CENTERS FOR DISEASE CONTROL AND PREVENTION

ICD-10 Coordination and Maintenance Committee Meeting
September 10-11, 2019
Diagnosis Agenda
Part 2 of 2

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

Diagnosis Topics:

Contents

ICD-10 TIMELINE ONLY

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

September 10-11, 2019	ICD-10 Coordination and Maintenance Committee Meeting. Those who wish to attend the ICD-10 Coordination and Maintenance Committee meeting **must have registered for the meeting online by September 2, 2019.** You must bring an official form of picture identification (such as a driver's license) in order to be admitted to the building. In compliance to The Real ID Act, enacted in 2005, (http://www.dhs.gov/real-id-enforcement-brief) the following states/territories: Maine, Minnesota, Missouri, Montana and Washington State **will not** gain access into any Federal Agencies using the **above states** driver's license or ID. This means CMS visitors from these states/territories will need to provide alternative proof of identification (**such as a passport**) to gain entrance into Baltimore-based and Bethesda CMS buildings, as well as the Humphrey Building in Washington.
September 2019	Webcast of the September 10-11, 2019 ICD-10 Coordination and Maintenance Committee meeting will be posted on the CMS webpage as follows: https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html
October 1, 2019	**New and revised ICD-10-CM and ICD-10-PCS codes go into effect along with DRG changes. Final addendum available on web pages as follows:** **Diagnosis addendum –** https://www.cdc.gov/nchs/icd/icd10cm.htm **Procedure addendum –** https://www.cms.gov/Medicare/Coding/ICD10/
October 11, 2019	**Deadline for receipt of public comments on proposed new codes discussed at the September 10-11, 2019 ICD-10 Coordination and Maintenance Committee meetings for implementation on April 1, 2020.**

November 2019	Any new ICD-10 codes required to capture new technology or new diseases that will be implemented on the following April 1 will be announced. Information on any new codes to be implemented April 1, 2020 will be posted on the following websites: https://www.cdc.gov/nchs/icd/icd10cm.htm https://www.cms.gov/Medicare/Coding/ICD10/
November 8, 2019	**Deadline for receipt of public comments on proposed new codes and revisions discussed at the September 10-11, 2019 ICD-10 Coordination and Maintenance Committee meetings for implementation on October 1, 2020.**
December 6, 2019	**Deadline for requestors: Those members of the public requesting that topics be discussed at the March 17-18, 2020 ICD-10 Coordination and Maintenance Committee meeting must have their requests submitted to CMS for procedures and NCHS for diagnoses by this date.**
February 2020	Tentative agenda for the Procedure part of the March 17, 2020 ICD-10 Coordination and Maintenance Committee meeting posted on CMS webpage as follows: https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html Tentative agenda for the Diagnosis part of the March 18, 2020 ICD-10 Coordination and Maintenance Committee meeting posted on NCHS homepage as follows: https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm Federal Register notice of March 17-18, 2020 ICD-10 Coordination and Maintenance Committee Meeting will be published.
February 6, 2020	**On-line registration opens for the March 17-18, 2020 ICD-10 Coordination and Maintenance Committee meeting at:** https://www.cms.gov/apps/events/default.asp
March 6, 2020	Because of increased security requirements, **those wishing to attend the March 17-18, 2020** ICD-10 Coordination and Maintenance Committee meeting are required to register for the meeting online at: https://www.cms.gov/apps/events/default.asp **Attendees must register online by March 6, 2020; failure to do so may result in lack of access to the meeting**.

March 17-18, 2020	ICD-10 Coordination and Maintenance Committee Meeting.
March 2020	Webcast of the March 17-18, 2020 ICD-10 Coordination and Maintenance Committee meeting will be posted on the CMS webpage as follows: https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html
April 1, 2020	Any new ICD-10 codes to capture new diseases or technology will be implemented on April 1, 2020.
April 17, 2020	**Deadline for receipt of public comments on proposed new codes and revisions discussed at the March 17-18, 2020 ICD-10 Coordination and Maintenance Committee meetings for implementation on October 1, 2020**.
April 2020	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 finalized FY 2021 ICD-10-CM diagnosis and ICD-10-PCS procedure codes to date. It will also include proposed revisions to the MS-DRG system based on ICD-10-CM/PCS codes on which the public may comment. The proposed rule can be accessed at: https://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/AcuteInpatientPPS/index.html?redirect=/AcuteInpatientPPS/IPPS/list.asp
June 2020	Final addendum posted on web pages as follows: Diagnosis addendum - https://www.cdc.gov/nchs/icd/icd10cm.htm Procedure addendum - https://www.cms.gov/Medicare/Coding/ICD10/index.html
June 12, 2020	**Deadline for requestors: Those members of the public requesting that topics be discussed at the September 2020 ICD-10 Coordination and Maintenance Committee meeting, tentatively scheduled for September 8-9, 2020, must have their requests submitted to CMS for procedures and NCHS for diagnoses.**
August 1, 2020	Hospital Inpatient Prospective Payment System final rule to be published in the Federal Register as mandated by Public Law 99-509. This rule will also include links to all the final codes to be implemented on October 1, 2020.

	This rule can be accessed at: https://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/AcuteInpatientPPS/index.html
August 2020	Tentative agenda for the Procedure part of the September 2020 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 part of the September 8-9, 2020 ICD-10 Coordination and Maintenance Committee meeting will be posted on the NCHS webpage at - https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm Federal Register notice for the September 2020 ICD-10 Coordination and Maintenance Committee meeting will be published. This will include the tentative agenda.
August 3, 2020	**On-line registration opens for the September 2020 ICD-10 Coordination and Maintenance Committee meeting at:** https://www.cms.gov/apps/events/default.asp
September 3, 2020	Because of increased security requirements, those wishing to attend the September 2020 ICD-10 Coordination and Maintenance Committee meeting must register for the meeting online at: https://www.cms.gov/apps/events/default.asp **Attendees must register online by September 3, 2020; failure to do so may result in lack of access to the meeting.**
September 8-9, 2020 (Tentative date)	ICD-10 Coordination and Maintenance Committee Meeting. Those who wish to attend the ICD-10 Coordination and Maintenance Committee meeting **must have registered for the meeting online by September 3, 2020.** You must bring an official form of picture identification (such as a driver's license) in order to be admitted to the building.
September 2020	Webcast of the September 8-9, 2020 ICD-10 Coordination and Maintenance Committee meeting will be posted on the CMS webpage as follows: https://www.cms.gov/Medicare/Coding/ICD10/C-and-M-Meeting-Materials.html

October 1, 2020	New and revised ICD-10-CM and ICD-10-PCS codes go into effect along with DRG changes. Final addendum available on web pages as follows: **Diagnosis addendum –** https://www.cdc.gov/nchs/icd/icd10cm.htm **Procedure addendum –** https://www.cms.gov/Medicare/Coding/ICD10/
October 9, 2020	**Deadline for receipt of public comments on proposed new codes discussed at the September 8-9, 2020 ICD-10 Coordination and Maintenance Committee meetings for implementation on April 1, 2021.**
November 2020	Any new ICD-10 codes required to capture new technology that will be implemented on the following April 1 will be announced. Information on any new codes to be implemented April 1, 2021 will be posted on the following websites: https://www.cdc.gov/nchs/icd/icd10cm.htm https://www.cms.gov/Medicare/Coding/ICD10/
November 9, 2020	**Deadline for receipt of public comments on proposed new codes and revisions discussed at the September 8-9, 2020 ICD-10 Coordination and Maintenance Committee meetings for implementation on October 1, 2021.**

Webcast and Dial-In Information for Listen-only Participants

• Day 1: September 10, 2019: The meeting will begin promptly at 9:00 AM ET and will end at 5:00 PM ET. Lunch will be held from 12:30 PM to 1:30 PM. The meeting will be webcast via CMS at http://www.cms.gov/live/.
• Day 2: September 11, 2019: The meeting will begin promptly at 9:00 AM ET and will end at 5:00 PM ET. Lunch will be held from 12:30 PM to 1:30 PM. The meeting will be webcast via CMS at http://www.cms.gov/live/.
• Toll-free dial-in access is available for listen-only participants who cannot join the webcast:

Day 1-September 10, 2019: Phone: 1-877-267-1577; Meeting ID: 995 481 510.
Day 2-September 11, 2019: Phone: 1-877-267-1577; Meeting ID: 995 481 510.
We encourage you to join early, as the number of phone lines is limited.

In-Person Attendance
• Day 1: September 10, 2019: The meeting is being held in the CMS Auditorium. The meeting time is listed above. By your attendance, you are giving consent to the use and distribution of your name, likeness and voice during the meeting. You are also giving consent to the use and distribution of any personally identifiable information that you or others may disclose about you during the meeting. Please do not disclose personal health information.

There will be a WebEx option for this meeting. In-person attendees and those participating via WebEx may ask questions, as time permits. Remaining questions, as well as questions from those attending the meeting via the webcast may be submitted via the CMS ICD-10 Procedure Code Request mailbox at ICDProcedureCodeRequest@cms.hhs.gov.

• Day 2: September 11, 2019: The meeting is being held in the CMS Auditorium. The meeting time is listed above. By your attendance, you are giving consent to the use and distribution of your name, likeness and voice during the meeting. You are also giving consent to the use and distribution of any personally identifiable information that you or others may disclose about you during the meeting. Please do not disclose personal health information.

Note: Proposals for diagnosis code topics are scheduled for September 11, 2019 and will be led by the Centers for Disease Control (CDC). Please visit CDCs website for the Diagnosis agenda located at the following address: http://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm

Registration to attend meeting in-person:
Information on registering online to attend the meeting in-person can be found at: http://www.cms.hhs.gov/apps/events/ ***If participating via the webcast or dialing in, and not attending in-person, you do NOT need to register on-line for the meeting.** For questions about the registration process, please contact Mady Hue at 410-786-4510 or marilu.hue@cms.hhs.gov or Noel Manlove at 410-786-5161 or noel.manlove@cms.hhs.gov.
Updated Security Information for In-person Attendees:

Beginning June 1, 2018, Federal Protective Services (FPS) has implemented new security screening procedures at all CMS Baltimore locations to align with national screening standards. Please allow extra time to clear security prior to the beginning of the meeting.
Employees, contractors and visitors must place **all items** in bins for screening, including:
• Any items in your pockets
• Belts, hats, jackets & coats (not suit jackets or sport coats)
• Purses, laptop computers & cell phones
• Larger items (e.g. computer bags) can be placed directly onto the conveyer.

In the event the metal detector beeps when you walk through:
• A security guard will run a hand-held metal detector over you. If the metal detector doesn't alarm, you're cleared to enter.
• If the hand-held metal detector alarms, the guard will pat down the area of the body where the metal detector alarmed.
• If footwear alarms, it will need to be removed and placed in a bin for x-ray screening.
• Employees using a mobility aid (e.g. wheelchair, motorized scooter) will be screened using a hand-held metal detector and/or pat-down.

If you believe that you have a disability that will cause you to require reasonable accommodation to comply with the new process, please contact **reasonableaccommodationprogram@cms.hhs.gov** as soon as possible.

Contact Information
Mailing address:

National Center for Health Statistics
ICD-9-CM Coordination and Maintenance Committee
3311 Toledo Road
Hyattsville, Maryland 20782
Fax: (301) 458-4022

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

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

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 a 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.

Cytokine Release Syndrome (CRS)

In August 2017, the U.S. Food and Drug Administration (FDA) approved the first Chimeric Antigen Receptor T (CAR-T) Cell Therapy product, Novartis' Kymriah® (tisagenlecleucel), for certain pediatric and young adult patients with relapsed or refractory B cell acute lymphoblastic leukemia. While this therapy may represent a last chance for patients who have exhausted other treatment options, it is not without side effects or known complications—primarily Cytokine Release Syndrome (CRS.

Cytokine Release Syndrome (CRS) is a condition that may occur after treatment with some types of immunotherapy, such as monoclonal antibodies and Chimeric Antigen Receptor T (CAR-T) Cell therapy. It is the most common reaction after CAR-T Cell therapy.

This syndrome is caused by a large, rapid release of cytokines into the blood from immune cells affected by the immunotherapy. Cytokines are immune substances that have many different actions in the body.

In most patients, the symptoms are mild to moderate in severity and are managed easily. Signs and symptoms of cytokine release syndrome include fever, nausea, headache, rash, rapid heartbeat, low blood pressure, and trouble breathing. When cytokines are released into circulation, a range of symptoms can result, including low-grade constitutional symptoms, or a high-grade syndrome associated with life-threatening multi-organ dysfunction.
However, some patients may experience severe, life-threatening reactions that result from massive release of cytokines. Severe reactions occur more commonly during the first infusion in patients with hematologic malignancies who have not received prior chemotherapy; severe reactions are marked by their rapid onset and the acuity of associated symptoms including but not limited to: signs of fluid overload (including pulmonary and hepatic edema) and rash, associated with hematopoietic stem cells engraftment following a transplant of bone marrow, stem cells or other hematopoietic tissues.

Massive cytokine release is an oncologic emergency, and special precautions must be taken to prevent life-threatening complications. It is the sudden exuberant growth of these cells and the ensuing cytokine release that results in this condition. Without intensive clinical support patients may succumb to this syndrome.

NCHS has received multiple code proposal regarding CRS. This proposal represents a starting point to capture information on this clinical condition and more specific codes may be needed in the future. Two options are being presented for consideration. Of note this proposal is supported by the Alliance of Dedicated Cancer Centers (ADCC) with preference of option #2. NCHS recommends option #1.

TABULAR MODIFICATIONS

Option #1

D89 Other disorders involving the immune mechanism, not elsewhere classified

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

D89.81 Graft-versus-host disease

Code first underlying cause, such as:
complications of transplanted organs and tissue (T86.-)
complications of blood transfusion (T80.89)

Use additional code to identify associated manifestations, such as:
desquamative dermatitis (L30.8)
diarrhea (R19.7)
elevated bilirubin (R17)
hair loss (L65.9)

D89.810 Acute graft-versus-host disease
D89.811 Chronic graft-versus-host disease
D89.812 Acute on chronic graft-versus-host disease
D89.813 Graft-versus-host disease, unspecified

D89.82 Autoimmune lymphoproliferative syndrome [ALPS]

New Code D89.83 Cytokine release syndrome

Cytokine release syndrome (CRS)

Code first underlying cause, such as:
complications of transplanted organs and tissue (T86.89-)

Use additional code to identify associated manifestations

D89.89 Other specified disorders involving the immune mechanism, not elsewhere classified

Excludes1:human immunodeficiency virus disease (B20)

D89.9 Disorder involving the immune mechanism, unspecified

Option #2

New subcategory	D89.83 Cytokine release syndrome Cytokine release syndrome (CRS) Code first underlying cause, such as: complications of transplanted organs and tissue (T86.89-) Use additional code to identify associated manifestations
New code	D89.831 Cytokine release syndrome (CRS) grade 1
New code	D89.832 Cytokine release syndrome (CRS) grade 2
New code	D89.833 Cytokine release syndrome (CRS) grade 3
New code	D89.834 Cytokine release syndrome (CRS) grade 4
New code	D89.835 Cytokine release syndrome (CRS) grade 5 Cytokine release syndrome (CRS) death
New code	D89.839 Cytokine release syndrome (CRS), grade unspecified

Electric Scooter and Other Micro-Mobility Devices

The American College of Surgeons Committee on Trauma, Atrium Health, UNC Highway Safety Research Center, and the Vision Zero San Francisco Injury Prevention Research Collaborative are proposing capture of electric scooters (e-scooters) as "pedestrian conveyances" (V00-V09), as they are closest in form and function to other types of transportation in which the rider is standing and maintains a high center of gravity (e.g. skateboard and rollerblades). 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.

We feel that due to the widespread media attention surrounding injuries related to e-scooters and other micro-mobility devices (particularly in communities with rideshares), clinicians will be familiar with these devices and will, therefore, document their involvement in the clinical notes. Studies have identified e-scooter injuries using keywords ("e-scooter", "Lime", "Bird", etc.) present in the medical record and/or in syndromic surveillance data (e.g. chief complaint and triage note). Medical coders are already experienced in correctly classifying a wide range of transportation-related injuries using information documented by clinicians. Accurate injury surveillance is critical to the successful development, implementation and evaluation of prevention initiatives. Urban transportation's rapidly expanding micro-mobility movement and escalating competition between rideshare companies.

The American College of Surgeons Committee on Trauma, Atrium Health, UNC Highway Safety Research Center, and the Vision Zero San Francisco Injury Prevention Research Collaborative are requesting the following new ICD-10-CM External cause codes to differentiate injuries related to e-scooters and other ultralight micro-mobility devices from injuries related to other types of pedestrian conveyances.

TABULAR MODIFICATIONS

V00 Pedestrian conveyance accident

V00.0 Pedestrian on foot injured in collision with pedestrian conveyance

New subcategory	V00.03 Pedestrian on foot injured in collision with standing micro-mobility pedestrian conveyance
New code	V00.031 Pedestrian on foot injured in collision with rider of standing electric scooter
New code	V00.038 Pedestrian on foot injured in collision with rider of other standing pedestrian conveyance
Add	Pedestrian on foot injured in collision with rider of

segway

Add — Pedestrian on foot injured in collision with rider of hoverboard

V00.8 Accident on other pedestrian conveyance

New subcategory — V00.84 Accident with standing micro-mobility pedestrian conveyance

New code — V00.841 Fall from standing electric scooter

New code — V00.842 Pedestrian on standing electric scooter colliding with stationary object

New code — V00.848 Other accident with standing micro-mobility pedestrian conveyance

Add — Accident with segway

Add — Accident with hoverboard

V01 Pedestrian injured in collision with pedal cycle

V01.0 Pedestrian injured in collision with pedal cycle in nontraffic accident

New subcategory — V01.03 Pedestrian on standing micro-mobility pedestrian conveyance injured in collision with pedal cycle in nontraffic accident

New code — V01.031 Pedestrian on standing electric scooter injured in collision with pedal cycle in nontraffic accident

New code — V01.038 Pedestrian on other standing pedestrian conveyance injured in collision with pedal cycle in nontraffic accident

Add — Pedestrian on segway injured in collision with pedal cycle in nontraffic accident

Add — Pedestrian on hoverboard injured in collision with pedal cycle in nontraffic accident

V01.1 Pedestrian injured in collision with pedal cycle in traffic accident

New subcategory — V01.13 Pedestrian on standing micro-mobility pedestrian conveyance injured in collision with pedal cycle in traffic accident

New code V01.131 Pedestrian on standing electric scooter injured in collision with pedal cycle in traffic accident

New code V01.138 Pedestrian on other standing pedestrian conveyance injured in collision with pedal cycle in traffic accident

Add Pedestrian on segway injured in collision with pedal cycle in traffic accident

Add Pedestrian on hoverboard injured in collision with pedal cycle in traffic accident

V01.9 Pedestrian injured in collision with pedal cycle, unspecified whether traffic or nontraffic accident

New subcategory V01.93 Pedestrian on standing micro-mobility pedestrian conveyance injured in collision with pedal cycle, unspecified whether traffic or nontraffic accident

New code V01.931 Pedestrian on standing electric scooter injured in collision with pedal cycle, unspecified whether traffic or nontraffic accident

New code V01.938 Pedestrian on other standing pedestrian conveyance injured in collision with pedal cycle, unspecified whether traffic or nontraffic accident

Add Pedestrian on segway injured in collision with pedal cycle, unspecified whether traffic or nontraffic accident

Add Pedestrian on hoverboard injured in collision with pedal cycle, unspecified whether traffic or nontraffic accident

V02 Pedestrian injured in collision with two- or three-wheeled motor vehicle

V02.0 Pedestrian injured in collision with two- or three-wheeled motor vehicle in nontraffic accident

New subcategory V02.03 Pedestrian on standing micro-mobility pedestrian conveyance injured in collision with two- or three-wheeled motor vehicle in nontraffic accident

New code	V02.031	Pedestrian on standing electric scooter injured in collision with two- or three-wheeled motor vehicle in nontraffic accident
New code	V02.038	Pedestrian on other standing micro-mobility pedestrian conveyance injured in collision with two- or three-wheeled motor vehicle in nontraffic accident
Add		Pedestrian on segway injured in collision with two- or three-wheeled motor vehicle in nontraffic accident
Add		Pedestrian on hoverboard injured in collision with two-or three wheeled motor vehicle in nontraffic accident

V02.1 Pedestrian injured in collision with two- or three-wheeled motor vehicle in traffic accident

New subcategory	V02.13	Pedestrian on standing micro-mobility pedestrian conveyance injured in collision with two- or three-wheeled motor vehicle in traffic accident
New code	V02.131	Pedestrian on standing electric scooter injured in collision with two- or three-wheeled motor vehicle in traffic accident
New code	V02.138	Pedestrian on other standing micro-mobility pedestrian conveyance injured in collision with two- or three-wheeled motor vehicle in traffic accident
Add		Pedestrian on segway injured in collision with two- or three-wheeled motor vehicle in traffic accident
Add		Pedestrian on hoverboard injured in collision with two-or three wheeled motor vehicle in traffic accident

V02.9 Pedestrian injured in collision with two- or three-wheeled motor vehicle, unspecified whether traffic or nontraffic accident

New subcategory V02.93 Pedestrian on standing micro-mobility pedestrian conveyance injured in collision with two- or three-wheeled motor vehicle, unspecified whether traffic or nontraffic accident

New code V02.931 Pedestrian on standing electric scooter injured in collision with two- or three wheeled motor vehicle, unspecified whether traffic or nontraffic accident

New code V02.938 Pedestrian on other standing pedestrian conveyance injured in collision with two- or three wheeled motor vehicle, unspecified whether traffic or nontraffic accident

Add Pedestrian on segway injured in collision with two- or three wheeled motor vehicle, unspecified whether traffic or nontraffic accident

Add Pedestrian on hoverboard injured in collision with two-three-wheeled motor vehicle, unspecified whether traffic or nontraffic accident

V03 Pedestrian injured in collision with car, pick-up truck or van

V03.0 Pedestrian injured in collision with car, pick-up or van in nontraffic accident

New subcategory V03.03 Pedestrian on standing micro-mobility pedestrian conveyance injured in collision with car, pick-up or van in nontraffic accident

New code V03.031 Pedestrian on standing electric scooter injured in collision with car, pick-up or van in nontraffic accident

New code V03.038 Pedestrian on other standing micro-mobility pedestrian conveyance injured in collision with car, pick-up or van in nontraffic accident

Add Pedestrian on segway injured in collision with car, pick-up or van in nontraffic accident

Add Pedestrian on hoverboard injured in collision with car, pick-up or van in nontraffic accident

V03.1 Pedestrian injured in collision with car, pick-up or van in traffic accident

New subcategory — V03.13 Pedestrian on standing micro-mobility pedestrian conveyance injured in collision with car, pick-up or van in traffic accident

New code — V03.131 Pedestrian on standing electric scooter injured in collision with car, pick-up or van in traffic accident

New code — V03.138 Pedestrian on other standing micro-mobility pedestrian conveyance injured in collision with car, pick-up or van in traffic accident

Add — Pedestrian on segway injured in collision with car, pick-up or van in traffic accident

Add — Pedestrian on hoverboard injured in collision with car, pick-up or van in traffic accident

V03.9 Pedestrian injured in collision with car, pick-up or van, unspecified whether traffic or nontraffic accident

New subcategory — V03.93 Pedestrian on standing micro-mobility pedestrian conveyance injured in collision with car, pick-up or van, unspecified whether traffic or nontraffic accident

New code — V03.931 Pedestrian on standing electric scooter injured in collision with car, pick-up or van, unspecified whether traffic or nontraffic accident

New code — V03.938 Pedestrian on other standing pedestrian conveyance injured in collision with car, pick-up or van, unspecified whether traffic or nontraffic accident

Add — Pedestrian on segway injured in collision with car, pick-up or van, unspecified whether traffic or nontraffic accident

Add — Pedestrian on hoverboard injured in collision with car, pick-up or van, unspecified whether traffic or nontraffic accident

V04 Pedestrian injured in collision with heavy transport vehicle or bus

V04.0 Pedestrian injured in collision with heavy transport vehicle or bus in nontraffic accident

New subcategory
V04.03 Pedestrian on standing micro-mobility pedestrian conveyance injured in collision with heavy transport vehicle or bus in nontraffic accident

New code
V04.031 Pedestrian on standing electric scooter injured in collision with heavy transport vehicle or bus in nontraffic accident

New code
V04.038 Pedestrian on other standing micro-mobility pedestrian conveyance injured in collision with heavy transport vehicle or bus in nontraffic accident

Add
Pedestrian on segway injured in collision with heavy transport vehicle or bus in nontraffic accident

Add
Pedestrian on hoverboard injured in collision with heavy transport vehicle or bus in nontraffic accident

V04.1 Pedestrian injured in collision with heavy transport vehicle or bus in traffic accident

New subcategory
V04.13 Pedestrian on standing micro-mobility pedestrian conveyance injured in collision with heavy transport vehicle or bus in traffic accident

New code
V04.131 Pedestrian on standing electric scooter injured in collision with heavy transport vehicle or bus in traffic accident

New code
V04.138 Pedestrian on other standing micro-mobility pedestrian conveyance injured in collision with heavy transport vehicle or bus in traffic accident

Add
Pedestrian on segway injured in collision with heavy transport vehicle or bus in traffic accident

Add
Pedestrian on hoverboard injured in collision with

heavy transport vehicle or bus in traffic accident

V04.9 Pedestrian injured in collision with heavy transport vehicle or bus in traffic accident, unspecified whether traffic or nontraffic accident

New subcategory

V04.93 Pedestrian on standing micro-mobility pedestrian conveyance injured in collision with heavy transport vehicle or bus, unspecified whether traffic or nontraffic accident

New code

V04.931 Pedestrian on standing electric scooter injured in collision with heavy transport vehicle or bus, unspecified whether traffic or nontraffic accident

New code

V04.938 Pedestrian on other standing pedestrian conveyance injured in collision with heavy transport vehicle or bus, unspecified whether traffic or nontraffic accident

Add

Pedestrian on segway injured in collision with heavy transport vehicle or bus, unspecified whether traffic or nontraffic accident

Add

Pedestrian on hoverboard injured in collision with heavy transport vehicle or bus, unspecified whether traffic or nontraffic accident

V05 Pedestrian injured in collision with railway train or railway vehicle

V05.0 Pedestrian injured in collision with railway train or railway vehicle in nontraffic accident

New subcategory

V05.03 Pedestrian on standing micro-mobility pedestrian conveyance injured in collision with railway train or railway vehicle in nontraffic accident

New code

V05.031 Pedestrian on standing electric scooter injured in collision with railway train or railway vehicle in nontraffic accident

New code

V05.038 Pedestrian on other standing micro-mobility pedestrian conveyance injured in collision with railway train or railway vehicle in nontraffic accident

Add		Pedestrian on segway injured in collision with railway train or railway vehicle in nontraffic accident
Add		Pedestrian on hoverboard injured in collision with railway train or railway vehicle in nontraffic accident
	V05.1	Pedestrian injured in collision with railway train or railway vehicle in traffic accident
New subcategory	V05.13	Pedestrian on standing micro-mobility pedestrian conveyance injured in collision with railway train or railway vehicle in traffic accident
New code	V05.131	Pedestrian on standing electric scooter injured in collision with railway train or railway vehicle in traffic accident
New code	V05.138	Pedestrian on other standing micro-mobility pedestrian conveyance injured in collision with railway train or railway vehicle in traffic accident
Add		Pedestrian on segway injured in collision with railway train or railway vehicle in traffic accident
Add		Pedestrian on hoverboard injured in collision with railway train or railway vehicle in traffic accident
	V05.9	Pedestrian injured in collision with railway train or railway vehicle in traffic accident, unspecified whether traffic or nontraffic accident
New subcategory	V05.93	Pedestrian on standing micro-mobility pedestrian conveyance injured in collision with railway train or railway vehicle, unspecified whether traffic or nontraffic accident
New code	V05.931	Pedestrian on standing electric scooter injured in collision with railway train or railway vehicle, unspecified whether traffic or nontraffic accident
New code	V05.938	Pedestrian on other standing pedestrian conveyance injured in collision with railway train or railway vehicle, unspecified whether traffic or nontraffic accident

Add		Pedestrian on segway injured in collision with railway train or railway vehicle, unspecified whether traffic or nontraffic accident
Add		Pedestrian on hoverboard injured in collision with railway train or railway vehicle, unspecified whether traffic or nontraffic accident

V06 Pedestrian injured in collision with other nonmotor vehicle

V06.0 Pedestrian injured in collision with other nonmotor vehicle in nontraffic accident

New subcategory	V06.03	Pedestrian on standing micro-mobility pedestrian conveyance injured in collision with other nonmotor vehicle in nontraffic accident
New code	V06.031	Pedestrian on standing electric scooter injured in collision with other nonmotor vehicle in nontraffic accident
New code	V06.038	Pedestrian on other standing micro-mobility pedestrian conveyance injured in collision with other nonmotor vehicle in nontraffic accident
Add		Pedestrian on segway injured in collision with other nonmotor vehicle in nontraffic accident
Add		Pedestrian on hoverboard injured in collision with other nonmotor vehicle in nontraffic accident

V06.1 Pedestrian injured in collision with other nonmotor vehicle in traffic accident

New subcategory	V06.13	Pedestrian on standing micro-mobility pedestrian conveyance injured in collision with other nonmotor vehicle in traffic accident
New code	V06.131	Pedestrian on standing electric scooter injured in collision with other nonmotor vehicle in traffic accident
New code	V06.138	Pedestrian on other standing micro-mobility pedestrian conveyance injured in collision with other nonmotor vehicle in traffic accident

Add		Pedestrian on segway injured in collision with other nonmotor vehicle in traffic accident
Add		Pedestrian on hoverboard injured in collision with other nonmotor vehicle in traffic accident

V06.9 Pedestrian injured in collision with other nonmotor vehicle, unspecified whether traffic or nontraffic accident

New subcategory	V06.93	Pedestrian on standing micro-mobility pedestrian conveyance injured in collision with other nonmotor vehicle, unspecified whether traffic or nontraffic accident
New code	V06.931	Pedestrian on standing electric scooter injured in collision with other nonmotor vehicle, unspecified whether traffic or nontraffic accident
New code	V06.938	Pedestrian on other standing pedestrian conveyance injured in collision with other nonmotor vehicle, unspecified whether traffic or nontraffic accident
Add		Pedestrian on segway injured in collision with other nonmotor vehicle, unspecified whether traffic or nontraffic accident
Add		Pedestrian on hoverboard injured in collision with other nonmotor, unspecified whether traffic or nontraffic accident

Friedreich Ataxia

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 (Cook 2017). 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 (Cook 2017; Kniffin 2013; Campuzano 1996). 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 (Cook 2017).

Friedreich ataxia is inherited in an autosomal recessive manner. Most cases of Friedreich ataxia are caused by mutation in the gene encoding frataxin (FXN), located on chromosome 9q21.11 (Kniffin 2013). The majority of patients, about 96%, have two GAA triplet repeat expansions in intron 1 on the FXN gene alleles; most remaining patients (about 4%) are compound heterozygotes with a GAA repeat expansion on one FXN allele and a point mutation or whole gene deletion on the other FXN allele (Corben 2014). In general, patients with one allele of a GAA triplet repeat of <500 have a later age of onset of FA (Castaldo 2008). Genetic testing is available to confirm a clinical diagnosis of FA.

In ICD-9-CM, there was a specific code 334.0, Friedreich ataxia; however, in the ICD-10-CM, Friedreich ataxia was grouped with certain other cerebellar ataxias, under code G11.1, Early-onset cerebellar ataxia. This grouping combines a number of distinct disorders, which do not present with the same clinical course, and can be important to differentiate clinically. Several related problems, including cardiomyopathy and diabetes, are unique to FA as compared to other cerebellar ataxias. A specific code for FA will benefit tracking it, as well as better estimating its true prevalence. It will also facilitate identifying those who have FA, and differentiating them from those with other types of early-onset cerebellar ataxia. This in turn will facilitate implementation and evaluation of clinical management guidelines for FA. Some new treatments are expected to be available within the near future, and improved tracking can help assure that new treatments are made available promptly to those affected. Drug treatments for FA are likely to be specific for it, and thus not relevant to the treatment of other ataxia conditions included under G11.1.

The Friedreich's Ataxia Research Alliance (FARA), a patient advocacy organization, has requested that a specific ICD-10-CM code be created for Friedreich ataxia, with support from a number of clinicians in neurology and pediatrics, with experience treating Friedreich ataxia.

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TABULAR MODIFICATIONS

G11 Hereditary ataxia

G11.1 Early-onset cerebellar ataxia

Delete	~~Early-onset cerebellar ataxia with essential tremor~~	
Delete	~~Early-onset cerebellar ataxia with myoclonus [Hunt's ataxia]~~	
Delete	~~Early-onset cerebellar ataxia with retained tendon reflexes~~	
Delete	~~Friedreich's ataxia (autosomal recessive)~~	
Delete	~~X-linked recessive spinocerebellar ataxia~~	
New code	G11.10	Early-onset cerebellar ataxia, unspecified
New code	G11.11	Friedreich ataxia Autosomal recessive Friedreich ataxia Friedreich ataxia with retained reflexes
New code	G11.19	Other early-onset cerebellar ataxia
Add		Early-onset cerebellar ataxia with essential tremor
Add		Early-onset cerebellar ataxia with myoclonus [Hunt's ataxia]
Add		Early-onset cerebellar ataxia with retained tendon reflexes
Add		X-linked recessive spinocerebellar ataxia

Gastric Intestinal Metaplasia

Gastric cancer is the fourth most common cancer worldwide and the second leading cause of cancer deaths. It afflicts approximately 26,000 Americans yearly. Gastric intestinal metaplasia (IM) is an important precursor lesion to gastric cancer. Currently, there is no ICD-10-CM unique code for gastric IM. A similar precursor lesion for esophageal cancer, Barrett's esophagus (also known as esophageal intestinal metaplasia) has a unique code (K22.7-).

It is believed risk for progression onto gastric cancer is highest among patients with diffuse gastric IM (which involves both antrum and body), and European guidelines use presence of diffuse gastric IM as a marker of higher risk. Gastric IM is categorized histopathologically into incomplete and complete types. Endoscopic gastric mapping to define extent of IM should be done for patients with incomplete IM to rule out dysplasia or adenocarcinoma.

The American Gastroenterological Association (AGA) is requesting new codes to contribute to epidemiologic understanding and subsequent development of appropriate surveillance guidelines in the United States.

TABULAR MODIFICATIONS

K31 Other diseases of stomach and duodenum

New subcategory	K31.A	Gastric intestinal metaplasia without dysplasia
Add		Intestinal metaplasia
New code		K31.A0 Gastric intestinal metaplasia without dysplasia, unspecified site
New code		K31.A1 Gastric intestinal metaplasia without dysplasia, involving the antrum
New code		K31.A2 Gastric intestinal metaplasia without dysplasia, involving the body (corpus)
New code		K31.A3 Gastric intestinal metaplasia without dysplasia, involving the fundus
New code		K31.A4 Gastric intestinal metaplasia without dysplasia, involving the cardia
New subcategory	K31.B	Gastric intestinal metaplasia with dysplasia
New code		K31.B0 Gastric intestinal metaplasia with dysplasia, unspecified
New code		K31.B1 Gastric intestinal metaplasia with low grade dysplasia
New code		K31.B2 Gastric intestinal metaplasia with high grade dysplasia

Hypereosinophilic Syndromes and Other Eosinophil Diseases

The hypereosinophilic syndromes and other eosinophil diseases were presented at the March 2019 meeting. This proposal modifies that original proposal, based on comments from that meeting. The text below is repeated from the previous proposal.

Hypereosinophilic syndromes (HES) are a clinically and pathogenically distinct group of disorders characterized in part by elevated blood eosinophil count and eosinophil-mediated end-organ damage. A request for new codes for certain hypereosinophilic syndromes has been received, from the American Partnership for Eosinophilic Disorders (APFED), a 501(c)3 non-profit patient advocacy group, and the International Eosinophil Society, Inc. (IES), an organization of scientists and clinicians interested in the eosinophil and its roles in health and disease.

The clinical manifestation of HES varies according to the target organ systems affected, most commonly cutaneous, pulmonary, gastrointestinal, cardiac, and nervous system tissues. The diagnosis of HES is made based on an elevated eosinophil count ($\geq 1.50 \times 10^9$/L) on at least 2 occasions, associated with evidence of eosinophil-induced organ damage or tissue infiltration after other secondary causes of hypereosinophilia such as allergic, parasitic, and non-hematological malignant disorders have been excluded. The disease burden of HES is due to both acute disease activity as well as chronic end organ damage and fibrosis.

Considerable progress has been made in the pathogenesis and treatment of different HES diseases, with development of kinase inhibitors, biologics, and other therapies, with various therapies specific for particular disorders. Specific ICD codes for these specific HES diseases will facilitate tracking and review of the management of HES patients by providing a more detailed context for understanding disease natural history and optimal therapy.

Myeloid Hypereosinophilic Syndrome (MHES)

MHES is defined as HES with features more typical of myeloproliferative disorders, including increased serum vitamin B_{12} and tryptase levels, chromosomal abnormalities, anemia and/or thrombocytopenia, hepatomegaly, splenomegaly, and circulating leukocyte precursors. Approximately half of MHES is caused by a specific fusion gene (FIP1L1-PDGFRA), associated with a deletion at 4q12. An additional population of approximately 10-20% of MHES is associated with other fusion genes or mutations (involving PDGFRA, PDGFRB, FGFR1, or JAK2). Additionally, a minority of MHES patients have no characterized mutation.

Compared to other HES diseases, MHES has a higher frequency of cardiac involvement and evolution into a more aggressive myeloproliferative disorder, both of which lead to shortened survival. This may be identified as chronic eosinophilic leukemia. MHES is typically resistant to corticosteroid therapy, and response to other treatments can depend on the MHES subtype (e.g., FIP1L1-PDGFRA positive MHES is highly responsive to low dose imatinib).

Lymphocytic Variant Hypereosinophilic Syndrome (LHES)

LHES is defined by the presence of clonal or aberrant T lymphocytes that produce Th2 cytokines, such as interleukin-5, that drive eosinophilia. Skin is the most commonly involved organ system, but other organs are frequently affected. Compared to Idiopathic Hypereosinophilic Syndrome (IHES; described below), LHES often requires higher doses of corticosteroid and off-label use of biologic or other immunosuppressive therapy to achieve an acceptable clinical response. LHES may progress into lymphoma in 5-25% of patients.

Idiopathic Hypereosinophilic Syndrome (IHES)

Prior to the discovery of the specific HES diseases noted above, all HES was considered idiopathic, IHES. It still accounts for approximately 70% of HES and is not associated with the laboratory findings that define MHES and LHES. As its name implies, the etiology of IHES is unknown. Corticosteroids are the first line treatment for IHES.

Episodic Angioedema with Eosinophilia (EAE), also known as Gleich's Syndrome

EAE (Gleich's Syndrome) was first described in 1984 as a cyclic disorder characterized by recurrent episodes of fever, angioedema, weight gain and peripheral eosinophilia with a frequency of every 4-6 weeks. It is a rare eosinophilic disorder with multilineage cell cycling with unknown prevalence. EAE has traditionally been considered a variant of HES; patients presenting with this disorder have a unique and unusual clinical phenotype distinct from other hypereosinophilic syndromes. Patients have an aberrant T cell phenotype, increase in polyclonal immunoglobulin M and frequently have a clonal T cell population in the blood. Some patients with this disorder have developed organ-system involvement and lymphoma.

EAE is rare, although it is felt that many patients may go undiagnosed or are treated as HES prior to receiving a definitive diagnosis. Based on the number of cases seen at a major referral center for HES, EAE likely makes up <1% of patients with HES. Disease onset may occur in childhood though presentation is more common in adulthood and is associated with delays in diagnosis due to the unusual nature of the presentation. Untreated patients will often have diuresis and resolution of symptoms in between cycles; however, symptoms may be severe enough to warrant daily glucocorticoid treatment.

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Drug rash with eosinophilia and systemic symptoms (DRESS) syndrome

Drug rash with eosinophilia and systemic symptoms (DRESS) syndrome is a severe, life-threatening drug reaction characterized by widespread skin rash accompanied by marked systemic symptoms including fever, lymphadenopathy, facial edema, and maculopapular rash. Systemic involvement can also include hepatitis and interstitial pneumonia. Severe cardiac involvement may be present, with eosinophilic myocarditis. Less frequently, other organ manifestations such as nephritis or pancreatitis are present. Because DRESS is triggered by both long-term and short-term drug exposure, it is critical to seek and identify the culprit drugs in the time prior to eruption.

DRESS syndrome has been attributed to a synergistic interaction of lymphocyte activation, drug metabolic enzyme defects with accumulation of drug metabolites, eosinophil activation and viral infection reactivation (human herpesvirus-6, cytomegalovirus, Epstein–Barr virus) in persons with genetic susceptibility in association with certain human leukocyte antigen (HLA) class I alleles. Autoantibody formation and autoimmune diseases, including type I diabetes mellitus, autoimmune thyroid disease, sclerodermoid graft-versus-host disease, systemic lupus erythematosus and bullous pemphigoid may occur up to four years after resolution (and are attributed to depletion of regulatory T-cells upon recovery from DRESS syndrome).

Modifications of immune presentation of endogenous and self-proteins induce the inflammatory responses that triggers systemic clinical and biological signs. For example, abacavir (a reverse transcriptase inhibitor) binds non-covalently to the HLA groove modifying the self-peptide repertoire presented in the groove. Another drug is carbamazepine, an anticonvulsant medication, and susceptibility to carbamazepine reactivity is found in patients with HLA-B*15:02 variant. The

mechanism of T-cell activation induced by carbamazepine is supposed to be the same as that described for abacavir. Allopurinol induces reactions in patients with HLA-B*58:01.These HLA associations probably share a similar T-cell immune activation mechanism that depends on the culprit drug/HLA interaction and HLA/peptide repertoire presentation. The identification of risk-associated HLA variants opens new avenues for physicians by using patient selection based on HLA typing.

In summary, DRESS is a systemic drug reaction in which eosinophil activation is driven by an immunological response directed against viral reactivation and a culprit drug. Eosinophils infiltrate organs in response to chemokines, in synergy with IL-5, and toxic granule protein release represents a key factor of tissue damage.

Prevalence of DRESS may be estimated based on the number of doses of agents administered, and has been estimated to have an overall population risk of between 1 in 1000 and 1 in 10,000 drug exposures. However, the total overall prevalence across all agents is not known, as some agents may only present with DRESS rarely. The prognosis and recovery are generally good, although death is possible in the acute phase of the disease (mortality rate of about 10% in some studies, but lower in others). While scoring systems exist for the diagnosis of DRESS, and international registries such as RegiSCAR were created to track cases, the creation of a specific ICD code for DRESS would enhance ability to track it, along with the ability to assess pharmacogenetics, detect new culprit agents, and assess important immunopathogenic questions regarding etiology, impact of genetic background, and potential for cross-reactive medications after the development of DRESS.

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TABULAR MODIFICATIONS

C93 Monocytic leukemia

C93.1 Chronic myelomonocytic leukemia
Add Code also, if applicable, eosinophilia (D72.18)

D72 Other disorders of white blood cells

D72.1 Eosinophilia

New code D72.10 Eosinophilia, unspecified

New subcategory D72.11 Hypereosinophilic syndrome [HES]

New code D72.110 Idiopathic hypereosinophilic syndrome [IHES]

New code D72.111 Lymphocytic Variant Hypereosinophilic Syndrome [LHES]
Add Lymphocyte Variant Hypereosinophilia

Add Code also if applicable any associated lymphocytic neoplastic disorder

New code D72.112 Myeloid hypereosinophilic syndrome [MHES]
Add Eosinophilia associated with myeloid or lymphoid neoplasms and abnormalities of FGFR1, JAK2, PDGFRA, or PDGFRB

Add Code also if applicable any associated neoplastic disorder, such as:
Add Chronic eosinophilic leukemia (C94.8-)
Add (Chronic) myeloproliferative disease (D47.1)

New code D72.118 Other hypereosinophilic syndrome
Add Episodic Angioedema with Eosinophilia
Add Gleich's Syndrome

New code D72.119 Hypereosinophilic syndrome [HES], unspecified

New code	D72.12 Drug rash with eosinophilia and systemic symptoms syndrome
Add	DRESS syndrome
Add	Use additional code for adverse effect, if applicable, to identify drug (T36-T50 with fifth or sixth character 5)
New code	D72.18 Eosinophilia in diseases classified elsewhere
Add	Code first the underlying disease, such as:
Add	Chronic myelomonocytic leukemia (C93.1-)
New code	D72.19 Other eosinophilia
Add	Familial eosinophilia
Add	Hereditary eosinophilia

INDEX MODIFICATIONS

Leukemia, leukemic C95.9-

Add - chronic eosinophilic (see also Syndrome, hypereosinophilic, myeloid) C94.8-

Immunodeficiency Status

The American Academy of Pediatrics (AAP) proposes that new codes be created to indicate when a patient is immunocompromised. This topic was presented at past Coordination and Maintenance meeting (March 2017, March 2018 & March 2019). The Academy submits this revised proposal, based on stakeholder comments, for reconsideration.

An immunocompromised status is a state in which a person's immune system is immunosuppressed or weakened. Individuals who are immunocompromised are less capable of battling infections because the immune system response is not functioning properly. Treating a patient who is immunocompromised poses more risks and challenges, therefore, it is very important to be able to identify a patient with this status.

These individuals are more prone to serious infections, opportunistic infections and other types of complications. A patient may be immunocompromised due to a specific clinical condition such as HIV, AIDS, certain cancers and genetic disorders. There are also external factors such as treatment with certain medications or exposure to radiation therapy, or a combination of both clinical conditions and external factors.

There are circumstances where a patient may be immune competent because of improvement of an underlying condition that can affect the immune system, but become immunocompromised because of an acute illness, new treatment or medication, e.g. bone marrow transplant with a fever.

A patient whose immune system is suppressed because of illness or external factors generally requires greater resource utilization. These patients are at increased risk because of fevers, non-environmental hypothermia, or injury thus requiring more interventions such as laboratory testing and medications than those with normally functioning immune systems.

Clinicians routinely document in the medical record when a patient's immune system may be compromised by using terms such as "immunodeficiency," "immunosuppressed" or "immunocompromised." Conditions within category D80-D89, Certain disorders involving the immune mechanism, do not indicate that a patient is immunocompromised and are generally specific to the type of immune deficiency. The codes D84.8, Other specified immunodeficiencies and D89.89, Other specified disorders involving the immune mechanism, not elsewhere classified, are not specific enough to capture the details as to why a patient's immune system status is compromised and which places the patient at greater health risks.

The following tabular modifications are being proposed:

TABULAR MODIFICATIONS

D84 Other immunodeficiencies

D84.8 Other specified immunodeficiencies

New code D84.81 Immunodeficiency due to conditions classified elsewhere
Add Code first underlying condition, such as:

Add chromosomal abnormalities (Q90-Q99)
Add diabetes mellitus (E08-E13)
Add malignant neoplasms (C00-C96)

Add **Excludes1**: combined immunodeficiencies (D81.-)
Add common variable immunodeficiency (D83.-)
Add defects in the complement system (D84.1)
Add human immunodeficiency virus [HIV] disease (B20)
Add immunodeficiency associated with other major defects (D82.-)
Add immunodeficiency with predominantly antibody defects (D80.-)
Add lymphocyte function antigen-1 [LFA-1] defect (D84.0)

New sub-subcategory D84.82 Immunodeficiency due to drugs and external ~~causes~~

New code D84.821 Immunodeficiency due to drugs
Add Immunodeficiency due to (current or past) medication

Add Use additional code for adverse effect if applicable, to identify adverse effect of drug (T36-T50 with fifth or six character 5)

Add Use additional code, if applicable, for associated long term (current) drug therapy drug or medication such as:
Add Long term (current) drug therapy systemic steroids (Z79.52)
Add Other long term (current) drug therapy (Z79.899)

New code D84.822 Immunodeficiency due to external causes
Add Code also radiological procedure and radiotherapy (Y84.2)

Add Use additional code for external cause such as:
exposure to ionizing radiation (W88)
other contact with and (suspected)exposures hazardous to health (Z77)

New code D84.89 Other immunodeficiencies

D84.9 Immunodeficiency, unspecified
Add Immunocompromised NOS
Add Immunodeficient NOS
Add Immunosuppressed NOS

Chapter 19

Injury, poisoning and certain other consequences of external causes (S00-T88)

Poisoning by, adverse effects of and underdosing of drugs, medicaments and biological substances (T36-T50)

Excludes2: abuse and dependence of psychoactive substances (F10-F19)
abuse of non-dependence-producing substances (F55.-)
drug reaction and poisoning affecting newborn (P00-P96)
Add immunodeficiency due to drugs (D84.821)
pathological drug intoxication (inebriation) (F10-)

Intracranial Hypotension and Cerebrospinal Fluid Leak

Previous proposals to create new codes for intracranial hypotension have been presented at the March 2018 and March 2019 ICD-10 Coordination and Maintenance Committee meeting. This proposal shows some modifications to the March 2019 proposal, based on comments from that prior presentation. Some information from that proposal is duplicated here, but the reader is directed to see the previous proposal for full details.
[Link: https://www.cdc.gov/nchs/data/icd/Topic-packet-March-2019-Part-2Vs3.pdf, see p. 30]

Intracranial hypotension results from a loss of cerebrospinal fluid volume, and it is an under-recognized and under-diagnosed central nervous system disorder. Intracranial hypotension is most often associated with a cerebrospinal fluid leak at the level of the spine and is not causally associated with cerebrospinal fluid leaks arising from the skull base.

The causes of intracranial hypotension include:

- Spontaneous cerebrospinal fluid leaks at the level of the spine (most under-recognized category)
- Iatrogenic holes or defects in the spinal dura from:
  - Intentional diagnostic or therapeutic spinal dural punctures
  - Inadvertent spinal dural puncture during epidural injection procedures
  - Inadvertent or intentional spinal durotomies during spinal or other surgeries
- Over-drainage of cerebrospinal fluid shunting devices
- Traumatic spinal dural tears or defects resulting in spinal cerebrospinal fluid leaks

It is clinically important to differentiate between spontaneous CSF leaks and other CSF leaks, and also between cranial CSF leaks and spinal CSF leaks.

TABULAR MODIFICATIONS

Note: changes below show the changes proposed in March 2019 at G96.0 and G96.1, along with new or modified changes being proposed now, in bold. Other changes proposed in March 2019, including those directly for intracranial hypotension, are not shown, as there are no further changes being proposed to those codes.

G96 Other disorders of central nervous system

G96.0 Cerebrospinal fluid leak

Add (modified Sep. 2019) Code also if applicable:
Add (modified Sep. 2019) head injury (S00.- to S09.-)
Add (modified Sep. 2019) intracranial hypotension (G96.81-)
Excludes1: cerebrospinal fluid leak from spinal puncture (G97.0)

New code G96.00 Cerebrospinal fluid leak, unspecified

New code G96.01 Cranial cerebrospinal fluid leak, spontaneous
Cerebrospinal fluid leak from skull base
CSF otorrhea
CSF rhinorrhea
otorrhea due to CSF leak
rhinorrhea due to CSF leak

New code G96.02 Spinal cerebrospinal fluid leak, spontaneous
Cerebrospinal fluid leak from spine

New code (Sep. 2019) G96.08 Other cranial cerebrospinal fluid leak
Other cranial CSF leak

New code (Sep. 2019) G96.09 Other spinal cerebrospinal fluid leak
Other spinal CSF leak

G96.1 Disorders of meninges, not elsewhere classified

G96.11 Dural tear

Add (modified Sep. 2019) Code also intracranial hypotension, if applicable (G96.81-)

Pediatric Feeding Disorder

Pediatric Feeding Disorder (PFD) can be described as impaired oral intake that is not age-appropriate, and is associated with medical, nutritional, feeding skill, and/or psychosocial dysfunction. Regardless of whether PFD is associated with problems in body function and structure, individuals with PFD experience limitations. These may include not being able to feed effectively which leads to participation restrictions or modifications in childcare, school and other environments that involve mealtime interactions. Pediatric feeding disorders can profoundly impact a child's physical, social, emotional, and/or cognitive function, and increase caregiver stress.

In children, feeding occurs in the context of the caregiver-child dyad. A disruption in any of these systems places a child at risk for a feeding disorder and associated complications. Often, more than one system is disrupted, contributing to the development and persistence of pediatric feeding disorders. Hence, effective assessment and treatment of pediatric feeding disorders require the involvement of multiple disciplines.

PFD is most frequently seen in young children, but can affect children of all ages. Age-appropriate feeding was chosen as the reference standard for oral intake. This refers to the progressive acquisition of feeding skills in the infant and child to enable progression from breast or bottle feeding to self-feeding a variety of age-appropriate table foods. Children with developmental delays may have feeding behaviors that are appropriate for their level of development but not their age; these children may have PFD if this is associated with activity limitation and/or participation restriction.

Four important domains underlie PFD: medical, nutritional, feeding skills and psychosocial.
For each domain, impairments that can lead to PFD, and potential interactions among health conditions, personal factors, and environmental factors, resulting in disability are discussed. PFD, in turn can cause dysfunction in each of the domains.

Symptoms must be present daily for at least 2 weeks; acute illness, once resolved, is associated with spontaneous improvement in feeding. Consistent with accepted norms, PFD can be classified into acute (< 3 months' duration) and chronic (≥ 3 months' duration). Acute PFD may be triggered by medical conditions, such as esophagitis or a choking episode. Chronic PFD has myriad causes.

By promoting usage of a consistent, comprehensive, interdisciplinary terminology that encompasses both physiologic impairment and function, this definition has the potential to facilitate interdisciplinary collaboration; promote educational curricula to train practitioners; promote research investigating best practices; allow comparison of outcomes between studies and clinical programs; partner with policymakers, so that PFD is a qualifying diagnosis for early intervention services under part C of the Individuals with Disabilities Education Act; and stimulate changes to the ICD-10-CM codes to allow for better tracking and care for these patients. These criteria aim to create a platform for change to ensure infants and children with PFD receive the best care possible and that the families receive the broadest community support available.

A single diagnostic term and code number would enable practitioners and researchers to better characterize the needs of heterogeneous patient populations, facilitate inclusion of all relevant disciplines in treatment planning, and allow the health-care team to use the common, precise terminology necessary to advance clinical practice and research.

The American Academy of Pediatrics, with support from its Committee on Nutrition and Section on Gastroenterology Hepatology and Nutrition are requesting the following tabular updates:

TABULAR MODIFICATION

F98 Other behavioral and emotional disorders with onset usually occurring in childhood and adolescence

Excludes2: breath-holding spells (R06.89)
gender identity disorder of childhood (F64.2)
Kleine-Levin syndrome (G47.13)
obsessive-compulsive disorder (F42-)
sleep disorders not due to a substance or known physiological condition (F51.-)

F98.2 Other feeding disorders of infancy and childhood Excludes1: feeding difficulties (R63.3)

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

F98.21 Rumination disorder of infancy

New code F98.22 Pediatric feeding disorder (PFD)

F98.29 Other feeding disorders of infancy and early childhood

Pulmonary Eosinophilic Diseases

The pulmonary eosinophilic diseases were presented at the March 2019 meeting. This proposal is modified based on comments, including a change to use the subcategory J82.8. For clarity, eosinophilia generally means elevated blood levels of eosinophils, while pulmonary eosinophilia refers to infiltration of eosinophils into the lungs. Eosinophilia and pulmonary eosinophilia may be present at the same time, or may not be, so it has been proposed that the Excludes1 note at D72.1 be changed to an Excludes2 note, to enable use of codes that could overlap. Also, there is a separate proposal now to expand at D72.1, which would create a number of specific codes that could potentially coexist with pulmonary eosinophilic diseases, which adds further impetus for changing this note. The text below is from the March 2019 meeting.

New codes for certain pulmonary eosinophilic diseases have been requested by the American Partnership for Eosinophilic Disorders (APFED), a 501(c)3 non-profit patient advocacy group, and the International Eosinophil Society, Inc. (IES), an organization of scientists and clinicians interested in the eosinophil and its roles in health and disease.

Eosinophilic Pneumonia

There are 2 types of eosinophilic pneumonia - acute and chronic. While both are characterized by eosinophil invasion of the lung tissue, they are quite different from one another and are described below.

Acute Eosinophilic Pneumonia

Acute eosinophilic pneumonia (AEP) is classified as a form of eosinophilic lung disease, one of a large group of interstitial lung diseases. AEP is different from chronic eosinophilic pneumonia (CEP), which is marked by slower progression, lack of progression to acute respiratory failure, frequent relapses and is often associated with asthma.

AEP is characterized by rapidly progressive respiratory failure, usually of less than one-month duration, often leading to the need for intensive care and mechanical ventilation. Approximately two-thirds of individuals may require mechanical ventilation. Chest imaging usually shows abnormalities throughout the lung fields. Finding eosinophils in lung washings from bronchoalveolar lavage (BAL) is key in the diagnosis of AEP. BAL fluid in individuals with AEP reveals abnormally high levels of eosinophils (greater than 25%). Associated symptoms are nonspecific and can include fever, cough, difficulty breathing (dyspnea) and chest pain. Less common symptoms include fatigue, muscle pain (myalgia), joint aches, and abdominal discomfort or pain.

While respiratory failure is a common feature of AEP, this disease is quite responsive to therapy with corticosteroids, making recognition and diagnosis critical. Within the medical literature, the dose and duration of corticosteroid therapy has varied greatly, with a recent series suggesting that just two-weeks of treatment is sufficient. Following treatment, long term prognosis is excellent.

The causes of AEP are not entirely clear, though it has been noted to be associated with new onset of cigarette smoking or an increase in smoking. Other case series of AEP in the literature include a series in U.S military personnel in Iraq and another in 9/11 rescue workers exposed to dust from the World Trade Center. Men are more commonly affected than women. The cause of AEP is unknown (idiopathic).

Researchers believe that AEP develops due to an unidentified triggering agent that causes the body to produce extra eosinophils and recruit them specifically to the lung. The exact reason for the overproduction and accumulation of eosinophils is unknown. Additional reports in the medical literature have linked some cases of AEP to the use of a number of medications. Drug-induced cases have been linked to minocycline, daptomycin, and velafaxine, an antidepressant, and others (www.pneumotox.com). Several environmental factors including occupational exposures have been shown to trigger AEP including dust and smoke. It is unlikely that a single environmental factor causes AEP. Most likely, multiple factors are necessary for the development of the disorder, with association of a triggering condition in a predisposed individual. The triggering factor in AEP can be different from one individual to another.

Chronic Eosinophilic Pneumonia

Chronic Eonsinophilic Pneumonia (CEP) has been a well-established distinct entity in pulmonary medicine for decades, yet it does not have its own diagnostic ICD code. CEP is different from acute eosinophilic pneumonia (AEP), which is marked by rapid onset, the absence of asthma, a greater potential for acute respiratory failure and no relapse following treatment. As the name implies, CEP is a more chronic, indolent condition than AEP and is characterized by progressive shortness of breath and abnormalities on chest imaging (CT scan and chest x-ray) that often are located in the periphery of the lungs. Bronchoscopy with the finding of increased eosinophils on airway washings confirms the diagnosis. Eosinophils also increase in the bloodstream (peripheral blood eosinophilia). Common symptoms include shortness of breath (dyspnea), cough, fatigue, night sweats, low grade fevers, and unintended weight loss. Symptoms can be very similar to those seen in asthma, including the development of wheezing. In fact, a diagnosis of asthma may precede the development of CEP.

CEP is generally treated with a prolonged course of corticosteroids lasting several months or more, with relapses common during attempts to taper corticosteroids. Like many eosinophilic diseases, the exact inciting causes are unknown. Researchers believe that CEP may develop due to an unidentified, nonspecific triggering agent that causes the body to produce excess eosinophils. The exact reason for the overproduction and accumulation of eosinophils is unknown. CEP tends to recur and many individuals will relapse at some point, especially when therapy is not maintained. A relapse can occur as much as 10 years or more after the initial episode. Some individuals eventually develop severe asthma. In some cases, individuals with CEP have developed a related disorder known as Churg-Strauss syndrome, now renamed eosinophilic granulomatosis with polyangiitis, or EGPA, suggesting that there may be an overlap between these two disorders. The disorder can occur in individuals of any age, but is extremely rare in childhood. The peak incidence is during the fifth decade. CEP occurs twice as often in women than men.

Eosinophilic Asthma

Currently, diagnostic codes classify asthma based on severity, persistence, and the presence of exacerbation. However, our state of the art understanding of the immune mechanisms underlying asthma has defined a subset of asthma that is characterized by the presence of a certain level of eosinophils in the circulation and in the airways. In the United States, an estimated 25.7 million people have some form of asthma, with 15 percent of these people having severe asthma that is difficult to control with standard medications. Eosinophilic asthma is considered a leading cause of severe asthma, affecting 50 to 60 percent of people with the severe form of the disease.

Eosinophilic asthma can be difficult to treat and may have a detrimental effect on an individual's quality of life. It does not generally respond well to inhaled corticosteroids, even at high doses. Until recently, oral corticosteroids were the standard treatment for eosinophilic asthma. Although many people respond well to these medications, they are not generally successful at controlling this disease long term. Some patients become dependent on the oral corticosteroids. A number of biologic treatments have been approved by the US FDA to treat eosinophilic asthma, and can reduce the frequency of eosinophilic asthma attacks. Patients with eosinophilic asthma have been found in several pivotal clinical trials to benefit from these therapies that specifically target eosinophils by blocking a molecule known as IL-5 or its receptor. In fact, these drugs reduce asthma exacerbations regardless of the cause and are oral steroid sparing. Their benefit on lung function is modest. With these circumstances shaping the current scientific, clinical, and pragmatic landscape, eosinophilic asthma has now become a well-accepted and widespread diagnostic entity.

The connection between eosinophils and asthma has been recognized and cited in research since 1889. Eosinophilic asthma is more prominent in adults. Adult asthma usually affects more women than men, but eosinophilic asthma affects men and women at about the same rate. As the level of eosinophils increases, inflammation and other symptoms of asthma become more severe. The primary symptom is shortness of breath rather than the more traditional wheezing in non-eosinophilic asthma. People with eosinophilic asthma may also experience chronic sinus infections, nasal polyps, eosinophilic otitis media, an inner ear infection, and aspirin-exacerbated respiratory disease. The symptoms of eosinophilic asthma differ from classic asthma and, in fact, more closely resemble those of chronic pulmonary obstructive disorder (COPD).

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TABULAR MODIFICATIONS

D72 Other disorders of white blood cells

D72.1 Eosinophilia

Delete ~~Excludes1: Löffler's syndrome (J82)~~
~~pulmonary eosinophilia (J82)~~

Add Excludes2: Löffler's syndrome (J82.89)
pulmonary eosinophilia (J82.-)

J82 Pulmonary eosinophilia, not elsewhere classified

Delete ~~Allergic pneumonia~~
Delete ~~Eosinophilic asthma~~
Delete ~~Eosinophilic pneumonia~~
Delete ~~Löffler's pneumonia~~
Delete ~~Tropical (pulmonary) eosinophilia NOS~~
Delete Excludes1: ~~pulmonary eosinophilia due to aspergillosis (B44.-)~~
Delete ~~pulmonary eosinophilia due to drugs (J70.2-J70.4)~~
Delete ~~pulmonary eosinophilia due to specified parasitic infection (B50-B83)~~
Delete ~~pulmonary eosinophilia due to systemic connective tissue disorders (M30-M36)~~
Add Excludes2: pulmonary eosinophilia due to aspergillosis (B44.-)
Add pulmonary eosinophilia due to drugs (J70.2-J70.4)
Add pulmonary eosinophilia due to specified parasitic infection (B50-B83)
Add pulmonary eosinophilia due to systemic connective tissue disorders (M30-M36)

New subcategory J82.8 Pulmonary eosinophilia, not elsewhere classified

New code J82.81 Chronic eosinophilic pneumonia
Eosinophilic pneumonia, NOS

New code J82.82 Acute eosinophilic pneumonia

New code J82.83 Eosinophilic asthma

New code J82.89 Other pulmonary eosinophilia, not elsewhere classified
Add Allergic pneumonia
Add Löffler's pneumonia
Ad Tropical (pulmonary) eosinophilia NOS

INDEX MODIFICATIONS

Eosinophilia …
Revise - infiltrative – see Eosinophilia, pulmonary ~~J82~~
Revise - Löffler's J82.89
Revise - pulmonary NEC J82.89
Add - - acute J82.82
Add - - asthmatic J82.83
Add - - chronic J82.81
Revise - tropical (pulmonary) J82.89

Infiltrate, infiltration

- lung R91.8
Revise - - eosinophilic – see Eosinophilia, pulmonary ~~J82~~
- pulmonary R91.8
Revise - - with eosinophilia – see Eosinophilia, pulmonary ~~J82~~

Löffler's
Revise - eosinophilia J82.89
Revise - pneumonia J82.89
Revise - syndrome (eosinophilic pneumonitis) J82.89

Pneumonia
Revise - allergic ~~(eosinophilic)~~ (see also Pneumonitis, hypersensitivity) J82.89
- broncho-, bronchial …
Revise - - allergic ~~(eosinophilic)~~ (see also Pneumonitis, hypersensitivity) J82.89
Revise - eosinophilic J82.81
Add - - acute J82.82
Add - - chronic J82.81

Pneumonitis …
Revise - eosinophilic J82.81
Add - - acute J82.82
Add - - chronic J82.81

Syndrome …
Revise - PIE (pulmonary infiltration with eosinophilia) (see also Eosinophilia, pulmonary) J82.89
Revise - Weingarten's (tropical eosinophilia) J82.89

Sickle Cell Disease

The American Academy of Pediatrics (AAP) requests tabular modifications for sickle cell disorders to identify patients without major complications but who are in crisis. This proposal was presented at the September 2016 and March 2018 Coordination & Maintenance (C&M) meeting. However, based on additional public comments, the proposal has been revamped and being presented for reconsideration. N bn

Currently in ICD-10-CM, patients with sickle cell vasoocclusive crisis not associated with acute chest syndrome or splenic sequestration are coded as "with crisis, unspecified". In the majority of these encounters, the vasoocclusive pain crisis is the problem that requires medical intervention as other major complications may not be present. Therefore, the vasoocclusive pain is considered inherent and not a manifestation.

There is currently no adequate means to track patients with other types of complications in addition to acute chest syndrome and splenic sequestration. Other complication include but not limited to acute gall bladder involvement, priapism or fever. Cerebral infarcts are a major complication in patients with sickle cell disease. According to the CDC, about 10% of children with sickle cell disease will have a symptomatic stroke.

In addition, within the current code set there is only one code to identify patients with sickle cell-thalassemia, D57.4. However, there are two distinct types of sickle cell-thalassemia, sickle cell-thalassemia beta zero (HbS-β^0) and sickle cell-thalassemia beta plus (HbS-β^+). They are clinically very different.

HbS-β^0 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.

To be able to better track each of these unique conditions, the Academy request new codes and also request previously proposed codes to the sickle cell categories be added as shown in this revised proposal.

TABULAR MODIFICATIONS

D57 Sickle-cell disorders

Use additional code for any associated fever (R50.81)

Excludes1: other hemoglobinopathies (D58.-)

D57.0 Hb-SS disease with crisis

Revise — Sickle-cell disease ~~NOS~~ with crisis

Hb-SS disease with vasooclusive pain

D57.00 Hb-SS disease with crisis, unspecified

Add — Hb-SS disease with (painful) crisis NOS

Add — Hb-SS disease with vasoocclusive pain NOS

D57.01 Hb-SS disease with acute chest syndrome

D57.02 Hb-SS disease with splenic sequestration

New code — D57.03 Hb-SS disease with cerebral vascular involvement

Add — Code also, if applicable: cerebral infarction (I63)

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

Add — Use additional code to identify complications, such as:

Add — cholelithiasis (K80)

Add — priapism (N48.32)

D57.2 Sickle-cell/Hb-C disease

Hb-SC disease

Hb-S/Hb-C disease

D57.20 Sickle-cell/Hb-C disease without crisis

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

D57.211 Sickle-cell/Hb-C disease with acute chest syndrome

D57.212 Sickle-cell/Hb-C disease with splenic sequestration

New code D57.213 Sickle-cell/Hb-C disease with cerebral vascular involvement

Add Code also, if applicable: cerebral infarction (I63)

New code D57.218 Sickle-cell/Hb-C disease with crisis with other specified complication

Add Use additional code to identify complications, such as:

Add cholelithiasis (K80)

Add priapism (N48.32)

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

Sickle-cell/Hb-C disease with (painful) crisis NOS

Add Sickle-cell/Hb-C disease with vasoocclusive pain NOS

D57.4 Sickle-cell thalassemia

Sickle-cell beta thalassemia

Thalassemia Hb-S disease

D57.40 Sickle-cell thalassemia without crisis

Microdrepanocytosis

Sickle-cell thalassemia NOS

Revise D57.41 Sickle-cell thalassemia, <u>unspecified</u>, with crisis

Add Sickle-cell thalassemia with (painful) crisis NOS

Revise Sickle-cell thalassemia with vasoocclusive pain <u>NOS</u>

Revise D57.411 Sickle-cell thalassemia, <u>unspecified,</u> with acute chest syndrome

Revise D57.412 Sickle-cell thalassemia, <u>unspecified</u>, with splenic sequestration

New code	D57.413 Sickle-cell/Hb-C disease with cerebral vascular involvement
Add	Code also, if applicable cerebral infarction (I63)
New code	D57.418 Sickle-cell thalassemia with crisis with other specified complication
Add	Use additional code to identify complications, such as:
Add	cholelithiasis (K80)
Add	priapism (N48.32)
Revise	D57.419 Sickle-cell thalassemia, unspecified, with crisis,
Add	Sickle-cell thalassemia with (painful) crisis NOS
Add	Sickle-cell thalassemia with vasoocclusive pain
Add	Sickle-cell thalassemia with crisis NOS
New code	**D57.42 Sickle cell-thalassemia beta zero without crisis**
Add	**Sickle cell β^0 without crisis**
Add	**HbS-β^0 without crisis**
New sub-category	**D57.43 Sickle-cell thalassemia beta zero with crisis**
New code	**D57.431 Sickle cell-thalassemia beta zero with acute chest syndrome**
New code	**D57.432 Sickle-cell thalassemia beta zero with splenic sequestration**
New code	**D57.433 Sickle cell-thalassemia beta zero with cerebral vascular involvement**
Add	**Code also, if applicable cerebral infarction (I63)**

New code **D57.438 Sickle cell-thalassemia beta zero with crisis with other specified complication**

Add **Use additional code to identify complications, such as:**

Add **cholelithiasis (K80)**

Add **priapism (N48.32)**

New code **D57.439 Sickle-cell thalassemia beta zero with crisis, unspecified**

Add **Sickle-cell thalassemia beta zero with (painful) crisis NOS**

Add **Sickle-cell thalassemia beta zero with vasoocclusive pain**

New code **D57.44 Sickle cell-thalassemia beta plus without crisis**

Add **Sickle cell β^+ without crisis**

Add **HbS-β^+ without crisis**

New sub-category **D57.45 Sickle cell-thalassemia beta plus with crisis**

New code **D57.451 Sickle cell-thalassemia beta plus with acute chest syndrome**

New code **D57.452 Sickle-cell thalassemia beta plus with splenic sequestration**

New code **D57.453 Sickle cell-thalassemia beta plus with cerebral vascular involvement**

Add **Code also, if applicable cerebral infarction (I63)**

New code **D57.458 Sickle cell-thalassemia beta plus with crisis with other specified complication**

Add **Use additional code to identify complications, such as:**

Add **cholelithiasis (K80)**

Add **priapism (N48.32)**

New code **D57.459 Sickle-cell thalassemia beta plus with crisis, unspecified**

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

Add **Sickle-cell thalassemia beta plus with vasoocclusive pain**

D57.8 Other sickle-cell disorders

Hb-SD disease

Hb-SE disease

D57.81 Other sickle-cell disorders with crisis

D57.811 Other sickle-cell disorders with acute chest syndrome

D57.812 Other sickle-cell disorders with splenic sequestration

New code D57.813 Other sickle-cell disorders with cerebral vascular involvement

Add Code also, if applicable: cerebral infarction (I63)

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

Add Use additional code to identify complications, such as:

Add cholelithiasis (K80)

Add priapism (N48.32)

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

Other sickle-cell disorders with crisis NOS

Add Other sickle-cell disorders with vassooclusive pain NOS

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Stage 3 Chronic Kidney Disease

Kidney disease is often a silent disease, and is asymptomatic just prior to kidney failure and the need for kidney replacement therapy. Chronic Kidney Disease (CKD) is therefore diagnosed on the basis of laboratory tests.

The National Kidney Foundation's Kidney Disease Outcome Quality Initiative (NKFKDOQI) published the Guideline for Chronic Kidney Disease (CKD): Evaluation, Classification, and Stratification in 2002. The KDOQI guideline defined CKD as an abnormality of kidney structure or function that is present for three or more months, regardless of cause or specific clinical presentation and proposed a staging system based on the level of glomerular filtration rate (GFR). The NKFKDOQI guideline has been well accepted by the medical and public health communities and has led to much change in clinical practice within the primary care, nephrology and other specialty communities. ICD-9-CM codes were revised to reflect this definition and staging system in Oct 2005 and have been widely used. The changes were carried over into ICD-10-CM, which became effective October 1, 2015.

The National Kidney Foundation, supported by the Renal Physicians Association and the American Society of Nephrology propose the addition of two new codes at N18.3, Chronic Kidney Disease Stage 3 (moderate), to capture additional stage 3 detail.

TABULAR MODIFICATIONS

N18 Chronic kidney disease (CKD)

N18.1 Chronic kidney disease, stage 1

N18.2 Chronic kidney disease, stage 2 (mild)

New subcategory N18.3 Chronic kidney disease, stage 3 (moderate)

New code N18.30 Chronic kidney disease, stage 3 unspecified

New code N18.31 Chronic kidney disease, stage3a

New code N18.32 Chronic kidney disease, stage 3b

Tarlov Cyst

This topic was originally presented at the September 2017 Coordination and Maintenance (C&M) meeting. The requestor proposed a new code for Tarlov cyst at category G54, Nerve root and plexus disorders. Based on public comments received, input from various clinical subject matter experts, as well consulting with the World Health Organization (WHO), the revised proposal is being presented for consideration.

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. Individuals may also be affected by multiple cysts of varying size in other sections of the spine, with less prevalence (Cervical 3%, Thoracic and Lumbar 6%). Eleven percent of reported cases have cysts imaged in more than one section of the spine.

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.

Diagnosis of Tarlov or perineural cyst is best confirmed by spine MRI imaging. In some cases, a diagnosis of a Tarlov cyst is made incidentally through MRI scan investigation undertaken for other reasons including pelvic pain, hip pain, abdominal pain, external genitalia and rectal pain, which can also be symptoms of Tarlov cysts. They are described as extradural meningeal cysts arising from the meninges.

WHO has provided guidance that this condition would be appropriately classified in ICD-10 and therefore in ICD-10-CM, at subcategory G96.1, Disorders of meninges, not elsewhere classified. The following proposed new code to identify this condition in ICD-10-CM aligns with and is consistent with the placement in ICD-11.

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National Organization for Rare Disorders (NORD) on Tarlov cysts. https://rarediseases.org/rare-diseases/tarlov-cysts/

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TABULAR MODIFICATIONS

	G96	Other disorders of central nervous system
New subcategory	G96.19	Other disorders of meninges, not elsewhere classified
New code	G96.191	Perineural cyst
Add		Tarlov cyst
Add		Sacral nerve root cyst
Add		Lumbar nerve root cyst
Add		Thoracic nerve root cyst
Add		Cervical nerve root cyst
New code	G96.198	Other disorders of meninges, not elsewhere classified

X-Linked Myotubular Myopathy and Other Congenital Myopathies

The congenital myopathies are a group of muscle disorders with onset generally at birth or in infancy. These typically cause weakness, with poor muscle tone or floppiness, difficulty breathing or feeding, and lagging behind in meeting normal developmental milestones such as turning over or sitting up (NINDS 2019). The congenital myopathies have traditionally been grouped broadly based on histopathological findings from muscle biopsy. These have specifically included three main groups, based on the presence of cores (core myopathy), central nuclei (centronuclear myopathy), or nemaline bodies (nemaline myopathy) (Ravenscroft 2018). However, there are also two other types, congenital fiber-type disproportion myopathy, and myosin storage myopathy (the latter also called hyaline body myopathy) (Cassandrini 2017; NINDS 2019; O'Neill 2010). Discovery of the genetic basis for various congenital myopathies, and work toward development of genetic therapies, are ongoing areas of research and development (Cassandrini 2017; Ravenscroft 2018).

Nemaline myopathy is the most common congenital myopathy (estimated incidence 1 in 50,000) (NINDS 2019; NLM 2015b). Infants usually have problems with breathing and feeding. Weakness can be throughout the body, but particularly involves proximal muscles such as the face, neck, trunk, and upper arms and thighs. Later, some skeletal problems may arise, such as scoliosis (curvature of the spine). In general, the weakness does not worsen during life. There are six types of nemaline myopathy, which are in order of decreasing severity: severe congenital, Amish, intermediate congenital, typical congenital, childhood-onset, and adult-onset. The more severe types generally have earlier age of onset, and the most severe can cause death in early childhood (NINDS 2019; NLM 2015b). Joint contractures may occur, including arthrogryposis multiplex congenital, in the severe congenital and Amish types (NORD 2015). Nemaline myopathy is most often caused by mutations in one of two genes, NEB (about 50%) or ACTA1 (about 15-25%), although a number of other genes can also be associated (NLM 2015b; Ravenscroft 2018). Most cases are recessive, although some are dominant (NLM 2015b). Nemaline myopathy is also called rod body disease or rod myopathy (NLM 2015b).

Centronuclear myopathy has two subtypes, X-linked myotubular myopathy (XLMTM) and autosomal centronuclear myopathy. In the medical literature, the term centronuclear myopathy is generally used for the autosomal forms of the disorder (both dominant and recessive), while the term myotubular myopathy is generally used for the X-linked form, XLMTM (Cassandrini 2017; NORD 2016). Distinguishing between XLMTM and the autosomal forms is essential, as the symptoms are usually more severe in XLMTM (NORD 2016).

XLMTM is rare, and occurs almost exclusively in males (estimated incidence 1 in 50,000 newborn males). It is caused by mutations in the *MTM1* gene, located on the X chromosome, and which encodes a protein called myotubularin. Myotubularin is an enzyme thought to be involved in the development, maintenance and function of muscle cells in skeletal muscle tissue (NLM 2014; NORD 2016). In XLMTM, weakness and floppiness are so severe that a mother may notice reduced movements of the baby in her womb during pregnancy (NINDS 2019). Infants with XLMTM are typically born with severe muscle weakness and decreased muscle tone, with the majority requiring chronic mechanical ventilation from birth (Beggs 2018). One study found about 46% died in the first 18 months of life, but found a median survival of 29 months (McEntagart 2002). There is no

approved specific treatment for XLMTM, and disease management now is primarily supportive. Most surviving infants are hospitalized about 90 days in the US, and on average patients spend 35 to 47% of their first year of life in the hospital (Beggs 2018; NORD 2016). Of the patients that survive the infantile period, most are severely incapacitated, require ventilator support, have severely impaired motor skills, need feeding support (including gastrostomy), and few live beyond early adolescence (Beggs 2018; McEntagart 2002). Research is ongoing toward development of therapies for XLMTM. Genetic therapy in both a mouse and a dog model of XLMTM have been found to be effective to improve strength, as well as prolonging the usually shortened lifespan (Beggs 2018; Ravenscroft 2018).

Autosomal centronuclear myopathy is rare, but the prevalence is unknown (NINDS 2019; NLM 2015a). It usually begins in infancy or early childhood with weakness of the arms and legs, droopy eyelids, and problems with eye movements, and weakness often gets worse with time (NINDS 2019). Onset can also be later, even to early adulthood (NLM 2015a). Some people with centronuclear myopathy may have mild to severe breathing problems, related to the weakness of the muscles needed for breathing (NLM 2015a). Centronuclear myopathy is most often inherited in an autosomal dominant fashion, or more rarely as a recessive trait (Cassandrini 2017). It is most often caused by mutations in the *DNM2*, *BIN1*, or *TTN* gene (NLM 2015a). When centronuclear myopathy is caused by mutations in the *DNM2* gene, inheritance is autosomal dominant; rarely, *BIN1* gene mutations may also be autosomal dominant (NLM 2015a). Centronuclear myopathy caused by *TTN* gene mutations and most cases caused by *BIN1* gene mutations are autosomal recessive. Cases of centronuclear myopathy caused by other genes are typically autosomal recessive, although are autosomal dominant (NLM 2015a).

The core myopathies, central core disease and multiminicore disease, are both thought to be uncommon or rare, although the incidence is not known (NLM 2007a; NLM 2007b). Central core disease varies in the severity of problems and the degree of worsening over time; usually, there is mild floppiness in infancy, delayed milestones, and moderate limb weakness, which do not worsen much over time. Children with central core disease may have life-threatening reactions of malignant hyperthermia to general anesthesia (NINDS 2019; NLM 2007a). Multiminicore disease has several different subtypes, with most causing severe weakness of the limbs and scoliosis. Often breathing difficulties also occur. Some children have weakened eye movements. It also may be called minicore disease or multicore disease (NINDS 2019; NLM 2007b).

Other congenital myopathies, which include congenital fiber-type disproportion myopathy and myosin storage myopathy, are rare and do not have known incidence rates (NLM 2013; NLM 2016). Congenital fiber-type disproportion (CFTD) myopathy begins with floppiness, limb and facial weakness, and breathing problems (NINDS 2019). It is diagnosed based on a relative hypotrophy of type 1 muscle fibers compared to type 2 fibers seen on skeletal muscle biopsy; however, these findings are not specific but are found in many different myopathic and neuropathic conditions (Kniffin 2009). Thus, CFTD is a diagnosis of exclusion, and the term CFTD should be reserved for those cases in which no secondary cause can be found, while it has been recommended that the term fiber size disproportion be used for the nonspecific finding (Kniffin 2009). Myosin storage myopathy, or hyaline body myopathy, is characterized by myosin protein clumps being seen on muscle biopsy; the symptoms are quite variable (NINDS 2019; NLM 2013). Myosin storage myopathy may cause

those affected to start walking late, and some may have breathing problems. It generally worsens only slowly, if at all (NLM 2013).

Arthrogryposis multiplex congenita refers to the development of multiple joint contractures in more than one area prior to birth (GARD 2015). It is a physical symptom, which can be associated with many different medical conditions, including congenital myopathies (GARD 2015; NORD 2019). Thus, it should more appropriately be excluded from the congenital myopathies in a way that allows coding of both conditions if appropriate (i.e., using an Excludes2 note).

Specific codes are being proposed for nemaline myopathy, X-linked myotubular myopathy, other centronuclear myopathy, and for other congenital myopathy (other to include central core disease, multiminicore disease, congenital fiber-type disproportion, and myosin storage myopathy), as well as a code for unspecified congenital myopathy.

Requests for creation of specific ICD-10-CM codes for X-linked myotubular myopathy (XLMTM), centronuclear myopathy (CNM), and certain other congenital myopathies, have been received from Audentes Therapeutics, the Joshua Frase Foundation, and MTM-CNM Family Connection, Inc. This proposal has been modified from those original requests. As for many rare diseases, it is believed that XLMTM is underdiagnosed and that anticipated approval of new treatments will help increase disease awareness, screening, and diagnosis. It is also believed that early diagnosis and treatment of XLMTM may be able to improve patient outcomes, and patient and caregiver quality of life. Treatments that prolong life for XLMTM and other congenital myopathies will be expected thus also to increase the prevalence. Creation of specific ICD-10-CM codes would enable improved tracking of these disorders, as well as enabling easier assessment of cases where there are associated diagnoses present, and where particular procedures are necessary related to a specific diagnosis. This would be of particular benefit for a diagnosis such as XLMTM, which in many cases requires long hospitalizations and related care and surgical procedures.

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TABULAR MODIFICATIONS

G71.2 Congenital myopathies

Delete ~~Central core disease~~
Delete ~~Fiber-type disproportion~~
Delete ~~Minicore disease~~
Delete ~~Multicore disease~~
Delete ~~Myotubular (centronuclear) myopathy~~
Delete ~~Nemaline myopathy~~

Revise: Excludes2~~1~~: arthrogryposis multiplex congenita (Q74.3)

New code G71.20 Congenital myopathy, unspecifed

Add			Fiber-type disproportion
New code	G71.21		Nemaline myopathy
New sub-subcategory	G71.22		Centronuclear myopathy
New code		G71.220	X-linked myotubular myopathy
Add			Myotubular (centronuclear) myopathy
New code		G71.228	Other centronuclear myopathy
Add			Autosomal centronuclear myopathy
Add			Autosomal dominant centronuclear myopathy
Add			Autosomal recessive centronuclear myopathy
Add			Centronuclear myopathy, NOS
New code	G71.29		Other congenital myopathy
Add			Central core disease
Add			Congenital fiber-type disproportion
Add			Minicore disease
Add			Multicore disease
Add			Multiminicore disease

INDEX MODIFICATIONS

	Disease …
Add	- rod body G71.21
	Disproportion
Revise	- fiber-type G71.20
Add	- - congenital G71.29
	Myopathy …
Add	- hyaline body G71.29
Add	- myosin storage G71.29
Add	- rod (body) G71.21

ICD-10-CM TABULAR OF DISEASES - PROPOSED ADDENDA

All proposed effective October 1, 2020

B20 Human immunodeficiency virus [HIV] disease
Add Excludes1: acute HIV infection syndrome (Z21)
asymptomatic human immunodeficiency virus [HIV] infection status (Z21)
exposure to HIV virus (Z20.6)
inconclusive serologic evidence of HIV (R75)

B48 Other mycoses, not elsewhere classified
B48.4 Penicillosis
Add Talaromycosis

D21 Other benign neoplasms of connective and other soft tissue
D21.6 Benign neoplasm of connective and other soft tissue of trunk, unspecified
Revise Benign neoplasm of connective and other soft tissue **of** back NOS

D61 Other aplastic anemias and other bone marrow failure syndromes
Delete ~~Excludes1: neutropenia (D70.-)~~
Add Excludes2: neutropenia (D70.-)

Metabolic disorders (E70-E88)
Revise Excludes1: Ehlers-Danlos syndrome**s** (Q79.6-)

E88 Other and unspecified metabolic disorders
E88.0 Disorders of plasma-protein metabolism, not elsewhere classified
Revise ~~Excludes1: disorder of lipoprotein metabolism (E78.-)~~
Excludes2: disorder of lipoprotein metabolism (E78.-)

M35 Other systemic involvement of connective tissue
M35.7 Hypermobility syndrome
Revise Excludes1: Ehlers-Danlos syndrome**s** (Q79.6-)

G92 Toxic encephalopathy
Toxic encephalitis
Toxic metabolic encephalopathy
Revise Code first, if applicable, drug induced (T36-T50) or use (T51-T65) to identify toxic agent
Delete ~~Code first (T51-T65) to identify toxic agent~~

G93 Other disorders of brain
G93.2 Benign intracranial hypertension
Add Pseudotumor
Add Excludes1: obstructive hydrocephalus (G91.1)

H65 Nonsuppurative otitis media
Revise Use additional code, if applicable, to identify:

I46 Cardiac arrest
Delete ~~Excludes1: cardiogenic shock (R57.0)~~
Add Excludes2: cardiogenic shock (R57.0)

Acute upper respiratory infections (J00-J06)
Delete Excludes1: ~~influenza virus with other respiratory manifestations (J09.X2, J10.1, J11.1)~~

J00 Acute nasopharyngitis [common cold]
Add Excludes1: influenza virus with other respiratory manifestations (J09.X2, J10.1, J11.1)

J02 Acute pharyngitis
J02.9 Acute pharyngitis, unspecified
Add Excludes1: influenza virus with other respiratory manifestations (J09.X2, J10.1, J11.1)

J03 Acute tonsillitis
J03.9 Acute tonsillitis, unspecified
Add Excludes1: influenza virus with other respiratory manifestations (J09.X2, J10.1, J11.1)
J04 Acute laryngitis and tracheitis
Add Code also if present such as:
Add Influenza due to identified novel influenza A virus with other respiratory manifestations (J09.X2)
Add Influenza due to other identified influenza virus with other respiratory manifestations (J10.1)
Add Influenza due to unidentified influenza virus with other respiratory manifestations (J11.1)

J05 Acute obstructive laryngitis [croup] and epiglottitis
Add Code also if present such as:
Add Influenza due to identified novel influenza A virus with other respiratory manifestations (J09.X2)
Add Influenza due to other identified influenza virus with other respiratory manifestations (J10.1)
Add Influenza due to unidentified influenza virus with other respiratory manifestations (J11.1)

J06 Acute upper respiratory infections of multiple and unspecified sites
Add Excludes1: influenza virus with other respiratory manifestations (J09.X2, J10.1, J11.1)

J10 Influenza due to other identified influenza virus
Add Influenza A (non-novel)
Add Influenza B
Add Influenza C

J70 Respiratory conditions due to other external agents
J70.5 Respiratory conditions due to smoke inhalation
Delete ~~Smoke inhalation NOS~~
Add Code first smoke inhalation (T59.81-)
Delete ~~Excludes1:smoke inhalation due to chemicals, gases, fumes and vapors (J68.9)~~
Add Excludes2:smoke inhalation due to chemicals, gases, fumes and vapors (J68.9)

K56 Paralytic ileus and intestinal obstruction without hernia
K56.6 Other and unspecified intestinal obstruction
K56.60 Unspecified intestinal obstruction
Delete ~~Excludes1:intestinal obstruction due to specified condition-code to condition~~

K57 Diverticular disease of intestine
K57.0 Diverticulitis of small intestine with perforation and abscess
Delete ~~Diverticulitis of small intestine with peritonitis~~
K57.2 Diverticulitis of large intestine with perforation and abscess
Delete ~~Diverticulitis of colon with peritonitis~~
K57.4 Diverticulitis of both small and large intestine with perforation and abscess
Delete ~~Diverticulitis of both small and large intestine with peritonitis~~
K57.8 Diverticulitis of intestine, part unspecified, with perforation and abscess
Delete ~~Diverticulitis of intestine NOS with peritonitis~~

K62 Other diseases of anus and rectum
K62.7 Radiation proctitis
Revise Use additional code to identify the type of radiation ~~(W90.-)~~ W88.- or radiation therapy (Y84.2)

K80 Cholelithiasis
K80.4 Calculus of bile duct with cholecystitis
Any condition listed in K80.5 with cholecystitis (with cholangitis)
Use additional code if applicable for associated gangrene of gallbladder (K82.A1), or perforation of gallbladder (K82.A2)
Add Codes also fistula of bile duct (K83.3)

L57 Skin changes due to chronic exposure to nonionizing radiation
Revise Use additional code to identify the source of the ultraviolet radiation (W89, W90)

L98 Other disorders of skin and subcutaneous tissue, not elsewhere classified
L98.4 Non-pressure chronic ulcer of skin, not elsewhere classified
Revise Excludes2:varicose ulcer (I83.0- ~~I82.2~~ I82.3-)

M00 Pyogenic arthritis
M00.0 Staphylococcal arthritis and polyarthritis
Delete ~~Excludes2: infection and inflammatory reaction due to internal joint prosthesis (T84.5-)~~
M00 Pyogenic arthritis
Add Excludes2: infection and inflammatory reaction due to internal joint prosthesis (T84.5-)
M00.0 Staphylococcal arthritis and polyarthritis

Cleft lip and cleft palate (Q35-Q37)
Delete ~~Excludes1: Robin's syndrome (Q87.0)~~
Add Excludes2: Robin's syndrome (Q87.0)

M22 Disorder of patella
Delete Excludes~~1~~: ~~traumatic dislocation of patella (S83.0-)~~
Add Excludes2: traumatic dislocation of patella (S83.0-)

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)

M30 Polyarteritis nodosa and related conditions
M30.1 Polyarteritis with lung involvement [Churg-Strauss]
Add Eosinophilic granulomatosis with polyangiitis [EGPA]

O99 Other maternal diseases classifiable elsewhere but complicating pregnancy, childbirth and the puerperium
O99.3Mental disorders and diseases of the nervous system complicating pregnancy, childbirth and the puerperium

O99.34Other mental disorders complicating pregnancy, childbirth, and the puerperium
Revise Conditions in F01-F09, and F20-~~F99~~F52 and F54-F99

P04 Newborn affected by noxious substances transmitted via placenta or breast milk
P04.8 Newborn affected by other maternal noxious substances
Revise P04.89 Newborn affected by other maternal noxious substances

Q82 Other congenital malformations of skin
Q82.8 Other specified congenital malformations of skin
Revise Excludes1: Ehlers-Danlos syndrome**s** (Q79.6-)

R53 Malaise and fatigue
R53.1 Weakness
Asthenia NOS
Delete Excludes1: ~~muscle weakness (M62.8-)~~
Add muscle weakness (generalized) (M62.81)

R75 Inconclusive laboratory evidence of human immunodeficiency virus [HIV]
Nonconclusive HIV-test finding in infants
Add Excludes1: acute HIV infection syndrome (Z21)
asymptomatic human immunodeficiency virus [HIV] infection status (Z21)
human immunodeficiency virus [HIV] disease (B20)

S83 Dislocation and sprain of joints and ligaments of knee
Delete Excludes1: ~~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 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)

W89 Exposure to man-made visible and ultraviolet light
Delete ~~Excludes1: exposure to sunlight (X32)~~
Add Excludes2: exposure to sunlight (X32)

W90 Exposure to other nonionizing radiation
Delete Excludes1: exposure to sunlight (X32)
Add Excludes2: exposure to sunlight (X32)

Y92 Place of occurrence of the external cause
Y92.00 Unspecified non-institutional (private) residence as the place of occurrence of the external cause
Revise Y92.002 Bathroom of unspecified non-institutional (private) residence ~~single-family (private) house~~ as the place of occurrence of the external cause

Z20 Contact with and (suspected) exposure to communicable diseases
Z20.6 Contact with and (suspected) exposure to human immunodeficiency virus [HIV]
Add Excludes1: acute HIV infection syndrome (Z21)
asymptomatic human immunodeficiency virus [HIV]
HIV infection status (Z21)

Z21 Asymptomatic human immunodeficiency virus [HIV] infection status
Add Acute HIV infection syndrome
HIV positive NOS

Z68 Body mass index [BMI]
Delete Z68.1 Body mass index ~~(~~BMI~~)~~ 19.9 or less, adult
Add Z68.1 Body mass index [BMI] 19.9 or less, adult

Delete Z68.2 Body mass index ~~(~~BMI~~)~~ 20-29, adult
Add Z68.2 Body mass index [BMI] 20-29, adult
Delete Z68.20 Body mass index ~~(~~BMI~~)~~ 20.0-20.9, adult
Add Z68.20 Body mass index [BMI] 20.0-20.9, adult
Delete Z68.21 Body mass index ~~(~~BMI~~)~~ 21.0-21.9, adult
Add Z68.21 Body mass index [BMI] 21.0-21.9, adult
Delete Z68.22 Body mass index ~~(~~BMI~~)~~ 22.0-22.9, adult
Add Z68.22 Body mass index [BMI] 22.0-22.9, adult
Delete Z68.23 Body mass index ~~(~~BMI~~)~~ 23.0-23.9, adult
Add Z68.23 Body mass index [BMI] 23.0-23.9, adult
Delete Z68.24 Body mass index ~~(~~BMI~~)~~ 24.0-24.9, adult
Add Z68.24 Body mass index [BMI] 24.0-24.9, adult
Delete Z68.25 Body mass index ~~(~~BMI~~)~~ 25.0-25.9, adult
Add Z68.25 Body mass index [BMI] 25.0-25.9, adult
Delete Z68.26 Body mass index ~~(~~BMI~~)~~ 26.0-26.9, adult
Add Z68.26 Body mass index [BMI] 26.0-26.9, adult
Delete Z68.27 Body mass index ~~(~~BMI~~)~~ 27.0-27.9, adult
Add Z68.27 Body mass index [BMI] 27.0-27.9, adult

Delete		Z68.28	Body mass index ~~(~~BMI~~)~~ 28.0-28.9, adult
Add		Z68.28	Body mass index [BMI] 28.0-28.9, adult
Delete		Z68.29	Body mass index ~~(~~BMI~~)~~ 29.0-29.9, adult
Add		Z68.29	Body mass index [BMI] 29.0-29.9, adult
Delete	Z68.3		Body mass index ~~(~~BMI~~)~~ 30-39, adult
Add	Z68.3		Body mass index [BMI] 30-39, adult
Delete		Z68.30	Body mass index ~~(~~BMI~~)~~ 30.0-30.9, adult
Add		Z68.30	Body mass index [BMI] 30.0-30.9, adult
Delete		Z68.31	Body mass index (BMI) 31.0-31.9, adult
Add		Z68.31	Body mass index (BMI) 31.0-31.9, adult
Delete		Z68.32	Body mass index (BMI) 32.0-32.9, adult
Add		Z68.32	Body mass index (BMI) 32.0-32.9, adult
Delete		Z68.33	Body mass index (BMI) 33.0-33.9, adult
Add		Z68.33	Body mass index (BMI) 33.0-33.9, adult
Delete		Z68.34	Body mass index (BMI) 34.0-34.9, adult
Add		Z68.34	Body mass index (BMI) 34.0-34.9, adult
Delete		Z68.35	Body mass index ~~(~~BMI~~)~~ 35.0-35.9, adult
Add		Z68.35	Body mass index [BMI] 35.0-35.9, adult
Delete		Z68.36	Body mass index ~~(~~BMI~~)~~ 36.0-36.9, adult
Add		Z68.36	Body mass index [BMI] 36.0-36.9, adult
Delete		Z68.37	Body mass index ~~(~~BMI~~)~~ 37.0-37.9, adult
Add		Z68.37	Body mass index [BMI] 37.0-37.9, adult
Delete		Z68.38	Body mass index ~~(~~BMI~~)~~ 38.0-38.9, adult
Add		Z68.38	Body mass index [BMI] 38.0-38.9, adult
Delete		Z68.39	Body mass index ~~(~~BMI~~)~~ 39.0-39.9, adult
Add		Z68.39	Body mass index [BMI] 39.0-39.9, adult
Delete	Z68.4		Body mass index ~~(~~BMI~~)~~ 40 or greater, adult
Add	Z68.4		Body mass index [BMI] 40 or greater, adult
Delete		Z68.41	Body mass index ~~(~~BMI~~)~~ 40.0-44.9, adult
Add		Z68.41	Body mass index [BMI] 40.0-44.9, adult
Delete		Z68.42	Body mass index ~~(~~BMI~~)~~ 45.0-49.9, adult
Add		Z68.42	Body mass index [BMI] 45.0-49.9, adult
Delete		Z68.43	Body mass index ~~(~~BMI~~)~~ 50-59.9, adult
Add		Z68.43	Body mass index [BMI] 50-59.9, adult
Delete		Z68.44	Body mass index ~~(~~BMI~~)~~ 60.0-69.9, adult
Add		Z68.44	Body mass index [BMI] 60.0-69.9, adult
Delete		Z68.45	Body mass index ~~(~~BMI~~)~~ 70 or greater, adult
Add		Z68.45	Body mass index [BMI] 70 or greater, adult
Delete	Z68.5		Body mass index ~~(~~BMI~~)~~ pediatric
Add	Z68.5		Body mass index [BMI] pediatric
Delete		Z68.51	Body mass index ~~(~~BMI~~)~~ pediatric, less than 5th percentile for age
Add		Z68.51	Body mass index [BMI] pediatric, less than 5th percentile for age

Delete Z68.52 Body mass index ~~(~~BMI~~)~~ pediatric, 5th percentile to less than 85th percentile for age
Add Z68.52 Body mass index [BMI] pediatric, 5th percentile to less than 85th percentile for age
Delete Z68.53 Body mass index ~~(~~BMI~~)~~ pediatric, 85th percentile to less than 95th percentile for age
Add Z68.53 Body mass index [BMI] pediatric, 85th percentile to less than 95th percentile for age
Delete Z68.54 Body mass index ~~(~~BMI~~)~~ pediatric, greater than or equal to 95th percentile for age
Add Z68.54 Body mass index [BMI] pediatric, greater than or equal to 95th percentile for age

Z79 Long term (current) drug therapy
Includes: long term (current) drug use for prophylactic purposes
Code also any therapeutic drug level monitoring (Z51.81)
Delete Excludes2: ~~long term (current) use of oral antidiabetic drugs (Z79.84)~~
Delete ~~long term (current) use of oral hypoglycemic drugs (Z79.84)~~

Z79.4 Long term (current) use of insulin
Add Excludes1: long term (current) use of oral antidiabetic drugs (Z79.84)
Add long term (current) use of oral hypoglycemic drugs (Z79.84)

Z88 Allergy status to drugs, medicaments and biological substances
Revise Z88.1 Allergy status to other antibiotic agents ~~status~~
Revise Z88.2 Allergy status to sulfonamides ~~status~~
Revise Z88.3 Allergy status to other anti-infective agents ~~status~~
Revise Z88.4 Allergy status to anesthetic agent ~~status~~
Revise Z88.5 Allergy status to narcotic agent ~~status~~
Revise Z88.6 Allergy status to analgesic agent ~~status~~
Revise Z88.7 Allergy status to serum and vaccine ~~status~~
Revise Z88.8 Allergy status to other drugs, medicaments and biological substances ~~status~~
Revise Z88.9 Allergy status to unspecified drugs, medicaments and biological substances ~~status~~

ICD-10-CM Proposed Tabular Addenda
Part 2
All proposed effective October 1, 2020

I70 Atherosclerosis

I70.2 Atherosclerosis of native arteries of the extremities

Use additional code, if applicable, to identify chronic total occlusion of artery of extremity (I70.92)

Excludes2: atherosclerosis of bypass graft of extremities (I70.30-I70.79)

I70.22 Atherosclerosis of native arteries of extremities with rest pain
Add Critical limb ischemia of native arteries of extremities with rest pain
Add Chronic limb-threatening ischemia of native arteries of extremities with rest pain
Add Critical limb ischemia NOS of native arteries of extremities
Add Chronic limb-threatening ischemia NOS of native arteries of extremities

I70.23 Atherosclerosis of native arteries of right leg with ulceration
Add Critical limb ischemia of native arteries of right leg with ulceration
Add Chronic limb-threatening ischemia of native arteries of right leg with ulceration

I70.24 Atherosclerosis of native arteries of left leg with ulceration
Add Critical limb ischemia of native arteries of left leg with ulceration
Add Chronic limb-threatening ischemia of native arteries of left leg with ulceration

I70.26 Atherosclerosis of native arteries of extremities with gangrene
Add Critical limb ischemia of native arteries of extremities with gangrene
Add Chronic limb-threatening ischemia of native arteries of extremities with gangrene

I70.3 Atherosclerosis of unspecified type of bypass graft(s) of the extremities
Use additional code, if applicable, to identify chronic total occlusion of artery of extremity (I70.92)
Excludes1: embolism or thrombus of bypass graft(s) of extremities (T82.8-)

I70.32 Atherosclerosis of unspecified type of bypass graft(s) of the extremities with rest pain

Add Critical limb ischemia of unspecified type of bypass graft(s) of the extremities with rest pain

Add Chronic limb-threatening ischemia of unspecified type of bypass graft(s) of the extremities with rest pain, right leg

Add Critical limb ischemia NOS of unspecified type of bypass graft(s) of the extremities

Add Chronic limb-threatening ischemia NOS of unspecified type of bypass graft(s) of the extremities

I70.33 Atherosclerosis of unspecified type of bypass graft(s) of the right leg with ulceration

Add Critical limb ischemia of unspecified type of bypass graft(s) of the right leg with ulceration

Add Chronic limb-threatening ischemia of unspecified type of bypass graft(s) of the right leg with ulceration

I70.34 Atherosclerosis of unspecified type of bypass graft(s) of the left leg with ulceration

Add Critical limb ischemia of unspecified type of bypass graft(s) of the left leg with ulceration

Add Chronic limb-threatening ischemia of unspecified type of bypass graft(s) of the left leg with ulceration

I70.36 Atherosclerosis of unspecified type of bypass graft(s) of the extremities with gangrene

Add Critical limb ischemia of unspecified type of bypass graft(s) of the extremities with gangrene

Add Chronic limb-threatening ischemia of unspecified type of bypass graft(s) of the extremities with gangrene

I70.4 Atherosclerosis of autologous vein bypass graft(s) of the extremities
Use additional code, if applicable, to identify chronic total occlusion of artery of extremity (I70.92)

I70.42 Atherosclerosis of autologous vein bypass graft(s) of the extremities with rest pain

Add Critical limb ischemia of autologous vein bypass graft(s) of the extremities with rest pain

Add Chronic limb-threatening ischemia of autologous vein bypass graft(s) of the extremities with rest pain

Add Critical limb ischemia NOS of autologous vein bypass graft(s) of the extremities

Add Chronic limb-threatening ischemia NOS of autologous vein bypass graft(s) of the extremities

Includes: any condition classifiable to I70.41-

I70.43 Atherosclerosis of autologous vein bypass graft(s) of the right leg with ulceration

Add Critical limb ischemia of autologous vein bypass graft(s) of the right leg with ulceration

Add Chronic limb-threatening ischemia of autologous vein bypass graft(s) of the right leg with ulceration

I70.44 Atherosclerosis of autologous vein bypass graft(s) of the left leg with ulceration

Add Critical limb ischemia of autologous vein bypass graft(s) of the left leg with ulceration

Add Chronic limb-threatening ischemia of autologous vein bypass graft(s) of the left leg with ulceration

I70.46 Atherosclerosis of autologous vein bypass graft(s) of the extremities with gangrene

Add Critical limb ischemia of autologous vein bypass graft(s) of the extremities with gangrene

Add Chronic limb-threatening ischemia of autologous vein bypass graft(s) of the extremities with gangrene

I70.5 Atherosclerosis of nonautologous biological bypass graft(s) of the extremities

Use additional code, if applicable, to identify chronic total occlusion of artery of extremity (I70.92)

I70.52 Atherosclerosis of nonautologous biological bypass graft(s) of the extremities with rest pain

Add Critical limb ischemia of nonautologous biological bypass graft(s) of the extremities with rest pain

Add Chronic limb-threatening ischemia of nonautologous biological bypass graft(s) of the extremities with rest pain

Add Critical limb ischemia NOS of nonautologous biological bypass graft(s) of the extremities

Add Chronic limb-threatening ischemia NOS of nonautologous biological bypass graft(s) of the extremities

I70.53 Atherosclerosis of nonautologous biological bypass graft(s) of the right leg with ulceration

Add Critical limb ischemia of nonautologous biological bypass graft(s) of the right leg with ulceration

Add Chronic limb-threatening ischemia of nonautologous biological bypass graft(s) of the right leg with ulceration

Includes: any condition classifiable to I70.511 and I70.521

Use additional code to identify severity of ulcer (L97.-)

I70.54 Atherosclerosis of nonautologous biological bypass graft(s) of the left leg with ulceration

Add Critical limb ischemia of nonautologous biological bypass graft(s) of the left leg with ulceration

Add Chronic limb-threatening ischemia of nonautologous biological bypass graft(s) of the left leg with ulceration

Includes: any condition classifiable to I70.512 and I70.522

Use additional code to identify severity of ulcer (L97.-)

I70.56 Atherosclerosis of nonautologous biological bypass graft(s) of the extremities with gangrene

Add Critical limb ischemia of nonautologous biological bypass graft(s) of the extremities with gangrene

Add Chronic limb-threatening ischemia of nonautologous biological bypass graft(s) of the extremities with gangrene

Includes: any condition classifiable to I70.51-, I70.52-, I70.53-, I70.54-, I70.55

Use additional code to identify the severity of any ulcer (L97.-, L98.49-), if applicable

I70.6 Atherosclerosis of nonbiological bypass graft(s) of the extremities
Use additional code, if applicable, to identify chronic total occlusion of artery of extremity (I70.92)

I70.62 Atherosclerosis of nonbiological bypass graft(s) of the extremities with rest pain
Add Critical limb ischemia of nonbiological bypass graft(s) of the extremities with rest pain
Chronic limb-threatening ischemia of nonbiological bypass graft(s) of the extremities with rest pain
Add Critical limb ischemia NOS of nonbiological bypass graft(s) of the extremities
Add Chronic limb-threatening ischemia NOS of nonbiological bypass graft(s) of the extremities
Includes: any condition classifiable to I70.61-

I70.63 Atherosclerosis of nonbiological bypass graft(s) of the right leg with ulceration
Add Critical limb ischemia of nonbiological bypass graft(s) of the right leg with ulceration
Add Chronic limb-threatening ischemia of nonbiological bypass graft(s) of the right leg with ulceration
Includes: any condition classifiable to I70.611 and I70.621
Use additional code to identify severity of ulcer (L97.-)

I70.64 Atherosclerosis of nonbiological bypass graft(s) of the left leg with ulceration
Add Critical limb ischemia of nonbiological bypass graft(s) of the left leg with ulceration
Add Chronic limb-threatening ischemia of nonbiological bypass graft(s) of the left leg with ulceration

I70.66 Atherosclerosis of nonbiological bypass graft(s) of the extremities with gangrene
Add Critical limb ischemia of nonbiological bypass graft(s) of the extremities with gangrene
Add Chronic limb-threatening ischemia of nonbiological bypass graft(s) of the extremities with gangrene

I70.7 Atherosclerosis of other type of bypass graft(s) of the extremities
Use additional code, if applicable, to identify chronic total occlusion of artery of extremity (I70.92)

I70.72 Atherosclerosis of other type of bypass graft(s) of the extremities with rest pain

Add Critical limb ischemia of other type of bypass graft(s) of the extremities with rest pain

Add Chronic limb-threatening ischemia of other type of bypass graft(s) of the extremities with rest pain

Add Critical limb ischemia NOS of other type of bypass graft(s) of the extremities

Add Chronic limb-threatening ischemia NOS of other type of bypass graft(s) of the extremities

I70.73 Atherosclerosis of other type of bypass graft(s) of the right leg with ulceration

Add Critical limb ischemia of other type of bypass graft(s) of the right leg with ulceration

Add Chronic limb-threatening ischemia of other type of bypass graft(s) of the right leg with ulceration

I70.74 Atherosclerosis of other type of bypass graft(s) of the left leg with ulceration

Add Critical limb ischemia of other type of bypass graft(s) of the left leg with ulceration

Add Chronic limb-threatening ischemia of other type of bypass graft(s) of the left leg with ulceration

I70.76 Atherosclerosis of other type of bypass graft(s) of the extremities with gangrene

Add Critical limb ischemia of other type of bypass graft(s) of the extremities with gangrene

Add Chronic limb-threatening ischemia of other type of bypass graft(s) of the extremities with gangrene

ICD-10-CM INDEX OF DISEASES - PROPOSED ADDENDA

All proposed effective October 1, 2020

Abnormal, abnormality, abnormalities- see also Anomaly
- diagnostic imaging
Revise - - skull ~~R93.09~~ R93.0

Abortion
Add - inevitable O03.4

Abscess
- intra-abdominal - see also Abscess, peritoneum K65.1
Revise - - postprocedural ~~T81.49~~ T81.43

Revise Arrhythmia (auricle)(cardiac)(juvenile)(nodal)
(reflex)~~(sinus)~~(supraventricular)(transitory)(ventricle) I49.9
Add - sinus I49.8

Arthritis, arthritic
Revise - spine - see also ~~Spondylopathy, inflammatory~~ Spondylosis

Buschke's
Revise - disease ~~B45.3~~ – see Cryptococcosis by site

Revise Busse-Buschke disease ~~B45.3~~ – see Cryptococcosis by site

Defect, defective Q89.9
Revise - speech – see Disorder, speech R47.9

Dermatitis (eczematous) L30.9
Add - desquamative L30.8

Disorder (of) - see also Disease
- speech R47.9
Add - - slurring R47.81

Disease
- lung J98.4
Revise - - obstructive (chronic) ~~J43.9~~ J44.9

Diverticulitis (acute) K57.92
- intestine K57.92
- - with

Revise - - - abscess, perforation ~~or peritonitis~~ K57.80
- - large K57.32
- - - with
Revise - - - - abscess, perforation ~~or peritonitis~~ K57.20
- - - - small intestine K57.52
- - - - - with
Revise - - - - - - abscess, perforation ~~or peritonitis~~ K57.40
- - small K57.12
- - - with
Revise - - - - abscess, perforation ~~or peritonitis~~ K57.00

Embolism (multiple) (paradoxical) I74.9
Revise - postoperative, ~~postprocedural~~ postprocedural

Enthesopathy (peripheral) M77.9
Revise - foot NEC ~~M77.9~~ M77.8
Revise - forearm ~~M77.9~~ M77.8
Revise - hand ~~M77.9~~ M77.8
Revise - multiple sites ~~M77.9~~ M77.8
Revise - pelvis ~~M77.9~~ M77.8
Revise - shoulder ~~M77.9~~ M77.8
Delete ~~- specified site NEC M77.9~~
Revise - upper arm ~~M77.9~~ M77.8

Fistula
- bile duct (common) (hepatic) K83.3
Revise - - with calculus, stones – see also Calculus, bile duct (K83.3)

Fracture, traumatic (abduction) (adduction) (separation) (see also Fracture, pathological) T14.8
Add - buckle – see Fracture by site torus

Infarct, infarction
- myocardium, myocardial (acute) (with stated duration of 4 weeks or less) I21.9
Revise - - - transmural ~~I22.9~~ I21.3

Inhalation
Revise - gases, fumes, or vapors ~~NEC~~ T59.9-
Revise - - specified agent NEC - see Table of Drugs and Chemicals, by substance T59.89-
Revise - smoke ~~J70.5~~ T59.81-
Add - - with respiratory conditions J70.5
Revise - - - due to chemicals, gases, fumes and vapors J68.9
Revise - steam ~~- see Toxicity, vapors~~ T59.9-

Revise Impediment, speech – see also Disorder, speech R47.9

Injury
- deep tissue - see Contusion, by site
Revise - - meaning pressure ulcer - see Ulcer, pressure~~, unstageable, by site~~ L89 with final character .6
Revise Myelodysplastic syndrome (see also Syndrome, myelodysplastic) D46.9

Revise Pancreatitis (annular) (apoplectic) (calcareous) (edematous) (hemorrhagic) (malignant) ~~(recurrent)~~ (subacute) (suppurative) K85.90
Revise - recurrent ~~(chronic) K86.1~~
Add - - acute K85.50
Add - - chronic K86.1

Prominence, prominent
Revise - ischial spine or sacral promontory with disproportion (fetopelvic) O33.0
Delete ~~- with disproportion (fetopelvic) O33.0~~

Sequestration - see also Sequestrum
Revise - ~~disk~~ disc- see Displacement, intervertebral ~~disk~~ disc

Speech
Revise - defect, disorder, disturbance, impediment – see Disorder, speech R47.9

Syndrome
- myelodysplastic D46.9
- - with
Add - - - multilineage dysplasia D46.A
Add - - - - with ringed sideroblasts D46.B
- postencephalitic F07.89
Revise - -post endometrial ablation N99.85 **Remove second dash**

Thrombosis
- mesenteric (artery) (with gangrene) (see also Infarct, intestine) K55.069
Revise - - vein (inferior) (superior) ~~K55.0~~ K55.0-

Revise Thymoma (benign) D15.0 (see also Neoplasm, thymus, by type)
Add - benign D15.0
Add - metaplastic C37
Add - microscopic D15.0
Add - type A C37
Add - type AB C37
Add - type B1 C37
Add - type B2 C37
Add - type B3 C37
Add - sclerosing C37

Proposed Index Addenda
Part 2
All proposed effective October 1, 2020

Arteriosclerosis, arteriosclerotic (diffuse) (obliterans) (of) (senile) (with calcification) I70.90
Add - with
Add - - chronic limb-threatening ischemia - *see* Arteriosclerosis, with critical limb ischemia
Add - - critical limb ischemia
Add - - - bypass graft I70.329
Add - - - - autologous vein graft I70.429
Add - - - - - leg I70.429
Add - - - - - - with
Add - - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.469
Add - - - - - - - rest pain (and intermittent claudication) I70.429
Add - - - - - - bilateral I70.423
Add - - - - - - - with
Add - - - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.463
Add - - - - - - - - rest pain (and intermittent claudication) I70.423
Add - - - - - - left I70.422
Add - - - - - - - with
Add - - - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.462
Add - - - - - - - - rest pain (and intermittent claudication) I70.422
Add - - - - - - - - ulceration (and intermittent claudication and rest pain) I70.449
Add - - - - - - - - - ankle I70.443
Add - - - - - - - - - calf I70.442
Add - - - - - - - - - foot site NEC I70.445
Add - - - - - - - - - heel I70.444
Add - - - - - - - - - lower leg NEC I70.448
Add - - - - - - - - - mid foot I70.444
Add - - - - - - - - - thigh I70.441
Add - - - - - - right I70.421
Add - - - - - - - with
Add - - - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.461
Add - - - - - - - - rest pain (and intermittent claudication) I70.421
Add - - - - - - - - ulceration (and intermittent claudication and rest pain) I70.439
Add - - - - - - - - - ankle I70.433
Add - - - - - - - - - calf I70.432
Add - - - - - - - - - foot site NEC I70.435
Add - - - - - - - - - heel I70.434
Add - - - - - - - - - lower leg NEC I70.438
Add - - - - - - - - - midfoot I70.434
Add - - - - - - - - - thigh I70.431
Add - - - - leg I70.329
Add - - - - - with

Add - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.369
Add - - - - - - rest pain (and intermittent claudication) I70.329
Add - - - - - bilateral I70.323
Add - - - - - - with
Add - - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.363
Add - - - - - - - rest pain (and intermittent claudication) I70.323
Add - - - - - left I70.322
Add - - - - - - with
Add - - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.362
Add - - - - - - - rest pain (and intermittent claudication) I70.322
Add - - - - - - - ulceration (and intermittent claudication and rest pain) I70.349
Add - - - - - - - - ankle I70.343
Add - - - - - - - - calf I70.342
Add - - - - - - - - foot site NEC I70.345
Add - - - - - - - - heel I70.344
Add - - - - - - - - lower leg NEC I70.348
Add - - - - - - - - midfoot I70.344
Add - - - - - - - - thigh I70.341
Add - - - - - right I70.321
Add - - - - - - with
Add - - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.361
Add - - - - - - - rest pain (and intermittent claudication) I70.321
Add - - - - - - - ulceration (and intermittent claudication and rest pain) I70.339
Add - - - - - - - - ankle I70.333
Add - - - - - - - - calf I70.332
Add - - - - - - - - foot site NEC I70.335
Add - - - - - - - - heel I70.334
Add - - - - - - - - lower leg NEC I70.338
Add - - - - - - - - midfoot I70.334
Add - - - - - - - - thigh I70.331
Add - - - - nonautologous biological graft I70.529
Add - - - - - leg I70.529
Add - - - - - - with
Add - - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.569
Add - - - - - - - rest pain (and intermittent claudication) I70.529
Add - - - - - - bilateral I70.523
Add - - - - - - - with
Add - - - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.563
Add - - - - - - - - rest pain (and intermittent claudication) I70.523
Add - - - - - - left I70.522
Add - - - - - - - with
Add - - - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.562
Add - - - - - - - - rest pain (and intermittent claudication) I70.522
Add - - - - - - - - ulceration (and intermittent claudication and rest pain) I70.549
Add - - - - - - - - - ankle I70.543

Add - - - - - - - - - calf I70.542
Add - - - - - - - - - foot site NEC I70.545
Add - - - - - - - - - heel I70.544
Add - - - - - - - - - lower leg NEC I70.548
Add - - - - - - - - - midfoot I70.544
Add - - - - - - - - - thigh I70.541
Add - - - - - - right I70.521
Add - - - - - - - with
Add - - - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.561
Add - - - - - - - - rest pain (and intermittent claudication) I70.521
Add - - - - - - - - ulceration (and intermittent claudication and rest pain) I70.539
Add - - - - - - - - - ankle I70.533
Add - - - - - - - - - calf I70.532
Add - - - - - - - - - foot site NEC I70.535
Add - - - - - - - - - heel I70.534
Add - - - - - - - - - lower leg NEC I70.538
Add - - - - - - - - - midfoot I70.534
Add - - - - - - - - - thigh I70.531
Add - - - - nonbiological graft I70.629
Add - - - - - leg I70.629
Add - - - - - - with
Add - - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.669
Add - - - - - - - rest pain (and intermittent claudication) I70.629
Add - - - - - - bilateral I70.623
Add - - - - - - - with
Add - - - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.663
Add - - - - - - - - rest pain (and intermittent claudication) I70.623
Add - - - - - - left I70.622
Add - - - - - - - with
Add - - - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.662
Add - - - - - - - - rest pain (and intermittent claudication) I70.622
Add - - - - - - - - ulceration (and intermittent claudication and rest pain) I70.649
Add - - - - - - - - - ankle I70.643
Add - - - - - - - - - calf I70.642
Add - - - - - - - - - foot site NEC I70.645
Add - - - - - - - - - heel I70.644
Add - - - - - - - - - lower leg NEC I70.648
Add - - - - - - - - - midfoot I70.644
Add - - - - - - - - - thigh I70.641
Add - - - - - - right I70.621
Add - - - - - - - with
Add - - - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.661
Add - - - - - - - - rest pain (and intermittent claudication) I70.621
Add - - - - - - - - ulceration (and intermittent claudication and rest pain) I70.639
Add - - - - - - - - - ankle I70.633

Add - - - - - - - - - calf I70.632
Add - - - - - - - - - foot site NEC I70.635
Add - - - - - - - - - heel I70.634
Add - - - - - - - - - lower leg NEC I70.638
Add - - - - - - - - - midfoot I70.634
Add - - - - - - - - - thigh I70.631
Add - - - - specified graft NEC I70.729
Add - - - - - leg I70.729
Add - - - - - - with
Add - - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.769
Add - - - - - - - rest pain (and intermittent claudication) I70.729
Add - - - - - - bilateral I70.723
Add - - - - - - - with
Add - - - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.763
Add - - - - - - - - rest pain (and intermittent claudication) I70.723
Add - - - - - - left I70.722
Add - - - - - - - with
Add - - - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.762
Add - - - - - - - - rest pain (and intermittent claudication) I70.722
Add - - - - - - - - ulceration (and intermittent claudication and rest pain) I70.749
Add - - - - - - - - - ankle I70.743
Add - - - - - - - - - calf I70.742
Add - - - - - - - - - foot site NEC I70.745
Add - - - - - - - - - heel I70.744
Add - - - - - - - - - lower leg NEC I70.748
Add - - - - - - - - - midfoot I70.744
Add - - - - - - - - - thigh I70.741
Add - - - - - - right I70.721
Add - - - - - - - with
Add - - - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.761
Add - - - - - - - - rest pain (and intermittent claudication) I70.721
Add - - - - - - - - ulceration (and intermittent claudication and rest pain) I70.739
Add - - - - - - - - - ankle I70.733
Add - - - - - - - - - calf I70.732
Add - - - - - - - - - foot site NEC I70.735
Add - - - - - - - - - heel I70.734
Add - - - - - - - - - lower leg NEC I70.738
Add - - - - - - - - - midfoot I70.734
Add - - - - - - - - - thigh I70.731
Add - - - leg I70.229
Add - - - - - with
Add - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.269
Add - - - - - - rest pain (and intermittent claudication) I70.229
Add - - - - bilateral I70.223
Add - - - - - with

Add - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.263
Add - - - - - - rest pain (and intermittent claudication) I70.223
Add - - - - left I70.222
Add - - - - - with
Add - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.262
Add - - - - - - rest pain (and intermittent claudication) I70.222
Add - - - - - - ulceration (and intermittent claudication and rest pain) I70.249
Add - - - - - - - ankle I70.243
Add - - - - - - - calf I70.242
Add - - - - - - - foot site NEC I70.245
Add - - - - - - - heel I70.244
Add - - - - - - - lower leg NEC I70.248
Add - - - - - - - midfoot I70.244
Add - - - - - - - thigh I70.241
Add - - - - right I70.221
Add - - - - - with
Add - - - - - - gangrene (and intermittent claudication, rest pain, and ulcer) I70.261
Add - - - - - - rest pain (and intermittent claudication) I70.221
Add - - - - - - ulceration (and intermittent claudication and rest pain) I70.239
Add - - - - - - - ankle I70.233
Add - - - - - - - calf I70.232
Add - - - - - - - foot site NEC I70.235
Add - - - - - - - heel I70.234
Add - - - - - - - lower leg NEC I70.238
Add - - - - - - - midfoot I70.234
Add - - - - - - - thigh I70.231

- aorta I70.0
- arteries of extremities - *see* Arteriosclerosis, extremities
Add - - with
Add - - - chronic limb-threatening ischemia - *see* Arteriosclerosis, with critical limb ischemia
Add - - - critical limb ischemia - *see* Arteriosclerosis, with critical limb ischemia
- brain I67.2
- bypass graft
Add - - with
Add - - - chronic limb-threatening ischemia - *see* Arteriosclerosis, with critical limb ischemia
Add - - - critical limb ischemia - *see* Arteriosclerosis, with critical limb ischemia
- - coronary - *see* Arteriosclerosis, coronary, bypass graft
- - extremities - *see* Arteriosclerosis, extremities, bypass graft

- extremities (native arteries) I70.209
Add - - with
Add - - - chronic limb-threatening ischemia - *see* Arteriosclerosis, with critical limb ischemia
Add - - - critical limb ischemia - *see* Arteriosclerosis, with critical limb ischemia
- - bypass graft I70.309

Add - - - with
Add - - - - chronic limb-threatening ischemia - *see* Arteriosclerosis, with critical limb ischemia
Add - - - - critical limb ischemia - *see* Arteriosclerosis, with critical limb ischemia

Ischemia, ischemic I99.8
Add - limb, critical - *see* Arteriosclerosis, with critical limb ischemia
Add - limb-threatening, chronic - *see* Arteriosclerosis, with critical limb ischemia