ICD-10 Coordination and Maintenance Committee Meeting
September 13-14, 2022
Diagnosis Agenda

Zoom Webinar and Dial-In Information
• This meeting will be conducted via Zoom Webinar. The URL to join the Zoom Webinar, the password, and the call-in numbers are the same for both days of the meeting.

Meeting details for each day are as follows:

• Day 1: September 13, 2022: The meeting will begin promptly at 9:00 AM ET and will end at 5:00 PM ET. Lunch will be held from 12:30 PM to 1:30 PM.

• Day 2: September 14, 2022: The meeting will begin promptly at 9:00 AM ET and will end at 5:00 PM ET. Lunch will be held from 12:30 PM to 1:30 PM.

To minimize feedback to the maximum extent possible, join the meeting using only ONE of the options listed below:

Option 1: Remote participants (attendees wishing to both view slides and ask questions during the Q&A portions of the meeting) must join the Zoom Webinar via the web. To join this Zoom Webinar conference from a PC, MAC, iPad, iPhone or Android device as well as, connect to the audio portion of the conference:
Click the following URL:

https://cms.zoomgov.com/j/1618774054?pwd=RzlGenRwL09TbHZKVEcrNnJDRi90QT09
Passcode: 703819

Option 2: Dial-in access is available for listen-only participants. Listen-only participants are participants who wish to only listen to the meeting and do not wish to comment or ask questions during the Q&A portions of the meeting.
ICD-10 Coordination and Maintenance Committee Meeting  
September 13-14-2022

1. From your phone, dial U.S.*: 669-254-5252 or 646-828-7666 or 833-568-8864 (Toll Free)  
2. Enter the webinar ID: 161 877 4054  
   *If dialing in from outside of the U.S., visit https://cms.zoomgov.com/u/ahSKgrsLu for a list of Zoom International Dial-in Numbers.

**Option 3**: To join this Zoom Webinar conference from an H.323/SIP room system:  
1. From your room system, dial 161.199.138.10 (US West) or 161.199.136.10 (US East)  
2. Enter the webinar ID: 161 877 4054  
   Passcode: 703819

   SIP: 1618774054@sip.zoomgov.com  
   Passcode: 703819

If you experience technical difficulties during the meeting, please contact Marvelyn Davis for assistance at marvelyn.davis1@cms.hhs.gov or 410-786-2580 Option 7.

Those participating in the Zoom Webinar may ask questions during the Q&A portions of the meeting using the “Raise Your Hand” feature. If time does not permit you to comment or ask a question during the Q&A session, you may submit comments and questions at any time using the “Q&A” feature. All comments and questions submitted using the “Q&A” feature, along with CDC’s responses to them, will be posted as soon as possible after the meeting on the CDC web page located at: ICD - ICD-10-CM - Coordination and Maintenance Committee (cdc.gov). Remaining questions may be submitted via the NCHS mailbox at nchsicd10CM@cdc.gov.

*Please note that registration is not required to attend the Zoom Webinar.
Welcome and announcements

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

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The University of Colorado, Division of Rheumatology

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ICD-10 TIMELINE

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

September 13-14, 2022  The September 2022 ICD-10 Coordination and Maintenance Committee Meeting.

September 2022  Recordings and slide presentations of the September 13-14, 2022 ICD-10 Coordination and Maintenance Committee Meeting will be posted on the following web pages:

   Diagnosis code portion of the recording and related materials–
   https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm

   Procedure code portion of the recording and related materials–

October 1, 2022  New and revised ICD-10-CM and ICD-10-PCS codes go into effect along with MS-DRG changes. Final addendum available on web pages as follows:

   Diagnosis addendum –

   Procedure addendum –
   https://www.cms.gov/medicare/coding/icd10

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

November 2022  Any new ICD-10 codes required to capture new diseases or technology that will be implemented on the following April 1 will be announced. Information on any new codes to be implemented April 1, 2023, will be posted on the following websites:
November 14, 2022
Deadline for receipt of public comments on proposed new codes and revisions discussed at the September 13-14, 2022 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2023.

December 2, 2022
Deadline for requestors: Those members of the public requesting that topics be discussed at the March 7-8, 2023 ICD-10 Coordination and Maintenance Committee Meeting must have their requests submitted to CMS for procedures and to NCHS for diagnoses by this date.

Procedure code requests should be directed to CMS at https://mearis.cms.gov. Diagnosis code requests should be directed to NCHS at nchsicd10cm@cdc.gov.

Requestors should indicate if they are submitting their code request for consideration for an October 1, 2023, implementation date or an April 1, 2024, implementation date.

January 2023
The ICD-10 Coordination and Maintenance Committee will make efforts to accommodate the requested implementation date for each request submitted, however, the Committee will determine which requests will be presented for consideration for an October 1, 2023 implementation date or an April 1, 2024 implementation date.

Federal Register notice for the March 7-8, 2023 ICD-10 Coordination and Maintenance Committee Meeting will be published. This will include the tentative agenda.

February 2023
Tentative agenda for the Procedure portion of the March 7, 2023 ICD-10 Coordination and Maintenance Committee Meeting posted on CMS webpage as follows:

ICD-10 Coordination and Maintenance Committee Meeting
September 13-14-2022

Tentative agenda for the Diagnosis portion of the March 8, 2023, ICD-10 Coordination and Maintenance Committee Meeting posted on NCHS homepage as follows:

https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm

**February 1, 2023**
ICD-10 MS-DRG Grouper software and related materials posted on CMS webpage at: https://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/AcuteInpatientPPS/MS-DRG-Classifications-and-Software

**February 1, 2023**
Any updates to the ICD-10-CM and ICD-10-PCS Coding Guidelines will be posted on the following websites:


https://www.cms.gov/medicare/coding/icd10

**February 1, 2023**
All ICD-10-CM and ICD-10-PCS code update files (includes April 1 update and full files from prior October 1) will be posted on the following websites:


https://www.cms.gov/medicare/coding/icd10

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

**March 2023**
Recordings and slide presentations of the March 7-8, 2023 ICD-10 Coordination and Maintenance Committee Meeting will be posted on the following web pages:

**Diagnosis code portion of the recording and related materials**–
https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm

**Procedure code portion of the recording and related materials**–
<table>
<thead>
<tr>
<th>Date</th>
<th>Event</th>
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<tr>
<td>April 1, 2023</td>
<td>Any new ICD-10 codes will be implemented on April 1, 2023.</td>
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<tr>
<td>April 7, 2023</td>
<td><strong>Deadline for receipt of public comments on proposed new codes and revisions discussed at the March 7-8, 2023 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2023.</strong></td>
</tr>
<tr>
<td>April 2023</td>
<td>Notice of Proposed Rulemaking to be published in the Federal Register as mandated by Public Law 99-509. This notice will include references to the FY 2024 ICD-10-CM diagnosis and ICD-10-PCS procedure codes finalized to date. It will also include proposed revisions to the MS-DRG system based on ICD-10-CM/PCS codes on which the public may comment. The proposed rule can be accessed at: <a href="https://www.cms.gov/medicare/medicare-fee-for-service-payment/acuteinpatientpps">https://www.cms.gov/medicare/medicare-fee-for-service-payment/acuteinpatientpps</a></td>
</tr>
<tr>
<td>May 5, 2023</td>
<td><strong>Deadline for receipt of public comments on proposed new codes and revisions discussed at the March 7-8, 2023 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on April 1, 2024.</strong></td>
</tr>
<tr>
<td></td>
<td><strong>Deadline for receipt of public comments on proposed new diagnosis codes and revisions discussed at the March 7-8, 2023 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2024.</strong></td>
</tr>
<tr>
<td>May/June 2023</td>
<td>Final addendum posted on web pages as follows:</td>
</tr>
<tr>
<td></td>
<td><strong>Diagnosis addendum</strong> -</td>
</tr>
<tr>
<td></td>
<td><strong>Procedure addendum</strong> -</td>
</tr>
</tbody>
</table>
June 9, 2023  Deadline for requestors: Those members of the public requesting that topics be discussed at the September 12-13, 2023 ICD-10 Coordination and Maintenance Committee Meeting, must have their requests submitted to CMS for procedures and NCHS for diagnoses.

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

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

August 1, 2023  Hospital Inpatient Prospective Payment System final rule expected to be published in the Federal Register as mandated by Public Law 99-509. This rule will also include links to all the final codes to be implemented on October 1, 2023.

This rule can be accessed at:
https://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/AcuteInpatientPPS/index.html

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

Tentative agenda for the Diagnosis portion of the September 13, 2023, ICD-10 Coordination and Maintenance Committee Meeting will be posted on the NCHS webpage at –
https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm

September 12-13, 2023  The September 2023 ICD-10 Coordination and Maintenance Committee Meeting is anticipated to be fully virtual by zoom and dial-in. Those who wish to attend must participate via Zoom Webinar or by dialing in.
September 2023

Recordings and slide presentations of the September 12-13, 2023 ICD-10 Coordination and Maintenance Committee Meeting will be posted on the following web pages:

**Diagnosis code portion of the recording and related materials**–
https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm

**Procedure code portion of the recording and related materials**–

October 1, 2023

New and revised ICD-10-CM and ICD-10-PCS codes go into effect along with MS-DRG changes. Final addendum available on web pages as follows:

**Diagnosis addendum** –

**Procedure addendum** –
https://www.cms.gov/medicare/coding/icd10

October 13, 2023

Deadline for receipt of public comments on proposed new codes discussed at the September 12-13, 2023 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on April 1, 2024.

November 2023

Any new ICD-10 codes that will be implemented on the following April 1 will be announced. Information on any new codes to be implemented April 1, 2024 will be posted on the following websites:


https://www.cms.gov/medicare/coding/icd10

November 15, 2023

Deadline for receipt of public comments on proposed new codes and revisions discussed at the September 12-13, 2023 ICD-10 Coordination and Maintenance Committee Meeting being considered for implementation on October 1, 2024.
Contact Information
Mailing address:

National Center for Health Statistics
ICD-9-CM Coordination and Maintenance Committee
3311 Toledo Road
Hyattsville, Maryland 20782
Fax: (301) 458-4045

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
Herman Thurman (301) 458-4282
Continuing Education Credits

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

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

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

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

AHIMA credential-holders may claim 1 CEU per 60 minutes of attendance at an educational program. Maintain documentation about the program for verification purposes in the event of an audit. A program does not need to be pre-approved by AHIMA, nor does a CEU certificate need to be provided, 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.
Abnormal rheumatoid arthritis-related immunological findings without current or prior diagnosis of clinically-apparent inflammatory arthritis

Rheumatoid arthritis (RA) is a well-known autoimmune condition that is characterized by the presence of inflammatory arthritis (IA). Furthermore, in up to 80% of individuals with RA there are also abnormalities of circulating biomarkers including but not limited to the autoantibodies rheumatoid factor (RF) and antibodies to cyclic citrullinated proteins (anti-CCP).

There are established diagnostic classification criteria for RA that incorporate several features including the presence and distribution of joint tenderness and swelling, autoantibodies and inflammatory markers, duration of symptoms and in some criteria the presence of imaging evidence of joint damage. However, while these criteria are commonly used in clinical research of RA, in routine clinical practice RA is commonly diagnosed and treated even if the classification criteria are not met. Indeed, in most current clinical practices, ICD-10-CM for RA are applied based on a clinical diagnosis and not necessarily fulfillment of established classification criteria.

The current paradigm for the diagnosis and treatment of RA is for a clinician to identify joint findings that are determined to be clinically-apparent IA, diagnosis that as RA based on clinical, laboratory and radiographic features, and initiate treatment. Furthermore, this is the typical clinical situation when the existing ICD-10-CM codes for RA (e.g., M60, M50.) are applied.

However, it is now well-established that RA-related immunologic tests such as RF and ACPA/anti-CCP can be present in individuals in absence of and prior to the appearance of IA, and predictive of future onset of clinical RA. Furthermore, individuals who have abnormal RA-related immunologic tests without IA are identified in growing numbers in clinical care. There are current recommendations for medical follow-up and lifestyle changes (e.g., smoking cessation) that can be applied to these individuals. In addition, the predictive ability of RF and ACPA for future clinical RA has underpinned multiple clinical observational studies and prevention trials in RA.

While there are ICD-10-CM codes that can be used to designate clinical RA, RF and anti-CCP positivity, there is not currently a clear way in the existing ICD-10-CM to code individuals who may exhibit RA-related biomarkers but not have clinically-apparent IA. As such, the introduction of a new code to accurately designate an individual who has abnormal RA-related immunologic test will facilitate clinical designation and care of these individuals, as well as facilitate clinical research.

This proposal was submitted by The University of Colorado, Division of Rheumatology and is supported by the American College of Rheumatology (ACR).
ICD-10 Coordination and Maintenance Committee Meeting
September 13-14-2022

References


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<thead>
<tr>
<th>Code</th>
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<tr>
<td>R76</td>
<td>Other abnormal immunological findings in serum</td>
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<td>R76.8</td>
<td>Other specified abnormal immunological findings in serum</td>
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<td>Raised level of immunoglobulins NOS</td>
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<td>New Code</td>
<td>Abnormal rheumatoid arthritis-related immunological findings without current or prior diagnosis of clinically-apparent inflammatory arthritis</td>
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<td>Add</td>
<td>Other specified abnormal immunological findings in serum</td>
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<tr>
<td>New code</td>
<td>Raised level of immunoglobulins NOS</td>
</tr>
</tbody>
</table>
Acinetobacter Baumannii Infections

Acinetobacter are commonly found in the environment, like in soil and water. Acinetobacter baumannii accounts for most Acinetobacter infections in humans. It is a gram-negative bacteria which can cause infections in the blood, urinary tract, lungs, or in wounds in different parts of the body.\textsuperscript{1,2} It can also “colonize” or live in a patient without causing infections or symptoms, especially in respiratory secretions or open wounds.\textsuperscript{1}

Antibiotic resistance is common in Acinetobacter baumannii, and carbapenem resistance is a challenging threat to hospitalized patients. In 2017, carbapenem-resistant Acinetobacter caused an estimated 8,500 infections in hospitalized patients and 700 estimated deaths in the United States.\textsuperscript{1,3}

This proposal is based on internal CDC review.

References
1. CDC. Acinetobacter in Healthcare Settings. Centers for Disease Control and Prevention, National Center for Emerging and Zoonotic Infectious Diseases (NCEZID), Division of Healthcare Quality Promotion (DHQP). https://www.cdc.gov/hai/organisms/acinetobacter.html
2. CDC. Multi-site Gram-negative Surveillance Initiative. Centers for Disease Control and Prevention, National Center for Emerging and Zoonotic Infectious Diseases (NCEZID), Division of Healthcare Quality Promotion (DHQP). https://www.cdc.gov/hai/eip/mugsi.html

TABULAR MODIFICATIONS

A41 Other sepsis
  A41.5 Sepsis due to other Gram-negative organisms
    New code A41.54 Sepsis due to Acinetobacter baumannii
    Use additional code, if applicable, for resistance to carbapenem (Z16.13)

Add B96 Other bacterial agents as the cause of diseases classified elsewhere
  B96.8 Other specified bacterial agents as the cause of diseases classified elsewhere
    New code B96.83 Acinetobacter baumannii as the cause of diseases classified elsewhere
    Use additional code, if applicable, for resistance to carbapenem (Z16.13)

J15 Bacterial pneumonia, not elsewhere classified
  J15.6 Pneumonia due to other Gram-negative bacteria
<table>
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<th>Description</th>
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<tr>
<td>New code</td>
<td>J15.61</td>
<td>Pneumonia due to Acinetobacter baumannii</td>
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<tr>
<td>Add</td>
<td></td>
<td>Use additional code, if applicable, for resistance to carbapenem (Z16.13)</td>
</tr>
<tr>
<td>New code</td>
<td>J15.69</td>
<td>Pneumonia due to other Gram-negative bacteria</td>
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<tr>
<td>Add</td>
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<td>Pneumonia due to other aerobic Gram-negative bacteria</td>
</tr>
<tr>
<td>Add</td>
<td></td>
<td>Pneumonia due to Serratia marcescens</td>
</tr>
<tr>
<td>Z16</td>
<td></td>
<td>Resistance to antimicrobial drugs</td>
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<tr>
<td>Z16.1</td>
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<td>Resistance to beta lactam antibiotics</td>
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<tr>
<td>New code</td>
<td>Z16.13</td>
<td>Resistance to carbapenem</td>
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<tr>
<td>Z22</td>
<td></td>
<td>Carrier of infectious disease</td>
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<tr>
<td>Z22.3</td>
<td></td>
<td>Carrier of other specified bacterial diseases</td>
</tr>
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<td>New sub-subcategory</td>
<td>Z22.34</td>
<td>Carrier of Acinetobacter baumannii</td>
</tr>
<tr>
<td>New code</td>
<td>Z22.340</td>
<td>Carrier of carbapenem-resistant Acinetobacter baumannii</td>
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<tr>
<td>New code</td>
<td>Z22.341</td>
<td>Carrier of carbapenem-sensitive Acinetobacter baumannii</td>
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<tr>
<td>New code</td>
<td>Z22.349</td>
<td>Carrier of Acinetobacter baumannii, unspecified</td>
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<td>Z22.35</td>
<td>Carrier of Enterobacterales</td>
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<td>Carrier of E. coli</td>
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<tr>
<td>Add</td>
<td></td>
<td>Carrier of K. pneumoniae</td>
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<tr>
<td>New code</td>
<td>Z22.350</td>
<td>Carrier of carbapenem-resistant Enterobacterales</td>
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<td>New code</td>
<td>Z22.358</td>
<td>Carrier of other Enterobacterales</td>
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<td>Carrier of carbapenem-sensitive Enterobacterales</td>
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<tr>
<td>Add</td>
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<td>Carrier of extended-spectrum beta-lactamase producing Enterobacterales</td>
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<tr>
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<td>Carrier of ESBL-producing Enterobacterales</td>
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<tr>
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<td>Z22.359</td>
<td>Carrier of Enterobacterales, unspecified</td>
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Acute HIV Infection Syndrome and HIV Pre-Exposure Prophylaxis (PrEP)

HIV disease has been a significant public health issue for many years, and despite improvements, there is a need for continuing efforts to reduce the burden of HIV disease. Acute HIV infection syndrome is the initial infection with HIV, which involves symptoms, but is appropriate to differentiate from AIDS, or later HIV disease.

Pre-exposure prophylaxis (PrEP) for HIV is recommended for people at risk of HIV to prevent them from getting HIV from sex or injection drug use. There are needs to increase awareness of pre-exposure prophylaxis among some at risk groups, to enable wider use, and reduce the burden of HIV disease.

A request for a specific code for pre-exposure prophylaxis (PrEP) for HIV was received from a private individual, referencing the needs for tracking this. Acute HIV infection syndrome is a code in the WHO ICD-10, and this proposal would add it to ICD-10-CM. This proposal is based on internal NCHS review of these issues, and CDC HIV expert review is supportive.

References

TABULAR MODIFICATIONS

B20  Human immunodeficiency virus [HIV] disease

Add  Excludes1: acute HIV infection syndrome (B23.0)

New Category  B23  Human immunodeficiency virus [HIV] disease resulting in other conditions

New code  B23.0  Acute HIV infection syndrome

Z29  Encounter for other prophylactic measures

Z29.8  Encounter for other specified prophylactic measures

New code  Z29.81  Encounter for HIV pre-exposure prophylaxis

Add  Code also, if applicable, risk factors for HIV, such as:
Add  contact with and (suspected) exposure to human immunodeficiency virus [HIV] (Z20.6)
Add  high risk sexual behavior (Z72.5-)

New code  Z29.89  Encounter for other specified prophylactic measures
Age-Related Osteoporosis with Current Pathological Fracture, Pelvis

Autologous bone graft is used in a variety of orthopedic and maxillofacial procedures. The iliac crest of the pelvis is the most common site of autologous bone graft harvesting. In patients with osteoporosis, pathological fracture of the iliac crest can occur, either during or after the bone graft harvesting. Published series and reviews have reported incidence rates of up to 1%. This complication may be associated with significant pain and delayed postoperative mobility, although it typically does not require surgical intervention.

Osteoporosis with pathological fracture of pelvis is currently coded as “osteoporosis with current pathological fracture, femur”. This is anatomically incorrect. Codes specific to pelvis are needed to accurately identify this condition. Clinically, treatment of an osteoporotic fracture of the pelvis would differ significantly from treatment of an osteoporotic fracture of the femur. Since ICD-10-CM codes are used for a variety of purposes, including clinical decision making, statistical analysis, patient safety measures, etc., it is important that codes accurately identify the clinical condition they describe.

The Agency for Healthcare Research and Quality (AHRQ) is requesting creation of new codes for “age-related osteoporosis with current pathological fracture of pelvis”, and “other osteoporosis with current pathological fracture of pelvis”. These would allow more accurate reporting of the location of osteoporotic fractures.

References


## TABULAR MODIFICATIONS

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<td>M80</td>
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<td><strong>New code</strong></td>
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<tr>
<td>M80.0B9</td>
<td>Age-related osteoporosis with current pathological fracture, unspecified pelvis</td>
</tr>
</tbody>
</table>

| M80.8  | Other osteoporosis with current pathological fracture |
|        | **New subcategory**                                   |
| M80.8B | Other osteoporosis with current pathological fracture, pelvis |
|        | **New code**                                          |
| M80.8B1| Other osteoporosis with current pathological fracture, right pelvis |
|        | **New code**                                          |
| M80.8B2| Other osteoporosis with current pathological fracture, left pelvis |
|        | **New code**                                          |
| M80.8B9| Other osteoporosis with current pathological fracture, unspecified pelvis |
Alagille Syndrome

Alagille syndrome is a rare genetic disorder that primarily affects the liver. However, this syndrome can affect multiple organ systems of the body including the cardiovascular system, skeletal system, eyes, and kidneys.

The specific symptoms and severity of Alagille syndrome can vary greatly from one person to another, even within the same family. Some individuals may have mild forms of the disorder while others may have more serious forms.

Common symptoms, which often develop during the first three months of life, include blockage of the flow of bile from the liver (cholestasis), yellowing of the skin and mucous membranes (jaundice), poor weight gain and growth, severe itching (pruritus) and pale, loose stools. Additional symptoms include heart murmurs, congenital heart defects, vertebral (back bone) differences, thickening of the ring that normally lines the cornea in the eye (posterior embryotoxon) and distinctive facial features.

Most people with Alagille syndrome have mutations in one copy of the JAG1 gene. A small percentage (less than 1 percent) of patients have mutations of the NOTCH2 gene. These mutations are inherited as autosomal dominant traits, however in about half of cases the mutation occurs as a new change in the individual and was not inherited from a parent. The current estimated incidence of ALGS is between 1:30,000 and 1:45,000 with no difference in gender (https://www.ncbi.nlm.nih.gov/books/NBK1273/).

This condition is currently classified (an inclusion term) under code, Q44.7 Other congenital malfunctions of the liver, which also includes accessory liver, congenital absence of liver congenital hepatomegaly and congenital malformation of liver NOS.

A specific code for Alagille syndrome will effectively enable meeting the needs of clinical practice, patient and provider education, and epidemiology research for a condition for which the medical and scientific information and public health implications have been rapidly evolving. This request will allow for clearer identification of patients with Alagille Syndrome.

This proposal was submitted from the Global Liver Institute (GLI).
TABULAR MODIFICATIONS

Q44  Congenital malformations of gallbladder, bile ducts and liver

Q44.7  Other congenital malformations of liver

Add    Code also, if applicable, associated malformations affecting other systems

Delete    Accessory liver
Delete    Alagille's syndrome
Delete    Congenital absence of liver
Delete    Congenital hepatomegaly
Delete    Congenital malformation of liver NOS

New code    Q44.70 Other congenital malformation of liver, unspecified
Add    Congenital malformation of liver, NOS

New code    Q44.71 Alagille syndrome
Add    Alagille-Watson syndrome

New code    Q44.79 Other congenital malformations of liver
Add    Accessory liver
Add    Congenital absence of liver
Add    Congenital hepatomegaly
Anal Fistula

This topic was presented at the March 2022 Coordination and Maintenance meeting. Based on public comments, revisions to the proposal have been made for reconsideration. An anal fistula is an inflammatory tract or connection between the surface of the anal canal and, most frequently, the perianal skin or perineum, typically evolving from an anal abscess. The disease has significant implications for a patient’s quality of life, as symptoms range from pain and discharge to fecal incontinence.

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

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

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

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

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

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

TABULAR MODIFICATIONS

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<td>or rectal fistula (K60.4-) or anorectal fistula (K60.5-)</td>
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| New subcategory | K60.4  | Rectal fistula |
| Add |  | Excludes1: congenital fistula (Q43.6) |

| New sub-subcategory | K60.41  | Rectal fistula, simple |
| Add |  | Low intersphincteric rectal fistula |
| Add |  | Superficial rectal fistula |
| New code | K60.411  | Rectal fistula, simple, persistent |
| Add |  | Rectal fistula, simple, chronic |
| New code | K60.412  | Rectal fistula, simple, recurrent |
| New code | K60.419  | Rectal fistula, simple, unspecified |

| New sub-subcategory | K60.42  | Rectal fistula, complex |
| Add |  | Extrasphincteric rectal fistula |
| Add |  | Intersphincteric rectal fistula |
| Add |  | Suprasphincteric rectal fistula |
| Add |  | Transsphincteric rectal fistula |
| New code | K60.421  | Rectal fistula, complex, persistent |
| Add |  | Rectal fistula, complex, chronic |
New code       K60.422 Rectal fistula, complex, recurrent
New code       K60.429 Rectal fistula, complex, unspecified

New code       K60.49 Other rectal fistula
Add                  Rectal fistula NOS

New subcategory    K60.5 Anorectal fistula
Add                Excludes1: congenital fistula (Q43.6)

New sub-subcategory K60.51 Anorectal fistula, simple
Add                  Low intersphincteric anorectal fistula
Add                  Superficial anorectal fistula

New code       K60.511 Anorectal fistula, simple, persistent
Add                  Anorectal fistula, simple, chronic

New code       K60.512 Anorectal fistula, simple, recurrent
New code       K60.519 Anorectal fistula, simple, unspecified

New sub-subcategory K60.52 Anorectal fistula, complex
Add                Extrasphincteric anorectal fistula
Add                Intersphincteric anorectal fistula
Add                Suprasphincteric anorectal fistula
Add                Transsphincteric anorectal fistula

New code       K60.521 Anorectal fistula, complex, persistent
Add                  Anorectal fistula, complex, chronic

New code       K60.522 Anorectal fistula, complex, recurrent
New code       K60.529 Anorectal fistula, complex, unspecified

New code       K60.59 Other anorectal fistula
Add                  Anorectal fistula NOS

**INDEX MODIFICATIONS**

Fistula (cutaneous) L98.8
Add                  - anorectal (infectional) K60.5-
Revise              - anus, anal *(recurring)* (infectional) K60.3-
Add                  - rectal (infectional) K60.4-
Anuria, Oliguria and Hepatorenal Syndrome in Complicating the Puerperium

The National Center for Health Statistics (NCHS) received a request to clarify coding related to the current code of O90.4, postpartum acute kidney failure. Currently, anuria, oliguria and hepatorenal syndrome complicating the puerperium are classified together under code O90.4.

The NCHS determined it was best to create separate codes for hepatorenal syndrome complicating the puerperium and create a new code for other postpartum acute kidney failure to include puerperal anuria and oliguria.

Anuria, complicating the puerperium, refers to the lack or no production of urine. Oliguria, complicating the puerperium, refers to low urine output. Hepatorenal syndrome (HRS) is a kidney impairment in decompensated liver disease characterized by decreased renal perfusion caused by severe splanchnic vasodilatation in the absence of shock, renal disease, or nephrotoxic effects of medications or other substances.

The American College of Obstetricians and Gynecologists (ACOG) has reviewed and supports the proposal.

**TABULAR MODIFICATIONS**

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<tr>
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<td>Puerperal oliguria</td>
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<table>
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<tr>
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<th>Description</th>
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<td>Revise</td>
<td>anuria and oliguria complicating the puerperium (O90.4-)</td>
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</table>
INDEX MODIFICATIONS

Anuria R34
Add - puerperal O90.49

Failure, failed
- renal N19
- - following
Revise - - - labor and delivery (acute) O90.49

Revise Hepatorenal syndrome following labor and delivery O90.41

Oliguria R34
Add - puerperal O90.49

Puerperal, puerperium (complicated by, complications)
- failure
Revise - - renal, acute O90.49
Revise - hepatorenal syndrome O90.41
- necrosis, liver (acute) (subacute) (conditions in subcategory K72.0) O26.63
Revise - - with renal failure O90.49
- renal
Revise - - failure O90.49
Revise - uremia (due to renal failure) O90.49

Syndrome
- hepatorenal K76.7
Revise - - following delivery O90.41
Revise - - postpartum, puerperal O90.41
Autosomal Dominant Hypocalcemia

The National Center for Health Statistics received a proposal requesting new ICD-10-CM codes for autosomal dominant hypocalcemia.

Autosomal dominant hypocalcemia, commonly abbreviated as ADH, is a genetic disorder of calcium metabolism mediated by hypoparathyroidism associated with impaired secretion of parathyroid hormone.

There are two types of ADH. ADH type 1 is caused by mutations in the gene \textit{CASR}. This gene codes for a protein called calcium-sensing receptor (CaSR). CaSR plays a key role in controlling parathyroid hormone, which in turn controls calcium metabolism. In ADH, the genetic mutation causes the body to sense and interpret even very low levels of calcium as normal and then prevent any increase to the amount of calcium in the blood. ADH type 2, which is rarer, is caused by mutations in the \textit{GNA11} gene, with similar effect.

Individuals with ADH may have no family history of the condition; in other cases the genetic mutation is familial. The disorder has an autosomal dominance pattern, meaning that one copy of the mutated gene is sufficient to cause the disorder. So, the child of a person with an autosomal dominant condition has a 50% chance of inheriting the condition.

Individuals with ADH may remain asymptomatic. Otherwise, the disorder causes a variety of symptoms which vary from mild, to severe, to debilitating. Symptoms and complications may impact multiple organ systems, particularly the musculoskeletal, nervous, and renal systems.

Acute symptoms commonly involve the muscles because calcium is essential to muscle contraction. For example, people with ADH often suffer from muscle spasms, especially of the hands and feet, muscle cramping and twitching, and paresthesia such as prickling and tingling. More severely, muscle symptoms include bronchospasm, which can lead to respiratory distress, as well as impaired cardiac contractility and arrhythmias.

In the nervous system, manifestations include seizures and abnormal movement, such as gait and coordination disorders. Calcium build-up and formation of calculi in the basal ganglia of the brain, the area which helps to control movement, is a known complication of ADH. Cognitive impairment, sometimes referred to as brain fog, can also arise.

People with ADH are at significantly increased risk of developing renal disorders, notably nephrolithiasis (kidney stones), nephrocalcinosis (permanent calcifications of the kidney tissue itself), and chronic kidney disease up to and including kidney failure. Hypercalciuria, abnormally high concentration of calcium in urine, is a frequent finding and contributes to formation of the kidney stones and calcifications and the impairment of kidney function. Abnormal blood levels of other minerals, such as hyperphosphatemia and hypomagnesemia, are common findings as well.

After identification of the hypocalcemia and related findings, the diagnosis of ADH is made by genetic testing. Median age at diagnosis is about 25 years old, although individuals with the most
severe symptoms tend to present earlier in life, including in infancy, while the disorder may be identified much later in others. ADH is seen equally in men and women. Prevalence of ADH in the US population is not precisely known but a recent analysis of genetic sequencing data coupled with laboratory data from electronic health records arrived at an estimated prevalence of 3.9 per 100,000.

Treatment of ADH focuses on addressing hypocalcemia and the associated symptoms. Individuals routinely take oral calcium supplements, though they may require IV calcium infusions on an urgent basis when acute symptoms are present. Prescription active vitamin D is also commonly used to improve calcium absorption, because hypoparathyroidism impairs the kidneys’ ability to convert the precursor form of vitamin D into its activated form.

Unfortunately, chronic use of calcium and active vitamin D can paradoxically cause worsening hypercalciuria, leading to calcifications in the kidneys and basal ganglia. This requires an ongoing balancing act in the level of treatment to address the symptoms without causing the complications. Due to the narrow range of parameters, patients often remain symptomatic despite currently available treatments. With the associated and significant risks of renal complications, treatment may be avoided in completely asymptomatic individuals.

People with ADH require life-long monitoring. In stable individuals, serum calcium is usually measured twice a year. However, this can be much more frequent when dosages of calcium and active vitamin D are being adjusted, as happens when patients are experiencing symptoms. A 24-hour urine is collected once a year to monitor creatinine, calcium excretion, and other indicators of renal function. As indicated, renal and basal ganglia imaging is periodically performed for calcifications.

In addition to ADH, new codes are being proposed for other types of hypoparathyroidism due to impaired parathyroid hormone secretion. These include secondary hypoparathyroidism, for example associated with neoplasm or thalassemia, and autoimmune hypoparathyroidism in which individuals produce anti-parathyroid gland antibodies.

New ICD-10-CM codes will provide coding specificity to identify individuals with autosomal dominant hypocalcemia for treatment, tracking, and research.

References
## TABULAR MODIFICATIONS

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E83 Disorders of mineral metabolism
Excludes1: dietary mineral deficiency (E58-E61)
parathyroid disorders (E20-E21)
vitamin D deficiency (E55.-)

E83.5 Disorders of calcium metabolism
Add Excludes1: autoimmune hypoparathyroidism (E20.812)
Add autosomal dominant hypocalcemia (E20.810)
chondrocalcinosis (M11.1-M11.2)
hungry bone syndrome (E83.31)
hyperparathyroidism (E21.0-E21.3)
Add secondary hypoparathyroidism in diseases classified
elsewhere (E20.811)
Bronchiolitis Obliterans and Bronchiolitis Obliterans Syndrome

Bronchiolitis obliterans and bronchiolitis obliterans syndrome (BOS) together with chronic lung allograft dysfunction had a prior proposal for ICD-10-CM code expansion, September 2021. Based on comments from that time, a revised proposal is re-presented. Please see the prior proposal for further clinical details.

In brief, bronchiolitis obliterans or obliterative bronchiolitis may occur for a number of reasons, in a clinical syndrome characterized by airflow limitation not reversible with inhaled bronchodilators which may be associated with progressive dyspnea. BOS may occur following lung transplant, with fibrosis involving terminal and respiratory bronchioles; it may also follow stem cell transplant with chronic graft-versus-host-disease. Chronic lung allograft dysfunction may involve BOS, or restrictive allograft syndrome (RAS), or a mix of these, and potentially other clinical issues.

TABULAR MODIFICATIONS

Revise Chronic lower respiratory diseases (J40-J47) (J40-J4A)

J42 Unspecified chronic bronchitis

Add Excludes1: bronchiolitis obliterans (J44.81)

New subcategory J44.8 Other specified chronic obstructive pulmonary disease

New code J44.81 Bronchiolitis obliterans and bronchiolitis obliterans syndrome

Add Code first, if applicable:
Add complication of bone marrow transplant (T86.09)
Add complication of stem cell transplant (T86.5)
Add heart-lung transplant rejection (T86.31)
Add lung transplant rejection (T86.810)
Add other complications of heart-lung transplant (T86.39)
Add other complications of lung transplant (T86.818)

Add Code also, if applicable, associated conditions, such as:
Add chronic graft-versus-host disease (D89.811)
Add chronic lung allograft dysfunction (J4A.-)
Add chronic respiratory conditions due to chemicals, gases, fumes and vapors (J68.4)
New code  J44.89 Other specified chronic obstructive pulmonary disease
Add     Chronic asthmatic (obstructive) bronchitis
Add     Chronic emphysematous bronchitis

J4A  Chronic Lung Allograft Dysfunction
Add     Code first, if applicable:
Add     heart-lung transplant rejection (T86.31)
Add     lung transplant rejection (T86.810)
Add     other complications of heart-lung transplant (T86.39)
Add     other complications of lung transplant (T86.818)
Add     Code also, if applicable: bronchiolitis obliterans syndrome (J44.81)

New code  J4A.0 Restrictive allograft syndrome
Add     Note: for mixed chronic lung allograft dysfunction, code both restrictive allograft syndrome, J4A.0, and bronchiolitis obliterans syndrome, J44.81.

New code  J4A.8 Other chronic lung allograft dysfunction

New code  J4A.9 Chronic lung allograft dysfunction, unspecified

J68  Respiratory conditions due to inhalation of chemicals, gases, fumes and vapors

J68.4  Chronic respiratory conditions due to chemicals, gases, fumes and vapors
Delete     Emphysema (diffuse) (chronic) due to inhalation of chemicals, gases, fumes and vapors
Delete     Obliterative bronchiolitis (chronic) (subacute) due to inhalation of chemicals, gases, fumes and vapors
Delete     Pulmonary fibrosis (chronic) due to inhalation of chemicals, gases, fumes and vapors
Add     Use additional code, if applicable, for chronic conditions, such as:
Add     emphysema (J43.-)
Add     obliterative bronchiolitis (J44.81)
Add     pulmonary fibrosis (J84.10)
INDEX MODIFICATIONS

Asthma, asthmatic (bronchial) (catarrh) (spasmodic) J45.909
- revise - chronic obstructive J44.89
- revise - with chronic obstructive bronchitis J44.89
- revise - with chronic obstructive pulmonary disease J44.89

Bronchiolitis (acute) (infective) (subacute) J21.9
- revise - chronic (fibrosing) (obliterative) J44.89
- add - - obliterative J44.81
- revise - fibrosa obliterans J44.9 J44.81
- revise - obliterans J42 (see also Bronchiolitis, obliterative) J44.81
- add - - syndrome J44.81
- revise - obliterative (chronic) (subacute) (see also Bronchiolitis, obliterans) J44.9 J44.81

Bronchitis (diffuse) (fibrinous) (hypostatic) (infective) (membranous) J40
- - asthmatic J45.9
- - chronic J44.89
- chronic J42
- revise - - asthmatic (obstructive) J44.89
- revise - - emphysematous J44.89
- revise - - obliterans J44.9 see Bronchiolitis, obliterans
- revise - - obstructive J44.89
- revise - - with airways obstruction J44.89
- revise - emphysematous (obstructive) J44.89
- revise - obliterans (chronic) J44.9 see Bronchiolitis, obliterans
- revise - obstructive (chronic) (diffuse) J44.89
- revise - with obstruction (airway) (lung) J44.89

Disease, diseased - see also Syndrome
- lung J98.4
- - obstructive (chronic) J44.9
- revise - - with bronchitis J44.89

Dyspnea (nocturnal) (paroxysmal) R06.00
- asthmatic (bronchial) J45.909
- - with
- - - bronchitis J45.909
- revise - - - chronic J44.89

Obstruction, obstructed, obstructive
- airway J98.8
- revise - - with bronchitis (chronic) J44.89
Pneumatocele (lung) J98.4
   Revise - tension J44.9 J98.8

   Revise Vanishing lung J44.89
**Chronic Migraine with Aura**

The National Center for Health Statistics received a proposal from Wisconsin Physicians Service Corporation requesting a new ICD-10-CM code for chronic migraine with aura.

“Migraine is a common, multifactorial brain disorder with recurring disabling attacks of headache and associated features.” These features include migraine with and without auras. The international Classification of Headache Disorders, third edition defines Chronic Migraine as a headache occurring on 15 or more days a month for more than three months, which, on at least eight days a month has the features of migraine headache. Chronic headache criteria includes both migraine with and without aura.

A new ICD-10-CM code will provide coding specificity for individuals with chronic migraines with aura for treatment and research.

**References**


**TABULAR MODIFICATIONS**

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<td>Add</td>
<td>Excludes1: migraine with aura (G43.1-)</td>
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<td>G43.E0 Chronic migraine with aura, not intractable</td>
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<td>Add</td>
<td>Chronic migraine with aura, without refractory migraine</td>
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<td>G43.E01 Chronic migraine with aura, not intractable, with status migrainosus</td>
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<tr>
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<td>G43.E09 Chronic migraine with aura, not intractable, without status migrainosus</td>
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<td>Add</td>
<td>Chronic migraine with aura NOS</td>
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Coronary Microvascular Dysfunction

Approximately 112 million people globally are affected by angina\(^1\) and a significant proportion of these patients experience ischemia with non-obstructive coronary arteries (INOCA) and myocardial infarction with non-obstructive coronary arteries (MINOCA) due to pathologies in the microvasculature.\(^2\) The microcirculatory system is largely responsible for the regulation and distribution of blood flow to the myocardium and is composed of an extensive network of narrow vessels downstream from the epicardial arteries.\(^2\) Coronary microvascular dysfunction (CMD) is a condition that impacts the microvasculature by restricting microvascular flow and increasing microvascular resistance. Manifestations of CMD widely range in severity and presentation from angina to heart failure.\(^2,3\)

Due to the growing body of evidence on CMD and its clinical impact on patients, the 2021 Guideline for the Evaluation and Diagnosis of Chest Pain indicated that patients with INOCA benefit from an assessment of functional significance to help guide management of the condition.\(^4\) The guidelines also recommend testing practices that can aid in the diagnosis of microvascular flow abnormalities to support risk stratification.\(^4\) Additionally, studies have indicated that the presence of CMD is not benign, as it is associated with significantly higher rates of major adverse cardiac events.\(^5\) There is higher risk of CMD in women, those with hypertension, diabetes, and other insulin-resistant states.\(^4\)

A proposal to create specific ICD-10-CM codes for CMD was received from Abbott Laboratories, Inc., to ensure afflicted patients receive appropriate diagnosis and care.

References

**TABULAR MODIFICATIONS**

I20 Angina pectoris

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<td>Delete</td>
<td>Angina equivalent</td>
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<tr>
<td>Delete</td>
<td>Angina of effort</td>
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Delete  Coronary slow flow syndrome
Delete  Stenocardia
Delete  Stable angina

New code  I20.81 Angina pectoris with coronary microvascular dysfunction
Add  Angina pectoris with coronary microvascular disease

New code  I20.89 Other forms of angina pectoris
Add  Angina equivalent
Add  Angina of effort
Add  Coronary slow flow syndrome
Add  Stenocardia
Add  Stable angina

I21 Acute myocardial infarction

New code  I21.B Myocardial infarction with coronary microvascular dysfunction
Add  Myocardial infarction with coronary microvascular disease
Add  Myocardial infarction with nonobstructive coronary arteries [MINOCA] with microvascular disease

I24 Other acute ischemic heart diseases

I24.8 Other forms of acute ischemic heart disease

New code  I24.81 Acute presentation of coronary microvascular dysfunction
Add  Acute (presentation of) coronary microvascular disease

New code  I20.89 Other forms of acute ischemic heart disease

I25 Chronic ischemic heart disease

I25.8 Other forms of chronic ischemic heart disease

New code  I25.85 Chronic presentation of coronary microvascular dysfunction
Add  Chronic (presentation of) coronary microvascular disease
Dense Breast(s) on Mammography

The National Center for Health Statistics received a request for a new ICD-10-CM codes to identify dense breast(s) on mammography. Breasts are made up of lobules, ducts, fatty and fibrous connective tissue. Lobules are the small glands that produce milk, while ducts are the tiny tubes that carry the milk from the lobules to the nipple. Together, the lobules and ducts are referred to as glandular tissue. Fibrous tissue and fat give breasts their size and shape and hold the other structures in place.

Fibrous and glandular tissue are harder to see through on a mammogram, so the breast tissue may be called ‘dense’. Having dense breast tissue is common. Some women have more dense breast tissue. For most women, breasts become less dense with age. For others, there’s little change.

The Breast Imaging Reporting and Data System, called BI-RADS, is used to group different types of breast density. This system, developed by the American College of Radiology, helps clinicians to interpret and report specific mammogram findings. BI-RADS classifies breast density into four categories, as follows:

(a) Almost entirely fatty breast tissue, found in about 10% of women
(b) Scattered areas of dense glandular tissue and fibrous connective tissue (scattered fibroglandular breast tissue) found in about 40% of women
(c) Heterogeneously dense breast tissue with many areas of glandular tissue and fibrous connective tissue, found in about 40% of women
(d) Extremely dense breast tissue, found in about 10% of women

It is proposed to create new codes to allow code assignment to capture both the screening mammogram and the finding of dense breasts on the same encounter.

References
https://www.cdc.gov/cancer/breast/basic_info/dense-breasts.htm
https://www.cancer.gov/types/breast/breast-changes/dense-breasts
TABULAR MODIFICATIONS

R92 Abnormal and inconclusive findings on diagnostic imaging of breast
   R92.2 Inconclusive mammogram
      Delete Dense breasts NOS
      Inconclusive mammogram NEC
      Delete Inconclusive mammography due to dense breasts
      Inconclusive mammography NEC
      Add Code also, if applicable, density of breast (R92.3-)

New subcategory R92.3 Mammographic density found on imaging of breast
      Add Code also, if applicable, inconclusive mammogram (R92.2)

New code R92.30 Dense breasts, unspecified
      Add Dense breasts NOS
      Add Low density

New subcategory R92.31 Mammographic fatty tissue density of breast
      Add Breast Imaging Reporting and Data System (BI-RADS): A
      New code R92.311 Mammographic fatty tissue density of right breast
      New code R92.312 Mammographic fatty tissue density of left breast
      New code R92.313 Mammographic fatty tissue density of bilateral breasts

New subcategory R92.32 Mammographic fibroglandular density of breast
      Add Breast Imaging Reporting and Data System (BI-RADS): B
      New code R92.321 Mammographic fibroglandular density of right breast
      New code R92.322 Mammographic fibroglandular density of left breast
      New code R92.323 Mammographic fibroglandular density of bilateral breasts

New subcategory R92.33 Mammographic heterogeneous density of breast
      Add Breast Imaging Reporting and Data System (BI-RADS): C
      New code R92.331 Mammographic heterogeneous density of right breast
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New code R92.332 Mammographic heterogeneous density of left breast
New code R92.333 Mammographic heterogeneous density of bilateral breasts
New subcategory R92.34 Mammographic extreme density of breast
Add Breast Imaging Reporting and Data System (BI-RADS): D
New code R92.341 Mammographic extreme density of right breast
New code R92.342 Mammographic extreme density of left breast
New code R92.343 Mammographic extreme density of bilateral breasts

INDEX MODIFICATIONS

Breast - see also condition
Revise - dense R92.2 R92.3-
Revise Dense breasts R92.2 R92.3-
Density
Add - breast R92.3-

Findings, abnormal, inconclusive, without diagnosis - see also Abnormal
- mammogram NEC R92.8
Delete - - inconclusive result (due to dense breasts) R92.2

Inconclusive
Delete - mammogram (due to dense breasts) R92.2
Eating Disorders

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

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

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

The American Psychiatric Association has reviewed and supports this proposal.

TABULAR MODIFICATIONS

F50 Eating disorders
   Excludes1: anorexia NOS (R63.0)
      feeding problems of newborn (P92.-)
      polyphagia (R63.2)

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

F50.0 Anorexia nervosa
   Excludes1: loss of appetite (R63.0)
      psychogenic loss of appetite (F50.89)
   F50.00 Anorexia nervosa, unspecified

   Add Anorexia nervosa, restricting type
   Add Anorexia nervosa, restricting type, extreme
   Add Anorexia nervosa, restricting type, in full remission
   Add Anorexia nervosa, restricting type, in partial remission
   Add Anorexia nervosa, restricting type, mild
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Add  Anorexia nervosa, restricting type, moderate
Add  Anorexia nervosa, restricting type, severe

F50.02 Anorexia nervosa, binge eating/purging type
Excludes1: bulimia nervosa (F50.2)
Add  Anorexia nervosa, binge eating/purging type, extreme
Add  Anorexia nervosa, binge eating/purging type, in full remission
Add  Anorexia nervosa, binge eating/purging type, in partial remission
Add  Anorexia nervosa, binge eating/purging type, mild
Add  Anorexia nervosa, binge eating/purging type, moderate
Add  Anorexia nervosa, binge eating/purging type, severe

F50.2 Bulimia nervosa
Bulimia NOS
Hyperorexia nervosa
Excludes1: anorexia nervosa, binge eating/purging type (F50.02)
Add  Bulimia nervosa, restricting type, extreme
Add  Bulimia nervosa, restricting type, in full remission
Add  Bulimia nervosa, restricting type, in partial remission
Add  Bulimia nervosa, restricting type, mild
Add  Bulimia nervosa, restricting type, moderate
Add  Bulimia nervosa, restricting type, severe

F50.8 Other eating disorders
Delete  Excludes2: pica of infancy and childhood (F98.3)
F50.81 Binge eating disorder
Add  Binge eating disorder, extreme
Add  Binge eating disorder in full remission
Add  Binge eating disorder in partial remission
Add  Binge eating disorder, mild
Add  Binge eating disorder, moderate
Add  Binge eating disorder, severe

F50.82 Avoidant/restrictive food intake disorder
Add  Avoidant/restrictive food intake disorder, in remission

New code  F50.83 Pica in adults
Add  Pica in adults, in remission
Add  Excludes1: pica of infancy and childhood (F98.3)
New
F50.84 Rumination disorder in adults
Add
   Rumination in adults, in remission
   Excludes1: rumination disorder in infancy and childhood (F98.21)

Delete
F50.89 Other specified eating disorder
Pica in adults
Psychogenic loss of appetite

F98.2 Other feeding disorders of infancy and childhood
   Excludes2: anorexia nervosa and other eating disorders (F50.-)
      feeding difficulties (R63.3)
      feeding problems of newborn (P92.-)
      pica in infancy and childhood (F98.3)

Revise
F98.21 Rumination disorder of infancy and childhood
Add
   Excludes1: rumination disorder in adults (F50.84)

F98.3 Pica of infancy and childhood
Add
   Excludes1: pica in adults (F50.83)
Add
   Pica in infancy and childhood, in remission
**Extraocular Muscle Entrapment**

This is a representation from the September 2021 and March 2022 ICD-10 Coordination and Maintenance Committee Meetings of the Extraocular Muscle Entrapment proposal with the recommended modification to add “unspecified” to the sub-subcategory H50.68, Extraocular muscle entrapment. The modifications are in **bold**.

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

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

The National Center for Health Statistics received a proposal requesting for the creation of ICD-10-CM codes for extraocular muscle entrapment for coding specificity and research.

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

**References**


**TABULAR MODIFICATIONS**

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<th>Mechanical strabismus</th>
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Familial Adenomatous Polyposis

Familial adenomatous polyposis (FAP) is an inherited disorder which predisposes to colon cancer, and also is associated with development of large numbers of colon polyps. There are three types of FAP, classic, attenuated, and autosomal recessive. Both the classic and attenuated types are cause by mutations in the APC gene, while the autosomal recessive type is caused by mutations in the MUTYH gene (also called MUTYH-associated polyposis).

The autosomal recessive FAP is a milder type of familial adenomatous polyposis. People with the autosomal recessive type have fewer polyps than those with the classic type. Fewer than 100 polyps typically develop, rather than hundreds or thousands.

The genetic mutations affect cell ability to maintain normal growth and function. Cell overgrowth resulting from mutation leads to the colon polyps seen in familial adenomatous polyposis. While it is expected that most people with mutations in the APC gene will develop colorectal cancer, the number of polyps and the time frame in malignancy develops depend on the particular mutation in the gene.

People with the classic type of familial adenomatous polyposis may develop multiple colon polyps in their teenage years. Without colectomy, polyps will be expected to become malignant. The average age for colon cancer to develop in classic familial adenomatous polyposis is 39 years. In attenuated FAP, polyp growth is delayed, and the average age of colorectal cancer development is 55 years.

In people with classic FAP, the number of polyps increases with age and hundreds to thousands of polyps can develop in the colon. This may also be development of desmoid tumors, noncancerous or uncertain behavior growths. These are fibrous tumors, that may be provoked by colectomy, and may tend to recur after being surgically removed. In both classic and attenuated FAP, benign and malignant tumors may sometimes be found in other places in the body, including the duodenum, stomach, bones, skin, and other tissues; the combination of colon polyps and growths outside the colon may be referred to as Gardner syndrome.

The autosomal recessive FAP is a milder type of familial adenomatous polyposis. People with the autosomal recessive type have fewer polyps than those with the classic type. Fewer than 100 polyps typically develop, rather than hundreds or thousands.

Reference
MedlinePlus. Familial adenomatous polyposis. National Library of Medicine, NIH.
https://medlineplus.gov/genetics/condition/familial-adenomatous-polyposis/
TABULAR MODIFICATIONS

Option #1

Z15  Genetic susceptibility to disease
    Z15.8  Genetic susceptibility to other disease

New code  Z15.82  Familial adenomatous polyposis
Add        Code also associated clinical findings, such as:
Add        benign neoplasm of colon (D12.6)
Add        malignant neoplasm of colon (C18.-)

Option #2

D13  Benign neoplasm of other and ill-defined parts of digestive system
    D13.9  Benign neoplasm of ill-defined sites within the digestive system

New code  D13.91  Familial adenomatous polyposis
Add        Code also associated clinical findings, such as:
Add        benign neoplasm of colon (D12.6)
Add        malignant neoplasm of colon (C18.-)

New code  D13.99  Benign neoplasm of ill-defined sites within the digestive system
Family History of Adenomatous Polyps

The National Center for Health Statistics received a proposal requesting new ICD-10-CM codes for family history of adenomatous polyps.

It is important to identify adenomatous/serrated polyps (D12.-), or adenoma, current or by history, as they carry a high risk of cancer and are considered premalignant. About 70% colorectal cancers originate from adenomatous adenomas and 25% - 30% arise from (sessile) serrated polyps.¹ This is relevant to colorectal cancer screening and surveillance. The addition of the new code will provide more specific data for research and screening protocols.

New ICD-10-CM codes will provide coding specificity for family history of adenomatous polyps for individuals with this familial risk factor.

References:
¹ACG Clinical Guidelines: Colorectal Cancer Screening 2021: Official journal of the American College of Gastroenterology | ACG (lww.com)
²Task Force on Colorectal Cancer Recommends Colon Cancer Screening at 45 - Gastroenterologist San Antonio (gastroconsa.com)
³Understanding Your Pathology Report: Colon Polyps (Sessile or Traditional Serrated Adenomas) (cancer.org)

TABULAR MODIFICATIONS

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<td>Z83.71</td>
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<tr>
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<td>(Z83.71-.)</td>
</tr>
<tr>
<td>New code</td>
<td>Family history of adenomatous and serrated</td>
</tr>
<tr>
<td></td>
<td>polyps</td>
</tr>
<tr>
<td>Add</td>
<td>Conditions classifiable to D12.-</td>
</tr>
<tr>
<td>Add</td>
<td>Family history of tubular adenoma polyps</td>
</tr>
<tr>
<td>Add</td>
<td>Family history of tubulovillous adenoma polyps</td>
</tr>
<tr>
<td>Add</td>
<td>Family history of villous adenoma polyps</td>
</tr>
<tr>
<td>New code</td>
<td>Family history of hyperplastic colon polyps</td>
</tr>
<tr>
<td>New code</td>
<td>Other family colon polyps</td>
</tr>
<tr>
<td>Add</td>
<td>Family history of inflammatory colon polyps</td>
</tr>
<tr>
<td>New code</td>
<td>Family history of colon polyps, unspecified</td>
</tr>
<tr>
<td>Add</td>
<td>Family history of colon polyps NOS</td>
</tr>
</tbody>
</table>
Flank Anatomical Specificity

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

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

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

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

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

**TABULAR MODIFICATIONS**

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

L02.21 Cutaneous abscess of trunk

Revise L02.212 Cutaneous abscess of back [any part, except buttock and flank]
New code    L02.217 Cutaneous abscess of flank

L02.22 Furuncle of trunk
    Boil of trunk
    Folliculitis of trunk
Revised    L02.222 Furuncle of back [any part, except buttock and flank]
New code    L02.227 Furuncle of flank

L02.23 Carbuncle of trunk
Revise    L02.232 Carbuncle of back [any part, except buttock and flank]
New code    L02.237 Carbuncle of flank

L03 Cellulitis and acute lymphangitis
L03.3 Cellulitis and acute lymphangitis of trunk
    L03.31 Cellulitis of trunk
New code    L03.31A Cellulitis of flank

L03.32 Acute lymphangitis of trunk
New code    L03.32A Acute lymphangitis of flank

R10 Abdominal and pelvic pain
Excludes1: renal colic (N23)
Add    Excludes2: costovertebral (angle) tenderness (R39.85)
        dorsalgia (M54.-)
Add    flatulence and related conditions (R14.-)

R10.1 Pain localized to upper abdomen
Add    Excludes2: pain localized to lateral abdomen (R10.A-)
Add    pelvic and perineal pain (R10.2-)

R10.2 Pelvic and perineal pain
Add    Excludes2: pain localized to other parts of lower abdomen
        (R10.3-)
Add    pain localized to upper abdomen (R10.1-)

New code    R10.20 Pelvic and perineal pain unspecified side
New code    R10.21 Pelvic and perineal pain right side
New code    R10.22 Pelvic and perineal pain left side
New code    R10.23 Pelvic and perineal pain bilateral
New code  R10.24  Suprapubic pain

R10.3  Pain localized to other parts of lower abdomen
Add  Excludes2: pain localized to lateral abdomen R10.4-
Add  pelvic and perineal pain (R10.2-)

New subcategory  R10.A  Pain localized to lateral flank abdomen
Add  Latus pain
Add  Excludes2: pain localized to other parts of lower abdomen (R10.3-)
Add  pain localized to upper abdomen (R10.1-)
New code  R10.A0  Flank pain, unspecified side
New code  R10.A1  Flank pain, right side
New code  R10.A2  Flank pain, left side
New code  R10.A3  Flank pain, bilateral

R10.8  Other abdominal pain

New subcategory  R10.8A  Flank tenderness
New code  R10.8A1  Right flank tenderness
New code  R10.8A2  Left flank tenderness
New code  R10.8A3  Suprapubic tenderness

New code  R10.85  Abdominal pain of multiple sites
Add  Excludes1: abdominal rigidity NOS (R19.3)
Add  generalized abdominal pain associated with acute abdomen (R10.0)
Add  generalized abdominal pain NOS (R10.84)
Add  localized abdominal pain (R10.1-R10.4-)

R39  Other and unspecified symptoms and signs involving the genitourinary system

R39.8  Other symptoms and signs involving the genitourinary system
New code  R39.85  Costovertebral (angle) tenderness
Add  Excludes2: abdominal and pelvic pain (R10.-)

S30  Superficial injury of abdomen, lower back, pelvis and external genitals
S30.1  Contusion of abdominal wall and latus region
Delete  Contusion of flank
Delete  Contusion of groin
ICD-10 Coordination and Maintenance Committee Meeting
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New code S30.10 Contusion of abdominal wall and latus region, unspecified
New code S30.11 Contusion of abdominal wall and latus region
New code S30.12 Contusion of flank
New code S30.13 Contusion of groin

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

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

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

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

S30.85 Superficial foreign body of abdomen, lower back, pelvis, and external genitals
New code S30.85A Superficial foreign body of flank

S30.86 Insect bite (nonvenomous) of abdomen, lower back, pelvis, and external genitals
New code S30.86A Insect bite (nonvenomous) of flank

S30.87 Other superficial bite of abdomen, lower back, pelvis, and external genitals
New code S30.87A Other superficial bite of flank

S30.9 Unspecified superficial injury of abdomen, lower back, pelvis, and external genitals
New code S30.9A Unspecified superficial injury of flank
S31 Open wound of abdomen, lower back, pelvis and external genitals

S31.1 Open wound of abdominal wall without penetration into peritoneal cavity

S31.10 Unspecified open wound of abdominal wall without penetration into peritoneal cavity

New code S31.106 Unspecified open wound of abdominal wall, right flank without penetration into peritoneal cavity

New code S31.107 Unspecified open wound of abdominal wall, left flank without penetration into peritoneal cavity

New code S31.10A Unspecified open wound of abdominal wall, unspecified flank without penetration into peritoneal cavity

Add Open wound of abdominal wall of flank NOS without penetration into peritoneal cavity

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

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

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

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

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

S31.12 Laceration with foreign body of abdominal wall without penetration into peritoneal cavity
New code S31.126 Laceration with foreign body of abdominal wall, right flank without penetration into peritoneal cavity

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

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

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

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

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

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

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

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

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

New code S31.146 Puncture wound of abdominal wall with foreign body, right flank without penetration into peritoneal cavity
ICD-10 Coordination and Maintenance Committee Meeting
September 13-14-2022

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

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

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

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

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

New code
S31.157 Open bite of abdominal wall, left flank without penetration into peritoneal cavity

New code
S31.15A Open bite of abdominal wall, unspecified flank without penetration into peritoneal cavity

Add
Open bite of abdominal wall of flank NOS without penetration into peritoneal cavity

S31.6 Open wound of abdominal wall with penetration into peritoneal cavity

S31.60 Unspecified open wound of abdominal wall with penetration into peritoneal cavity

New code
S31.606 Unspecified open wound of abdominal wall, right flank with penetration into peritoneal cavity

New code
S31.607 Unspecified open wound of abdominal wall, left flank with penetration into peritoneal cavity

New code
S31.60A Unspecified open wound of abdominal wall, unspecified flank with penetration into peritoneal cavity

Add
Unspecified open wound of abdominal wall of flank NOS, with penetration into peritoneal cavity
S31.61 Laceration without foreign body of abdominal wall with penetration into peritoneal cavity

New code  S31.616 Laceration without foreign body of abdominal wall, right flank with penetration into peritoneal cavity

New code  S31.617 Laceration without foreign body of abdominal wall, left flank with penetration into peritoneal cavity

New code  S31.61A Laceration without foreign body of abdominal wall, unspecified flank with penetration into peritoneal cavity

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

S31.62 Laceration with foreign body of abdominal wall with penetration into peritoneal cavity

New code  S31.626 Laceration with foreign body of abdominal wall, right flank with penetration into peritoneal cavity

New code  S31.627 Laceration with foreign body of abdominal wall, left flank with penetration into peritoneal cavity

New code  S31.62A Laceration with foreign body of abdominal wall, unspecified flank with penetration into peritoneal cavity

Add  Laceration with foreign body of abdominal wall, flank NOS, with penetration into peritoneal cavity

S31.63 Puncture wound without foreign body of abdominal wall with penetration into peritoneal cavity
New code S31.636 Puncture wound of abdominal wall without foreign body, right flank with penetration into peritoneal cavity

New code S31.637 Puncture wound of abdominal wall without foreign body, left flank with penetration into peritoneal cavity

New code S31.63A Puncture wound of abdominal wall without foreign body, unspecified flank with penetration into peritoneal cavity

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

S31.64 Puncture wound with foreign body of abdominal wall with penetration into peritoneal cavity

New code S31.646 Puncture wound of abdominal wall with foreign body, right flank with penetration into peritoneal cavity

New code S31.647 Puncture wound of abdominal wall with foreign body, left flank with penetration into peritoneal cavity

New code S31.64A Puncture wound of abdominal wall with foreign body, unspecified flank with penetration into peritoneal cavity

Add Puncture wound of abdominal wall with foreign body, flank NOS, with penetration into peritoneal cavity

S31.65 Open bite of abdominal wall with penetration into peritoneal cavity

New code S31.656 Open bite of abdominal wall, right flank with penetration into peritoneal cavity
<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>New code</td>
<td>S31.657 Open bite of abdominal wall, left flank with penetration into peritoneal cavity</td>
</tr>
<tr>
<td>New code</td>
<td>S31.65A Open bite of abdominal wall, unspecified flank with penetration into peritoneal cavity</td>
</tr>
<tr>
<td>Add</td>
<td>Open bite of abdominal wall, flank NOS, with penetration into peritoneal cavity</td>
</tr>
</tbody>
</table>
Foreign Body Entering Into or Through a Natural Orifice

Foreign bodies can enter through natural body orifices. Some of which are benign and cause irritation (i.e. bead in the ear or nose). However other types of foreign bodies can have significant morbidity or mortality.

Button batteries can result in rapid caustic tissue injury with both acute and chronic complications. An extensive analysis of 8648 battery ingestion cases, 8161 were button batteries and 487 were cylindrical cells (AA, AAA). Of the button battery ingestions, 62.5% were in children under 6 years of age and 15.9% involved adults over 60 years old1. It has been reported that 12.6% of children under age 6 who ingested a 20 mm button battery suffered a major complication2. Ingestion of multiple magnets can cause serious conditions such as pinch the intestine walls quickly resulting in tissue necrosis and bowel perforation3.

Currently in ICD-10-CM, there is not a specific way to identify these more serious types of foreign bodies entering into or through a natural orifice. Instead of adding each of these types of foreign bodies to the existing codes in T16-T19, the American Academy of Pediatrics is requesting that the WHO ICD-10 code category W44, Foreign body entering into or through eye or natural orifice be incorporated in the ICD-10-CM classification structure.

The American Academy of Pediatrics are requesting the following tabular modifications.

REFERENCES

TABULAR MODIFICATIONS

Effects of foreign body entering through natural orifice (T15-T19)

Excludes2: foreign body accidentally left in operation wound (T81.5-)

foreign body in penetrating wound - See open wound by body region residual
foreign body in soft tissue (M79.5)

splinter, without open wound - See superficial injury by body region

Add Code also, if known, foreign body entering into or through a natural orifice (W44.-)
New category  W44 Foreign body entering into or through a natural orifice
Add  Excludes2: contact with sharp glass (W25)
Take with other sharp objects (W26)
foreign body or object entering through skin (W45)

Add  The appropriate 7th character is to be added to each code
from category W44
Add  A - initial encounter
Add  D - subsequent encounter
Add  S - sequela

New subcategory  W44.A Battery entering into or through a natural orifice
New code  W44.A0 Battery unspecified, entering into or through a
natural orifice
New code  W44.A1 Button battery entering into or through a natural
orifice
New code  W44.A9 Other batteries entering into or through a natural
orifice
New subcategory  W44.B Plastic entering into or through a natural orifice
New code  W44.B0 Plastic object unspecified, entering into or through a
natural orifice
New code  W44.B1 Plastic bead entering into or through a natural
orifice
New code  W44.B2 Plastic coin entering into or through a natural
orifice
New code  W44.B3 Plastic toy and toy part entering into or through a natural
orifice
New code  W44.B4 Plastic jewelry entering into or through a natural
orifice
New code  W44.B5 Plastic bottle entering into or through a natural
orifice
New code W44.B9 Other plastic object entering into or through a natural orifice

New subcategory W44.C Glass entering into or through a natural orifice

New code W44.C0 Glass unspecified, entering into or through a natural orifice

New code W44.C1 Sharp glass entering into or through a natural orifice

Add Glass shard entering into or through a natural orifice

New code W44.C2 Intact glass entering into or through a natural orifice

Add Intact glass bottle entering into or through a natural orifice

New subcategory W44.D Magnetic metal entering into or through a natural orifice

New code W44.D0 Magnetic metal object unspecified, entering into or through a natural orifice

New code W44.D1 Magnetic metal bead entering into or through a natural orifice

New code W44.D2 Magnetic metal coin entering into or through a natural orifice

New code W44.D3 Magnetic metal toy entering into or through a natural orifice

New code W44.D4 Magnetic metal jewelry entering into or through a natural orifice

New code W44.D9 Other magnetic metal objects entering into or through a natural orifice

New subcategory W44.E Non-magnetic metal entering into or through a natural orifice

New code W44.E0 Non-magnetic metal object unspecified, entering into or through a natural orifice
<table>
<thead>
<tr>
<th>New code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>W44.E1</td>
<td>Non-magnetic metal bead entering into or through a natural orifice</td>
</tr>
<tr>
<td>W44.E2</td>
<td>Non-magnetic metal coin entering into or through a natural orifice</td>
</tr>
<tr>
<td>W44.E3</td>
<td>Non-magnetic metal toy entering into or through a natural orifice</td>
</tr>
<tr>
<td>W44.E4</td>
<td>Non-magnetic metal jewelry entering into or through a natural orifice</td>
</tr>
<tr>
<td>W44.E9</td>
<td>Other non-magnetic metal objects entering into or through a natural orifice</td>
</tr>
<tr>
<td>Add</td>
<td>Bottle cap entering into or through a natural orifice</td>
</tr>
<tr>
<td>Add</td>
<td>Can lid entering into or through a natural orifice</td>
</tr>
<tr>
<td>Add</td>
<td>Pull tab entering into or through a natural orifice</td>
</tr>
<tr>
<td>W44.F1</td>
<td>Bezoar entering into or through a natural orifice</td>
</tr>
<tr>
<td>W44.F2</td>
<td>Rubber band entering into or through a natural orifice</td>
</tr>
<tr>
<td>W44.F3</td>
<td>Food entering into or through a natural orifice</td>
</tr>
<tr>
<td>W44.F4</td>
<td>Insect entering into or through a natural orifice</td>
</tr>
<tr>
<td>W44.F9</td>
<td>Other object of natural or organic material, entering into or through a natural orifice</td>
</tr>
<tr>
<td>W44.G0</td>
<td>Other non-organic objects unspecified, entering into or through a natural orifice</td>
</tr>
<tr>
<td>W44.G1</td>
<td>Audio device entering into or through a natural orifice</td>
</tr>
</tbody>
</table>
Add Ear buds
Add Hearing aids

New code W44.G2 Combination metal and plastic toy and toy part entering into or through natural orifice
New code W44.G3 Combination metal and plastic jewelry entering into or through a natural orifice
New code W44.G9 Other non-organic objects entering into or through a natural orifice

New subcategory W44.H Other sharp object entering into or through a natural orifice
New code W44.H0 Other sharp object unspecified, entering into or through a natural orifice
New code W44.H1 Needle entering into or through a natural orifice
Add Dart entering into or through a natural orifice
Add Hypodermic needle entering into or through a natural orifice
Add Safety pin entering into or through a natural orifice
Add Sewing needle entering into or through a natural orifice
New code W44.H2 Knife, sword or dagger entering into or through a natural orifice
New code W44.8 Other foreign body entering into or through a natural orifice
Add Foreign body NOS entering into or through a natural orifice
New code W44.9 Unspecified foreign body entering into or through a natural orifice
Frailty Risk Analysis Index

As the US population ages, the number of operations performed on elderly patients will increase. Frailty predicts postoperative mortality and morbidity more than age alone, thus presenting opportunities to identify the highest-risk surgical patients and improve their outcomes.

Studies have been done to determine if surgical outcomes of frail patient can be improved by facility-wide frailty screening and subsequent administrative review of perioperative surgical decision making. The Frailty Analysis Index can be performed during the pre-surgical care visit. Patients with high frailty scores (a Risk Analysis Index of 42 or more on a linear scale from zero to 81) must have a documented care plan that reflects their frailty-associated risks. Surgeons have two options: They can refer the selected patients for further evaluation by their primary care physician or they can use evidence-based protocols to prepare patients for surgery. Additionally, surgeons can document that the patient has been informed about his or her risk score and has engaged with the physician in a shared decision-making process. The result of that process is documented in a plan which might include deciding not to proceed with surgery.

Study findings revealed that this Frailty Screening Initiative (FSI) using Risk Analysis Index scores had a 3-fold survival benefit after controlling for age, frailty, and predicted mortality.

New specific ICD-10-CM codes will promote a more accurate assessment of the patient’s severity of illness and needs for care. The Regulatory Committee of the Association of Clinical Documentation Integrity Specialists (ACDIS) propose the following revisions:

References
https://jamanetwork.com/journals/jamasurgery/fullarticle/2587479
https://achp.org/prevent-unnecessary-surgery/
**ICD-10 Coordination and Maintenance Committee Meeting**
*September 13-14-2022*

**TABULAR MODIFICATIONS**

R54  Age-related physical debility

Delete  Frailty
Delete  Old age
Delete  Senescence
Delete  Senile asthenia
Delete  Senile debility

Excludes1: age-related cognitive decline (R41.81)
  sarcopenia (M62.84)
  senile psychosis (F03)
  senility NOS (R41.81)

New code  R54.8  Other age-related physical debility
Add  Frailty
Add  Old age
Add  Senescence
Add  Senile asthenia
Add  Senile debility

New subcategory  R54.A  Frailty risk
Add  Code first, underlying condition
New code  R54.A0 Frailty risk analysis index score unspecified
New code  R54.A1 Frailty risk analysis index score 0-41
New code  R54.A2 Frailty risk analysis index score 42-81
Immunoglobulin A Nephropathy (IgAN)

The Renal Physicians Association (RPA) is requesting a new ICD-10-CM code for Immunoglobulin A Nephropathy (IgAN), the most common form of glomerulonephropathy.\(^1\) The proposal was presented at the March and September 2021 ICD-10 Coordination and Maintenance meetings. Based on public comments from September 2021 C&M meeting, revised code titles are being presented for consideration and noted in bold.

IgAN affects approximately 2.5 per 100,000 persons worldwide. In the U.S., approximately 130 thousand patients have IgAN (incidence of 20-45 patients per million/year). In approximately 25% of patients with the condition, the nephropathy may progress to end-stage renal disease (ESRD) within 10-15 years.\(^\text{ii}\) It is estimated that IgAN accounts for up to 10% of all patients in need of renal replacement therapy for ESRD in western countries.\(^\text{iii}\) IgAN represents a particularly significant burden on the health care system because patients are usually relatively young when they reach ESRD. Also, the disease recurs in up to 60% of the patients who have received renal transplantation, though not all will develop clinically significant disease.\(^\text{iv}\)

IgAN is characterized by deposition of immune complexes containing Immunoglobulin A in the glomerulus and proliferation of mesangial cells.\(^\text{v,vi}\) The course of disease progression in IgAN can usually be predicted by clinical signs (hypertension, proteinuria, impaired renal function) and histologic lesions (extent of sclerosis and tubulointerstitial damage). Higher levels and longer duration of proteinuria are the strongest prognostic risk factors for disease progression.\(^\text{vii,viii}\) There are a number of specific therapies that are used in the treatment of IgAN patients.\(^\text{ix,x}\)

IgAN is diagnosed by renal biopsy.\(^\text{x}\) Immuno-fluorescence shows abundant deposition of IgA in the glomeruli, mainly in the mesangial region. The histological changes are variable but are dominated by mesangial proliferation and matrix expansion.\(^\text{xii}\) It is commonly diagnosed between the ages of 16 and 35 years, usually due to the discovery of micro- or macrohematuria not attributable to other causes, with or without proteinuria.

Specific coding for IgAN is critical for accurately identifying cases, allowing for etiology-related research, patient segmentation, and therapeutic selection. A recommendation for a revision to the ICD-10-CM coding for IgAN is in line with the consensus of a group of experts in renal pathology, nephrology, and complement biology and therapeutics, as well as IgAN patients. Feedback from this group suggests that current coding for IgAN is neither sufficient nor adequate for identifying and differentiating IgAN patients because:

1. Current codes do not distinguish IgAN from other glomerular lesions that may have different treatment pathways, and do not enable a clear understanding of the epidemiology of the disease.
2. The distinctions between the different types of glomerular lesions in current codes may not be precise enough to indicate the severity or course of IgA nephropathy.

Currently, IgAN cases are commonly coded as N02.8, Recurrent and persistent hematuria with other morphologic changes, however IgAN is a well-defined condition. This expansion is recommended for further specificity. The RPA supports these revisions.

References


## TABULAR MODIFICATIONS

N02 Recurrent and Persistent Hematuria

N02.B Recurrent and persistent immunoglobulin A nephropathy

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>N02.B1</td>
<td>Recurrent and persistent immunoglobulin A nephropathy with glomerular lesion</td>
</tr>
<tr>
<td>N02.B2</td>
<td>Recurrent and persistent immunoglobulin A nephropathy with focal and segmental glomerular lesion</td>
</tr>
<tr>
<td>Add</td>
<td>Recurrent and persistent immunoglobulin A nephropathy with focal and segmental hyalinosis or sclerosis</td>
</tr>
<tr>
<td>N02.B3</td>
<td>Recurrent and persistent immunoglobulin A nephropathy with diffuse membranoproliferative glomerulonephritis</td>
</tr>
<tr>
<td>N02.B4</td>
<td>Recurrent and persistent immunoglobulin A nephropathy with diffuse membranous glomerulonephritis</td>
</tr>
<tr>
<td>N02.B5</td>
<td>Recurrent and persistent immunoglobulin A nephropathy with diffuse mesangial proliferative glomerulonephritis</td>
</tr>
<tr>
<td>N02.B6</td>
<td>Recurrent and persistent immunoglobulin A nephropathy with diffuse mesangiocapillary glomerulonephritis</td>
</tr>
<tr>
<td>N02.B9</td>
<td>Other recurrent and persistent immunoglobulin A nephropathy</td>
</tr>
</tbody>
</table>
INDEX MODIFICATIONS

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

This is a representation from the March 2022 ICD-10 Coordination and Maintenance Committee Meeting of the Inappropriate Sinus Tachycardia (IST) proposal with the recommended title modification adding “as state” to the code title at I47.11, Inappropriate sinus tachycardia. The modification is in **bold**.

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

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

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

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

**References**


**TABULAR MODIFICATIONS**

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<tr>
<td>I47</td>
<td>Paroxysmal tachycardia</td>
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<td>Supraventricular tachycardia</td>
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<td>Junctional (paroxysmal) tachycardia</td>
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<tr>
<td>Delete</td>
<td>Nodal (paroxysmal) tachycardia</td>
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</table>

75
New code   I47.10  Supraventricular tachycardia, unspecified

New code   I47.11  Inappropriate sinus tachycardia, **as stated**

New code   I47.19  Other supraventricular tachycardia

Add   Atrial (paroxysmal) tachycardia

Add   Atrioventricular [AV] (paroxysmal) tachycardia

Add   Atrioventricular re-entrant (nodal) tachycardia [AVNRT]
         [AVRT]

Add   Junctional (paroxysmal) tachycardia

Add   Nodal (paroxysmal) tachycardia

Add   Supraventricular tachycardia
Insulin Resistant Syndrome

This is a representation from the September 2021 and March 2022 ICD-10 Coordination and Maintenance Committee Meetings of the Insulin Resistant Syndrome proposal with the recommended modification to add “syndrome” to the inclusion term Insulin resistance, Type B at E88.818, Other insulin resistance. The modification is in bold.

The National Institute of Health defines metabolic syndrome as the presence of at least 3 of the following traits (including the ones that are controlled by medication): large waist, elevated triglyceride level, reduced HDL cholesterol, increased blood pressure and elevated fasting blood glucose. Other names for metabolic syndrome are: Dysmetabolic syndrome, Hypertriglyceridemic waist, Insulin resistance syndrome, Obesity syndrome or Syndrome X.

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

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

This proposal is supported by the Office of Genomics Precision Public Health and American College of Medical Genetics and Genomics.

References
1. Orphanet: Insulin resistance syndrome type A. INSERM US14 -- ALL RIGHTS RESERVED
https://www.orpha.net/consor/cgi-bin/OC_Exp.php?Expert=2297

TABULAR MODIFICATIONS

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<tr>
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<td>Excludes1: histiocytosis X (chronic) (C96.6)</td>
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Intestinal Microbial Overgrowth

Intestinal microbial overgrowth includes several diseases including small intestinal bacterial overgrowth (SIBO), intestinal methanogen overgrowth (IMO) and small intestinal fungal overgrowth (SIFO). These diseases result from the overpopulation of bacteria, methanogenic archaea or fungi in the intestines and can lead to debilitating symptoms with significant effect on quality of life. Over the past 2 decades, there has been significant progress in the diagnosis and treatment of these diseases specially SIBO and IMO with various published practice guidelines including the American College of Gastroenterology (ACG).\textsuperscript{1-3}

Small intestinal bacterial overgrowth (SIBO): Common symptoms of SIBO include abdominal distention, abdominal bloating, diarrhea, and abdominal pain/discomfort. Malnutrition, weight loss, and anemia are seen in more severe cases.\textsuperscript{4} SIBO is estimated to affect at least 33\% of patients with unexplained GI symptoms.\textsuperscript{1} The gold standard for a diagnosis of SIBO is the presence of \( \geq 10^3 \) CFU/mL of jejunal aspirate by culture.\textsuperscript{2} Alternatively, SIBO can be diagnosed by a rise in exhaled hydrogen levels from baseline after consumption of a fermentable sugar substrate such as glucose or lactulose.\textsuperscript{2} Antibiotics remain the core treatment modality for SIBO. A systematic review of the literature found 23 trials addressing the role of antibiotics in SIBO.\textsuperscript{5} The most commonly used antibiotics were clindamycin, metronidazole, neomycin, rifaximin and tetracycline.\textsuperscript{1} Short term courses of elemental diets can also be a safe and effective alternative to antibiotics. Elemental diets are believed to be fully absorbed within the first few feet of small intestine, limiting the delivery of nutrients to the microbes in mid/distal small bowel and colon.\textsuperscript{6}

Intestinal methanogen overgrowth (IMO): Methanogens are classified under the kingdom of archaea and are not bacteria, hence, excessive archaea cannot be classified under bacterial overgrowth. Symptoms of IMO include abdominal bloating, distention, constipation and abdominal discomfort.\textsuperscript{7} Severity of symptoms directly correlates with the amount of methane produced by the gut methanogens.\textsuperscript{1} IMO is diagnosed by detection of exhaled methane levels >10 ppm during breath testing.\textsuperscript{1, 2, 7} In patients with IMO, single antibiotic therapy may not have the expected effectiveness. \textit{Methanobrevibacter smithii}, which is the main archaeon responsible for methane production in humans, is resistant to several antibiotics. In a retrospective study \textsuperscript{8}, subjects with IMO who received either neomycin or rifaximin alone did not have a substantial improvement. However, combination of neomycin with rifaximin decreased methane production in more than 80 \% of subjects, with a similar response in constipation-related symptoms. Similar findings were seen in prospective trials.\textsuperscript{9}

Small intestinal fungal overgrowth (SIFO) symptoms include abdominal pain, gas, bloating, fullness, belching, indigestion, and diarrhea. SIFO is diagnosed via small bowel aspiration and fungal cultures, and the choice of antifungal therapy is mainly driven by the culture and sensitivity results. Irrespective of geographic distribution, \textit{Candida albicans} and \textit{Candida glabrata} are the most common species isolated from the small bowel.\textsuperscript{10} Overgrowth of H\textsubscript{2}S-producing bacteria are also associated with abdominal pain and diarrhea.\textsuperscript{11}
Overall, intestinal microbial overgrowth has multiple objectively-defined subtypes which are diagnosed and treated differently. Currently, there are no codes dedicated to intestinal microbial overgrowth or its subtypes. Given this deficiency, physicians and coders commonly use: “A04.9: Bacterial intestinal infection, unspecified” (however this code relates to intestinal infections and not a microbial overgrowth), or “K63.8: Other specified diseases of intestine” which has an unacceptable specificity for microbial overgrowth.

This lack of specificity decreases the opportunity to use ICD-10-CM codes for accurate disease tracking. The following revisions are proposed to achieve sufficient clinical granularity for diagnosis of intestinal microbial overgrowth and further improve international classification, tracking, and surveillance.

Specific codes for intestinal microbial overgrowth are requested by Drs. Rezaie and Pimentel who are the lead authors of the North American consensus on breath testing to diagnose SIBO and IMO², and the American College of Gastroenterology (ACG) clinical guidelines for diagnosis and treatment of microbial overgrowth.¹

References
ICD-10 Coordination and Maintenance Committee Meeting  
September 13-14-2022

**TABULAR MODIFICATIONS**

K63 Other diseases of intestine

K63.8 Other specified diseases of intestine

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<td>K63.8222 Small intestinal bacterial overgrowth, hydrogen sulfide-subtype</td>
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<td>K63.8229 Small intestinal bacterial overgrowth, unspecified</td>
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<tbody>
<tr>
<td>K63.823 Small intestinal fungal overgrowth</td>
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Intrahepatic Cholestasis in Pregnancy

The American College of Obstetricians and Gynecologists (ACOG) is proposing to add new codes that allow for the reporting of Intrahepatic Cholestasis in Pregnancy (ICP) for each trimester.

ICP is a pregnancy-specific disorder where bile acid transport in the liver is altered. This leads to a build-up of bile acid in both the maternal circulation as well as the fetal amniotic fluid. There are both maternal and fetal risks of intrahepatic cholestasis of pregnancy. Maternal risks include an increased risk of pre-eclampsia and gestational diabetes. Fetal risks include preterm birth, increased respiratory distress after birth when matched to the same gestational age non ICP pregnancies, meconium staining of the amniotic fluid and stillbirth.

Intrahepatic cholestasis of pregnancy affects about 0.5% of pregnancies in the United States. The exact cause of ICP is unknown but is thought to be related to the increased hormones of pregnancy altering bile acid transport. About 15% of cases are linked with a genetic mutation, most commonly ABCB4. ICP most often occurs in the 2nd and 3rd trimesters of pregnancy, but cases have also been diagnosed as early as 5 weeks.

In the United States, the Society for Maternal-Fetal Medicine (SMFM) has a set of guidelines for the diagnosis and treatment of intrahepatic cholestasis of pregnancy. The main symptom of ICP is pruritus which is often on the hands and feet but can become generalized and severe. The syndrome is diagnosed by a total bile acid blood test which is elevated in ICP. The most often agreed-upon diagnosis level in the United States is 10 µmol/L. Other pre-pregnancy liver conditions should also be screened for with a thorough personal and family history as well as necessary laboratory evaluation prior to diagnosis. SMFM has outlined a specific set of treatment guidelines including fetal monitoring, medication treatment with ursodeoxycholic acid and appropriately timed infant delivery to prevent complications such as stillbirth. This disorder is a unique disease requiring its own specific management in pregnancy.

Intrahepatic cholestasis of pregnancy resolves after the peripartum period is over. Some patients might have underlying liver disorders that predisposed them to the development of intrahepatic cholestasis of pregnancy, but the disorder itself is pregnancy-specific.

There is a need for a specific code for intrahepatic cholestasis of pregnancy both to allow for the proper identification and treatment of these patients as well as being able to gather more information collectively about the outcomes of these pregnancies.

It has also been shown that some patients with intrahepatic cholestasis of pregnancy are at risk for later diseases. In looking at the long-term health of these patients, they are more likely to have other hepatobiliary diseases in the future including cholelithiasis, pancreatitis and need for cholecystectomy. There is also an increased risk of hypothyroidism. Given that there are some
life-long health concerns with the disorder, it would be beneficial to be able to track these patients in order to hopefully modify their lifestyle factors to possibly prevent further disease.

In summary, intrahepatic cholestasis of pregnancy is a pregnancy specific liver disorder with its own set of diagnostic criteria and implications for pregnancy as well as future health. Patients with this disorder would benefit by the creation of an ICD-10-CM code as the disorder differs from other liver disorders with which it is currently classified.

References:


TABULAR MODIFICATIONS

O26 Maternal care for other conditions predominantly related to pregnancy

O26.6 Liver and biliary tract disorders in pregnancy, childbirth and the puerperium

Use additional code to identify the specific disorder

Excludes2: hepatorenal syndrome following labor and delivery (O90.4)

O26.64 Intrahepatic cholestasis of pregnancy

New code    O26.641 Intrahepatic cholestasis of pregnancy, first trimester

New code    O26.642 Intrahepatic cholestasis of pregnancy, second trimester

New code    O26.643 Intrahepatic cholestasis of pregnancy, third trimester

New code    O26.649 Intrahepatic cholestasis of pregnancy, unspecified trimester
Leukodystrophies

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

This proposal was presented at the March 2022 Coordination and Maintenance Meeting. Based on public comment, changes have been made and resubmitted for reconsideration. Changes are noted in **bold**.

Leukodystrophies may present at any age from preterm infants and neonates to late adulthood and have been reported across all ethnicities and regions of the world. 30 years ago only a few leukodystrophies were recognized as distinct disease entities, but in the past 10 years over 400 genetically unique leukodystrophies have been reported.2 Even though most leukodystrophies are individually rare, as a group leukodystrophies affect close to 1 in 4,000 live births.3,4 Further, consensus work in the community has defined a group of leukodystrophies with unique genetic causes and well-studied, distinct clinical and pathophysiological features.

Currently there are only specific ICD-10-CM codes for six of the primary leukodystrophies (X-linked Adrenoleukodystrophy, ALD- E71.52x; Metachromatic leukodystrophy, MLD- E75.25; Krabbe disease- E75.23, Refsum’s disease- G60.1; Zellweger syndrome- E71.510; and E71.511 Neonatal Adrenoleukodystrophy). Otherwise, many leukodystrophies are indexed under a single ICD-10-CM code E75.29, Other sphingolipidosis. Prior to ICD-10-CM, there were not specific ICD codes for ALD, MLD, or Krabbe. The advent of specific ICD-10-CM codes for ALD, Krabbe, and MLD enabled clinical trials, newborn screening, and studies of racial disparities.6,7

The leukodystrophies proposed for unique ICD-10-CM codes all have unique genetic causes; distinct clinical courses and morbidities; and have different treatments- either currently available or in clinical trials. For example, VWM has a sputtering clinical course8 and has a clinical trial with the α-agonist guanabenz.9 In contrast, Canavan’s disease has an early rapid progression10 and potential treatment with antisense oligonucleotides.11

Creation of specific ICD-10-CM codes for this heterogenous, complex group of disorders known as leukodystrophies is critical for patient care, clinical trials, and research. The diversity in causes should be reflected by a diversity of codes to best represent these disorders. The importance of and difference between these leukodystrophy disorders can be seen in the codes already created for ICD-11. In ICD-11, Leukodystrophy has its own ICD-11 category, there are five new leukodystrophy codes, and there are also fourteen new leukodystrophy indices.
References:

TABULAR MODIFICATIONS

E74 Other disorders of carbohydrate metabolism

E74.0 Glycogen storage disease

E74.09 Other glycogen storage disease

Andersen disease
Hers disease
Tauri disease

Glycogen storage disease, types 0, IV, VI-XI
Hers disease
Liver phosphorylase deficiency
Muscle phosphofructokinase deficiency
Tauri disease
E75 Disorders of sphingolipid metabolism and other lipid storage disorders
   E75.2 Other sphingolipidosis
      Excludes1: adrenoleukodystrophy [Addison-Schilder] (E71.528)
      New code E75.27 Pelizaeus-Merzbacher disease
      New code E75.28 Canavan disease

E79 Disorders of purine and pyrimidine metabolism
   E79.8 Other disorders of purine and pyrimidine metabolism
      Delete Hereditary xanthinuria
      New code E79.81 Aicardi-Goutières syndrome
      New code E79.82 Hereditary xanthinuria
      New code E79.89 Other specified disorders of purine and pyrimidine metabolism

E88 Other and unspecified metabolic disorders
   E88.4 Mitochondrial metabolism disorders
      New code E88.43 Disorders of mitochondrial tRNA synthetases

      G11 Hereditary ataxia
      New code G11.5 Hypomyelination - hypogonadotropic hypogonadism - hypodontia
      Add 4H syndrome
      Add Pol III-related leukodystrophy
      New code G11.6 Leukodystrophy with vanishing white matter disease

      G23 Other degenerative diseases of basal ganglia
      New code G23.3 Hypomyelination with atrophy of the basal ganglia and cerebellum
      Add H-ABC

      G31 Other degenerative diseases of nervous system, not elsewhere classified
      New code G31.80 Leukodystrophy, unspecified
      New code G31.86 Alexander Disease

      G90 Disorders of autonomic nervous system
      New code G90.A LMNB1-related autosomal dominant leukodystrophy
G93 Other Disorders of the Brain

**G93.4 Other and unspecified encephalopathy**

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<td><strong>G93.42</strong> Megaloencephalic leukoencephalopathy with subcortical cysts</td>
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<tr>
<td>New code</td>
<td><strong>G93.43</strong> Leukoencephalopathy with calcifications and cysts</td>
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<tr>
<td>New code</td>
<td><strong>G93.44</strong> Adult-onset leukodystrophy with axonal spheroids</td>
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<tr>
<td>Add</td>
<td>Adult-onset leukoencephalopathy with axonal spheroids and pigmented glia</td>
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</table>
Lymphoma in Remission

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

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

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

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

References

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

TABULAR MODIFICATIONS

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

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

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

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

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

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

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

C81.9  Hodgkin lymphoma, unspecified  

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

C82  Follicular lymphoma  

C82.0  Follicular lymphoma grade I  

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

C82.1  Follicular lymphoma grade II  

New code  C82.1A  Follicular lymphoma grade II, in remission

C82.2  Follicular lymphoma grade III, unspecified  

New code  C82.2A  Follicular lymphoma grade III, unspecified, in remission

C82.3  Follicular lymphoma grade IIIa  

New code  C82.3A  Follicular lymphoma grade IIIa, in remission

C82.4  Follicular lymphoma grade IIIb  

New code  C82.4A  Follicular lymphoma grade IIIb, in remission
C82.5  Diffuse follicle center lymphoma
    New code C82.5A  Diffuse follicle center lymphoma, in remission

C82.6  Cutaneous follicle center lymphoma
    New code C82.6A  Cutaneous follicle center lymphoma, in remission

C82.8  Other types of follicular lymphoma
    New code C82.8A  Other types of follicular lymphoma, in remission

C82.9  Follicular lymphoma, unspecified
    New code C82.9A  Follicular lymphoma, unspecified, in remission

C83  Non-follicular lymphoma
    C83.0  Small cell B-cell lymphoma
        Lymphoplasmacytic lymphoma
        Nodal marginal zone lymphoma
        Non-leukemic variant of B-CLL
        Splenic marginal zone lymphoma
    New code C83.0A  Small cell B-cell lymphoma, in remission
    Add Lymphoplasmacytic lymphoma, in remission
    Add Nodal marginal zone lymphoma, in remission
    Add Non-leukemic variant of B-CLL, in remission
    Add Splenic marginal zone lymphoma, in remission

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

    C83.3  Diffuse large B-cell lymphoma
    New code C83.3A  Diffuse large B-cell lymphoma, in remission
    Add Anaplastic diffuse large B-cell lymphoma, in remission
    Add CD30-positive diffuse large B-cell lymphoma, in remission
    Add Centroblastic diffuse large B-cell lymphoma, in remission
    Add Diffuse large B-cell lymphoma, subtype not specified, in remission
    Add Immunoblastic diffuse large B-cell lymphoma, in remission
    Add Plasmablastic diffuse large B-cell lymphoma, in remission
    Add T-cell rich diffuse large B-cell lymphoma, in remission
C83.5 Lymphoblastic (diffuse) lymphoma

New code
Add
Add
Add
Add

C83.7 Burkitt lymphoma

New code
Add
Add

C83.8 Other non-follicular lymphoma

New code
Add
Add

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

New code

C84 Mature T/NK-cell lymphomas

C84.0 Mycosis fungoides

New code

C84.1 Sézary disease

New code

C84.4 Peripheral T-cell lymphoma, not classified

New code
Add
Add
Add

C84.6 Anaplastic large cell lymphoma, ALK-positive

Anaplastic large cell lymphoma, CD30-positive
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Add      Extranodal NK/T-cell lymphoma, nasal type with failed remission
New code       C86.01      Extranodal NK/T-cell lymphoma, nasal type, in remission

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

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

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

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

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

New code       C86.21      Enteropathy-type (intestinal) T-cell lymphoma, in remission
Add           Enteropathy associated T-cell lymphoma, in remission

C86.3 Subcutaneous panniculitis-like T-cell lymphoma

New code       C86.30      Subcutaneous panniculitis-like T-cell lymphoma not having achieved remission
Add           Subcutaneous panniculitis-like T-cell lymphoma NOS
Add           Subcutaneous panniculitis-like T-cell lymphoma with failed remission

New code       C86.31      Subcutaneous panniculitis-like T-cell lymphoma, in remission

C86.4 Blastic NK-cell lymphoma
Blastic plasmacytoid dendritic cell neoplasm (BPDCN)
New code C86.40 Blastic NK-cell lymphoma not having achieved remission
Add Blastic NK-cell lymphoma NOS
Add Blastic NK-cell lymphoma with failed remission
Add Blastic plasmacytoid dendritic cell neoplasm (BPDCN) NOS
Add Blastic plasmacytoid dendritic cell neoplasm (BPDCN) not having achieved remission
Add Blastic plasmacytoid dendritic cell neoplasm (BPDCN) with failed remission

New code C86.41 Blastic NK-cell lymphoma, in remission
Add Blastic plasmacytoid dendritic cell neoplasm (BPDCN), in remission

C86.5 Angioimmunoblastic T-cell lymphoma
Angioimmunoblastic lymphadenopathy with dysproteinemia (AILD)

New code C86.50 Angioimmunoblastic T-cell lymphoma not having achieved remission
Add Angioimmunoblastic lymphadenopathy with dysproteinemia (AILD) NOS
Add Angioimmunoblastic lymphadenopathy with dysproteinemia (AILD) not having achieved remission
Add Angioimmunoblastic lymphadenopathy with dysproteinemia (AILD) with failed remission
Add Angioimmunoblastic T-cell lymphoma NOS
Add Angioimmunoblastic T-cell lymphoma with failed remission

New code C86.51 Angioimmunoblastic T-cell lymphoma, in remission
Add Angioimmunoblastic lymphadenopathy with dysproteinemia (AILD), in remission

C86.6 Primary cutaneous CD30-positive T-cell proliferations
Lymphomatoid papulosis
Primary cutaneous anaplastic large cell lymphoma
Primary cutaneous CD30-positive large T-cell lymphoma

New code C86.60 Primary cutaneous CD30-positive T-cell proliferations not having achieved remission
Add Lymphomatoid papulosis NOS
Add Lymphomatoid papulosis not having achieved remission
Add Lymphomatoid papulosis with failed remission
Add Primary cutaneous anaplastic large cell lymphoma NOS
Add Primary cutaneous anaplastic large cell lymphoma not having achieved remission
Add  Primary cutaneous anaplastic large cell lymphoma with failed remission
Add  Primary cutaneous CD30-positive large T-cell lymphoma NOS
Add  Primary cutaneous CD30-positive large T-cell lymphoma not having achieved remission
Add  Primary cutaneous CD30-positive T-cell proliferations with failed remission

New code  C86.61 Primary cutaneous CD30-positive T-cell proliferations, in remission
Add  Lymphomatoid papulosis, in remission
Add  Primary cutaneous anaplastic large cell lymphoma, in remission
Add  Primary cutaneous CD30-positive large T-cell lymphoma, in remission

C88  Malignant immunoproliferative diseases and certain other B-cell lymphomas

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

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

New code  C88.01 Waldenström macroglobulinemia, in remission
Add  Lymphoplasmacytic lymphoma with IgM-production, in remission
Add  Macroglobulinemia (idiopathic) (primary), in remission
C88.2 Heavy chain disease
Franklin disease
Gamma heavy chain disease
Mu heavy chain disease

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

New code C88.21 Heavy chain disease, in remission
Add Franklin disease, in remission
Add Gamma heavy chain disease, in remission
Add Mu heavy chain disease, in remission

C88.3 Immunoproliferative small intestinal disease
Alpha heavy chain disease
Mediterranean lymphoma

New code C88.30 Immunoproliferative small intestinal disease not having achieved remission
Add Alpha heavy chain disease immunoproliferative small intestinal disease not having achieved remission
Add Alpha heavy chain disease NOS
Add Alpha heavy chain disease with failed remission
Add Immunoproliferative small intestinal disease NOS
Add Immunoproliferative small intestinal disease with failed remission
Add Mediterranean lymphoma NOS
Add Mediterranean lymphoma not having achieved remission
Add Mediterranean lymphoma with failed remission

New code C88.31 Immunoproliferative small intestinal disease, in remission
Add Alpha heavy chain disease, in remission
Add Mediterranean lymphoma, in remission
C88.4 Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue [MALT-lymphoma]
Add Lymphoma of bronchial-associated lymphoid tissue [BALT-lymphoma]
Delete Lymphoma of skin-associated lymphoid tissue [SALT-lymphoma]

New code C88.40 Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue [MALT-lymphoma] not having achieved remission
Add Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue [MALT-lymphoma] NOS
Add Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue [MALT-lymphoma] with failed remission
Add Lymphoma of bronchial-associated lymphoid tissue [BALT-lymphoma] NOS
Add Lymphoma of bronchial-associated lymphoid tissue [BALT-lymphoma] not having achieved remission
Add Lymphoma of bronchial-associated lymphoid tissue [BALT-lymphoma] with failed remission
Add Lymphoma of skin-associated lymphoid tissue [SALT-lymphoma] NOS
Add Lymphoma of skin-associated lymphoid tissue [SALT-lymphoma] not having achieved remission
Add Lymphoma of skin-associated lymphoid tissue [SALT-lymphoma] with failed remission

New code C88.41 Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue [MALT-lymphoma], in remission
Add Lymphoma of bronchial-associated lymphoid tissue [BALT-lymphoma], in remission
Add Lymphoma of skin-associated lymphoid tissue [SALT-lymphoma], in remission

C88.8 Other malignant immunoproliferative diseases

New code C88.80 Other malignant immunoproliferative diseases not having achieved remission
Add Other malignant immunoproliferative diseases NOS
Add Other malignant immunoproliferative diseases with failed remission
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Lysosome-Associated Membrane Protein 2 (LAMP2) Deficiency (Danon Disease)

Danon disease is an X-linked dominant metabolic disorder originally described as “lysosomal glycogen storage disease with normal acid maltase”\(^1\). Subsequently, a causative gene (\(LAMP2\)) was identified. Mutations in this gene result in a primary deficiency of lysosome-associated membrane protein 2, an intracellular protein that mediates endosomal-lysosomal fusion, an integral component of autophagy. The absence of intracellular LAMP2 protein in tissues/organs is associated with impaired autophagic and mitochondrial function and accumulation of intracellular debris resulting in functional impairment\(^2\).

Danon disease is a multi-systemic disorder and represents one of the most aggressive cardiomyopathies ever characterized, especially for male patients. There currently are no available disease-modifying therapies that mitigate progression to end stage heart failure and death. As a result, most male Danon patients do not live beyond adolescence or early adulthood in the absence of heart transplantation.

Danon disease involves the heart, skeletal muscles, central nervous system, and retina, although the non-cardiac manifestations are most frequently reported in males. Given the multisystemic presentation and rarity of Danon disease, proper identification and management of patients has been challenging. Disease heterogeneity is also apparent between males and females, who manifest significant clinical differences due to the X-linked inheritance of the disease\(^3\). The average age of initial symptoms in males is 12 years and the average age of death is 19 years\(^4\). The average age of initial symptoms in females is 28 and of death is 35 years\(^4\). Within each gender, there exist considerable variability of presentation, organ involvement, and rapidity of cardiac deterioration\(^4\).

Danon disease is diagnosed via genetic testing for pathogenic variations of the \(LAMP2\) gene\(^5\). As of 2019, at least 146 cases had been reported in the literature with molecular confirmation indicating Danon disease\(^6\). The true prevalence of Danon disease is not known. A crude estimate of prevalence can be based on extrapolation from studies of pediatric and adult hypertrophic cardiomyopathy (for which there is an overall prevalence of approximately 1:500 in the overall population); in these series, LAMP2 pathogenic variants have been identified as underlying approximately 1% - 4% of hypertrophic cardiomyopathy cases\(^4,7,8\) although there is still no consensus on these estimates\(^6,9\). These data yield an estimate of 10,000 – 25,000 total cases in the US. The volume of documented cases is rising with wider availability of genetic testing\(^10\) and genetically-targeted therapies in development, which may motivate more widespread diagnostic efforts.

Danon disease was originally classified as a glycogen storage disease (GSD), but is distinct from other GSDs, which are caused by a deficient enzyme of glycogen metabolism. Danon disease differs from other GSDs in terms of genetic cause, pathophysiology (i.e., it is a transport disorder, not a storage disorder), prevalence, symptoms, and treatment; this distinction also applies to Danon disease compared with other disorders in the E74.0 subcategory, as shown in the comparisons noted below.
Von Gierke disease is GSD type I, and involves gene GSDIA or GSDIB, and is autosomal recessive with incidence 1:100,000 births. Symptoms include low blood sugar, enlarged liver and kidneys, and treatment involves diet. The ICD-10-CM code is E74.01.

Pompe disease is GSD type II, involving gene GAA, autosomal recessive, incidence 1:40,000 births. Symptoms involve respiratory issues, muscle weakness, and hypertrophic cardiomyopathy (HCM); treatment involves enzyme replacement. The ICD-10-CM code is E74.02.

Danon disease is GSD type IIB, involving the gene LAMP2, X-linked dominant, prevalence 1:12,000 – 1:50,000 individuals. Symptoms include HCM, dilated cardiomyopathy (DCM), and musculoskeletal weakness. Treatment includes heart transplant.

Cori disease is GSD III, involving the gene PYGL, autosomal recessive, with incidence 1:100,000 births. Symptoms include low blood sugar, and enlarged liver; treatment involves diet. It is coded to E74.03.

Andersen disease is GSD type IV, involving the gene GBE, autosomal recessive, incidence 1:600,000 – 1:800,000 births. Symptoms include enlarged liver, fibrosis, and cirrhosis; treatment includes liver transplant. It is coded to E74.09 (not specific).

McArdle disease is GSD type V, involving gene PYGM, autosomal recessive, with prevalence 1:100,000 individuals. Symptoms include cramps and fatigue, and treatment involves diet and exercise. It is coded to E74.04.

No specific treatment is currently available for Danon disease\textsuperscript{11}. Current management focuses on surveillance and symptom palliation, with heart transplantation provided when clinically indicated. There is currently a Danon disease-directed gene therapy treatment under investigation; a registrational trial is anticipated over the coming year.

Danon disease has unique monitoring and management needed to provide optimal patient care (e.g., alcohol septal ablation for HCM, which is not recommended for Danon disease per the 2020 AHA/ACC HCM Guidelines, or a patient being evaluated for heart transplant for exercise intolerance because skeletal myopathy has not yet been diagnosed).

A specific ICD-10-CM code for Danon disease will facilitate patient identification for appropriate treatments as well as for clinical trials, and will facilitate better understanding of prevalence, onset, and disease progression, as well as supporting further research in the evaluation and treatment of Danon disease.

A proposal to create a specific code for Danon disease was received from University of Colorado, Adult Medical Genetics Program.

REFERENCES

**TABULAR MODIFICATIONS**

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MED13L Syndrome

The MED13L Foundation is requesting a unique International Classification of Disease, 10th Revision, Clinical Modification (ICD-10-CM) code specific for MED13L Syndrome.

MED13L Syndrome is a genetic disorder with a world-wide prevalence estimated at over 1 million individuals. The syndrome is “characterized by [moderate to severe] intellectual disability, heart malformation, and hypotonia.” Notably, MED13L Syndrome may present with severe cyanotic forms of congenital heart diseases and other congenital cardiac defects. In addition, MED13L Syndrome has been associated with mitochondrial dysfunction. The condition is also characterized by significant speech impairment and dysmorphic facial features. “Most children with the syndrome [also] . . . take longer to learn to sit and walk independently (delayed motor skills).” “Other features may include short stature, cleft palate, problems with coordination (ataxia), and recurrent seizures (epilepsy).”

MED13L Syndrome is an autosomal dominant monogenic disorder diagnosed in children associated with the mutation and disruption of function of the MED13L (Mediator Complex Subunit 13L) gene, which is located on chromosome 12. The MED13 gene is a component of the CDK8-kinase module, which can reversibly bind the Mediator complex, a multi-protein complex required for assembly and stabilization of the pre-initiation complex. Congenital malformations have been associated with thirty-two sub-units of the Mediator complex, and the complex is “essential for transcription initiation….The core function of Mediator is to transmit signals from various transcription factors to RNA polymerase II (Pol II) . . . . Binding of the CDK8-module to Mediator has been reported to prevent the association of Mediator with the Pol II pre-initiation complex, thus preventing transcription initiation and/or re-initiation.” As such, MED13L “plays a role in the control of cell growth, repression of cell cycle target genes, and cell cycle inhibition.” Reported cases of MED13L Syndrome are overwhelmingly de novo (i.e., spontaneous) mutations. Nevertheless, there are also reports of parental germline mosaicism (presence of mutation in some of the sperms/eggs) leading to more than one affected individual in the families. Approximately 92% of known cases involve de novo mutations, 3% involve hereditary mutations, and 5% of cases are of unknown inheritances. “Although [different] MED13L mutations have been associated with this syndrome, a causative mechanism(s) for the multiple facets of [the clinical presentation of] this disorder has not [yet] been established.”

A specific diagnosis code for MED13L Syndrome would provide research and public health institutions a more reliable way to track the epidemiology associated with MED13L Syndrome. Currently, clinicians cannot specifically characterize and report on MED13L Syndrome through existing diagnosis codes, which inhibits accurate data collection and
systematic tracking of incidence and outcomes. The result is MED13L Syndrome being categorized with other disorders that are phenotypically distinct.

Further, lack of a specific code can cause inaccurate coding. Indeed, a unique code is vital to allow epidemiologists and researchers to understand and reliably track the full prevalence of the condition, as well as associated rates of morbidity and mortality. This inhibits accurate reporting and can delay treatment—ultimately contributing to increased costs and burdens on the health system as a whole which impacts physicians, patients, and their families.

Due to deficits in identification and diagnosis—which are compounded by the absence of a unique diagnosis code—there are significant limitations on the current data about the incidence, prevalence, and epidemiology of MED13L Syndrome. Further, as noted, the estimated worldwide prevalence of MED13L Syndrome is over 1 million individuals. As such, although rare, MED13L Syndrome is significantly represented among birth cohorts relative to many other rare genetic conditions. It also presents in birth cohorts at comparable incidence rates as other rare conditions assigned ICD-10-CM codes, such as SYNGAP1.

Currently, there is no cure or pharmacologic treatment for MED13L Syndrome. Management of the syndrome generally involves speech, behavioral, and occupational therapy. Depending on the presentation of the syndrome, therapy could also involve treatment for epilepsy and management of congenital anomalies (e.g., congenital heart defects, cleft palates, etc.) if present.

Ongoing surveillance is typically part of effective management, including assessments to exclude for development of congenital heart defects, ophthalmologic examinations for eye problems, and monitoring for other clinical anomalies. The MED13L Foundation is currently leading an effort to develop therapeutics for MED13L Syndrome, including the repurposing of small molecule drugs, de novo small molecule and biologic drugs, and genetic medicines such as anti-sense oligonucleotides, gene therapy, and gene-editing constructs. An ICD-10-CM code is critical to the continued development of these programs, in addition to facilitating more reliable public health tracking for the condition.

References
3. NIH, Genetic and Rare Diseases information Center, MED13L haploinsufficiency syndrome, https://tinyurl.com/yc7xrbds (last visited Apr. 2, 2022) (citing Asadollahi et al.).

5. L.S. Blok et al., De novo mutations in MED13, a component of the Mediator complex, are associated with a novel neurodevelopmental disorder, 137 Human Genetics 375 (2018).


9. Asadollahi, Presentation: A literature review of MED13L-related syndrome, Institute of Medical Genetics, University of Zurich (n.d.).


**TABULAR MODIFICATIONS**

Q87 Other specified congenital malformation syndromes affecting multiple systems

Use additional code(s) to identify all associated manifestations

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

Excludes1: Zellweger syndrome (E71.510)
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<td>congenital malformations of cardiac septa (Q21-)</td>
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Membranous Nephropathy

The National Kidney Foundation has requested new ICD-10-CM codes for membranous nephropathy to simplify and clarify diagnostic coding.

Membranous nephropathy (MN), a common cause of nephrotic syndrome in adults, is characterized by thickening of the renal glomerular basement membrane (GBM), resulting from the accumulation of immune reactants in this structure. MN has a global incidence of 8-10 per million and approximately 12 cases per million in the U.S.

MN patients typically present with overt nephrotic syndrome or non-nephrotic proteinuria, commonly with edema and/or hypertension. Progression of nephrotic syndrome predicts higher risk of end-stage kidney disease (ESKD) and thrombo-embolic events. Half of MN patients who present with nephrotic syndrome have a worsening disease progression, and about 30% of MN patients may develop ESKD. In untreated patients, 40-50% progress to ESKD and will require dialysis or renal transplantation. These treatments are particularly burdensome on the patient and health care system, especially regarding transplantation, as the likelihood of MN recurrence following transplantation is common, leading to an increased risk of kidney failure and additional invasive therapies.

MN is caused by autoantibodies directed against the M-type phospholipase A2 receptor (PLA2R) that resides on glomerular podocytes, the cells which overlay the GBM. These antibodies react with podocyte PLA2R and initiate processes that lead to GBM thickening and glomerular injury in MN. Thus, MN is diagnosed by structural (renal biopsy) and/or serological (the presence in the blood or glomerulus of anti-PLA2R antibodies) criteria.

MN has been historically classified into two subtypes: “primary” and “secondary” forms of MN. 1) Primary MN comprises approximately 75-80% of cases. Light microscopy typically shows uniform thickening of the GBM in primary MN. The Jones silver methenamine stain shows “spikes” of GBM material extending between “holes” of unstained immune deposits in the GBM, which correspond to subepithelial deposits of IgG and C3 revealed by immunofluorescence microscopy. There are significant correlations between the anti-PLA2R antibody found in primary MN and disease outcomes. High titers of these antibodies have been associated with severe disease progression while low titers have been found to increase the likelihood of remission after kidney transplantation.

2) Other forms of MN comprise the remaining 20-25% of MN cases. Those cases with a recognizable underlying etiology (e.g., autoimmune diseases, diabetes, cancer, infections, non-steroidal anti-inflammatory drugs) are categorized as “secondary” MN. Light microscopy shows endocapillary hypercellularity in secondary MN, especially in cases of malignancy. Immunofluorescence reveals subepithelial deposits of IgG, including IgG1, IgG2, or IgG3 in secondary MN cases. Deposits of C3 and C1q are also present in secondary MN. As the presence of anti-PLA2R is often low in these secondary forms of MN, disease outcomes often vary, and typically follow the outcomes associated with the etiologic cause in this form.
Primary and secondary forms of MN have distinct treatment pathways. Initial treatment for primary MN involves antibody-targeted therapies, immunosuppressive agents, and supportive care for blood pressure control and proteinuria reduction. Initial treatment for secondary MN follows the treatment pathways of the underlying condition. Since treatment is often targeted at the etiologic cause in secondary MN, differential diagnosis is critical to distinguish between the two forms. To reflect the process and time it takes to achieve a specific diagnosis in MN, a code for unspecified MN is also necessary.

An advisory panel of expert stakeholders (including clinicians, researchers, and patient educators/advocates) have indicated that the term “membranous nephropathy” (MN) is generally used in clinical guidelines that describe this condition as well as being used in clinical practice (rather than the term “diffuse membranous glomerulonephritis”). This condition will generally present with either nephrotic syndrome or isolated proteinuria, and will be further classified as primary or secondary MN. Specific codes to identify these would enable tracking to be more clinically accurate, useful, and up-to-date with scientific advances. This proposal builds on a previous proposal submitted by the National Kidney Foundation in December 2021.

References
TABULAR MODIFICATIONS

N04  Nephrotic syndrome

N04.2  Nephrotic syndrome with diffuse membranous glomerulonephritis

New code  N04.20  Nephrotic syndrome with diffuse membranous glomerulonephritis, unspecified
Add  Membranous nephropathy NOS with nephrotic syndrome

New code  N04.21  Primary membranous nephropathy with nephrotic syndrome
Add  Idiopathic membranous nephropathy with nephrotic syndrome

New code  N04.22  Secondary membranous nephropathy with nephrotic syndrome
Add  Code first, if applicable, other disease or disorder or poisoning causing membranous nephropathy
Add  Use additional code, if applicable, for adverse effect of drug causing membranous nephropathy

New code  N04.29  Other nephrotic syndrome with diffuse membranous glomerulonephritis

N06  Isolated proteinuria with specified morphological lesion

N06.2  Isolated proteinuria with diffuse membranous glomerulonephritis

New code  N06.20  Isolated proteinuria with diffuse membranous glomerulonephritis, unspecified
Add  Membranous nephropathy, NOS
Add  Excludes1: membranous nephropathy with nephrotic syndrome (N04.20)
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<td>Other isolated proteinuria with diffuse membranous glomerulonephritis</td>
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**Myelin Oligodendrocyte Glycoprotein Antibody Disease**

Myelin oligodendrocyte glycoprotein (MOG) antibody disease (MOG-AD) is an inflammatory demyelinating condition of the central nervous system (CNS) characterized by a monophasic or relapsing course of neurological dysfunction, which does not meet the typical criteria for multiple sclerosis (MS) or other known neuroinflammatory conditions, and occurs in the presence of serum MOG antibodies detected using specific cell-based assays.\(^1\) MOG-AD is often associated with neuromyelitis optica spectrum disorder (NMOSD). Although the clinical course and presentation for these diseases can be similar, there are some critical differences in pathophysiology, prognoses and outcomes. Recognition and identification of MOG-AD versus other neuroinflammatory conditions, such as NMOSD, is critical to ensure appropriate clinical evaluation, treatment plans and optimal outcomes for MOG-AD patients. It has been proposed that a specific ICD-10-CM code for MOG-AD will facilitate research and data collection as well as improve diagnosis and patient care. This proposal is based on a submission from Jonathan D. Santoro, MD, Division of Neurology, Department of Pediatrics, Children's Hospital Los Angeles, Keck School of Medicine at USC and The MOG Project.

MOG-AD can occur in all decades of life, with a slight predominance in women and with median age of onset in the early to mid-thirties. The most common presenting feature is optic neuritis (ON), occurring in 54–61% of patients, followed by myelitis, acute disseminated encephalomyelitis (ADEM) or an ADEM-like presentation (e.g., brainstem attack).\(^1\) A relapsing course has been reported in 44–83% of patients and more commonly involves the optic nerve. MOG-positive ON is frequently bilateral and associated with optic nerve head swelling.\(^1\)

A large body of immunologic and clinical evidence has made clear that MOG-AD is a distinct entity.\(^2\) The autoantibodies in MOG-AD have a unique target and the disease has unique immunologic mechanisms that distinguish it from other conditions, such as aquaporin-4 (AQP-4) -positive NMOSD and MS.\(^3^4\) While MOG-AD was initially believed to represent a subset of patients with neuromyelitis optica (NMO), it is now clear that this is not the case. MOG is located on the surface of myelin sheaths and oligodendrocyte processes, whereas AQP-4 water channels, the target of seropositive NMO, are located on astrocytic foot processes.\(^5\) It has also been shown that about one-third of demyelinating lesions in patients with MOG-AD have MOG-dominant myelin loss, but relatively preserved oligodendrocytes. This differs from AQP-4-positive NMOSD lesions, which exhibit myelin-associated glycoprotein-dominant oligodendrogliopathy.\(^6\) In addition, complement activation plays a significant role in AQP-4-IgG-positive NMOSD and is a potential treatment target,\(^7^8\) but appears to be less important in MOG-AD.\(^6\) Most recently, a new neutrophil granulocyte-specific biomarker has been shown to have high sensitivity and specificity for rapid differentiation among patients with NMOSD, MOG-AD, and relapsing-remitting MS.\(^9\)

Differences in pathology of MOG-AD, MS, and AQP-4-positive NMOSD result in distinct clinical symptoms and prognoses. MOG-IgG optic neuritis has a higher likelihood of being recurrent, bilateral, associated with intervertebral disc edema, and have perineural enhancement around the optic nerve on magnetic resonance imaging in contrast to demyelinating optic neuritis arising from MS.\(^4\) In addition, the overall course of optic neuritis in MOG-AD differs from that in MS patients. MS patients with optic neuritis generally have unilateral involvement and occurrence only once
during the course of their disease, while involvement in MOG-AD is bilateral and relapsing. 
Recovery from attacks in MOG-AD is typically better than that seen in AQP-4-positive NMOSD. 
In addition, ADEM is a relatively common presentation in pediatric patients with MOG-AD, but is not seen in either MS or AQP-4-IgG-positive NMOSD. The prognoses for MOG-AD and AQP-4-IgG-positive NMOSD also differ, with much higher risk for fatality and permanent disability with the latter condition.

There is a very clear consensus in the clinical literature that MOG-AD is a distinct pathologic and clinical entity with a unique disease course and management requirements. Patients with MOG-AD exhibit unique characteristics. Although various data points such as clinical picture, severity and antibody titers are established, prognostic factors for MOG-AD are still lacking. For example, physicians have difficulty predicting whether a patient will relapse. For patients who do relapse, the consensus is to provide maintenance therapy, but there is currently no clinical consensus on what treatments are the most effective.

Inflammatory demyelinating CNS diseases are a heterogeneous group, and although the sensitivity and specificity of diagnostic criteria have improved, misdiagnosis is not infrequent and occurs in up to 10% of cases. Results from one study showed that 9% of patients with ADEM according to clinical criteria were misdiagnosed, since the pathology was similar to that of MS. This is particularly concerning, given that certain treatment protocols used for other similar disorders like MS have been found to exacerbate MOG-AD symptoms.

A study in the UK found an incidence rate for MOG-AD of about 3.4/1 000 000 person-years. This compared to a nationwide incidence rate of about 1.6/1 000 000 persons in Holland (from 2014 to 2017), with a higher rate in children (of 3.1/1 000 000, vs 1.3/1 000 000 in adults).

An individual code for MOG-AD will enable better identification and tracking of this distinct set of patients, which will advance the clinical understanding of MOG-AD, and subsequently enable improvement in the diagnostic and treatment paradigms and facilitate research and data collection. Increased awareness of MOG-AD will help to decrease diagnostic delay in these patients, which may potentially decrease the likelihood of permanent disability, such as blindness and paralysis. Furthermore, accurate identification is critical to facilitate research that will further elucidate the marked needs and characteristics of MOG-AD, as well as the development of effective treatment for MOG-AD.

References

TABULAR MODIFICATIONS

G37 Other demyelinating diseases of central nervous system

G37.8 Other specified demyelinating diseases of central nervous system

<table>
<thead>
<tr>
<th>New code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>G37.81</td>
<td>Myelin Oligodendrocyte Glycoprotein Antibody Disease</td>
</tr>
<tr>
<td>Add</td>
<td>Code also associated manifestations, such as:</td>
</tr>
<tr>
<td>Add</td>
<td>neuromyelitis optica (G36.0)</td>
</tr>
<tr>
<td>Add</td>
<td>noninfectious acute disseminated encephalomyelitis (G04.81)</td>
</tr>
</tbody>
</table>
Nontraumatic Coma Due to Underlying Condition

This is a representation from the September 2021 and March 2022 ICD-10 Coordination and Maintenance Committee Meetings of the Coma Due to Underlying Condition proposal with the recommended modification to add “nontraumatic” to the proposed code title R40.2A, Coma due to underlying condition. The modification is in bold.

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

R40.20, Unspecified Coma, is the only code available for coma in patients who do not have TBI but have conditions without combination codes describing the coma; for example, coma secondary to spontaneous brain hemorrhage.

TABULAR MODIFICATIONS

R40 Somnolence, stupor and coma

Excludes1: neonatal coma (P91.5)

somnolence, stupor and coma in diabetes (E08-E13)

somnolence, stupor and coma in hepatic failure (K72.-)

somnolence, stupor and coma in hypoglycemia (nondiabetic) (E15)

R40.2 Coma

Code first any associated:

fracture of skull (S02.-)

intracranial injury (S06.-)

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

New code R40.2A Nontraumatic coma due to underlying condition

Add Secondary coma

Add Code first underlying condition
Obesity in Children, Adolescents, and Adults

Obesity is a highly prevalent chronic disease with complex inflammatory and endocrinological pathophysiology.¹ The American Medical Association has recognized obesity as a disease since 2013.² Obesity is associated with serious health and social consequences.³ Childhood obesity is defined by a body mass index (BMI = body weight in kilograms divided by height in meters squared (kg/m²)) ≥95th percentile for age and sex.⁴ Currently, approximately 1 in 5 U.S. children have obesity.⁵ Furthermore, the proportion of children and adolescents 2-19 years with severe obesity (BMI ≥120% above the 95th percentile) in 2015-2018 was 7.6%.⁶ Obesity in childhood predisposes to obesity in both adolescence and adulthood.⁷

Obesity puts children and adolescents at risk for serious short- and long-term adverse health outcomes later in life, including cardiovascular disease (CVD), hypertension (HTN), dyslipidemia, insulin resistance, type 2 diabetes mellitus (T2DM), obstructive sleep apnea (OSA), obesity-related glomerulopathy (ORG), and non-alcoholic fatty liver disease (NAFLD).⁸-¹² In addition to physical and metabolic consequences, obesity in childhood and adolescence is associated with poor psychological and emotional health, stigmatization, bullying, increased stress, depressive symptoms, and low self-esteem.¹³ Significantly, the severity of comorbidities increases with increasing adiposity.⁸ Furthermore, stigmatization, depression and low self-esteem contribute to binge eating, social isolation, avoidance of health care services, and decreased physical activity.¹⁴

Obesity in childhood and adolescence is associated with increased health care utilization and costs.¹⁵ In an analysis of the National Inpatient Sample database from 2006-2016, the most common conditions that co-occur with a diagnosis of obesity and increase costs and utilization included mood disorders, asthma, and diabetes.¹⁵

Interventions to manage and treat childhood obesity consist of family-based, intensive and comprehensive lifestyle interventions.¹⁶ The United States Preventive Services Task Force (USPSTF) recognizes that weight management or lifestyle interventions that include nutritional and physical activity counseling and deliver ≥26 hours in a 6-month period as being effective in improving weight status among children and adolescents as well as cardiovascular risk factors.¹⁶ These interventions have been shown to be cost-effective with important health benefits to caregivers as well.¹⁶,¹⁷

Obesity in children and adolescents is determined by age- and gender-specific percentiles. Therefore, a child or adolescent may suffer from obesity at a lower BMI than an adult. For adults, the overweight range is from a BMI of 25.0 to <30. Obesity in adults is subdivided into the following: Class 1: BMI of 30 to < 35; Class 2: BMI of 35 to < 40; Class 3: BMI of 40 or higher (sometimes categorized as “severe” obesity).¹⁸ Obesity in children uses a classification system recognizing BMI ≥95th percentile as class I obesity, BMI ≥120% of the 95th percentile as class II obesity, and BMI ≥140% of the 95th percentile as class III obesity.¹⁹

Previous etiological hypotheses around obesity suggested an imbalance in caloric intake. This understanding is no longer accepted, and obesity is understood to be a complex, inflammatory
Therefore, identifying excess calories as the cause of obesity does not reflect current medical understanding.

Providers and medical associations recognize that children and adolescents with obesity, and their families, experience stigmatization that can negatively affect health-seeking behavior and outcomes. This stigmatization may come directly from providers and includes terminology such as “morbid” and assumptions that obesity is due to personal choices relating to caloric consumption. Using people-first language, e.g., “children with obesity” rather than “obese children,” is an important written and verbal communication strategy to avoid stigmatization. Use of language that may cause stigmatization, such as “morbid” or “excess calories,” or the use of non-people-first language may impact how providers document their services.

Accordingly, these difficulties may impact how obesity is diagnosed, classified, managed, and tracked over time in a clinical setting. Several studies have determined that current ICD codes have low sensitivity in identifying obesity and may impact clinical care by, for example, lowering referral rates to specialists and weight management programs. These shortcomings may also have serious implications for current and future health of the growing population of children and adolescents suffering from obesity.

Proposed changes to the ICD-10-CM obesity codes have been received from CDC, the Division of Nutrition, Physical Activity and Obesity of the National Center for Chronic Disease Prevention and Health Promotion with further input from additional obesity experts.

References

**TABULAR MODIFICATIONS**

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>E66</td>
<td>Overweight and obesity</td>
</tr>
</tbody>
</table>
|      | Code first obesity complicating pregnancy, childbirth and the puerperium, if applicable (O99.21-)
|      | Use additional code to identify body mass index (BMI), if known (Z68.-) |
| Delete | Excludes1: adiposogenital dystrophy (E23.6) |
|        | lipomatosi NOS (E88.2) |
|        | lipomatosi dolorosa [Dercum] (E88.2) |
|        | Prader-Willi syndrome (Q87.11) |
| Add   | Excludes2: adiposogenital dystrophy (E23.6) |
| Add   | lipomatosi NOS (E88.2) |
| Add   | lipomatosi dolorosa [Dercum] (E88.2) |
| Add   | Prader-Willi syndrome (Q87.11) |
| E66.0 | Obesity due to excess calories |
| Revise | E66.01 Extreme Morbid (severe) obesity due to excess calories |
| Revise | Excludes1: extreme morbid (severe) obesity with alveolar hypoventilation (E66.2) |
| Revise | E66.2 Extreme Morbid (severe) obesity with alveolar hypoventilation |
|      | Obesity hypoventilation syndrome (OHS) |
|      | Pickwickian syndrome |
E66.8 Other obesity

New sub-subcategory E66.81 Obesity in children and adolescents

Add Use additional code to identify body mass index (BMI), pediatric, if known (Z68.5-)

New code E66.811 Obesity in children and adolescents, class 1
New code E66.812 Obesity in children and adolescents, class 2
New code E66.813 Obesity in children and adolescents, class 3
Add Severe obesity in children and adolescents
New code E66.819 Obesity in children and adolescents, unspecified

New sub-subcategory E66.82 Obesity in adults

Add Use additional code to identify body mass index (BMI), adult, if known (Z68.1-Z68.45)

New code E66.821 Obesity in adults, class 1
New code E66.822 Obesity in adults, class 2
New code E66.823 Obesity in adults, class 3
Add Severe obesity (in adults)
New code E66.829 Obesity in adults, unspecified

New code E66.89 Other obesity
INDEX MODIFICATIONS

Obesity E66.9
Add - extreme (see also Obesity, severe) E66.823
Add - - with
Add - - - alveolar hypoventilation E66.2
Add - - - obesity hypoventilation syndrome (OHS) E66.2
Add - - due to excess calories E66.01
Revise - morbid (see also Obesity, severe) E66.823 E66.04

Revise - severe E66.823 E66.01
Add - - in adolescents E66.813
Add - - in adults E66.823
Add - - in children E66.813

Revise - specified type NEC E66.89
Phelan-McDermid Syndrome

Phelan-McDermid syndrome was recognized as a distinct diagnosis in the 1990’s with the first case report published in the medical literature in 1992. Often abbreviated as PMS, it is a genetic neurodevelopmental condition with multi-system manifestations.

The genetic cause of Phelan-McDermid syndrome has been established to be defects in a specific portion of the long arm of chromosome 22. In about 80% of individuals, the cause is a deletion in a segment of chromosome 22 known as q13.3. For this reason, Phelan-McDermid syndrome was initially named 22q13.3 deletion syndrome. Less commonly, in about 20% of cases, Phelan-McDermid syndrome is caused by pathogenic variants in the SHANK3 gene located at the distal long arm of chromosome 22.

Signs and symptoms of Phelan-McDermid syndrome are often seen in infancy and early childhood. Classically, these include low muscle tone in infants, delays in reaching developmental milestones, delayed speech, or inability to develop functional speech, and global developmental delay leading to varying degrees of intellectual disability. Certain minor dysmorphic features are also characteristic of Phelan-McDermid syndrome, such as elongation of the head, full brow, deep-set eyes, wide nasal bridge and bulbous nose, prominent ears, and large, fleshy hands.

As children with Phelan-McDermid syndrome age, other disorders and symptoms are often seen, although the specific symptoms vary with the individual and may occur only in certain subgroups or at specific stages in life. Common disorders and symptoms include seizures, gastrointestinal disorders such as cyclic vomiting and constipation, sleep difficulties, lymphedema, and neuropsychiatric illness. A significant minority have kidney abnormalities such as multicystic kidneys and vesicoureteral reflux. Of note, people with Phelan-McDermid syndrome often have decreased perception of pain as well as low levels of sweating, putting them at risk for unrecognized injury and overheating.

Phelan-McDermid syndrome is also known to have a strong association with autism. It is estimated that approximately 75-80% of people with Phelan-McDermid syndrome also have autism spectrum disorder. Conversely, it is estimated that approximately 1% of people with autism have Phelan-McDermid syndrome.

From its association with autism, it is believed that the prevalence of Phelan-McDermid syndrome is approximately 1 in 10,000 to 15,000 people. However, this is an extrapolation and may be understated because of the challenges in identifying the diagnosis distinctly in healthcare databases.

The diagnosis of Phelan-McDermid syndrome is made definitively by genetic testing. Chromosomal microarray analysis is the first line of testing and is required for diagnosis in most cases to detect a deletion. Whole exome/genome sequencing is required for detecting sequence variants in SHANK3. As technologies in genetic sequencing continue to become less costly and more widely used, the known prevalence of Phelan-McDermid syndrome is likely to continue to increase.
There is currently no treatment for the underlying genetic basis of Phelan-McDermid syndrome. Care is directed at managing the symptoms and risks for each affected individual. Families often see teams of specialists, including a neurologist, pediatrician or primary care physician, gastroenterologist, nephrologist, endocrinologist, and psychiatrist, as well as a speech-language pathologist and behavioral, occupational, and physical therapists.

There are currently three clinical trials related to Phelan-McDermid syndrome at various stages in the U.S. Three other treatment trials were recently completed in the U.S.

Given that the main cause of Phelan-McDermid syndrome is a deletion in chromosome 22, subcategory Q93.5 is an appropriate location for a new code. This is also consistent with WHO ICD-11, which categorizes Phelan-McDermid syndrome under code LD44, Deletions of the autosomes.

In addition to aiding in identifying the true prevalence of Phelan-McDermid syndrome, a specific ICD-10-CM code would enable tracking the disorder, calculating the public health impact, assist in clinical trials and retrospective research.

This proposal is submitted by the Phelan-McDermid Syndrome foundation.

References:


See also: https://pmsf.org/about-pms/
TABULAR MODIFICATIONS

Q93  Monosomes and deletions from the autosomes, not elsewhere classified

Q93.5 Other deletions of part of a chromosome

Q93.51 Angelman syndrome

New code  Q93.52 Phelan-McDermid syndrome
Add  22q13.3 deletion syndrome

Add  Use additional code(s) to identify any associated conditions, such as:
Add  autism spectrum disorder (F84.0)
Add  degree of intellectual disabilities (F70-F79)
Add  epilepsy and recurrent seizures (G40.-)
Add  lymphedema (I89.0)

Q93.59 Other deletions of part of a chromosome
Short Bowel Syndrome and Intestinal Failure

This topic was presented at the September 2020 Coordination and Maintenance Meeting. This proposal has been revised to divide short bowel syndrome (SBS) into those with and without colon in continuity with the residual small bowel. SBS with colon in continuity is when the colon has been anastomosed to residual small bowel. This includes ileocolonic and jejunocolonic anastomoses. SBS with no colon in continuity is when all colon has been resected, or otherwise is not in continuity with the residual small bowel. This includes mucus fistula, ileostomy, jejunostomy, duodenostomy patients and jejuno/ileo-rectal anastomosis that meet the definition for SBS.

Short bowel syndrome (SBS) is a condition that occurs when your body is unable to absorb enough nutrients from the foods you eat because you do not have enough small intestine. Short bowel syndrome is caused by the physical absence or loss of massive portions of intestine (typically to < 200 cm of residual intestine). Many, but not all individuals with SBS may also develop intestinal failure (IF), which is the inability to absorb enough nutrients and/or fluid necessary to maintain nutritional autonomy. Conversely, not all patients with IF suffer from SBS, but may have a myriad of different malabsorptive disorders. Treatment of patients with SBS and IF is complex, including the management of intravenous nutrition and fluids, and the prevention and treatment of nutrient deficiencies and dehydration. Additional complications of the disease can affect the liver, kidney, brain, and bones. It is likely that lack of knowledge and understanding of both SBS and IF has led to misclassification of these diseases under various identifiers, thereby commonly reported prevalence numbers may be either over- or under-representations of actual disease prevalence.

In patients with short bowel syndrome, the colon assumes a substantially greater role than normal with regard to both fluid and nutrient absorption than in individuals with a fully intact digestive tract. The colon absorbs fluid, electrolytes, some amino acids and medium chain triglycerides (MCT), but most importantly, through fermentation of unabsorbed carbohydrates by what is termed “carbohydrate salvage,” the colon becomes a factory for energy production through the production of short chain fatty acids. The amount of energy produced may amount to as much as 1000 kcal daily.

Individuals with short bowel syndrome who have no residual colon after resection have a poorer prognosis, respond to a lesser degree to dietary therapy, and are more likely to require long-term or permanent intravenous feeding than those individuals who have colon in continuity. This may relate largely to a decrease in glucagon-like peptide 2 (GLP-2) production in the former.

Creating a unique ICD-10-CM codes for SBS would facilitate better care via (a) exposing regional variations and areas for improvements in care, and (b) ultimately, enabling continuity of care by tracking patients across systems and optimizing standards of care.

Alan Buchman, MD, MSPH is proposing the following tabular modifications for better delineation in these conditions. Changes in bold.
TABULAR MODIFICATIONS

K90  Intestinal malabsorption

K90.8  Other intestinal malabsorption

New subcategory  K90.82 Short bowel syndrome

New code  K90.821 Short bowel syndrome with colon in continuity

New code  K90.822 Short bowel syndrome without colon in continuity

New code  K90.829 Short bowel syndrome, unspecified

New code  K90.83 Intestinal failure
**Sickle-Cell Retinopathy**

Sickle-cell disease is the most common inherited blood disorder. Sickle cell retinopathy is characterized by the blockage of outer retinal vessels, resulting in both non-proliferative and proliferative retinopathy, that can lead to complications such as vision impairment and blindness. Treatment of sickle cell retinopathy includes observation, retinal ablation, advanced retinal-vitreal surgery and the intravitreal injection of anti-VEGF biologic agents to control the proliferative retinopathy. The expanding use of intravitreal biologic agents is changing the prognosis for sickle-cell retinal disease.

Bevacizumab has been found to allow patients to maintain vision without the need for more invasive interventions such as vitrectomy to clear vitreous hemorrhage or to repair retinal detachment. At present there is no specific ICD-10-CM diagnostic code for proliferative sickle cell retinopathy. This absence makes it difficult to determine medical necessity of management. Specific reporting is also important from a public health perspective to be able to study the prevalence of this specific blinding disease in the US population. Retinal issues occur in the same eye or in fellow eyes of sickle-cell patients, either synchronously or asynchronously. Currently physicians and facilities use the following non-specific code, H35.2-, Other nondiabetic proliferative retinopathy.

There is no synonym identifying sickle cell retinal disease in the current ICD-10-CM. While including synonym(s) would be an improvement in ICD-10-CM there are several limitations to this approach. First, it would not allow the public health evaluation and study of this specific retinal disease. Second, it does not specify the proliferative form of the disease as compared with background retinopathy. Third, non-diabetic proliferative retinopathies have differing treatments based on ocular or systemic diagnosis. To that goal new diagnostic codes are being requested for ICD-10-CM.

The American Academy of Ophthalmology is proposing the following tabular modifications to report treatment accurately and to support the need for disease-specific reporting of sickle cell proliferative retinopathy.

**TABULAR MODIFICATIONS**

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<thead>
<tr>
<th>Code</th>
<th>Description</th>
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<tr>
<td>H35</td>
<td>Other retinal disorders</td>
</tr>
<tr>
<td>H35.2</td>
<td>Other non-diabetic proliferative retinopathy</td>
</tr>
<tr>
<td></td>
<td>Proliferative vitreo-retinopathy</td>
</tr>
<tr>
<td>Add</td>
<td>Thalassemia proliferative retinopathy</td>
</tr>
<tr>
<td>Add</td>
<td>Excludes2: Proliferative sickle-cell retinopathy (H36.82-)</td>
</tr>
</tbody>
</table>
H36  Retinal disorders in diseases classified elsewhere
Code first underlying disease, such as:
    lipid storage disorders (E75.-)
sickle-cell disorders (D57.-)

Excludes1: arteriosclerotic retinopathy (H35.0-)
    Diabetic retinopathy (E08.3-, E09.3-, E10.3-, E11.3-, E13.3-)

New subcategory   H36.8  Other retinal disorders in diseases classified elsewhere

New sub-subcategory H36.81  Non proliferative sickle-cell retinopathy
New code           H36.810 Non proliferative sickle cell retinopathy, unspecified eye
New code           H36.811 Non proliferative sickle cell retinopathy, right eye
New code           H36.812 Non proliferative sickle cell retinopathy, left eye
New code           H36.813 Non proliferative sickle cell retinopathy, bilateral

New sub-subcategory H36.82  Proliferative sickle-cell retinopathy
New code           H36.820 Proliferative sickle cell retinopathy, unspecified eye
New code           H36.821 Proliferative sickle cell retinopathy, right eye
New code           H36.822 Proliferative sickle cell retinopathy, left eye
New code           H36.823 Proliferative sickle cell retinopathy, bilateral

New code           H36.89  Other retinal disorders in diseases classified elsewhere
Add                 Retinal dystrophy in lipid storage disorders
Social Determinants of Health

This proposal was originally submitted by the Gravity Project (GP) and presented at the March 2021 and September 2021 and March 2022 ICD-10 Coordination and Maintenance (C&M) meetings. The American Academy of Pediatrics (AAP) also had several code requests that were presented at the September 2021 and March 2022 C&M meeting. Parts of the proposal were previously approved and will be implemented on October 1, 2022. The files are posted on the CDC webpage Comprehensive Listing ICD-10-CM Files (cdc.gov). This proposal will be considered for April 1, 2023, implementation.

Subsequently, we have received additional code requests from the GP and CMS, as well as modifications to AAP’s previously presented code request for Z91.1Patient's noncompliance with medical treatment and regimen expansion that have been incorporated in the proposal and are bold.

One of the five overarching goals of Healthy People 2030 is ‘attaining health literacy to improve the health and well-being of all”. Included in this goal is the adoption of the following definition. Personal health literacy is the degree to which individuals have the ability to find, understand, and use information and services to inform health-related decisions and actions for themselves and others

New ICD-10-CM codes are being requested at the T74.A- and the T76.A- previously expanded codes to include Child financial abuse, confirmed and Child financial abuse, suspected. There is also a request for inclusion terms under codes T74.3 and T76.3 to describe patients who identify that they have been threatened with harm. Receiving threats is a subtype of psychological abuse within the American Psychological Association.

The proposed Y07.0 Spouse or partner, perpetrator of maltreatment and neglect is being expanded as requested by comments from the March 2022 meeting.

A request for a new term under Z58 to cover basic necessities unavailable in the environment, Z58.81 Services unavailable in physical environment. Additional code requests were received to expand code Z59.1 Inadequate housing to further describe housing inadequacy.

Health Insurance Coverage, Healthy People 2030 includes several objectives that relate to improving the proportion of people with some form of health or dental insurance or reducing the proportion of people under 65 who are uninsured. Earlier this year, the Centers for Medicare and Medicaid Services (CMS) unveiled an initiative to reduce the uninsured rate among children and increase Medicaid enrollment for parents and pregnant people. Lack of insurance affects access to care and preventative services as well as an increase in mortality. Although one could say that the current code Z59.7 Insufficient social insurance and welfare support applies, our consensus statement is that it is not specific enough to support this
important use case. Social insurance is a broad-based term and welfare support can include different types of assistance programs outside of insurance coverage. The Gravity Project community determined that a specific code to identify insufficient health insurance coverage is needed to properly identify this situation.

New codes and revisions have been made to further describe circumstances related to food insecurity, housing instability, transportation needs, utility difficulties, and interpersonal safety to illuminate their impact on health outcomes. These health-related social needs (HRSNs), defined as individual-level, adverse social conditions that negatively impact a person’s health or healthcare, are significant risk factors associated with worse health outcomes as well as increased healthcare utilization. Assessment of HRSNs is an essential mechanism for capturing the interaction between social, community, and environmental factors associated with health status and health outcomes. While widespread interest in addressing HRSNs exists, action is inconsistent, with 92 percent of hospitals screening for one or more of the five HRSNs, but only 24 percent of hospitals screening for all five HRSNs. Utilization of screening tools to identify the burden of unmet HRSNs can be a helpful first step in identifying necessary community partners and connecting individuals to resources in their communities. For data collection of responses to screening tools, there is a need to better track data elements of the HRSN domains.

References


TABULAR MODIFICATIONS

T74 Adult and child abuse, neglect, and other maltreatment, confirmed

T74.3 Psychological abuse, confirmed
Bullying and intimidation, confirmed
Intimidation through social media, confirmed

Add
Target of threatened harm, confirmed

Add
Target of threatened physical violence, confirmed

Add
Target of threatened sexual abuse, confirmed

New
subcategory
T74.A Financial abuse, confirmed

New code
T74.A1 Adult financial abuse, confirmed

New code
T74.A2 Child financial abuse, confirmed
T76 Adult and child abuse, neglect, and other maltreatment, suspected
  T76.3 Psychological abuse, suspected
    Bullying and intimidation, suspected
    Intimidation through social media, suspected
  Add Target of threatened harm, suspected
  Add Target of threatened physical violence, suspected
  Add Target of threatened sexual abuse, suspected

New subcategory T76.A Financial abuse, suspected
New code T76.A1 Adult financial abuse, suspected
New code T76.A2 Child financial abuse, suspected

Y07 Perpetrator of assault, maltreatment and neglect
Note: Codes from this category are for use only in cases of confirmed abuse (T74.-)
Selection of the correct perpetrator code is based on the relationship between the perpetrator and the victim

Add Includes: perpetrator of verbal abuse

Y07.0 Spouse or partner, perpetrator of maltreatment and neglect
  Spouse or partner, perpetrator of maltreatment and neglect against spouse or partner

Y07.01 Husband, perpetrator of maltreatment and neglect
New code Y07.010 Husband, current, perpetrator of maltreatment and neglect
New code Y07.011 Husband, former, perpetrator of maltreatment and neglect

Y07.02 Wife, perpetrator of maltreatment and neglect
New code Y07.020 Wife, current, perpetrator of maltreatment and neglect
New code Y07.021 Wife, former, perpetrator of maltreatment and neglect

Y07.03 Male partner, perpetrator of maltreatment and neglect
Add Intimate or dating partner, perpetrator of maltreatment and neglect
<table>
<thead>
<tr>
<th>New code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Y07.030</td>
<td>Male partner, current, perpetrator of maltreatment and neglect</td>
</tr>
<tr>
<td>Y03.031</td>
<td>Male partner, former, perpetrator of maltreatment and neglect</td>
</tr>
<tr>
<td>Y07.04</td>
<td>Female partner, perpetrator of maltreatment and neglect</td>
</tr>
<tr>
<td>Add</td>
<td>Intimate or dating partner, perpetrator of maltreatment and neglect</td>
</tr>
<tr>
<td>Y07.040</td>
<td>Female partner, current, perpetrator of maltreatment and neglect</td>
</tr>
<tr>
<td>Y07.041</td>
<td>Female partner, former, perpetrator of maltreatment and neglect</td>
</tr>
<tr>
<td>Y07.05</td>
<td>Non-binary partner, perpetrator of maltreatment and neglect</td>
</tr>
<tr>
<td>Add</td>
<td>Gender non-conforming partner, perpetrator of maltreatment and neglect</td>
</tr>
<tr>
<td>Y07.050</td>
<td>Non-binary partner, current, perpetrator of maltreatment and neglect</td>
</tr>
<tr>
<td>Y07.051</td>
<td>Non-binary partner, former, perpetrator of maltreatment and neglect</td>
</tr>
<tr>
<td>Y07.4</td>
<td>Other family member, perpetrator of maltreatment or neglect</td>
</tr>
<tr>
<td>Y07.44</td>
<td>Child (biological, step, in-law, foster, adopted), perpetrator of maltreatment, and neglect</td>
</tr>
<tr>
<td>Add</td>
<td>Daughter, perpetrator of maltreatment, and neglect</td>
</tr>
<tr>
<td>Add</td>
<td>Non-binary child, perpetrator of maltreatment, and neglect</td>
</tr>
<tr>
<td>Add</td>
<td>Son, perpetrator of maltreatment, and neglect</td>
</tr>
<tr>
<td>Y07.45</td>
<td>Grandchild (biological, step, in-law, foster, adopted), perpetrator of maltreatment, and neglect</td>
</tr>
<tr>
<td>Add</td>
<td>Granddaughter, perpetrator of maltreatment, and neglect</td>
</tr>
<tr>
<td>Add</td>
<td>Grandson, perpetrator of maltreatment, and neglect</td>
</tr>
</tbody>
</table>
Add Non-binary grandchild, perpetrator of maltreatment, and neglect

New Code

Y07.46 Grandparent, perpetrator of maltreatment, and neglect

Add Grandfather, perpetrator of maltreatment, and neglect

Add Grandmother, perpetrator of maltreatment, and neglect

Add Non-binary grandparent, perpetrator of maltreatment, and neglect

New Code

Y07.47 Parental sibling, perpetrator of maltreatment and neglect

Add Aunt, perpetrator of maltreatment and neglect

Add Non-binary parental sibling, perpetrator of maltreatment or neglect

Add Uncle, perpetrator of maltreatment and neglect

Y07.49 Other family member, perpetrator of maltreatment and neglect

Y07.499 Other family member, perpetrator of maltreatment and neglect

Add Excludes2: parental sibling, perpetrator of maltreatment and neglect (Y07.47)

Y07.5 Non-family member, perpetrator of maltreatment and neglect

New Code

Y07.54 Acquaintance or friend, perpetrator of maltreatment and neglect

Z55 Problems related to education and literacy

New code

Z55.6 Problems related to health literacy

Add Difficulty understanding health related information

Add Difficulty understanding medication instructions

Add Problem completing medical forms

Z58 Problems related to physical environment

New subcategory

Z58.8 Other problems related to physical environment

New code

Z58.81 Basic services unavailable in physical environment
Add Unable to obtain utilities, due to inadequate physical environment
Add Unable to obtain internet service, due to unavailability in geographic area
Add Unable to obtain telephone service, due to unavailability in geographic area

New code Z58.89 Other problems related to physical environment

Z59 Problems related to housing and economic circumstances
Z59.1 Inadequate Housing
Delete Lack of heating
Delete Restriction of space
Delete Technical defects in home preventing adequate care
Delete Unsatisfactory surroundings

New code Z59.10 Inadequate housing, unspecified
Add Inadequate housing, NOS

New code Z59.11 Inadequate housing
Add Pest infestation
Add Restriction of space
Add Technical defects in home preventing adequate care
Add Unsatisfactory surroundings

New code Z59.12 Inadequate environmental temperature
Add Lack of air conditioning
Add Lack of heating

New code Z59.13 Inadequate utilities
Add Lack of electricity services
Add Lack of gas services
Add Lack of oil services
Add Lack of water services

Add Excludes2: lack of adequate food (Z59.4-)
Add other problems related to housing and economic circumstances (Z59.8-)

Z59.7 Insufficient social insurance and welfare support
Add Inadequate social and welfare insurance
Add Insufficient social and welfare insurance

New code Z59.70 Insufficient social insurance and welfare support, unspecified
New code  
Z59.71 Insufficient health insurance coverage  
Add  
No health insurance coverage  

New code  
Z59.79 Other insufficient social insurance and welfare support  

Z59.8 Other problems related to housing and economic circumstances  

Z59.81 Housing instability, housed  
Foreclosure on home loan  
Past due on rent or mortgage  
Unwanted multiple moves in the last 12 months  

Z59.811 Housing instability, housed, with risk of homelessness  
Imminent risk of homelessness  

Z59.812 Housing instability, housed, homelessness in past 12 months  

Z59.819 Housing instability, housed unspecified  
Add  
Excludes2: extreme poverty (Z59.5)  
Add  
financial insecurity (Z59.86)  
Add  
low income (Z59.6)  
Add  
material hardship, not elsewhere classified (Z59.87)  

Z59.82 Transportation insecurity  
Add  
Excludes2: unavailability and inaccessibility of health-care facilities (Z75.3)  

Revise  
Z59.87 Material hardship, due to limited financial resources, not elsewhere classified  
Revise  
Material deprivation due to limited financial resources  
Revise  
Unable to obtain adequate childcare due to limited financial resources  
Revise  
Unable to obtain adequate clothing due to limited financial resources  
Revise  
Unable to obtain adequate utilities due to limited financial resources  
Revise  
Unable to obtain basic needs, due to limited financial resources
Z60 Problems related to social environment
   Z60.4 Social exclusion and rejection
   Exclusion and rejection on the basis of personal
   characteristics, such as unusual physical appearance,
   illness or behavior
Add       Social isolation

Z62 Problems related to upbringing
   Z62.8 Other specified problems related to upbringing
      Z62.81 Personal history of abuse in childhood

New code   Z62.814 Personal history of child financial abuse
New code   Z62.815 Personal history of intimate partner abuse in childhood

Z91 Personal risk factors, not elsewhere classified
   Z91.1 Patient's noncompliance with medical treatment and regimen
Add       Code also, if applicable, to identify underdosing of specific
drug (T36-T50 with final character 6)

New sub-category Z91.14 Patient's other noncompliance with medication regimen
                  Patient's underdosing of medication
New code       Z91.141 Patient’s other noncompliance with medication regimen due to financial
                  hardship
New code       Z91.148 Patient’s other noncompliance with medication regimen for other reason
New sub-category Z91.15 Patient's noncompliance with renal dialysis
New code       Z91.151 Patient's noncompliance with renal dialysis due to financial hardship
New code       Z91.158 Patient's noncompliance with renal dialysis for other reason

Z91.4 Personal history of psychological trauma, not elsewhere classified
   Z91.41 Personal history of adult abuse

New code   Z91.413 Personal history of adult financial abuse
New code  

**Z91.414**  Personal history of adult intimate partner abuse

**INDEX MODIFICATIONS**

- Maltreatment
  - adult
  - - threatened abuse (harm)
  - Add - - - confirmed T74.31
  - Add - - - suspected T76.31
  - - child
  - Add - - threatened abuse (harm)
  - Add - - - confirmed T74.32
  - Add - - - suspected T76.32

- Threatened
  - Add - abuse (harm) – see Maltreatment
Wasting Disease (Syndrome) Due to Underlying Condition

This is a representation from the September 2021 and March 2022 ICD-10 Coordination and Maintenance Committee Meetings of the Wasting Disease (Syndrome) Due to Underlying Condition proposal with recommended modifications. The modifications are in **bold**.

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

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

The National Center for Health Statistics received a request to create an ICD-10-CM code for wasting disease (syndrome) due to underlying condition for coding specificity to aid in capturing severity of illness for mortality of the underlying conditions and to interrupt the physiological progress or pathways of the condition in hopes of supplying a better clinical outcome for the patients.

References


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**TABULAR MODIFICATIONS**

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>E88</td>
<td>Other and unspecified metabolic disorders</td>
</tr>
<tr>
<td>E88.A</td>
<td>Wasting disease (syndrome) due to underlying condition</td>
</tr>
</tbody>
</table>

**Add**

- Cachexia due to underlying condition
- Code first underlying condition
- Excludes1: cachexia NOS (R64)
- Excludes2: failure to thrive (R62.51, R62.7)
R64 Cachexia

Delete Wasting syndrome
Delete Code first underlying condition, if known
Add Exclude1: cachexia due to underlying condition (E88.A)
TABULAR MODIFICATIONS PROPOSED ADDENDA
All proposed effective October 1, 2024

A40  Streptococcal sepsis

Revise  Code first, if applicable, postprocedural streptococcal sepsis (T81.44-)
Add     sepsis due to central venous catheter (T80.211-)
        streptococcal sepsis during labor (O75.3)
Revise  streptococcal sepsis following abortion or ectopic or molar pregnancy
        (O03.37, O03.87, O04.87, O07.37, O08.0 O08.82)
Revise  streptococcal sepsis following immunization (T88.0-)
Revise  streptococcal sepsis following infusion, transfusion, or therapeutic
        injection (T80.211-, T80.22-, T80.29-)

A41  Other sepsis

Revise  Code first, if applicable, postprocedural sepsis (T81.44-)
Add     sepsis due to central venous catheter (T80.211-)
        sepsis during labor (O75.3)
Revise  sepsis following abortion, ectopic or molar pregnancy (O03.37, O03.87,
        O04.87, O07.37, O08.0 O08.82)
        sepsis following immunization (T88.0-)
Revise  sepsis following infusion, transfusion, or therapeutic injection (T80.211-,
        T80.22-, T80.29-)

A41.5  Sepsis due to other Gram-negative organisms

A41.52  Sepsis due to Pseudomonas

Revise  Pseudomonas aeruginosa  aeruginosa

C61  Malignant neoplasm of prostate

Revise  Use additional, if applicable, code to identify:
        hormone sensitivity status (Z19.1-Z19.2)
        rising PSA following treatment for malignant neoplasm of prostate
        (R97.21)

C92  Myeloid leukemia
Add    Code also, if applicable, pancytopenia (acquired) (D61.818)

C94  Other leukemias of specified cell type
    C94.8  Other specified leukemias
Add    Code also, if applicable, eosinophilia (D72.18)
D12 Benign neoplasm of colon, rectum, anus and anal canal

**Delete**

- Excludes1: benign carcinoid tumors of the large intestine, and rectum (D3A.02-)
- Delete: polyp of colon NOS (K63.5)

**Add**

- Excludes2: benign carcinoid tumors of the large intestine, and rectum (D3A.02-)
- Add: polyp of colon NOS (K63.5)

E13 Other specified diabetes mellitus

**Delete**

- Excludes2: type 2 diabetes mellitus (E11.-)

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

**Revise**

- SCAD deficiency

E72 Other disorders of amino-acid metabolism

- Cystathionine synthase deficiency

E87 Other disorders of fluid, electrolyte and acid-base balance

**Add**

- Excludes1: diabetes with hyperosmolarity (E08, E09, E11, E13 with ending .00 or .01)

F64 Gender identity disorders

**Delete**

- Gender identity disorder in adolescence and adulthood
- Gender dysphoria in adolescents and adults

**Add**

- Gender identity disorder in adolescence and adulthood
- Gender incongruence in adolescents and adults
- Transgender

**Add**

- Excludes1: gender identity disorder of childhood (F64.2)
F64.2  Gender identity disorder of childhood
Gender dysphoria in children
Gender incongruence of childhood
Add

F64.9  Gender identity disorder, unspecified
Gender incongruence, unspecified
Gender dysphoria, unspecified
Gender-role disorder NOS
Add

G32  Other degenerative disorders of nervous system in diseases classified elsewhere
G32.0  Subacute combined degeneration of spinal cord in diseases classified elsewhere

Code first underlying disease, such as:

Revise  vitamin B12 deficiency anemia, unspecified anemia (D51.9)
Revise  other dietary vitamin B12 deficiency anemia dietary (D51.3)
Revise  vitamin B12 deficiency anemia due to intrinsic factor deficiency pernicious (D51.0)

H49  Paralytic strabismus
H49.8  Other paralytic strabismus
H49.81  Kearns-Sayre syndrome

Revise  Use additional code Code also, if applicable, for other manifestations, such as:
heart block (I45.9)

Infections with a predominantly sexual mode of transmission (A50-A64)

Revise  Excludes1: human immunodeficiency virus [HIV] disease (B20) nonspecific and nongonococcal urethritis (N34.1)
Delete  nonspecific and nongonococcal urethritis (N34.1)
Reiter's disease (M02.3-)
Add  Excludes2: human immunodeficiency virus [HIV] disease (B20)

I25  Chronic ischemic heart disease
I25.1  Atherosclerotic heart disease of native coronary artery
I25.11  Atherosclerotic heart disease of native coronary artery with angina pectoris

Revise  I25.112  Atherosclerotic Atherosclerotic heart disease of native coronary artery with refractory angina pectoris
I43 Cardiomyopathy in diseases classified elsewhere

Code first underlying disease, such as:
- amyloidosis (E85.-)
- glycogen storage disease (E74.0-)

Revise

I71 Aortic aneurysm and dissection

I71.5 Thoracoabdominal aortic aneurysm, ruptured

Revise

I71.51 Supraceliac aneurysm of the thoracoabdominal aorta, ruptured

Revise

I71.52 Paravisceral aneurysm of the thoracoabdominal aorta, ruptured

I71.6 Thoracoabdominal aortic aneurysm, without rupture

Revise

I71.61 Supraceliac aneurysm of the thoracoabdominal aorta, without rupture

Revise

I71.62 Paravisceral aneurysm of the thoracoabdominal aorta, without rupture

J41 Simple and mucopurulent chronic bronchitis

Delete

Excludes1: chronic bronchitis NOS (J42)

Delete

chronic obstructive bronchitis (J44.-)

Add

Excludes2: chronic bronchitis NOS (J42)

Add

chronic obstructive bronchitis (J44.-)

J43 Emphysema

Excludes1: compensatory emphysema (J98.3)

Delete

emphysema with chronic (obstructive) bronchitis (J44.-)

Delete

emphysematous (obstructive) bronchitis (J44.-)

Add

Excludes2: emphysema with chronic (obstructive) bronchitis (J44.-)

Add

emphysematous (obstructive) bronchitis (J44.-)

J44 Other chronic obstructive pulmonary disease

Delete

Excludes1: bronchiectasis (J47.-)

Delete

emphysema without chronic bronchitis (J43.-)

Add

Excludes2: bronchiectasis (J47.-)

Add

emphysema without chronic bronchitis (J43.-)
ICD-10 Coordination and Maintenance Committee Meeting
September 13-14-2022

K51 Ulcerative colitis
K51.4 Inflammatory polyps of colon

Delete

Excludes1:
- adenomatous polyp of colon (D12.6)
- polyposis of colon (D12.6)
- polyps of colon NOS (K63.5)

Delete

Add

Excludes2:
- adenomatous polyp of colon (D12.6)
- polyposis of colon (D12.6)
- polyps of colon NOS (K63.5)

K63 Other diseases of intestine
K63.5 Polyp of colon

Delete

Excludes1:
- adenomatous polyp of colon (D12.-)
- inflammatory polyp of colon (K51.4-)
- polyposis of colon (D12.6)

Add

Excludes2:
- adenomatous polyp of colon (D12.6)
- inflammatory polyp of colon (K51.4-)
- polyposis of colon (D12.6)

K80 Cholelithiasis
K80.4 Calculus of bile duct with cholecystitis

Any condition listed in K80.5 with cholecystitis (with cholangitis)

Revise

Codes also, if applicable, fistula of bile duct (K83.3)

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

Revise

Excludes2:
- chondrocalcinosis (M11.1-, M11.2-)
- internal derangement of knee (M23.-)
- metastatic calcification (E83.5)
- ochronosis (E70.29)

Revise

M32 Systemic lupus erythematosus (SLE)
M32.1 Systemic lupus erythematosus with organ or system involvement
M32.19 Other organ or system involvement in systemic lupus erythematosus

Add

Use additional code(s) to identify organ or system involvement, such as encephalitis (G05.3)
N16  Renal tubulo-interstitial disorders in diseases classified elsewhere

Code first underlying disease, such as:
  brucellosis (A23.0-A23.9)
  cryoglobulinemia (D89.1)

Revise  glycogen storage disease (E74.0-)

N35  Urethral stricture

N35.8  Other urethral stricture
  N35.81  Other urethral stricture, male

Revise  N35.812 Other urethral bulbous urethral stricture, male

N81  Female genital prolapse

N81.6  Rectocele

Add  Excludes1: rectocele with prolapse of uterus (N81.2-N81.4)

Excludes2: perineocele (N81.81)
  rectal prolapse (K62.3)

Delete  rectocele with prolapse of uterus (N81.2-N81.4)

O11  Pre-existing hypertension with pre-eclampsia

Revise  Includes: conditions in O10 complicated by pre-eclampsia
  pre-eclampsia superimposed pre-existing hypertension

O26  Maternal care for other conditions predominantly related to pregnancy

O26.8  Other specified pregnancy related conditions
  O26.89  Other specified pregnancy related conditions

Add  Use additional code, if applicable, to identify specific condition such as insulin resistance (E88.81)

**Pregnancy, childbirth and the puerperium (O00-O9A)**

Revise  Use additional code, if applicable, from category Z3A, Weeks of gestation, to identify the specific week of the pregnancy, if known.

P29  Cardiovascular disorders originating in the perinatal period

P29.0  Neonatal cardiac failure

Add  Code also associated underlying condition

Q90  Down syndrome

Add  Code also associated physical condition(s), such as atrioventricular septal defect (Q21.2)
Revise     Use additional code(s) to identify any associated physical conditions and degree of intellectual disabilities (F70-F79)

S06     Intracranial injury
S06.3    Focal traumatic brain injury

Delete     Excludes1: any condition classifiable to S06.4-S06.6

Revise     Excludes2: focal cerebral edema (S06.1) any condition classifiable to S06.4-S06.6
Add        focal cerebral edema (S06.1)

T75     Other and unspecified effects of other external causes
T75.3    Motion sickness
         Airsickness
         Seasickness
         Travel sickness

Revise     Use additional external cause code to identify vehicle or type of motion (Y92.81-, Y93.5-)

W62     Contact with nonvenomous amphibians

Revise     Excludes1: contact with venomous amphibians (T63.81-R63.83)
           (T63.81-T63.83).

Z79     Long term (current) drug therapy
Z79.4    Long term (current) use of insulin

Excludes2: long-term (current) use of injectable non-insulin antidiabetic drugs (Z79.85)
           long term (current) use of oral antidiabetic drugs (Z79.84)
           long term (current) use of oral hypoglycemic drugs (Z79.84)
Add       long term (current) use of noninsulin injectable drug (Z79.84)

Z79.8    Other long term (current) drug therapy
Z79.84   Long term (current) use of oral hypoglycemic drugs
         Long term (current) use of oral antidiabetic drugs

Excludes2: long-term (current) use of injectable non-insulin antidiabetic drugs (Z79.85)
long term (current) use of insulin (Z79.4)
long term (current) use of noninsulin injectable drug (Z79.85)
INDEX MODIFICATION PROPOSED ADDENDA
All proposed effective October 1, 2024

Add          Burkholderia
Add          - cepacia A49.8
Add          - mallei A24.0
Add          - pseudomallei – see Melioidosis

Cachexia R64
Revise   - due to malnutrition R64-E43

Colibacillosis A49.8
Revise   - generalized – see also, Sepsis, Escherichia coli A41.50-A41.51

Complication(s) (from) (of)
- catheter (device) NEC -see also Complications, prosthetic device or implant
  - - epidural infusion T85.9
  - - - mechanical
Revise   - - - - malfunction T85.640 T85.690
  - - subdural infusion T85.9
  - - - mechanical
Revise   - - - - malfunction T85.640 T85.690

Dependence (on) (syndrome) F19.20
- drug NEC F19.20
- - psychoactive NEC F19.20
Add          - - in remission F19.21

Diabetes, diabetic (mellitus) (sugar) E11.9
- with
Revise   - - Kimmelsteil Kimmelstiel-Wilson disease E11.21
  - - due to drug or chemical E09.9
  - - - with
Revise   - - - Kimmelsteil Kimmelstiel-Wilson disease E09.21
  - - due to underlying condition E08.9
  - - - with
Revise   - - - - Kimmelsteil Kimmelstiel-Wilson disease E08.21
- specified type NEC E13.9
- - with
Revise - - - Kimmelsteil Kimmelstiel -Wilson disease E13.21

- type 1 E10.9
- - with
Revise - - - Kimmelsteil Kimmelstiel -Wilson disease E10.21

- type 2 E11.9
- - with
Revise - - - Kimmelsteil Kimmelstiel -Wilson disease E11.21

Disorder (of) -see also Disease
- bone M89.9
- - development and growth NEC M89.20
Revise - - - ilium M89.259 M89.28
Revise - - - ischium M89.259 M89.28
- eating (adult) (psychogenic) F50.9
Add - - specified NEC F50.89

Edema, edematous (infectious) (pitting) (toxic) R60.9
- lung J81.1
- - with heart condition or failure -see Failure, ventricular, left
Add - - newborn P29.0

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

Gastropathy K31.9
Add - specified NEC K31.89

Grief F43.21
Revise - prolonged F43.29 F43.81

Add Homocystinemia R79.83
Revise Homocystinemia, homocystinuria Homocystinuria E72.11

Revise Hyperosmolality - (see also, Diabetes, by type, with hyperosmolality) E87.0

Hypertrophy, hypertrophic
- bone M89.30
Revise - - ilium M89.359 M89.38
Revise - - ischium M89.359 M89.38
Add - - pubic ramus M89.38
Infection, infected, infective (opportunistic) B99.9
- bacterial NOS A49.9
  - as cause of disease classified elsewhere B96.89
Add - - - Cronobacter (sakazakii) B96.89
Add - - Cronobacter (sakazakii) B96.89
Add - - as cause of disease classified elsewhere B96.89
Add - - generalized A41.59

- Pseudomonas NEC A49.8
Add - - generalized A41.52
Add - Serratia NEC A49.8
Add - - as cause of disease classified elsewhere B96.89
Add - - generalized A41.53

Intolerance
- milk NEC K90.49
Revise ← orthostatic, chronic G90.A

Leukodystrophy E75.29
Add - metachromatic E75.25
Revise Lipodermatosclerosis (see also Insufficiency, venous) M79.3 -see Varix, leg, with, inflammation
Add - with
Add - - varicose veins -see Varix, leg, with, inflammation
Add - - - ulcerated -see Varix, leg, with, ulcer, with inflammation by site
Revise - ulcerated -see also Ulcer, by site -see Varix, leg, with, ulcer, with inflammation by site

Malfunction -see also Dysfunction
- catheter device NEC T85.618
  - infusion NEC T82.514
Revise - - cranial T85.640 (see also Complication(s), catheter, cranial infusion, mechanical) T85.690
Revise - - epidural T85.640 (see also Complication(s), catheter, cranial infusion, mechanical) T85.690
Revise - - intrathecal (see also Complication(s), catheter, cranial infusion, mechanical) T85.610 T85.690
Revise - - spinal (see also Complication(s), catheter, cranial infusion, mechanical) T85.610 T85.690
Revise - - subarachnoid (see also Complication(s), catheter, cranial infusion, mechanical) T85.610 T85.690
Revise - - subdural (see also Complication(s), catheter, cranial infusion, mechanical) T85.610 T85.690
Neuromyopathy G70.9
Revise - paraneoplastic (see also, Neoplasm, by site, if known) D49.9 [G13.0]

Neuropathy, neuropathic G62.9
Revise - paraneoplastic (sensorial) (Denny Brown) (see also, Neoplasm, by site, if known) D49.9 [G13.0]

Neutropenia, neutropenic (chronic) (genetic) (idiopathic) (immune) (infantile) (malignant) (pernicious) (splenic)
Add - specified NEC D70.8

Osteitis - see also Osteomyelitis
Revise - deformans (see also Paget’s disease, bone) M88.9
- - in (due to)
Revise - - - malignant neoplasm of bone (see also, Neoplasm, malignant, by site) C41.9 [M90.60]
Revise - - - neoplastic disease -(see also Neoplasm, by type and site) D49.9 [M90.60]
Revise - - - - carpus D49.9 D49.2 [M90.64-]
Revise - - - - clavicle D49.9 D49.2 [M90.61-]
Revise - - - - femur D49.9 D49.2 [M90.65-]
Revise - - - - fibula D49.9 D49.2 [M90.65-] [M90.66-]
Revise - - - - finger D49.9 D49.2 [M90.64-]
Revise - - - - humerus D49.9 D49.2 [M90.62-]
Revise - - - - ilium D49.9 D49.2 [M90.65-] [M90.68]
Revise - - - - ischium D49.9 D49.2 [M90.65-] [M90.68]
Revise - - - - metacarpus D49.9 D49.2 [M90.64-]
Revise - - - - metatarsus D49.9 D49.2 [M90.67-]
Revise - - - - multiple sites D49.9 D49.89 [M90.69]
Revise - - - - neck D49.9 D49.2 [M90.68]
Add - - - - pubic ramus D49.2 [M90.68]
Revise - - - - radius D49.9 D49.2 [M90.63-]
Revise - - - - rib D49.9 D49.2 [M90.68]
Revise - - - - scapula D49.9 D49.2 [M90.61-]
Revise - - - - skull D49.9 D49.2 [M90.68]
Revise - - - - tarsus D49.9 D49.2 [M90.67-]
Revise - - - - tibia D49.9 D49.2 [M90.66-]
Revise - - - - toe D49.9 D49.2 [M90.67-]
Revise - - - - ulna D49.9 D49.2 [M90.63-]
Revise - - - - vertebra D49.9 D49.2 [M90.68]

Osteoarthropathy (hypertrophic) M19.90
- specified type NEC M89.40
Revise - - ilium M89.459M89.48
Revise - - ischium M89.459M89.48
Add - - pubic ramus M89.48
Osteodystrophy Q78.9
Revise - ilium M89.559 M89.58
Revise - ischium M89.559 M89.58
Add - pubic ramus M89.58

Osteolysis M89.50
Revise - ilium M89.559 M89.58
Revise - ischium M89.559 M89.58
Add - pubic ramus M89.58

Osteomalacia M83.9
- vitamin-D-resistant in adults E83.31 [M90.8-]
Revise - - ilium E83.31 [M90.859] [M90.88]
Revise - - ischium E83.31 [M90.859] [M90.88]
Add - - pubic ramus E83.31 [M90.88]

Osteomyelitis (general) (infective) (localized) (neonatal) (purulent) (septic) (staphylococcal) (streptococcal)(suppurative) (with periostitis) M86.9 -
- chronic (or old) M86.60
- - with draining sinus M86.40
Revise - - - ilium M86.459 M86.48
Revise - - - ischium M86.459 M86.48
Add - - - pubic ramus M86.48
- - hematogenous NEC M86.50
Revise - - - ilium M86.559 M86.58
Revise - - - ischium M86.559 M86.58
Add - - - pubic ramus M86.58
- - - multifocal M86.30
Revise - - - - ilium M86.359 M86.38
Revise - - - - ischium M86.359 M86.38
Add - - - - pubic ramus M86.38

Osteonecrosis M87.9
- idiopathic aseptic M87.00
Add - - pubic ramus M87.050
- secondary NEC M87.30
- - due to
- - - drugs M87.10
Revise - - - - ilium M87.159 M87.150
Revise - - - - ischium M87.159 M87.150
Add - - - - pubic ramus M87.150
- hemoglobinopathy NEC D58.2 [M90.50]
  Revise  - ilium D58.2 [M90.55-] [M90.58]
  Revise  - ischium D58.2 [M90.55-] [M90.58]
  Add  - pubic ramus D58.2 [M90.58]

- trauma (previous) M87.20
  Revise  - ilium M87.25- M87.250
  Revise  - ischium M87.25- M87.250
  Add  - pubic ramus M87.250

- in
- caisson disease T70.3 [M90.50]
  Revise  - ilium T70.3 [M90.55-] [M90.58]
  Revise  - ischium T70.3 [M90.55-] [M90.58]
  Add  - pubic ramus T70.3 [M90.58]

Add  - pubic ramus M87.350

- specified type NEC M87.80
  Revise  - ilium M87.85- M87.850
  Revise  - ischium M87.85- M87.850
  Add  - pubic ramus M87.850

Osteopathy -see also Osteomyelitis, Osteonecrosis, Osteoporosis
- after poliomyelitis M89.60
  Revise  - ilium M89.659 M89.68
  Revise  - ischium M89.659 M89.68
  Add  - pubic ramus M89.68

Paget's disease
- bone M88.9
  Revise  - ilium M88.85- M88.88
  Revise  - ischium M88.85- M88.88
  Add  - pubic ramus M88.88

Revise  Pancolitis, ulcerative (chronic) (see also, Colitis) K51.00
  Add  - ulcerative (chronic) K51.00
  Revise  - with
  Revise  - complication K51.019
  Revise  - abscess K51.014
  Revise  - fistula K51.013
  Revise  - obstruction K51.012
  Revise  - rectal bleeding K51.011
  Revise  - specified complication NEC K51.018
Revise  Pellagra (alcoholic) (with polyneuropathy) E52
Add    - with
Add    - - polyneuropathy E52 [G63]

Rectocele
- female (without uterine prolapse) N81.6
- - with uterine prolapse N81.4
Add    - - - complete N81.3

Resistance, resistant (to)
Delete - complicating pregnancy O26.89
- insulin E88.81
Add    - - complicating pregnancy O26.89-

Schizophrenia, schizophrenic F20.9
- undifferentiated (type) F20.3
Revise - - chronic F20.9 F20.5

Sepsis (generalized) (unspecified organism) A41.9
Add    - Cronobacter A41.59
Add    - MRSA (Methicillin resistant Staphylococcus aureus) A41.02

Short, shortening, shortness
- stature (child) (hereditary) (idiopathic) NEC R62.52
- - due to
- - - genetic causes E34.329
Revise - - - - ACAN gene variant E34.828 E34.328
Revise - - - - aggrecan deficiency E34.828 E34.328
Revise - - - - NPR-2 gene variant E34.828 E34.328
Revise - - - - specified genetic cause NEC E34.828 E34.328

Syndrome -see also Disease
Revise - hyposmolality (see also, Diabetes, by type, with hyposmolarity) E87.0

Revise - Kimmelstiel Kimmelstiel -Wilson -see Diabetes, specified type, with Kimmelstiel Kimmelstiel -Wilson disease