sickle Cell Thalassemia as:
 - Thalassemia beta zero (D57.42-D57.43-) MCC for those "with Crisis"
 - Thalassemia beta plus (D57.44-D57.45-) MCC for those "with Crisis"

- There are two distinct types of sickle cell-thalassemia, sickle cell-thalassemia beta zero (HbS-β0) and sickle cell-thalassemia beta plus (HbS-β+). They are clinically very different.
- HbS-β0 is clinically similar to sickle cell-SS disease in terms of degree of frequency and severity of acute and chronic complications. The risk of stroke is similar. They both may be managed long term with medications such as hydroxyurea.
- On the other hand, HbS-β+ is significantly less severe with little or no anemia. The spectrum and severity of complications is less. It also carries a relative lower risk of stroke. Only a relatively few patients with this condition will require treatment with hydroxyurea.

- Sub-Category D59.1 was expanded to specify the type of autoimmune hemolytic anemia:
 - D59.10 Autoimmune hemolytic anemia, unspecified
 - D59.11 Warm autoimmune hemolytic anemia
 - D59.12 Cold autoimmune hemolytic anemia
 - D59.13 Mixed autoimmune hemolytic anemia
 - D59.19 Other autoimmune hemolytic anemia

CC Status

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

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- M80.0A Age-related osteoporosis with current pathological fracture, other site
- M80.8A Other osteoporosis with current pathological fracture, other site

CC Status

- Category N61 Inflammatory Disorders of Breast has been further expanded
 - N61.20 Granulomatous mastitis, unspecified breast
 - N61.21 Granulomatous mastitis, right breast
 - N61.22 Granulomatous mastitis, left breast
 - N61.23 Granulomatous mastitis, bilateral breast

CC/MCC Changes

| Code | Description |
|---------|---|
| A84.81 | Powassan virus disease |
| A84.89 | Other tick-borne viral encephalitis |
| D57.03 | Hb-S5 disease with cerebral vascular involvement |
| D57.09 | Hb-S5 disease with crisis with other specified complication |
| D57.213 | Sickle-cell/Hb-C disease with cerebral vascular involvement |
| D57.218 | Sickle-cell/Hb-C disease with crisis with other specified complication |
| D57.413 | Sickle-cell thalassemia, unspecified, with cerebral vascular involvement |
| D57.418 | Sickle-cell thalassemia, unspecified, with crisis with other specified complication |
| D57.431 | Sickle-cell thalassemia beta zero with acute chest syndrome |
| D57.432 | Sickle-cell thalassemia beta zero with splenic sequestration |
| D57.433 | Sickle-cell thalassemia beta zero with cerebral vascular involvement |
| D57.438 | Sickle-cell thalassemia beta zero with crisis with other specified complication |
| D57.439 | Sickle-cell thalassemia beta zero with crisis, unspecified |
| D57.451 | Sickle-cell thalassemia beta plus with acute chest syndrome |
| D57.452 | Sickle-cell thalassemia beta plus with splenic sequestration |
| D57.453 | Sickle-cell thalassemia beta plus with cerebral vascular involvement |
| D57.458 | Sickle-cell thalassemia beta plus with crisis with other specified complication |
| D57.459 | Sickle-cell thalassemia beta plus with crisis, unspecified |
| D57.813 | Other sickle-cell disorders with cerebral vascular involvement |
| D57.818 | Other sickle-cell disorders with crisis with other specified complication |

| Code | Description |
|---------|---|
| K20.81 | Other esophagitis with bleeding |
| K20.91 | Esophagitis, unspecified with bleeding |
| K21.01 | Gastro-esophageal reflux disease with esophagitis, with bleeding |
| N00.A | Acute nephritic syndrome with C3 glomerulonephritis |
| N01.A | Rapidly progressive nephritic syndrome with C3 glomerulonephritis |
| P91.821 | Neonatal cerebral infarction, right side of brain |
| P91.822 | Neonatal cerebral infarction, left side of brain |
| P91.823 | Neonatal cerebral infarction, bilateral |
| P91.829 | Neonatal cerebral infarction, unspecified side |
| U07.1 | COVID-19 |



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| Code | Description |
|---------|---|
| B60.00 | Babesiosis, unspecified |
| B60.01 | Babesiosis due to Babesia microti |
| B60.02 | Babesiosis due to Babesia duncani |
| B60.03 | Babesiosis due to Babesia divergens |
| B60.09 | Other babesiosis |
| D59.10 | Autoimmune hemolytic anemia, unspecified |
| D59.11 | Warm autoimmune hemolytic anemia |
| D59.12 | Cold autoimmune hemolytic anemia |
| D59.13 | Mixed type autoimmune hemolytic anemia |
| D59.19 | Other autoimmune hemolytic anemia |
| D84.81 | Immunodeficiency due to conditions classified elsewhere |
| D84.82 | Immunodeficiency due to drugs |
| D84.822 | Immunodeficiency due to external causes |
| D84.89 | Other immunodeficiencies |
| D89.833 | Cytokine release syndrome, grade 3 |
| D89.834 | Cytokine release syndrome, grade 4 |
| D89.835 | Cytokine release syndrome, grade 5 |

| Code | Description |
|---------|--|
| E70.81 | Aromatic L-amino acid decarboxylase deficiency |
| E70.89 | Other disorders of aromatic amino-acid metabolism |
| E74.810 | Glucose transporter protein type 1 deficiency |
| E74.818 | Other disorders of glucose transport |
| E74.819 | Disorders of glucose transport, unspecified |
| E74.89 | Other specified disorders of carbohydrate metabolism |
| F10.130 | Alcohol abuse with withdrawal, uncomplicated |
| F10.131 | Alcohol abuse with withdrawal delirium |
| F10.132 | Alcohol abuse with withdrawal with perceptual disturbance |
| F10.139 | Alcohol abuse with withdrawal, unspecified |
| F10.930 | Alcohol use, unspecified with withdrawal, uncomplicated |
| F10.931 | Alcohol use, unspecified with withdrawal delirium |
| F10.932 | Alcohol use, unspecified with withdrawal with perceptual disturbance |
| F10.939 | Alcohol use, unspecified with withdrawal, with unspecified |
| F11.13 | Opioid abuse with withdrawal |

| Code | Description |
|---------|--|
| F13.130 | Sedative, hypnotic or anxiolytic abuse with withdrawal, uncomplicated |
| F13.131 | Sedative, hypnotic or anxiolytic abuse with withdrawal delirium |
| F13.132 | Sedative, hypnotic or anxiolytic abuse with withdrawal with perceptual disturbance |
| F13.139 | Sedative, hypnotic or anxiolytic abuse with withdrawal, unspecified |
| F14.13 | Cocaine abuse, unspecified with withdrawal |
| F14.93 | Cocaine use, unspecified with withdrawal |
| F15.13 | Other stimulant abuse with withdrawal |
| F19.130 | Other psychoactive substance abuse with withdrawal, uncomplicated |
| F19.131 | Other psychoactive substance abuse with withdrawal delirium |
| F19.132 | Other psychoactive substance abuse with withdrawal with perceptual disturbance |
| F19.139 | Other psychoactive substance abuse with withdrawal, unspecified |
| G11.10 | Early-onset cerebellar ataxia, unspecified |
| G11.11 | Friedreich ataxia |
| G11.19 | Other early-onset cerebellar ataxia |
| G40.833 | Dravet syndrome, intractable, with status epilepticus |
| G40.834 | Dravet syndrome, intractable, without status epilepticus |

| Code | Description |
|---------|--|
| G71.20 | Congenital myopathy, unspecified |
| G71.21 | Nemaline myopathy |
| G71.220 | X-linked myotubular myopathy |
| G71.228 | Other centronuclear myopathy |
| G71.29 | Other congenital myopathy |
| G96.00 | Cerebrospinal fluid leak, unspecified |
| G96.01 | Cranial cerebrospinal fluid leak, spontaneous |
| G96.02 | Spinal cerebrospinal fluid leak, spontaneous |
| G96.08 | Other cranial cerebrospinal fluid leak |
| G96.09 | Other spinal cerebrospinal fluid leak |
| G97.83 | Intracranial hypotension following lumbar cerebrospinal fluid shunting |
| G97.84 | Intracranial hypotension following other procedure |
| J82.81 | Chronic eosinophilic pneumonia |
| J82.82 | Acute eosinophilic pneumonia |
| J82.83 | Eosinophilic asthma |
| J82.89 | Other pulmonary eosinophilia, not elsewhere classified |

ICD-10-CM Official Coding Guideline Changes

Official Coding Guideline I.B.14

- *Patient self-reported documentation may also be used to assign codes for social determinants of health, as long as the patient self-reported information is signed-off by and incorporated into the health record by either a clinician or provider.*

Official Coding Guidelines I.C.1.g

- 1) COVID-19 infection (infection due to SARS-CoV-2)

(a) Code Only Confirmed Cases

Code only a confirmed diagnosis of the 2019 novel coronavirus disease (COVID-19) as documented by the provider or documentation of a positive COVID19 test result. For a confirmed diagnosis, assign code U07.1, COVID-19.

This is an exception to the hospital inpatient guideline Section II, H. In this context, "confirmation" does not require documentation of a positive test result for COVID-19; the provider's documentation that the individual has COVID-19 is sufficient.

Official Coding Guidelines I.C.1.g

1) COVID-19 infection (infection due to SARS-CoV-2)

(a) Code Only Confirmed Cases

If the provider documents "suspected," "possible," "probable," or "inconclusive" COVID-19, do not assign code U07.1. Instead, code the signs and symptoms reported. See guideline I.C.1.g.1.g.

Official Coding Guidelines I.C.1.g

1) COVID-19 infection (infection due to SARS-CoV-2)

(b) Sequencing of Codes

When COVID-19 meets the definition of principal diagnosis, code U07.1, COVID-19, should be sequenced first, followed by the appropriate codes for associated manifestations, **except when another guideline requires that certain codes be sequenced first, such as obstetrics, sepsis, or transplant complications.**

Official Coding Guidelines I.C.1.g

1) COVID-19 infection (infection due to SARS-CoV-2)

(b) Sequencing of Codes

- o For a COVID-19 infection that progresses to sepsis, see Section I.C.1.d. Sepsis, Severe Sepsis, and Septic Shock
- o See Section I.C.15.s. for COVID-19 infection in pregnancy, childbirth, and the puerperium
- o See Section I.C.16.h. for COVID-19 infection in newborn For a COVID-19 infection in a lung transplant patient
- o See Section I.C.19.g.3.a. Transplant complications other than kidney.

Official Coding Guidelines I.C.1.g

- 1) COVID-19 infection (infection due to SARS-CoV-2)
 - (c) Acute Respiratory Manifestations of COVID-19
 - o When the reason for the encounter/admission is a respiratory manifestation of COVID-19, assign code U07.1, COVID-19, as the principal/first-listed diagnosis and assign code(s) for the respiratory manifestation(s) as additional diagnoses.
 - o The following conditions are examples of common respiratory manifestations of COVID-19.

Official Coding Guidelines I.C.1.g

- 1) COVID-19 infection (infection due to SARS-CoV-2)
 - (c) Acute Respiratory Manifestations of COVID-19
 - (i) Pneumonia
 - o For a patient with pneumonia confirmed as due to COVID-19, assign codes U07.1, COVID-19, and J12.89, Other viral pneumonia.

Official Coding Guidelines I.C.1.g

- 1) COVID-19 infection (infection due to SARS-CoV-2)
 - (c) Acute Respiratory Manifestations of COVID-19
 - (ii) Acute Bronchitis
 - o For a patient with acute bronchitis confirmed as due to COVID-19, assign codes U07.1, and J20.8, Acute bronchitis due to other specified organisms.
 - o Bronchitis not otherwise specified (NOS) due to COVID-19 should be coded using code U07.1 and J40, Bronchitis, not specified as acute or chronic.

Official Coding Guidelines I.C.1.g

- 1) COVID-19 infection (infection due to SARS-CoV-2)
 - (c) Acute Respiratory Manifestations of COVID-19
 - (iii) Lower Respiratory Infection
 - o If the COVID-19 is documented as being associated with a lower respiratory infection, not otherwise specified (NOS), or an acute respiratory infection, NOS, codes U07.1 and J22, Unspecified acute lower respiratory infection, should be assigned.
 - o If the COVID-19 is documented as being associated with a respiratory infection, NOS, codes U07.1 and J98.8, Other specified respiratory disorders, should be assigned.

Official Coding Guidelines I.C.1.g

- 1) COVID-19 infection (infection due to SARS-CoV-2)
 - (c) Acute Respiratory Manifestations of COVID-19
 - (iv) Acute Respiratory Distress Syndrome
 - For acute respiratory distress syndrome (ARDS) due to COVID-19, assign codes U07.1, and J80, Acute respiratory distress syndrome.

Official Coding Guidelines I.C.1.g

- 1) COVID-19 infection (infection due to SARS-CoV-2)
 - (c) Acute Respiratory Manifestations of COVID-19
 - (v) Acute Respiratory Failure
 - For acute respiratory failure due to COVID-19, assign code U07.1, and code J96.0-, Acute respiratory failure.

Official Coding Guidelines I.C.1.g

1) COVID-19 infection (infection due to SARS-CoV-2)

(d) Non-Respiratory Manifestations of COVID-19

When the reason for the encounter/admission is a non-respiratory manifestation (e.g., viral enteritis) of COVID-19, assign code U07.1, COVID-19, as the principal/first-listed diagnosis and assign code(s) for the manifestation(s) as additional diagnoses.

Official Coding Guidelines I.C.1.g

1) COVID-19 infection (infection due to SARS-CoV-2)

(e) Exposure to COVID-19

- o For asymptomatic individuals with actual or suspected exposure to COVID-19, assign code Z20.828, Contact with and (suspected) exposure to other viral communicable diseases.
- o For symptomatic individuals with actual or suspected exposure to COVID-19 and the infection has been ruled out, or test results are inconclusive or unknown, assign code Z20.828, Contact with and (suspected) exposure to other viral communicable diseases. See guideline I.C.21.c.1, Contact/Exposure, for additional guidance regarding the use of category Z20 codes.
- o If COVID-19 is confirmed, see guideline I.C.1.g.1.a.

Official Coding Guidelines I.C.1.g

1) COVID-19 infection (infection due to SARS-CoV-2)

(f) Screening for COVID-19

- o During the COVID-19 pandemic, a screening code is generally not appropriate. For encounters for COVID-19 testing, including preoperative testing, code as exposure to COVID-19 (guideline I.C.1.g.1.e).
- o Coding guidance will be updated as new information concerning any changes in the pandemic status becomes available.

Official Coding Guidelines I.C.1.g

1) COVID-19 infection (infection due to SARS-CoV-2)

(g) Signs and Symptoms without Definitive Diagnosis of COVID-19

For patients presenting with any signs/symptoms associated with COVID-19 (such as fever, etc.) but a definitive diagnosis has not been established, assign the appropriate code(s) for each of the presenting signs and symptoms such as:

- R05 Cough
- R06.02 Shortness of breath
- R50.9 Fever, unspecified

Official Coding Guidelines I.C.1.g

1) COVID-19 infection (infection due to SARS-CoV-2)

(g) Signs and Symptoms without Definitive Diagnosis of COVID-19

If a patient with signs/symptoms associated with COVID-19 also has an actual or suspected contact with or exposure to COVID-19, assign Z20.828, Contact with and (suspected) exposure to other viral communicable diseases, as an additional code.

Official Coding Guidelines I.C.1.g

1) COVID-19 infection (infection due to SARS-CoV-2)

(h) Asymptomatic Individuals Who Test Positive for COVID-19

- o For asymptomatic individuals who test positive for COVID-19, see guideline I.C.1.g.1.a.
- o Although the individual is asymptomatic, the individual has tested positive and is considered to have the COVID-19 infection.

Official Coding Guidelines I.C.1.g

1) COVID-19 infection (infection due to SARS-CoV-2)

- (i) Personal History of COVID-19

For patients with a history of COVID-19, assign code Z86.19, Personal history of other infectious and parasitic diseases.

Official Coding Guidelines I.C.1.g

1) COVID-19 infection (infection due to SARS-CoV-2)

- (j) Follow-up Visits After COVID-19 Infection Has Resolved

For individuals who previously had COVID-19 and are being seen for follow-up evaluation, and COVID-19 test results are negative, assign codes Z09, Encounter for follow-up examination after completed treatment for conditions other than malignant neoplasm, and Z86.19, Personal history of other infectious and parasitic diseases.

Official Coding Guidelines I.C.1.g

1) COVID-19 infection (infection due to SARS-CoV-2)

- (k) Encounter for Antibody Testing
 - o For an encounter for antibody testing that is not being performed to confirm a current COVID-19 infection, nor is a follow-up test after resolution of COVID-19, assign Z01.84, Encounter for antibody response examination.
 - o Follow the applicable guidelines above if the individual is being tested to confirm a current COVID-19 infection.
 - o For follow-up testing after a COVID-19 infection, see guideline I.C.1.g.1.j.

Official Coding Guidelines I.C.15.s

- During pregnancy, childbirth or the puerperium, when COVID-19 is the reason for admission/encounter, code O98.5-, Other viral diseases complicating pregnancy, childbirth and the puerperium, should be sequenced as the principal/first-listed diagnosis, and code U07.1, COVID-19, and the appropriate codes for associated manifestation(s) should be assigned as additional diagnoses. Codes from Chapter 15 always take sequencing priority.

Official Coding Guidelines I.C.15.s

- If the reason for admission/encounter is unrelated to COVID-19 but the patient tests positive for COVID-19 during the admission/encounter, the appropriate code for the reason for admission/encounter should be sequenced as the principal/first-listed diagnosis, and codes O98.5- and U07.1, as well as the appropriate codes for associated COVID-19 manifestations, should be assigned as additional diagnoses.

Official Coding Guidelines I.C.16.h

- For a newborn that tests positive for COVID-19, assign code U07.1, COVID-19, and the appropriate codes for associated manifestation(s) in neonates/newborns in the absence of documentation indicating a specific type of transmission.
- For a newborn that tests positive for COVID-19 and the provider documents the condition was contracted in utero or during the birth process, assign codes P35.8, Other congenital viral diseases, and U07.1, COVID-19.
- When coding the birth episode in a newborn record, the appropriate code from category Z38, Liveborn infants according to place of birth and type of delivery, should be assigned as the principal diagnosis.



DIABETES MELLITUS AND THE USE OF INSULIN, ORAL HYPOGLYCEMICS, AND INJECTABLE NON-INSULIN DRUGS

Official Coding Guidelines I.C.4.a.(3) & I.C.4.a.(6).(a)

- If the patient is treated with both insulin and an injectable non-insulin antidiabetic drug, assign codes Z79.4, Long-term (current) use of insulin, and Z79.899, Other long term (current) drug therapy.
- If the patient is treated with both oral hypoglycemic drugs and an injectable non-insulin antidiabetic drug, assign codes Z79.84, Long-term (current) use of oral hypoglycemic drugs, and Z79.899, Other long-term (current) drug therapy.



PSYCHOACTIVE SUBSTANCE USE, UNSPECIFIED

Official Coding Guidelines I.C.5.b.(3)

- As with all other unspecified diagnoses, the codes for unspecified psychoactive substance use (F10.9-, F11.9-, F12.9-, F13.9-, F14.9-, F15.9-, F16.9-, F18.9-, F19.9-) should only be assigned based on provider documentation and when they meet the definition of a reportable diagnosis (see Section III, Reporting Additional Diagnoses).
- These codes are to be used only when the psychoactive substance use is associated with a physical disorder included in chapter 5 (such as sexual dysfunction and sleep disorder), or a mental or behavioral disorder, and such a relationship is documented by the provider



HYPERTENSIVE HEART AND CHRONIC KIDNEY DISEASE

Official Coding Guidelines I.C.9.a.(3)

- For patients with both acute renal failure and chronic kidney disease, **the acute renal failure should also be coded. Sequence according to the circumstances of the admission/encounter**

Official Coding Guidelines I.C.10.e

- For patients presenting with condition(s) related to vaping, assign code U07.0, Vaping-related disorder, as the principal diagnosis.
- For lung injury due to vaping, assign only code U07.0.
- Assign additional codes for other manifestations, such as acute respiratory failure (subcategory J96.0-) or pneumonitis (code J68.0).
- Associated respiratory signs and symptoms due to vaping, such as cough, shortness of breath, etc., are not coded separately, when a definitive diagnosis has been established.
- However, it would be appropriate to code separately any gastrointestinal symptoms, such as diarrhea and abdominal pain.

Official Coding Guidelines I.C.15.k

Code O85 should not be assigned for sepsis following an obstetrical procedure (See Section I.C.1.d.5.b., Sepsis due to a postprocedural infection).

Official Coding Guidelines I.C.21.(c).(6)

- The observation codes are **primarily** to be used as a principal/**first-listed** diagnosis.
- **An observation code may be assigned as a secondary diagnosis code when the patient is being observed for a condition that is ruled out and is unrelated to the principal/first-listed diagnosis (e.g., patient presents for treatment following injuries sustained in a motor vehicle accident and is also observed for suspected COVID-19 infection that is subsequently ruled out).**

Other Index and Tabular Changes

ARTERIOSCLEROSIS

FFY2020

Arteriosclerosis, arteriosclerotic (diffuse) (obliterans)
 (soft) (scler) (with calcification) 178.50
 aorta 125.8
 arteries of extremities — see Arteriosclerosis, extremities
 brain 87.2
 bypass graft
 coronary — see Arteriosclerosis, coronary, bypass graft
 extremities — see Arteriosclerosis, extremities, bypass graft
 cardiac — see Disease, heart, ischemic, atherosclerotic
 cardiopathy — see Disease, heart, ischemic, atherosclerotic
 cardiovascular — see Hypertension, cardiovascular
 cardiovascular — see Disease, heart, ischemic, atherosclerotic
 carotid — see Arterio Occlusion, artery, carotid 85.2
 central nervous system 87.2
 cerebral 87.2
 cerebrovascular 87.2
 coronary (artery)
 due to
 calcified coronary lesion (sclerosis) 125.84
 lipid rich plaque 125.83
 bypass graft 125.818

FFY 2021

Arteriosclerosis, arteriosclerotic (diffuse) (obliterans)
 (soft) (scler) (with calcification) 178.50
 with
 Chronic Bicuspid-Aortic Valve Disease — see Arteriosclerosis, with critical limb ischemia
 cerebral (with hydrocephalus)
 bypass graft 178.518
 calcification
 autologous vein graft 178.429
 leg 178.428
 with
 gangrene (and intermittent claudication, rest pain, and ulcers)
 178.401
 rest pain (and intermittent claudication)
 femur 178.429
 bilateral 178.413
 gangrene (and intermittent claudication, rest pain, and ulcers)
 rest pain (and intermittent claudication)
 femur 178.429
 with
 gangrene (and intermittent claudication, rest pain, and ulcers)
 rest pain (and intermittent claudication)
 femur 178.429
 with
 gangrene (and intermittent claudication, rest pain, and ulcers)
 rest pain (and intermittent claudication)
 femur 178.429
 calcification (and intermittent claudication, rest pain)
 femur 178.428
 with
 gangrene (and intermittent claudication, rest pain, and ulcers)
 rest pain (and intermittent claudication)
 femur 178.429
 with
 gangrene (and intermittent claudication, rest pain, and ulcers)
 rest pain (and intermittent claudication)
 femur 178.429
 with
 gangrene (and intermittent claudication, rest pain, and ulcers)
 rest pain (and intermittent claudication)
 femur 178.429

FRACTURES

No Change Fracture, traumatic (abduction) (adduction) (separation) (see also Fracture, pathological) T14.8
 Add - buckle - see Fracture, by site, tons
 Add - metaphyseal - see Fracture, traumatic, by site, shaft

- Calculus of ureteropelvic junction has been added to the index and defaults to N20.1
- Emaciation (due to malnutrition) now indexes to E43 (instead of E41)
- Facet syndrome now defaults to M47.89- (other spondylosis)
- Central line infection now defaults to T80.211-
- Influenzas A, B, & C now have a direct index to J10-

No Change **Aplastic and other anemias and other bone marrow failure syndromes (D60-D64)**

No Change **D61 Other aplastic anemias and other bone marrow failure syndromes**

Delete **Excludes1: neutropenia (D70.-)** ←



Add **Excludes2: neutropenia (D70.-)** ←



No Change **D84.9 Immunodeficiency, unspecified**



Add **Immunocompromised NOS**



Add **Immunodeficient NOS**

Add **Immunosuppressed NOS**

No Change **E86 Volume depletion**
 Delete  **Excludes1:** hypovolemic shock NOS (R57.1)
 Add  **Excludes2:** hypovolemic shock NOS (R57.1)

No Change **I46 Cardiac arrest**
 Delete  **Excludes1:** cardiogenic shock (R57.0)
 Add  **Excludes2:** cardiogenic shock (R57.0)

No Change **M23 Internal derangement of knee**
 Delete  **Excludes1:** current injury - see injury of knee and lower leg (S80-S89)
 Delete recurrent dislocation or subluxation of joints (M24.4)
 Delete recurrent dislocation or subluxation of patella (M22.0-M22.1)
 Add  **Excludes2:** current injury - see injury of knee and lower leg (S80-S89)
 Add recurrent dislocation or subluxation of joints (M24.4)
 Add recurrent dislocation or subluxation of patella (M22.0-M22.1)

No Change **S93 Dislocation and sprain of joints and ligaments of knee**
 Delete  **Excludes1:** derangement of patella (M22.0-M22.3)
 Delete injury of patellar ligament (tendon) (S76.1-)
 Delete internal derangement of knee (M23.-)
 Delete old dislocation of knee (M24.36)
 Delete pathological dislocation of knee (M24.36)
 Delete recurrent dislocation of knee (M22.0)
 Add  **Excludes2:** derangement of patella (M22.0-M22.3)
 Add injury of patellar ligament (tendon) (S76.1-)
 Add internal derangement of knee (M23.-)
 Add old dislocation of knee (M24.36)
 Add pathological dislocation of knee (M24.36)
 Add recurrent dislocation of knee (M22.0)

| | |
|-----------|---|
| No Change | S06.3 Focal traumatic brain injury |
| Delete | ➔ Excludes1: focal cerebral edema (S06.1) |
| Add | ➔ Excludes2: focal cerebral edema (S06.1) |

On Your Own



https://www.timeshighereducation.com/sites/default/files/styles/the_breaking_news_image_style/public/stock-521611936.jpg?itok=yWaoXHYc

- Sub-Category D72.1 Eosinophilia was expanded
 - D72.10 Eosinophilia, unspecified
 - D72.110 Idiopathic hypereosinophilic syndrome [IHES]
 - D72.111 Lymphocytic variant hypereosinophilic syndrome [LHES]
 - D72.118 Other hypereosinophilic syndrome
 - D72.119 Hypereosinophilic syndrome [HES], unspecified
 - D72.12 Drug rash with eosinophilia and systemic symptoms syndrome
 - D72.18 Eosinophilia in diseases classified elsewhere
 - D72.19 Other eosinophilia

- Sub-Category G71.2 Congenital Myopathies was expanded
 - G71.20 Congenital myopathy, unspecified
 - G71.21 Nemaline myopathy
 - G71.22 Centronuclear myopathy
 - G71.220 X-linked myotubular myopathy
 - G71.228 Other centronuclear myopathy
 - G71.29 Other congenital myopathy

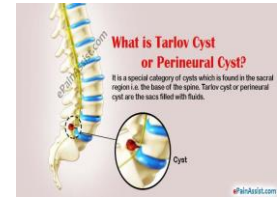
CC Status

```

G71.2 Congenital myopathies
  Centronuclear disease
  Fiber-type disproportion
  Minicore disease
  Multicore disease
  Myotubular (centronuclear) myopathy
  Nemaline myopathy
  [E02.00] arylglycosylase multiple congenite (G74.3)
  [E02.01] arylglycosylase multiple congenite (G74.3)
  [G71.20] Congenital myopathy, unspecified
  G71.21 Nemaline myopathy
  G71.22 Centronuclear myopathy
    G71.220 X linked myotubular myopathy
      Myotubular centronuclear myopathy
    G71.228 Other centronuclear myopathy
      Autosomal centronuclear myopathy
      Autosomal dominant centronuclear myopathy
      Autosomal recessive centronuclear myopathy
      Centronuclear myopathy, NOS
  G71.29 Other congenital myopathy
    Centronuclear disease
    Minicore disease
    Multicore disease
    Multifasciculation disease
    
```

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- Sub-Category G96.19 Other Disorders of Meninges NEC has been expanded
 - G96.191 Perineural cyst
 - G96.198 Other disorders of meninges, not elsewhere classified



- Tarlov or perineural cysts are cerebrospinal fluid-filled sacs that most often affect nerve roots of the spine, especially near the sacral region.
- The cyst can grow in size eventually compressing adjacent nerve roots or nerves contained within the cyst. Multiple systems symptomatology can occur depending upon the size and specific location of the cyst and due to progressive nerve damage and organ dysfunction.

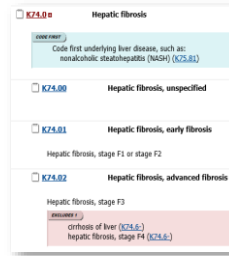
- Symptoms caused by Tarlov cysts include pain in the area of the affected nerves, paresthesias (numbness, burning, tingling and altered sensation), severe muscle spasms and cramping, leading to muscle atrophy, chronic headaches, and bladder, bowel and sexual dysfunction.
- The exact cause of Tarlov cysts is unknown; however, there is some clinical evidence that symptoms developed following trauma, and possible connective tissue disorders (Marfan's, Ehlers-Danlos, Loeys-Dietz, Lupus, Sjogren's, etc.) that predispose the patient to developing this type of spinal nerve root cyst.

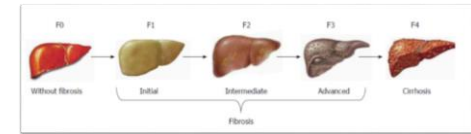
- Category J82 Pulmonary Eosinophilia NEC has been expanded
 - J82.81 Chronic eosinophilic pneumonia
 - J82.82 Acute eosinophilic pneumonia
 - J82.83 Eosinophilic asthma
 - J82.89 Other pulmonary eosinophilia, not elsewhere classified

CC Status

- Sub-Category J87.17 Other interstitial pulmonary disease with fibrosis in diseases classified elsewhere has been expanded
 - J84.170 *Interstitial lung disease with progressive fibrotic phenotype in diseases classified elsewhere*
 - J84.178 *Other interstitial pulmonary diseases with fibrosis in diseases classified elsewhere*
- *These are manifestation codes and cannot be sequenced as PDX

- Sub-Category K74.0 Hepatic Fibrosis has been expanded
 - K74.00 Hepatic fibrosis, unspecified
 - K74.01 Hepatic fibrosis, early fibrosis
 - K74.02 Hepatic fibrosis, advanced fibrosis





<https://www.wjnet.com/1007-9327/full/v21/i41/11552.htm>

- Advanced fibrosis due to NASH is associated with increased risk of liver-related complications, including liver-related mortality, as well overall mortality.
- Patients with advanced fibrosis due to NASH also have a reduced quality of life. They have increased risk of liver cancer, and increased risk of hospitalization.
- Assessment of hepatic fibrosis has traditionally depended on liver biopsy, and that is the gold standard.
- However, there are now non-invasive tests that can be used for assessment of the stages of hepatic fibrosis. These will be helpful in detecting and differentiating early and advanced fibrosis.

- Twenty-one new codes have been created for several joint related disorders by adding "other specified site" or "other specified joint"
 - e.g., rheumatoid arthritis, osteoarthritis, dislocation
- Sub-Category M26.6 Temporomandibular Joint Disorders has been expanded
 - M26.64- Arthritis of temporomandibular joint
 - M26.65- Arthropathy of temporomandibular joint

- Subcategory M92.5, Juvenile osteochondrosis of tibia and fibula, has been expanded to distinguish juvenile osteochondrosis (Blount's Disease vs Osgood-Schlatter Disease)
- Unspecified juvenile osteochondrosis (M92.50-)
- Juvenile osteochondrosis of proximal tibia (M92.51-)
- Juvenile osteochondrosis of tibia tubercle (M92.52-)
- Other juvenile osteochondrosis of tibia and fibula (M92.59-)
- Sixth characters at each of the codes identify laterality (i.e. unspecified leg, right leg, left leg and bilateral).

- The two conditions, Blount Disease and Osgood-Schlatter are very dissimilar both in character, prognosis and treatment.
- Blount disease is a growth disorder of the tibia (shin bone) that causes the lower leg to angle inward, resembling a bowleg which occurs in growing children.
- Osgood-Schlatter disease is a characteristic of soreness and swelling at the tibial tuberosity, which occurs in adolescence.

- New codes at categories N00 to N07 to identify C3 glomerulopathy:
 - N00.A Acute nephritic syndrome with C3 glomerulonephritis
 - N01.A Rapidly progressive nephritic syndrome with C3 glomerulonephritis
 - N02.A Recurrent and persistent hematuria with C3 glomerulonephritis
 - N03.A Chronic nephritic syndrome with C3 glomerulonephritis
 - N04.A Nephrotic syndrome with C3 glomerulonephritis
 - N05.A Unspecified nephritic syndrome with C3 glomerulonephritis
 - N06.A Isolated proteinuria with C3 glomerulonephritis
 - N07.A Hereditary nephropathy, not elsewhere classified with C3 glomerulonephritis

- Glomerulonephritis is classified into pathogenic types, which have been defined by the classification forms seen in renal biopsy.
- One of these types is C3 glomerulopathy. A newly classified, uncommon kidney disorder characterized by the deposition of complement component 3 (C3) within the glomeruli.
- C3G is comprised of two distinct clinical subtypes: C3 glomerulonephritis (C3GN) and dense deposit disease (DDD).
- While DDD had a specific ICD-10-CM code, there were none for C3G or the subtype C3GN.

- Granulomatous mastitis is a rare, chronic, inflammatory condition of the breast with unknown etiology that affects women of child-bearing age.
- It can be mistaken radiographically and clinically for breast cancer and due to its rarity can cause a delay in establishing a definitive diagnosis and subsequent initiation of treatment.
- Furthermore, granulomatous mastitis has a progressive clinical course with multiple recurrences.

- Category R51 Headache has been expanded
 - R51.0 Headache with orthostatic component, not elsewhere classified
 - R51.9 Headache, unspecified
- Also known as positional headache, postural headache or orthostatic headache.

- Sub-Category Y77.1 Therapeutic (nonsurgical) and rehabilitative ophthalmic devices associated with adverse incidents has been expanded
 - Y77.11 Contact lens associated with adverse incidents
 - Y77.19 Other therapeutic (nonsurgical) and rehabilitative ophthalmic devices associated with adverse incidents

• **New codes:**

- A84.81 Powassan virus disease **MCC**
- B60.00 Babesiosis, unspecified **CC**
- B60.01 Babesiosis due to *Babesia microti* **CC**
- B60.02 Babesiosis due to *Babesia duncani* **CC**
- B60.03 Babesiosis due to *Babesia divergens* **CC**
- B60.09 Other babesiosis **CC**

- Powassan (POW) virus disease is a tick-borne zoonosis caused by a bite of an infected tick, mostly *Ixodes scapularis* (deer tick).
- Most POW cases have occurred in the Northeastern and Great Lakes regions of the United States during the spring, summer, and mid-fall when ticks and humans are most active
- Although many infected persons may not develop symptoms immediately due to a long incubation period of 1 week to 1 month, Powassan virus disease is considered a serious disease that usually results in encephalitis and/or meningitis and may lead to death.
- Symptoms can include fever, headache, vomiting, weakness, confusion, loss of coordination, speech difficulties, and seizures.
- Only 21 cases were reported in 2018, but this is an increase from the 6 that were reported in 2009.

- Human babesiosis is a tick-borne zoonosis caused by intra-erythrocytic protozoa of the genus *Babesia*.
- Babesiosis can also be transmitted by transfusion of blood and blood components collected from an infected donor.
- Although the majority of U.S. babesiosis cases are caused by *B. microti*, which is prevalent in the Northeast and upper Midwest, other *Babesia* species such as *B. duncani*, *B. divergens*, and other species have been implicated in transmission in multiple U.S. states.
- On January 24, 2019, FDA approved a nucleic acid amplification test for donor screening, to detect specific *Babesia* species: *B. microti*, *B. duncani*, *B. divergens*, and *B. venatorum*

CC Status

- **New Codes**
 - E70.81 Aromatic L-amino acid decarboxylase deficiency
 - E70.89 Other disorders of aromatic amino-acid metabolism
- **Aromatic L-amino acid decarboxylase (AADC) deficiency is a rare genetic, autosomal-recessive disorder resulting in an inborn error of neurotransmitter biosynthesis.**
- **Common signs and symptoms of AADC deficiency include hypotonia, hypokinesia, hypertonia, dystonia, oculogyric crisis, developmental delay/failure to thrive, ptosis, and excessive sweating.**

CC Status

- **New Codes:**
 - E74.810 Glucose transporter protein type 1 deficiency
 - E74.818 Other disorders of glucose transport
 - E74.819 Disorders of glucose transport, unspecified
 - E74.89 Other specified disorders of carbohydrate metabolism

CC Status

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

• New Codes:

- G11.10 Early-onset cerebellar ataxia, unspecified
- G11.11 Friedreich ataxia
- G11.19 Other early-onset cerebellar ataxia

CC Status

- Friedreich ataxia (FA/FRDA) is a multi-system neurological disorder characterized by progressive symptoms of gait and balance instability, impaired coordination affecting all muscles, dysarthria, scoliosis, loss of sensation in the arms and legs, cardiomyopathy and arrhythmia, diabetes, and hearing and vision loss.
- While FA is a multi-systemic disease, brain development and cognitive functioning are at least for the most part preserved, although there may be some subtle cognitive deficits in some cases.
- FA is one of the most common forms of inherited ataxia, affecting about 1 in 50,000 people on average, although it may be as common as 1 in 20,000 in some populations, and much less common in other populations.
- It particularly affects Caucasians, especially those originating from southwestern Europe. It is estimated to affect thousands of people in the U.S. Most people with FA present between the ages 10 to 16 years, although symptoms may start early by age 5, or later, after the age of 25.

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• New Codes

- G40.42 Cyclin-dependent kinase-like 5 deficiency disorder
- CDKL5 Deficiency Disorder is a developmental encephalopathy caused by pathogenic variants in the gene *CDKL5*.
- It is a unique disorder that presents with early infantile onset refractory epilepsy, hypotonia, developmental intellectual and motor disabilities, and cortical visual impairment.
- It also causes autonomic problems and gastrointestinal dysfunction that range from oral adversity, swallow dysfunction, gastroesophageal reflux and constipation.
- Over time, children affected can develop scoliosis.

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• New Codes

- G40.833 Dravet syndrome, intractable, with status epilepticus
- G40.834 Dravet syndrome, intractable, without status epilepticus

CC Status

- Dravet syndrome, previously known as severe myoclonic epilepsy in infancy (SMEI), is a genetic encephalopathy that presents in the first year of life.

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• Hereditary Corneal Dystrophy

- 28 new codes in subcategory H18.5, Hereditary corneal dystrophies, have been expanded to identify laterality (right eye, left eye, bilateral and unspecified eye)

• Irregular Eye Movements

- Code H55.81 revised
 - From: Saccadic eye movements
 - To: Deficient saccadic eye movements
- New code: H55.82 Deficient smooth pursuit eye movements

• Sub-Category K59.8 Other Specified Functional Intestinal Disorders has been expanded

- K59.81 Ogilvie syndrome
- K59.89 Other specified functional intestinal disorders

- Ogilvie syndrome is a rare, acquired disorder characterized by abnormalities affecting the involuntary, rhythmic muscular contractions within the colon.
- Ogilvie syndrome is also known as acute colonic pseudo-obstruction (ACPO).
- Symptoms of Ogilvie syndrome are similar to other forms of intestinal pseudo-obstruction and can include nausea, vomiting, abdominal colic and constipation.
- The symptoms mimic those of mechanical blockage of the colon, but no such physical obstruction is present.
- Distention of the colon in Ogilvie syndrome can potentially lead to serious, life-threatening complications including the formation of a hole in the wall of the colon or lack of blood flow to the colon.
- Ogilvie syndrome is usually associated with an underlying disorder, trauma or surgery. Non-operative trauma, infection and heart disease are common conditions associated with Ogilvie syndrome.
- Ogilvie syndrome can be managed with conservative treatment, but if unrecognized and untreated can lead to serious, potentially life-threatening complications.
- It is not the same as chronic intestinal pseudo-obstruction (CIP), a similar, but distinct disorder.

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<https://encrypted-tbn0.gstatic.com/images?q=tbn%3AANd9GcRLhTKLVdffeOwLgD2yuitb6kgwpTgijXjCA&usqp=CAU>

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