APPENDIX 100

DIAGNOSES APPENDIX
CRITERIA FOR CLINICAL AND OTHER EVENTS

V. 1.3

NOVEMBER-14-2013
INTRODUCTION

The Appendix Merger Cross Network Working Group was formed in the summer of 2009 to review the current diagnoses appendices used by IMPAACT and ACTG with the purpose of blending the current appendices into one document. The Working Group divided into three subgroups to handle this task: Clinical, Network, and Coding.

The Clinical subgroup, comprised of clinicians from both IMPAACT and ACTG, reviewed the current criteria from each of the appendices, and revised as needed to accommodate standards of practice domestically and internationally. For AIDS-defining criteria, both the Centers for Disease Control (CDC) and World Health Organization (WHO) documentation were taken into consideration.

The Network subgroup, comprised of clinicians from all DAIDS-sponsored networks, reviewed the revisions submitted by the Clinical subgroup for a broader perspective.

The Coding group was charged with developing a coding assignment scheme that is unique to Appendix 100. The coding scheme includes a six (6) digit code where the first two digits signify the appendix section starting with the number eleven (11); the next three (3) digits are sequential numbers for each diagnosis starting with 001 within each section; the last digit signifies the level of evidence for each diagnosis. The levels of evidence are confirmed=1, probable=2, and no distinction between confirmed and probable=8.
**APPENDIX 100 - Diagnoses Appendix**

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# APPENDIX 100 - Diagnoses Appendix

## I. CARDIOVASCULAR DISEASES

### ANEURYSM, AORTIC

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<th>Status</th>
<th>Description</th>
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<tbody>
<tr>
<td>110011</td>
<td>CONFIRMED</td>
<td>Radiographic or surgical evidence of an aortic aneurysm</td>
</tr>
</tbody>
</table>

### ANEURYSM, specify location other than aorta

<table>
<thead>
<tr>
<th>Code</th>
<th>Status</th>
<th>Description</th>
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<tbody>
<tr>
<td>110021</td>
<td>CONFIRMED</td>
<td>Abnormal widening or ballooning of a portion of a blood vessel</td>
</tr>
</tbody>
</table>

### ANGINA PECTORIS

<table>
<thead>
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<th>Code</th>
<th>Status</th>
<th>Description</th>
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<tbody>
<tr>
<td>110031</td>
<td>CONFIRMED</td>
<td>Both of the following:</td>
</tr>
<tr>
<td></td>
<td></td>
<td>1. History of chest discomfort associated with exertion or excitement and alleviated with rest. May be described as pain but more frequently as heaviness, pressure, squeezing, or choking sensation. May radiate to the left shoulder, down the arm, back neck, or jaw. and</td>
</tr>
<tr>
<td></td>
<td></td>
<td>2. A report of at least one of the following:</td>
</tr>
<tr>
<td></td>
<td></td>
<td>a. Electrocardiograph consistent with acute ischemia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>or</td>
</tr>
<tr>
<td></td>
<td></td>
<td>b. Stress test findings consistent with ischemia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>or</td>
</tr>
<tr>
<td></td>
<td></td>
<td>c. Angiogram or other imaging test of the coronary arteries demonstrating significant occlusion(s), and other etiologies of the presenting signs and symptoms are unlikely</td>
</tr>
<tr>
<td>110032</td>
<td>PROBABLE</td>
<td>All of the following:</td>
</tr>
<tr>
<td></td>
<td></td>
<td>1. History of chest discomfort associated with exertion or excitement and alleviated with rest. May be described as pain but more frequently as heaviness, pressure, squeezing or choking sensation. May radiate to the left shoulder, down the arm, back neck, or jaw. and</td>
</tr>
<tr>
<td></td>
<td></td>
<td>2. Absence of ECG, stress test, angiogram, or other imaging test demonstrating ischemia or coronary artery disease and</td>
</tr>
<tr>
<td></td>
<td></td>
<td>3. Absence of another cause of pain</td>
</tr>
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</table>

### ANGINA PECTORIS, UNSTABLE

<table>
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</tr>
</thead>
<tbody>
<tr>
<td>110041</td>
<td>CONFIRMED</td>
<td>All of the following:</td>
</tr>
<tr>
<td></td>
<td></td>
<td>1. History of chest discomfort associated with exertion or excitement and alleviated with rest. May be described as pain but more frequently as heaviness, pressure, squeezing or choking sensation. May radiate to the left shoulder, down the arm, back neck, or jaw. Symptoms not controlled with directed therapy.</td>
</tr>
</tbody>
</table>
APPENDIX 100 - Diagnoses Appendix

2. Absence of myocardial infarction (see acute myocardial infarction)
and
3. Absence of myocarditis, pericarditis, or other explanation for chest pain
and
4. A report of at least one of the following:
   a. Electrocardiograph consistent with acute ischemia
or
   b. Stress test findings consistent with ischemia
or
   c. Angiogram or other imaging test of the coronary arteries demonstrating significant occlusion(s) and other etiologies of the presenting signs and symptoms are unlikely

ARRHYTHMIA, INSIGNIFICANT, specify the arrhythmia type

110051 CONFIRMED
Disorder of heart rate or rhythm that is not life-threatening and does not require medical intervention

ARRHYTHMIA, SIGNIFICANT, specify the arrhythmia type

110061 CONFIRMED
At least one of the following:
1. A cardiac arrhythmia present on an ECG or rhythm strip causing or with the potential to cause clinically significant hemodynamic consequences
or
2. Inducible or reproducible arrhythmia on electrophysiologic testing (EP) with or without implantation of an automated cardiac defibrillator

Specific types of rhythm disturbances of interest include but are not limited to:
   Atrial fibrillation, supraventricular tachyarrhythmia (SVT), Heart block (second – third degree only), Multifocal atrial tachycardia (MAT), Ventricular tachycardia, Ventricular fibrillation, Torsades de Pointe, and Tachy-brady syndrome.

CARDIOMYOPATHY

110071 CONFIRMED
At least one of the following:
1. Left or right ventricular diastolic/systolic dimensions greater than or equal to (≥) two standard deviations (SD) from the mean for body surface area
or
2. Abnormal fractional shortening index greater than or equal to (≥) standard deviations (SD) from the mean
or
3. Evidence of diastolic or systolic dysfunction on echocardiogram

CARDIOMYOPATHY, HIV-ASSOCIATED, SYMPTOMATIC
110081 CONFIRMED
All of the following:
1. Absence of ischemic heart disease, nutritional deficiency or other clear cause of cardiomyopathy such as drug-induced, alcohol-induced or evidence of another virus causing disease
   
   and

2. Systolic dysfunction confirmed by echocardiography, cardiac catheterization, or appropriate imaging modality

NOTE: It may not be possible to discern the etiology of cardiomyopathy.

CARDIOMYOPATHY, ISCHEMIC

110091 CONFIRMED
Evidence of diastolic or systolic dysfunction as defined above with proven coronary artery disease by angiography

CONGESTIVE HEART FAILURE

110101 CONFIRMED
Clinical signs and symptoms compatible with left- or right-sided heart failure (e.g., paroxysmal nocturnal dyspnea, rales of S3 on auscultation, jugular venous distention) without an alternative explanation with at least one of the following:
1. Hemodynamic measurements, radionucleotide ventriculography, echocardiogram, cardiac catheterization, or multiple gated acquisition scan showing a decreased ejection fraction of less than (<) 45% (percent)
   or
2. Echocardiogram, cardiac catheterization or other studies showing evidence of increased left atrial pressure or tight heart failure
   or
3. Elevated levels of Brain Natriuretic Peptide (BNP) or pro-BNP

110102 PROBABLE
Clinical signs and symptoms compatible with left- or right-sided heart failure (e.g., paroxysmal nocturnal dyspnea, rales of S3 on auscultation, jugular venous distention) without an alternative explanation with both of the following:
1. Chest x-ray or other imaging study showing evidence of congestive heart failure, including cardiac enlargement
   
   and

2. Documentation of treatment for congestive heart failure
CORONARY ARTERY DISEASE (CAD)

110111  CONFIRMED
At least one of the following:
1. A written report in the medical record documenting both of the following:
   a. Myocardial ischemia or coronary artery disease
   and
   b. Use of medications given to treat or prevent angina (e.g., nitrates, beta blockers, or
calcium channel blockers)

or
2. A procedure report, hospital discharge summary, or other medical record from the
hospitalization during which a procedure was performed for treatment of coronary artery
disease (such as coronary artery bypass graft, coronary artery stent implant, coronary
arterectomy, or any percutaneous coronary intervention)

or
3. A consultation NOTE from the participant’s cardiologist or clinical doctor documenting
the occurrence of the procedure

110112  PROBABLE
A written report in the medical record documenting both of the following:
1. Myocardial ischemia or coronary artery disease

2. No use of medications given to treat or prevent angina (e.g., nitrates, beta blockers, or
calcium channel blockers)

CORONARY REVASCULARIZATION

110121  CONFIRMED
At least one of the following:
1. A procedure report, hospital discharge summary, or other medical record from the
hospitalization during which a procedure was performed for treatment of coronary artery
disease (such as coronary artery bypass graft, coronary artery stent implant, coronary
arterectomy, and any percutaneous coronary intervention)

or
2. A consultation NOTE from the participant’s cardiologist documenting the occurrence of
the procedure

EMBOLISM, PULMONARY

110131  CONFIRMED
1. Both of the following:
   a. Symptoms compatible with pulmonary embolism, such as shortness of breath, chest
   pain, or hemoptysis
   and
b. Results consistent with a diagnosis of pulmonary embolism on pulmonary angiography, helical CT, ventilation-perfusion scan, or other comparable imaging studies

or
2. A diagnosis of pulmonary embolism on autopsy

110132 PROBABLE
All of the following:
1. Symptoms compatible with pulmonary embolism, such as shortness of breath, chest pain, or hemoptysis

and
2. Results consistent with a diagnosis of deep venous thrombosis on venography, ultrasound, or other comparable imaging studies

and
3. If a chest x-ray is performed, it does not suggest an alternative etiology for the above symptoms.

HYPERTENSION, CHRONIC, IN PREGNANCY

110141 CONFIRMED
At least one of the following:
1. Blood pressure persistently greater than or equal to (≥) 140/90 mm Hg that began prior to pregnancy or in the first 20 weeks of pregnancy

or
2. On anti-hypertension medication at the onset of pregnancy

NOTE: See also CHRONIC HYPERTENSION IN PREGNANCY in the Perinatal/Pregnancy section.

HYPERTENSION, PREGNANCY-INDUCED

110151 CONFIRMED
Both of the following:
1. Blood pressure persistently greater than or equal to (≥) 140/90 mm Hg without proteinuria

and
2. Onset is after the first 20 weeks gestation with no hypertension prior to pregnancy

NOTE: See also PREGNANCY-INDUCED HYPERTENSION in the Perinatal/Pregnancy section.

HYPERTENSION IN PERSONS LESS THAN (<) 18 YEARS OF AGE

110161 CONFIRMED
Average systolic or average diastolic blood pressure greater than (> the 95th percentile for age, gender, and height measured on at least three occasions
HYPERTENSION IN PERSONS GREATER THAN OR EQUAL TO (≥) 18 YEARS OF AGE

NOTE: The diagnosis of hypertension should be made by the study participant’s clinician and not diagnosed solely on the blood pressure measurements obtained during research visits unless the research visits are also considered the clinical visits.

110171 CONFIRMED
At least one of the following:
1. A clinical diagnosis of hypertension is based on the average diastolic blood pressure greater than (> 90 mm Hg or systolic blood pressure of greater than (> 140 mm Hg in an adult not taking antihypertensive medications and not acutely ill. Based on the average of two or more readings taken at each of two or more visits after the first elevated blood pressure was obtained.
   or
2. Antihypertensive treatment or a regimen of diet and exercise prior to starting antihypertensive medication recommended or initiated. This includes initial treatment with diuretics to control the hypertension.

HYPERTENSION, PULMONARY

110181 CONFIRMED
Right Heart Catheterization with mean pulmonary artery (Protocol Amendments) pressure greater than or equal to (≥) 25 mm Hg with wedge pressure less than or equal to (≤) 15 mm Hg.

110182 PROBABLE
At least one of the following:
1. Echocardiogram with estimated systolic PA pressure greater than (> 40 mm Hg
   or
2. Right ventricular dilatation

HYPERTROPHY, LEFT VENTRICULAR

110191 CONFIRMED
Echocardiographic or other equivalent imaging demonstrating enlargement of left ventricle muscle mass

110192 PROBABLE
A report of an electrocardiograph indicating findings consistent with diagnosis of left ventricular hypertrophy. For example, Cornell criteria require the sum of the amplitude of R wave in avL lead and S waved I V3 lead is greater than (> 28 in males or 20 in females.
HYPOTENSION

110201 CONFIRMED
Systolic blood pressure less than (<) 90 mm Hg in persons greater than (> or equal to) 18 years of age or lower than the 95th percentile for age, gender, and height in those less than or equal to (≤) 18 years of age.

MYOCARDIAL INFARCTION, ACUTE (SYMPTOMATIC)

110211 CONFIRMED
At least one of the following:
1. Rise or fall of cardiac biomarkers (e.g., troponin), with at least one value above 99th percentile of upper reference limit (URL) with at least one of the following:
   a. occurrence of a compatible clinical syndrome, including symptoms (such as chest pain) consistent with myocardial ischemia
   or
   b. ECG changes indicative of new ischemia (new ST-changes or new left bundle branch block (LBBB)) or development of pathological Q waves on the ECG
   or
   c. Imaging evidence of new loss of viable myocardium or new regional wall motion abnormality.
   or
2. Sudden unexpected cardiac death involving cardiac arrest before biomarkers are obtained or before a time when biomarkers appear, along with at least one of the following:
   a. New ST-changes or new LBBB
   or
   b. Evidence of fresh thrombus on coronary angiography or at autopsy
   or
3. In patients with percutaneous coronary interventions and normal baseline troponin, increases in troponin of three times the 99th percentile of URL PLUS at least one of the following:
   a. New pathological Q-waves or new LBBB
   or
   b. Angiographically documented new graft or native artery occlusion
   or
   c. Imaging evidence of new loss of viable myocardium

110212 PROBABLE
Occurrence of a compatible clinical syndrome, including symptoms (such as chest pain) consistent with myocardial infarction with at least one of the following:
1. Development of (1) evolving new Q waves or (2) evolving ST elevation, preferably based on at least two ECGs taken during the same hospital admission
   or
2. In patients with coronary artery bypass grafting and normal baseline troponin, increases in troponin of five times the 99th percentile of URL
MYOCARDIAL INFARCTION, SILENT (found at routine ECG or on hospital ECGs)

110221 CONFIRMED
Cardiology report of electrocardiograph indicating findings consistent with myocardial infarction. For example, new Q wave present in two (2) or more contiguous leads and with either duration greater than or equal to (≥) 40 msec (milliseconds) or amplitude greater than (> ¼) R wave.

MYOCARDITIS

110231 CONFIRMED
Clinical syndrome compatible with myocardial injury (e.g., chest pain) in the absence of myocardial infarction, ischemia, or trauma confirmed by electrocardiogram or appropriate imaging studies.

PERICARDITIS

110241 CONFIRMED
Inflammation of the pericardium causing severe substernal chest pain; proven by ECG or radiologic tests.

PERIPHERAL ARTERY DISEASE (PAD)

NOTE: Intermittent claudication is symptom of peripheral artery disease. Report only diagnoses of PAD using this diagnosis code. Symptoms in the absence of a clinician’s diagnosis should be reported on a sign and symptom form.

110251 CONFIRMED
Compatible clinical signs and symptoms (e.g., intermittent claudication, femoral bruit, decreased peripheral pulses, change in color or temperature of limb suggesting peripheral arterial disease) with at least one of the following:
1. Positive results on diagnostic imaging studies (e.g., doppler ultrasound, contrast arteriography, or MRI arteriography)
2. Ankle brachial pressure index less than (<) 0.90 in non-diabetics

110252 PROBABIL
Compatible clinical signs and symptoms (e.g., intermittent claudication, femoral bruit, decreased peripheral pulses, or change in color or temperature of limb suggesting peripheral arterial disease)
STROKE, HEMORRHAGIC

110261 CONFIRMED

1. Both of the following:
   a. Demonstrable lesion compatible with an acute hemorrhagic stroke on a CT or MRI by at least one of the following:
      1. Blood in subarachnoid space or intraparenchymal hemorrhage by CT scan. (Intraparenchymal blood must be dense and not mottled-mixed hyperdensity and hypodensity.)
      or
      2. Bloody spinal fluid by lumbar puncture. (Bloody CSF means greater than (>1) 100 cells/mm³ (cubic millimeter.) The LP is thought to be non-traumatic and counts in the last tube are similar to those in the first tube (no clearing) or xanthochromia when the specimen is spun down.)
      or
      3. Surgical evidence of hemorrhage as cause of clinical syndrome and
   b. Acute onset with a clinically compatible course, including unequivocal objective findings of a localizing neurologic deficit
      or
   2. Stroke diagnosed as cause of death at autopsy

110262 PROBABLE

All of the following:

1. Demonstrable lesion compatible with an acute hemorrhagic stroke on a CT or MRI by at least one of the following:
   a. Blood in subarachnoid space or intraparenchymal hemorrhage by CT scan. (Intraparenchymal blood must be dense and not mottled-mixed hyperdensity and hypodensity.)
   or
   b. Bloody spinal fluid by lumbar puncture. (Bloody CSF means greater than (>1) 100 cells/mm³ (cubic millimeter.) The LP is thought to be non-traumatic and counts in the last tube are similar to those in the first tube (no clearing) or xanthochromia when the specimen is spun down.)
   or
   c. Surgical evidence of hemorrhage as cause of clinical syndrome
   and
   2. At least one of the following:
      a. Positive lumbar puncture compatible with subarachnoid hemorrhage
      or
      b. Death certificate or death NOTE from medical record listing stroke as cause of death

NOTE: See also STROKE, HEMORRHAGIC in the Neurological Disorders section.
STROKE, ISCHEMIC INFARCTION

110271 CONFIRMED
   1. Both of the following:
      a. Demonstrable lesion compatible with an acute stroke with ischemic infarction on a CT or MRI by at least one of the following:
         1. Focal brain deficit without CT or LP evidence of blood, except mottled cerebral pattern. Either decreased density by CT in a compatible location or a negative CT or none done.
         or
         2. Surgical evidence of ischemic infarction
   and
      b. Acute onset with a clinically compatible course including unequivocal objective findings of a localizing neurologic deficit
   or
   2. Stroke diagnosed as cause of death at autopsy

110272 PROBABLE
   All of the following:
   1. Demonstrable lesion compatible with an acute stroke with ischemic infarction on a CT or MRI by at least one of the following:
      a. Focal brain deficit without CT or LP evidence of blood, except mottled cerebral pattern. Either decreased density by CT in a compatible location or a negative CT or none done.
      or
      b. Surgical evidence of ischemic infarction
   and
      2. At least one of the following:
         a. Positive lumbar puncture compatible with subarachnoid hemorrhage
         or
         b. Death certificate or death NOTE from medical record listing stroke as cause of death

NOTE: See also STROKE, ISCHEMIC INFARCTION in the Neurological Disorders section.

STROKE, UNKNOWN TYPE

110281 CONFIRMED
   1. Both of the following:
      a. Demonstrable lesion compatible with an acute stroke on a CT or MRI with inadequate information to categorize as hemorrhagic or ischemic infarction
         and
      b. Acute onset with a clinically compatible course, including unequivocal objective findings of a localizing neurologic deficit
         or
   2. Stroke diagnosed as cause of death at autopsy

NOTE: See also STROKE, UNKNOWN TYPE in the Neurological Disorders section.
THROMBOSIS, DEEP VEIN (DVT), SYMPTOMATIC

110291 CONFIRMED
Diagnosis of DVT by ultrasound, MRI, helical computerized tomography (CT), or other acceptable diagnostic method

110292 PROBABLE
1. All of the following:
   a. An elevated D-dimer test OR abnormal plethysmography
   and
   b. A score on the Wells Clinical Prediction Rule for DVT of greater than or equal to (≥) three points
   and
   c. No alternative diagnosis as likely as or greater than that of deep venous thrombosis

or

2. All of the following:
   a. Clinical presentation consistent with DVT (e.g., swelling, pain, and tenderness in extremity)
   and
   b. A score on the Wells Clinical Prediction Rule for DVT of greater than or equal to (≥) three points
   and
   c. No alternative diagnosis as likely as or greater than that of deep vein thrombosis

Wells Clinical Prediction Rule for DVT
One point for each of the following:
Active cancer (treatment ongoing or within previous six months, or palliative)
Paralysis, paresis, or plaster immobilization of lower extremities
Recently bedridden for more than three days, or major surgery, within 4 weeks
Localized tenderness along distribution of the deep venous system
Entire leg swollen
Calf swelling by more than three cm (centimeter) when compared with the asymptomatic leg
(measured 10cm (centimeter) below tibial tuberosity)
Pitting edema (greater in the symptomatic leg)
Collateral superficial veins (non-varicose)

NOTE: See also THROMBOSIS, DEEP VEIN (DVT), SYMPTOMATIC in the Hematologic Diseases section.
TRANSIENT ISCHEMIC ATTACK

110301  CONFIRMED  
Both of the following:
1. One or more episodes of focal neurologic deficit lasting more than 30 seconds and no longer than 24 hours with rapid evolution of the symptoms to the maximal deficit in less than (<) five minutes with complete resolution and no immediately preceding head trauma  
and  
2. There should be no evidence of clonic jerking, conjugate eye deviation, prolonged Jacksonian march, scintillating scotoma, or headache with nausea and vomiting

NOTE: Discovery of an infarct by CT in a location compatible with the symptoms, even if the symptoms cleared in less than (<) 24 hours, shall be diagnosed as a stroke.

VALVULAR HEART DISEASE

110311  CONFIRMED  
Abnormal opening or closing of a heart valve, specify valvular lesion

CARDIAC ABNORMALITY, OTHER, specify abnormality
When reporting other Cardiovascular System disease/disorders, use the following guidelines for confirmed and probable diagnoses:

119001  Confirmed diagnosis criteria may include either of the following:  
1. A compatible clinical syndrome plus any diagnostic evidence that supports the diagnosis  
or  
2. There may be the diagnostic test confirmation of the diagnosis without a clinical syndrome. Examples of diagnostic tests are: Microbiologic; cytologic, or histologic evidence that supports the diagnosis or any currently-approved diagnostic test.

119002  Probable diagnosis criteria may include one of the following:  
1. A compatible clinical syndrome  
or  
2. One or more test results from procedures generally accepted as supportive of the diagnosis in standard medical practice  
or  
3. Initiation or recommendation of specific therapy when appropriate

CARDIOMYOPATHY, OTHER (e.g., drug-induced, alcohol related, viral, unknown or other)

119011  CONFIRMED  
Systolic dysfunction confirmed by electrocardiography, cardiac catheterization, or appropriated imaging modality
II. CONGENITAL/BIRTH DEFECTS/GENETIC CONDITIONS

NOTE: All of the conditions listed in this section are considered “CONFIRMED” unless otherwise stated.

120011 ANOMALIES OF THE EAR, CONGENITAL, specify anomaly
120021 ANOMALIES OF THE EYE, CONGENITAL, specify anomaly
120031 ANOMALIES OF THE NOSE, CONGENITAL, specify anomaly

120041 CLEFT LIP
120051 CLEFT PALATE

If the study participant has both a cleft lip and a cleft palate, report each of these as a separate diagnosis.

CYSTIC FIBROSIS

120071 CONFIRMED
Systemic inherited disease of the exocrine glands that affects the lung and digestive systems. Confirmed by pilocarpine iontophoresis of sweat greater than or equal to (≥) 60 mEq/L (milliequivalents/liter).

120072 PROBABLE
All of the following:
1. Compatible syndrome
2. Pilocarpine iontophoresis of sweat test less than (<) 60 mEq/L (milliequivalents/liter)

NOTE: See also CYSTIC FIBROSIS in the Neonatal section.

120081 CUTANEOUS DEFECTS, specify (for example, skin dimples, brachial cleft and thyroglossal, or supernumery nipples)

120091 DIAPHRAGMATIC HERNIA, CONGENITAL, hemidiaphragm/absence of diaphragm

120101 DOWN SYNDROME, Trisomy 21

120121 FETAL ALCOHOL SYNDROME

120131 GASTROINTESTINAL, anatomical defect, specify

120141 GENITOURINARY, MALE, anatomical defect, specify

120151 GENITOURINARY, FEMALE, anatomical defect, specify
<table>
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<th>Code</th>
<th>Diagnosis</th>
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<tr>
<td>120171</td>
<td>GLYCOGEN STORAGE DISEASE, congenital, specify</td>
</tr>
<tr>
<td>120181</td>
<td>HEART DEFECTS, anatomical, specify</td>
</tr>
<tr>
<td>120191</td>
<td>INFANT OF DIABETIC MOTHER</td>
</tr>
<tr>
<td>120201</td>
<td>INBORN ERRORS OF METABOLISM</td>
</tr>
<tr>
<td>120211</td>
<td>MUSCULOSKELETAL ABNORMALITY, congenital, specify. Includes absence and duplication.</td>
</tr>
<tr>
<td>120221</td>
<td>NEURAL TUBE DEFECT, congenital, includes Spina Bifida, specify defect</td>
</tr>
<tr>
<td>120231</td>
<td>PIGMENT DISORDERS, congenital (e.g., albinism or café au lait spots), specify size and location</td>
</tr>
<tr>
<td>120241</td>
<td>PYLORIC STENOSIS</td>
</tr>
<tr>
<td>120261</td>
<td>TRISOMIES - TRISOMY, specify (excludes DOWN SYNDROME)</td>
</tr>
<tr>
<td>120271</td>
<td>TURNER SYNDROME</td>
</tr>
<tr>
<td>120281</td>
<td>VASCULAR LESIONS (e.g., port wine, nevi, and hemangiomas)</td>
</tr>
<tr>
<td>129001</td>
<td>CNS ANATOMICAL DEFECT, other, specify (excludes Neural tube)</td>
</tr>
<tr>
<td>129011</td>
<td>ENDOCRINE BIRTH DEFECT, other, specify</td>
</tr>
<tr>
<td>129021</td>
<td>GENITOURINARY DEFECT, other, specify (includes ambiguous genitalia)</td>
</tr>
<tr>
<td>129031</td>
<td>RESPIRATORY BIRTH DEFECT, other, specify</td>
</tr>
<tr>
<td>129041</td>
<td>OTHER BIRTH DEFECT, specify</td>
</tr>
</tbody>
</table>

Includes genetic or congenital conditions not already specified elsewhere, chromosomal abnormalities other than Down Syndrome, Trisomies, or Turner Syndrome
III. DERMATOLOGIC CONDITIONS

130018 ACNE

An inflammatory skin condition characterized by superficial skin eruptions typically occurring on the face, neck, or back

130028 ACRODERMATITIS

A skin condition characteristic to children that may be accompanied by mild symptoms of fever and malaise; may also be associated with hepatitis B infection, Epstein-Barr virus (EBV) infection, coxsackievirus A16, parainfluenza virus and other viral infections. Lesions are brownish-red or copper-colored papules that are flat topped and firm. The rash may appear as a linear string of papules with symmetrical distribution.

ALOPECIA

130031 CONFIRMED

Complete loss of hair within a defined area but may include the total body

ATOPIC DERMATITIS/ECZEMA, specify atopic dermatitis or eczema

130041 CONFIRMED

Dermatitis of unknown etiology characterized by itching and scratching in an individual with inherently irritable skin

EOSINOPHILIC PUSTULAR FOLLICULITIS

130051 CONFIRMED

Superficial inflammation of the hair follicles with eosinophils

130068 ERYTHEMA MULTIFORME

A moderate to severe skin disorder characterized by a rash which may be bullous or painful, resulting from an allergic reaction

ERYTHEMA TOXICUM NEONATORUM

130091 CONFIRMED

A benign condition in up to 50% (percent) of normal newborns characterized by a central whitish to yellowish white papule surrounded by reddened skin which may be present for a few hours to days. Eosinophils are present on histological examination of a skin scraping from affected areas.

130108 ERYTHRODERMA
A rash which may have target lesions, plaques, or other erythematous lesions

HEMANGIOMA MULTIPLE

130111 CONFIRMED
Vascular lesions of the skin, which are erythematous and may involve underlying tissue planes; specify size and locations

HEMANGIOMA SINGLE

130121 CONFIRMED
Vascular lesion of the skin, which is erythematous and may involve underlying tissue planes

HYPERSENSITIVITY REACTION- DRUG RELATED (E.G., ABACAVIR, NEVIRIPINE), (not RASH - DRUG RELATED; not STEVENS-JOHNSON SYNDROME)

130131 CONFIRMED
An acute reaction that follows the initiation of a new drug (e.g., abacavir) that is associated with a combination of any of the following:
1. Nonspecific rash

   or

2. Fever

   or

3. Abdominal pain

   or

4. Malaise

   or

5. Headache

Symptoms typically resolve when offending agent is discontinued.

ICHTHYOSIS

130141 CONFIRMED
Inherited skin disorder presenting with dry scaly skin most severe on extremities, classically may include fine palmar lines
<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Code</th>
<th>Status</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>LICE (PEDICULOSIS)</strong>, specify either lice or pediculosis</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>130151</td>
<td></td>
<td>CONFIRMED</td>
<td>At any site, proven by direct visualization of ectoparasites or by microscopy</td>
</tr>
<tr>
<td>130152</td>
<td></td>
<td>PROBABLE</td>
<td>Suspected lice at any site responding to specific pediculosis therapy</td>
</tr>
<tr>
<td><strong>LICHER NITIDUS</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>130161</td>
<td></td>
<td>CONFIRMED</td>
<td>Skin disorder characterized by chronic itching and tiny flesh colored to pink raised persistent papules generally occurring on abdomen, flexar surface of palms, and genitalia</td>
</tr>
<tr>
<td><strong>LICHEN PLANUS</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>130171</td>
<td></td>
<td>CONFIRMED</td>
<td>Recurring disorder of skin and mucous membranes resulting in inflammation, itching, and distinctive skin lesions; typically of whitish coloration</td>
</tr>
<tr>
<td><strong>PITYRIASIS ALBA</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>130181</td>
<td></td>
<td>CONFIRMED</td>
<td>A common skin disorder similar to mild eczema with round or oval colorless finely scaled patches of skin; most often presenting on the cheeks</td>
</tr>
<tr>
<td><strong>PITYRIASIS ROSEA</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>130191</td>
<td></td>
<td>CONFIRMED</td>
<td>Classic skin rash with herald rash; often in typical “Christmas tree” pattern</td>
</tr>
<tr>
<td><strong>PSORIASIS</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>130201</td>
<td></td>
<td>CONFIRMED</td>
<td>Common inflammatory skin condition characterized by frequent episodes of redness, itching, and thick dry silvery scales on the skin</td>
</tr>
</tbody>
</table>
### RASH-DRUG RELATED (not HYPERSENSITIVITY REACTION; not STEVENS-JOHNSON SYNDROME)

<table>
<thead>
<tr>
<th>Code</th>
<th>Status</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>130212</td>
<td>PROBABLE</td>
<td>This is a clinical diagnosis without definitive criteria; other etiologies must be considered unlikely (e.g., viral infections, syphilis, or contact dermatitis). Macular or papular lesions, often pruritic, which may result in post-inflammatory hyper- or hypopigmentation in the affected areas.</td>
</tr>
</tbody>
</table>

### STEVENS-JOHNSON SYNDROME (not Rash-Drug Related; not Hypersensitivity Reaction)

<table>
<thead>
<tr>
<th>Code</th>
<th>Status</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>130221</td>
<td>CONFIRMED</td>
<td>Severe adverse drug reaction resulting in involvement of skin and mucous membranes; desquamation may occur. Other organ systems may also be involved; fever is common, potentially fatal.</td>
</tr>
</tbody>
</table>

### SCABIES

<table>
<thead>
<tr>
<th>Code</th>
<th>Status</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>130231</td>
<td>CONFIRMED</td>
<td>Superficial infection causing a pruritic rash typically occurring in skin folds and exacerbated by heat or hot water; ectoparasites documented by microscopy</td>
</tr>
<tr>
<td>130232</td>
<td>PROBABLE</td>
<td>Superficial infection causing a pruritic rash typically occurring in skin folds and exacerbated by heat or hot water; no organisms seen or formal examination not performed; however, there is a positive response to treatment</td>
</tr>
</tbody>
</table>

### SEBORRHEIC DERMATITIS

<table>
<thead>
<tr>
<th>Code</th>
<th>Status</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>130241</td>
<td>CONFIRMED</td>
<td>A papulosquamous disorder patterned on the sebum-rich areas of the scalp, face, and trunk. It is characterized by loose, greasy or dry, white to yellowish scales, with or without associated reddened skin. The severity varies from mild dandruff to exfoliative erythroderma.</td>
</tr>
</tbody>
</table>

### VITILIGO

<table>
<thead>
<tr>
<th>Code</th>
<th>Status</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>130251</td>
<td>CONFIRMED</td>
<td>Loss of the pigment of the skin in a defined area; may be idiopathic or secondary to post-inflammatory condition.</td>
</tr>
</tbody>
</table>
WARTS, COMMON FLAT (thought to be due to non-oncogenic subtypes of human papilloma virus)

130261 CONFIRMED
Characteristic lesions which may occur anywhere but commonly occur on hands and feet ("plantar warts"). Biopsy of lesion positive by DNA hybridization or nucleic acid amplification test, or histopathology suggestive of human papilloma infection of the skin.

130262 PROBABLE
Typically made as a clinical diagnosis. May occur anywhere, but commonly occur on hands and feet ("plantar warts").

139008 OTHER SKIN DISEASE/DISORDER/RASH, specify diagnosis
Non-specific skin conditions not listed in this appendix; may be associated with or caused by another medical disorder
IV. GASTROINTESTINAL DISORDERS

APPENDICITIS

140011 CONFIRMED
Clinical diagnosis confirmed by surgical or histological findings

CHOLANGITIS/CHOLECYSTITIS, BACTERIAL, specify either cholangitis or cholecystitis

140021 CONFIRMED
Both of the following:
1. Ascending infection of the biliary tree with biliary colic, jaundice, spiking fevers with chills, and partial obstruction to the flow of bile
   and
2. Positive blood culture or biliary culture

140022 PROBABLE
Both of the following:
1. Ascending infection of the biliary tree with biliary colic, jaundice, spiking fevers with chills, and partial obstruction to the flow of bile
   and
2. Biliary or blood cultures negative or not done

CHOLELITHIASIS

140031 CONFIRMED
Presence of stones in the gall bladder by diagnostic testing including any of the following: x-ray, cholecystiogram, gall bladder ultrasound, or radioisotope scan

GASTROESOPHAGEAL REFLUX

140041 CONFIRMED
Regurgitation of gastric contents into the esophagus, possibly causing inflammation determined by endoscopic, esophageal pH monitoring, or radiographic tests

140042 PROBABLE
Both of the following:
1. Regurgitation of gastric contents into the esophagus, possibly causing inflammation
   and
2. Confirmatory tests not done
INTUSSUSCEPION

140051 CONFIRMED
Invagination of one segment of intestine into a segment of distal intestine as demonstrated by one or more of the following:
1. Findings at surgery
   or
2. Radiologic findings, as documented, by either air or liquid contrast enema; or by demonstration of an abdominal mass by abdominal ultrasound with specific characteristic features (target or doughnut sign on transverse scan AND a pseudo-kidney or sandwich sign on longitudinal scan) that is CONFIRMED to be reduced by hydrostatic enema on a post-reduction (repeat) ultrasound
   or
3. Findings at autopsy

140052 PROBABLE
Intussusception suspected but not CONFIRMED by surgery, radiology, or autopsy

PANCREATITIS, CLINICAL/SYMPTOMATIC

140061 CONFIRMED
All of the following:
1. Clinical or symptomatic pancreatitis is defined by the symptoms of nausea, vomiting, or abdominal pain of any duration
   and
2. Associated with greater than or equal to (≥) Grade 3 elevations of lipase (greater than (>)) 3.0 times upper limit of normal (ULN))
   and
3. Without other non-pancreatic diagnoses to reasonably account for the presentation with or without radiographic evaluation.

140062 PROBABLE
Both of the following:
Clinical diagnosis only; testing technology not available to determine diagnosis
1. Clinical symptoms greater than or equal to (≥) Grade 3
   and
2. No radiographic evaluation available
PANCREATITIS, CHEMICAL/ASYMPTOMATIC

140071 **CONFIRMED**
Chemical or asymptomatic pancreatitis is defined as persistent (twodeterminations, two weeks apart) elevations in lipase greater than or equal to (≥) Grade 3

140072 **PROBABLE**
Clinical diagnosis only; testing technology not available to determine diagnosis. **Both of the following:**
1. Clinical symptoms greater than or equal to (≥) Grade 3
   and
2. No radiographic evaluation available

PEPTIC ULCER DISEASE - either gastric or duodenal

140081 **CONFIRMED**
**Both of the following:**
1. Inflammation or ulceration of the lining of the stomach by radiographic or endoscopic means
   and
2. Etiology by H. pylori urease test on biopsy specimen or by urease breath test or histologic test

140082 **PROBABLE**
**Both of the following:**
1. Clinical syndrome consistent with inflammation of the lining of the stomach, confirmatory
   and
2. Tests not diagnostic or not done
GASTROINTESTINAL DISORDERS, OTHER

GASTRIC DISORDER, OTHER, not listed in the Appendix, specify disorder
When reporting other gastric disorders, use the following guidelines for confirmed and probable diagnoses:

149001 Confirmed diagnosis criteria may include one of the following:
1. A compatible clinical syndrome plus any diagnostic evidence that supports the diagnosis
   or
2. There may be the diagnostic test confirmation of the diagnosis without a clinical syndrome. Examples of diagnostic tests are: Microbiologic; cytologic, or histologic evidence that supports the diagnosis or any currently-approved diagnostic test.

149002 Probable diagnosis criteria may include one of the following:
1. A compatible clinical syndrome
   or
2. One or more test results from procedures generally accepted as supportive of the diagnosis in standard medical practice
   or
3. Initiation or recommendation of specific therapy when appropriate
V. GENITOURINARY/SEXUALLY TRANSMITTED DISEASES

BALANITIS, specify organism if identified

150011 CONFIRMED
Clinical diagnosis of inflammation of the glans of the penis

CHANCROID, specify site

150021 CONFIRMED PATHOGEN
All of the following:
1. Painful genital ulcer with tender, suppurative inguinal adenopathy
2. Exclusion of other genital ulcer infections (for example, syphilis, herpes simplex, or lymphogranuloma venereum)
3. Culture or Gram stain of genital ulcer or lymph node aspirate consistent with Haemophilus ducreyi organisms

150022 NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Painful genital ulcer with tender, suppurative inguinal adenopathy
2. Exclusion of other genital ulcer infections (for example, syphilis, herpes simplex, or lymphogranuloma venereum)
3. Cultures not done or not diagnostic

CHLAMYDIA TRACHOMATIS, specify urethritis or cervicitis; see LGV also

150031 CONFIRMED PATHOGEN
All of the following:
1. Clinical syndrome of urethritis, vaginitis, or cervicitis made by an experienced practitioner
2. Exclusion of gonorrheal disease as cause
3. At least one of the following:
   a. Positive culture or nucleic acid amplification test on urethral, vaginal, or cervical swabs as appropriate
   or
   b. Positive urine nucleic acid amplification test
150032  NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Clinical syndrome of urethritis, vaginitis, or cervicitis made by an experienced practitioner
   and
2. Exclusion of gonorrheal disease as cause
   and
3. Cultures or nucleic acid amplification tests negative or not done

DYSMENORRHEA, PRIMARY

150042  NO PATHOGEN CONFIRMED (PROBABLE)
Painful menstrual cramps, no underlying abnormality; usually appears within two years of menarche

DYSMENORRHEA, SECONDARY

150052  NO PATHOGEN CONFIRMED (PROBABLE)
Painful menstrual cramps, due to underlying pathology; usual onset beyond two years of menarche

AMENORRHEA, PRIMARY

150062  NO PATHOGEN CONFIRMED (PROBABLE)
No onset of menses beyond the age at which menarche normally occurs based on age, Tanner staging, and age of maternal menarche

AMENORRHEA, SECONDARY OR OLIGOMENORRHEA, specify either secondary amenorrhea or oligomenorrhea

150072  NO PATHOGEN CONFIRMED (PROBABLE)
Cessation of established menses for greater than (> ) three months

LYMPHOGRANULOMA VENEREUM

150081  CONFIRMED PATHOGEN
All of the following:
1. Tender, suppurative inguinal or femoral adenopathy, typically unilateral
   and
2. Exclusion of other genital ulcer infections (syphilis, herpes simplex, or chancroid); can also cause hemorrhagic proctocolitis among men and women who engage in anal intercourse
   and
3. Positive culture, immunofluorescence, or nucleic acid amplification tests for Chlamydia trachomatis from lymph node aspirate
150082  NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Tender, suppurative inguinal or femoral adenopathy; typically unilateral
   and
2. Exclusion of other genital ulcer infections (syphilis, herpes simplex, or chancroid); can also cause hemorrhagic proctocolitis among men and women who engage in anal intercourse
   and
3. Specific tests negative or not done

NEISSERIA GONORRHEA, specify body site

150091  CONFIRMED PATHOGEN
All of the following:
1. Clinical syndrome of urethritis, vaginitis, salpingitis, or cervicitis made by an experienced practitioner. Rectal and pharyngeal disease may be asymptomatic. Disseminated gonococcal disease may involve the skin and joints, and rarely cause endocarditis or meningitis.
   and
2. At least one of the following:
   a. Positive culture or nucleic acid amplification test on urethral, vaginal, or cervical swabs as appropriate
   or
   b. Positive urine nucleic acid amplification test
   or
   c. Positive cultures from rectal or pharyngeal swabs, or from joint fluid or blood

150092  NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Clinical syndrome of urethritis, vaginitis, salpingitis, or cervicitis made by an experienced practitioner. Rectal and pharyngeal disease may be asymptomatic. Disseminated gonococcal disease may involve the skin and joints, and rarely cause endocarditis or meningitis.
   and
2. Cultures or amplification tests negative or not done

ORCHITIS, specify etiology

150102  PROBABLE
Inflammation of the testes, clinical diagnosis

PELVIC INFLAMMATORY DISEASE (PID) (includes SALPINGITIS, TUBO-OVARIAN ABSCESS), specify which form
150111 CONFIRMED PATHOGEN
All of the following:
1. Clinical diagnosis made by an experienced practitioner
   and
2. Positive culture for specific organism (e.g., Chlamydia or Neisseria gonorrhoeae) from material obtained by laparoscopy or cul de sac aspiration, or cervical/vaginal diagnostic test in a woman with appropriate symptoms

150112 NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Clinical diagnosis made by an experienced practitioner
   and
2. Negative cultures in a woman with appropriate symptoms

NOTE: See also PELVIC INFLAMMATORY DISEASE in Infectious Disease section.

SYphilis, congenital

150121 CONFIRMED PATHOGEN
Diagnosis defined by having at least one of the following:
1. Demonstration of Treponema pallidum by dark field microscopy or specific fluorescent antibody stains from lesions of a newborn infant or from placenta, umbilical cord, or fetal autopsy
   or
2. Newborn infant with a nontreponemal test titer four-fold greater than (>) the mother’s titer using the same test preferably in the same laboratory
   or
3. Newborn infant who has a reactive treponemal test for syphilis and at least one of the following:
   a. Abnormal physical examination (including, but not limited to, hepatosplenomegaly, rash, or snuffles)
   or
   b. Abnormal long bone radiographs with characteristic epiphyseal and metaphyseal changes
   or
   c. Positive CSF VDRL
   or
   d. Abnormal CSF cell count or protein in the absence of other explanations
   or
   e. Reactive IgM EIA or IgM 19S-FTA-ABS test
   or
4. A child greater than (>2) years of age with a reactive nontreponemal test for syphilis and stigmata of untreated congenital syphilis (e.g., interstitial keratitis, eighth cranial nerve deafness, anterior bowing of shins, frontal bossing, mulberry molars, peg-shaped notched central incisors (Hutchinson teeth), saddle nose, rhagades, or symmetric, painless knee swelling (Clutton joints)
Diagnosis defined by having at least one of the following:
1. A stillbirth at greater than (> 20 weeks gestation or greater than (> 500 grams fetal weight to a woman with untreated or inadequately treated syphilis, i.e., any non-penicillin regimen during pregnancy or a penicillin regimen administered less than (< 30 days before delivery

or
2. A newborn infant born to a mother with untreated or inadequately treated syphilis, i.e., any non-penicillin regimen during pregnancy or a penicillin regimen administered less than (< 30 days before delivery; serologic tests negative or not done, but penicillin therapy initiated

NOTE: See also SYPHILIS, CONGENITAL in the Neonatal section.

SYPHILIS, POSTNATAL, includes primary, secondary, & latent syphilis; specify which one

Clinically compatible case (e.g., may include, depending upon stage, chancre, mucocutaneous lesions, rash, or asymptomatic) with at least one of the following:
1. Demonstration of Treponema pallidum by darkfield microscopy or specific fluorescent antibody stains from lesions

or
2. Both reactive nontreponemal serologic test (VDRL or RPR) and reactive treponemal test (FTA-ABS, MHA-TP, or EIA)

or
3. Past history of syphilis and new four-fold increase in nontreponemal serologic tests (VDRL or RPR)

or
4. Positive treponemal tests (FTA-ABS, MHA-TP, or EIA) and evidence of end organ dysfunction (e.g., gumma, aortic aneurysm)

or
5. Neurosyphilis - See MENINGITIS, SYPHILIS

All of the following:
1. Clinically compatible case (e.g., may include, depending upon stage, chancre, mucocutaneous lesions, rash, or asymptomatic)

and
2. Negative serologic tests or tests not done

and
3. Penicillin therapy initiated
TRICHOMONIASIS

150141  CONFIRMED PATHOGEN
All of the following:
1. Clinical syndrome of vaginal discharge, sometimes malodorous, with vulvovaginitis and itching, although may be asymptomatic in many women. Microbiologic cause is Trichomonas vaginalis.

and
2. Positive microscopic examination of wet-mount preparation of discharge (culture and immunodiagnostic rapid tests are only sometimes available)

150142  NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Clinical diagnosis as above, made by an experienced practitioner

and
2. Wet-mount preparation microscopy negative or not done

150158  TRICHOMONAS VAGINALIS

URETHRITIS

150161  CONFIRMED PATHOGEN, specify pathogen
All of the following:
1. Clinical diagnosis of mucopurulent urethral discharge or pyuria

and
2. No urinary tract infection

and
3. Positive cultures or nucleic acid amplification tests for Neisseria gonorrhoeae or Chlamydia trachomatis

150162  NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Clinical diagnosis of mucopurulent urethral discharge, or pyuria in the absence of urinary tract infection

and
2. Cultures or nucleic acid amplification tests for Neisseria gonorrhoeae or Chlamydia trachomatis negative or not done

150171  WARTS (EXCLUDING ORAL), specify site

NOTE: See also PAPILLOMATOSIS in the Infectious Disease Section.
REPRODUCTIVE SYSTEM DISEASE/DISORDER, OTHER, specify gender (male or female) and diagnosis

When reporting other reproductive system disease/disorders, use the following guidelines for confirmed and probable diagnoses:

159001 Confirmed diagnosis criteria may include at least one of the following:
1. A compatible clinical syndrome plus any diagnostic evidence that supports the diagnosis
   or
2. There may be the diagnostic test confirmation of the diagnosis without a clinical syndrome. Examples of diagnostic tests are: Microbiologic; cytologic, or histologic evidence that supports the diagnosis or any currently-approved diagnostic test.

159002 Probable diagnosis criteria may include at least one of the following:
1. A compatible clinical syndrome
   or
2. One or more test results from procedures generally accepted as supportive of the diagnosis in standard medical practice
   or
3. Initiation or recommendation of specific therapy when appropriate

VENEREAL DISEASE, OTHER, specify diagnosis

NOTE: When reporting other venereal disease, use the following guidelines for confirmed and probable diagnoses:

159011 Confirmed diagnosis criteria may include at least one of the following:
1. A compatible clinical syndrome plus any diagnostic evidence that supports the diagnosis
   or
2. There may be the diagnostic test confirmation of the diagnosis without a clinical syndrome. Examples of diagnostic tests are: Microbiologic; cytologic, or histologic evidence that supports the diagnosis or any currently-approved diagnostic test.

159012 Probable diagnosis criteria may include at least one of the following:
1. A compatible clinical syndrome
   or
2. One or more test results from procedures generally accepted as supportive of the diagnosis in standard medical practice
   or
3. Initiation or recommendation of specific therapy when appropriate
VI. HEMATOLOGIC DISEASES

ANEMIA, APLASTIC

160011 CONFIRMED
Diminished or absence of all hematopoietic precursors in the bone marrow

ANEMIA, PURE RED CELL APLASIA

160021 CONFIRMED
Failure of the bone marrow to make red blood cells with maturation arrest of red blood cell precursors, as demonstrated by bone marrow biopsy

ANEMIA, AUTOIMMUNE HEMOLYTIC
NOTE: Excludes HEMOLYTIC ANEMIA OF THE NEWBORN

160031 CONFIRMED
Hemolytic anemia as a result of autoantibodies as identified by both of the following:
1. Positive direct Coombs test
2. Evidence of hemolysis

ANEMIA, G6PD DEFICIENCY, HEMOLYTIC

160041 CONFIRMED
Hemolytic anemia that is often precipitated by medications, usually in association with low G6PD enzyme levels in plasma. Common drugs initiating hemolysis include dapsone, primaquine, quinidine, quinine, sulfonamides, and nitrofurantoin.

ANEMIA, IRON DEFICIENCY

160051 CONFIRMED
All of the following:
1. Microcytic anemia
2. At least one of the following:
   a. Low or absent iron stores as indicated by low serum iron concentration, high serum transferrin, and high transferrin binding capacity (low percent saturation of transferrin)
   b. Low serum ferritin

160052 PROBABLE
Microcytic anemia without testing of iron stores and no other explanation for the anemia
ANEMIA, MEGALOBLASTIC, resulting from vitamin B12 or folate deficiency; specify either “vitamin B12” or “folate deficiency” in description

160061 CONFIRMED
Macrocystosis with reduced serum B12 or folate levels

160062 PROBABLE
Macrocystosis with hypersegmented polys, no further testing done

COAGULATION DISORDER, DISSEMINATED INTRAVASCULAR (DIC)

160071 CONFIRMED
Evidence of bleeding with low platelets and microangiopathic hemolytic anemia in the appropriate clinical setting (including, but not limited to, sepsis or trauma) in association with findings of thrombin generation (as indicated by reduced plasma fibrinogen levels) and increased fibrinolysis (as indicated by elevated fibrin degradation products or D-dimer levels in plasma)

COAGULATION DISORDERS, HEREDITARY, HEMOPHILIA, FACTOR VIII DEFICIENCY

160081 CONFIRMED
Hereditary inability to clot blood confirmed by Factor VIII analysis

COAGULATION DISORDERS, HEREDITARY, HEMOPHILIA, FACTOR IX DEFICIENCY

160091 CONFIRMED
Hereditary inability to clot blood confirmed by Factor IX analysis

COAGULATION DISORDERS, HEREDITARY, VON WILLEBRAND DISEASE

160101 CONFIRMED
Inherited disorder of clotting confirmed by laboratory testing

ABO HEMOLYTIC DISEASE OF THE NEWBORN

160111 CONFIRMED
Mediated by maternal antibody as identified by a positive direct Coombs test

NOTE: See also ABO HEMOLYTIC DISEASE OF THE NEWBORN in the Neonatal section.
RH HEMOLYTIC DISEASE OF THE NEWBORN

160121 CONFIRMED
Mediated by maternal antibody as identified by a positive direct Coombs test

NOTE: See also RH HEMOLYTIC DISEASE OF THE NEWBORN in the Neonatal section.

HEMOGLOBINOPATHY, ALPHA (α) THALASSEMIA

160131 CONFIRMED
Inherited hemoglobinopathy leading to anemia. Severe Alpha thalassemia is confirmed by electrophoresis.

HEMOGLOBINOPATHY, BETA (β) THALASSEMIA

160141 CONFIRMED
Inherited hemoglobinopathy leading to anemia and extramedullary hematopoiesis. Severe Beta thalassemia is confirmed by electrophoresis.

HEMOGLOBINOPATHY, SICKLE CELL DISEASE

160151 CONFIRMED
Inherited hemoglobinopathy leading to sickle shaped red blood cells confirmed by hemoglobin electrophoresis

HEMOGLOBINOPATHY, SICKLE CELL TRAIT

160161 CONFIRMED
Single gene inherited condition of red blood cells confirmed by hemoglobin electrophoresis

IMMUNE (changed from IDIOPATHIC) THROMBOCYTOPENIC PURPURA, specify primary or secondary
PRIMARY IS WITHOUT ANY DEFINABLE etiology
SECONDARY IS WITH DEFINABLE etiology

160171 CONFIRMED
All of the following:
1. Isolated low platelet count secondary to accelerated platelet destruction by antiplatelet antibodies
   
   and

2. The diagnosis is based on history, physical examination, blood count, and blood film and bone marrow biopsy
   
   and

3. Other probable causes of thrombocytopenia must be excluded (e.g. infiltrative diseases of the bone marrow, disseminated intravascular coagulation)
THROMBOPHLEBITIS, OVARIAN VEIN

160181 CONFIRMED
Clinical diagnosis, documented by imaging studies such as ultrasound, x-ray, or MRI

THROMBOPHLEBITIS, SEPTIC PELVIC

160191 CONFIRMED
Clinical diagnosis, documented by imaging studies such as ultrasound, x-ray, or MRI and positive blood cultures

THROMBOSIS, DEEP VEIN, ASYMPTOMATIC

160201 CONFIRMED
DVT found incidentally by diagnostic imaging in a patient without symptoms of DVT

THROMBOSIS, DEEP VEIN (DVT), SYMPTOMATIC

110291 CONFIRMED
Diagnosis of DVT by ultrasound, MRI, helical computerized tomography (CT), or other acceptable diagnostic method

110292 PROBABLE
1. All of the following:
   a. An elevated D-dimer test OR abnormal plethysmography and
   b. A score on the Wells Clinical Prediction Rule for DVT of greater than or equal to (≥) three points and
   c. No alternative diagnosis as likely as or greater than that of deep venous thrombosis or
2. All of the following:
   a. Clinical presentation consistent with DVT (e.g., swelling, pain, and tenderness in extremity) and
   b. A score on the Wells Clinical Prediction Rule for DVT of ≥ three points and
   c. No alternative diagnosis as likely as or greater than that of deep vein thrombosis
Wells Clinical Prediction Rule for DVT

One point for each of the following:
- Active cancer (treatment ongoing or within previous six months, or palliative)
- Paralysis, paresis, or plaster immobilization of lower extremities
- Recently bedridden for more than three days, or major surgery, within 4 weeks
- Localized tenderness along distribution of the deep venous system
- Entire leg swollen
- Calf swelling by more than three cm (centimeter) when compared with the asymptomatic leg (measured 10cm (centimeter) below tibial tuberosity)
- Pitting edema (greater in the symptomatic leg)
- Collateral superficial veins (non-varicose)


NOTE: See also THROMBOSIS, DEEP VEIN (DVT), SYMPTOMATIC in the Cardiovascular section.

169008 ANEMIA, OTHER with or without hemolysis, specify cause if known

HEMOGLOBINOPATHY, OTHER, not otherwise listed in this document, specify.

169011 CONFIRMED
Confirmed by electrophoresis

HEMATOLOGIC DISEASE, OTHER

169021 CONFIRMED
Such as thrombotic thrombocytopenia purpura (TTP), hereditary spherocytosis, or hereditary elliptocytosis
### VII. HEPATOBILIARY DISORDERS

#### CIRRHOSIS

<table>
<thead>
<tr>
<th>Code</th>
<th>Status</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>170011</td>
<td>CONFIRMED</td>
<td>Biopsy of liver consistent with hepatic cirrhosis</td>
</tr>
<tr>
<td>170012</td>
<td>PROBABLE</td>
<td>No biopsy performed but extrahepatic manifestations suggestive of cirrhosis (spider angioma, splenomegaly, esophageal varices, palmar erythema, or thrombocytopenia, etc.)</td>
</tr>
</tbody>
</table>

#### HEPATITIS A, ACUTE

<table>
<thead>
<tr>
<th>Code</th>
<th>Status</th>
<th>Pathogen</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>170021</td>
<td>CONFIRMED</td>
<td>PATHOGEN</td>
<td>Hepatic inflammation diagnosed within six months of having a known normal (asymptomatic) liver or chronic stable hepatitis</td>
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<tr>
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<td></td>
<td>Hepatic inflammation is defined by at least one of the following:</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>1. Aminotransferase elevation of at least five times the upper limit of normal (ULN)</td>
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<td></td>
<td>or</td>
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<td></td>
<td></td>
<td></td>
<td>2. Seropositive for IgM antibody to HAV (Acute Hepatitis A)</td>
</tr>
</tbody>
</table>

**NOTE:** See also HEPATITIS in the Infectious Disease section.

#### HEPATITIS B, ACUTE

<table>
<thead>
<tr>
<th>Code</th>
<th>Status</th>
<th>Pathogen</th>
<th>Description</th>
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</thead>
<tbody>
<tr>
<td>170031</td>
<td>CONFIRMED</td>
<td>PATHOGEN</td>
<td>Hepatic inflammation diagnosed within six months of having a known normal (asymptomatic) liver or chronic stable hepatitis</td>
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<tr>
<td></td>
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<td></td>
<td>Hepatic inflammation is defined by at least one of the following:</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>1. Aminotransferase elevation of at least five times the upper limit of normal (ULN)</td>
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<td></td>
<td>or</td>
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<td></td>
<td></td>
<td></td>
<td>2. Seropositive for IgM antibody to HBc with presence of new HBsAg (acute hepatitis B)</td>
</tr>
</tbody>
</table>

**NOTE:** See also HEPATITIS in the Infectious Disease Section

#### HEPATITIS C, ACUTE

<table>
<thead>
<tr>
<th>Code</th>
<th>Status</th>
<th>Pathogen</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>170041</td>
<td>CONFIRMED</td>
<td>PATHOGEN</td>
<td>Hepatic inflammation diagnosed within six months of having a known normal (asymptomatic) liver or chronic stable hepatitis</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>Hepatic inflammation is defined by at least one of the following:</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>1. Aminotransferase elevation of at least five times ULN</td>
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<td>or</td>
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<td></td>
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<td></td>
<td>2. Evidence of HCV viremia without HCV antibody</td>
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<td>or</td>
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<td>3. Documented seroconversion within six months to anti-HCV with an aminotransferase elevation above the normal level (Acute Hepatitis C)</td>
</tr>
</tbody>
</table>

**NOTE:** See also HEPATITIS in the Infectious Disease Section
HEPATITIS, ACUTE

170052 NO PATHOGEN CONFIRMED (PROBABLE)
Hepatic inflammation diagnosed within six months of having a known normal (asymptomatic) liver or chronic stable hepatitis
Hepatic inflammation is defined by **at least one of the following:**
1. Aminotransferase elevation of at least five times ULN
**or**
2. Other causes of hepatitis have been ruled out if possible where none of the following are identified:
   a. Seropositive for IgM antibody to HAV (Acute Hepatitis A)
   **or**
   b. IgM antibody to HBc with presence of new HBsAg (Acute Hepatitis B)
   **or**
   c. Evidence of HCV viremia without HCV antibody or documented seroconversion within six months to anti-HCV with an aminotransferase elevation above the normal level (Acute Hepatitis C)
   **or**
   d. Elevated antibody to Hepatitis E in association with travel to or residence in an endemic area (Acute Hepatitis E)

**NOTE:** See also HEPATITIS in the Infectious Disease Section

170068 HEPATITIS, DRUG-INDUCED, specify drug

170078 HEPATITIS, ETIOLOGY UNKNOWN

HEPATITIS, NEONATAL

170082 NO PATHOGEN CONFIRMED (PROBABLE)
Characterized by elevated transaminases 1.5 times ULN with or without clinical findings such as jaundice, hepatomegaly, and hepatic failure

**NOTE:** See also HEPATITIS, NEONATAL in the Neonatal Disorders section.

HEPATIC STEATOSIS (FATTY LIVER), specify either hepatic steatosis or fatty liver

170091 CONFIRMED
Fatty infiltration **NOTEd** on liver biopsy

170092 PROBABLE
Enlarged with radiographic imaging (CT, MRI) suggestive of fatty infiltration
GASTRO-HEPATOBILIARY DISEASES, OTHER, specify diagnosis
When reporting other gastro-hepatobiliary diseases, use the following guidelines for confirmed and probable diagnoses:

179001 Confirmed diagnosis criteria may include one of the following:
   1. A compatible clinical syndrome plus any diagnostic evidence that supports the diagnosis
   or
   2. There may be the diagnostic test confirmation of the diagnosis without a clinical syndrome. Examples of diagnostic tests are: Microbiologic; cytologic, or histologic evidence that supports the diagnosis or any currently-approved diagnostic test.

179002 Probable diagnosis criteria may include one of the following:
   1. A compatible clinical syndrome
   or
   2. One or more test results from procedures generally accepted as supportive of the diagnosis in standard medical practice
   or
   3. Initiation or recommendation of specific therapy when appropriate

GASTROINTESTINAL DISEASE, OTHER, specify diagnosis
When reporting other gastrointestinal disease, use the following guidelines for confirmed and probable diagnoses:

179011 Confirmed diagnosis criteria may include one of the following:
   1. A compatible clinical syndrome plus any diagnostic evidence that supports the diagnosis
   or
   2. There may be the diagnostic test confirmation of the diagnosis without a clinical syndrome. Examples of diagnostic tests are: Microbiologic; cytologic, or histologic evidence that supports the diagnosis or any currently-approved diagnostic test.

179012 Probable diagnosis criteria may include one of the following:
   1. A compatible clinical syndrome
   or
   2. One or more test results from procedures generally accepted as supportive of the diagnosis in standard medical practice
   or
   3. Initiation or recommendation of specific therapy when appropriate
HEPATIC DISORDER, OTHER, specify disorder
When reporting other hepatic disorders, use the following guidelines for confirmed and probable diagnoses:

179021 Confirmed diagnosis criteria may include one of the following:
   1. A compatible clinical syndrome plus any diagnostic evidence that supports the diagnosis
      or
   2. There may be the diagnostic test confirmation of the diagnosis without a clinical
      syndrome. Examples of diagnostic tests are: Microbiologic; cytologic, or histologic
      evidence that supports the diagnosis or any currently-approved diagnostic test.

179022 Probable diagnosis criteria may include one of the following:
   1. A compatible clinical syndrome
      or
   2. One or more test results from procedures generally accepted as supportive of the diagnosis
      in standard medical practice
      or
   3. Initiation or recommendation of specific therapy when appropriate

LIVER DISEASE, OTHER, specify type

179031 CONFIRMED
   At least one of the following:
   1. Development of abnormal liver enzymes (ALT or AST) or alkaline phosphatase
      or
   2. Elevation in bilirubin in a study participant with previously normal tests
      or
   3. Further increases (to grade greater than (>) 3) in a study participant with chronic abnormal
      levels
VIII. IMMUNE RECONSTITUTION INFLAMMATORY SYNDROME (IRIS)

NOTE: The case definition portion of this section is in the format from the ACTG Definition of Immune Reconstitution Inflammatory Syndrome (IRIS) document, which is available on the ACTG website.

180018 IMMUNE RECONSTITUTION INFLAMMATORY SYNDROME (IRIS)

All of the following:
1. Initiation, reintroduction, or change in antiretroviral therapy/regimen or therapy for opportunistic infections (OI)
and
2. Evidence of at least one of the following:
   a. Increase in CD4+ cell count as defined by greater than or equal to (≥) 50 cells/mm³ (cubic millimeter) or a greater than or equal to (≥) two-fold rise in CD4+ cell count
   or
   b. Decrease in the HIV-1 viral load of greater than (> ) 0.5 log10
   or
   c. Weight gain or other investigator-defined signs of clinical improvement in response to initiation, reintroduction, or change of either antiretroviral therapy/regimen or opportunistic infection (OI) therapy
and
3. Symptoms or signs consistent with an infectious or inflammatory condition
and
4. These symptoms or signs cannot be explained by a newly acquired infection, the expected clinical course of a previously recognized infectious agent, or the side effects of medications.
and
5. For purposes of data collection, the infectious/inflammatory condition must be attributable to a specific pathogen or condition.

If the study participant is being evaluated for an inflammatory condition at a time that is less than (<) 4 weeks after initiation, reintroduction, or change in antiretroviral therapy/regimen or OI therapy, items 2a through 2c above are not required.

Refer to the “Criteria for the Diagnosis of Specific Immune Reconstitution Inflammatory Syndromes (IRIS)” section in the ACTG Definition of Immune Reconstitution Inflammatory Syndrome (IRIS) document for specific Opportunistic Infection (OI) and non-pathogen condition diagnosis criteria. Document is available on the ACTG Network website (www.actgnetwork.org).

SPECIFIC IRIS CASE DEFINITIONS

NOTE: All references listed in brackets for the following section can be found in the ACTG Definition of Immune Reconstitution Inflammatory Syndrome (IRIS) document.
1. Cytomegalovirus [CMV] [Ophthalmologic only]

180021 Confirmed CMV IRIS in patients with a prior history of CMV retinitis:
Previous CMV retinitis diagnosis by ACTG criteria and improvement of signs/symptoms with anti-CMV therapy, with the subsequent development of ocular inflammatory changes in uveovitreal tract, lens, or retina, with or without associated visual changes, as documented by an experienced ophthalmologist.

180022 Probable CMV IRIS in patients with a prior history of CMV retinitis:
Previous CMV retinitis diagnosis by ACTG criteria and improvement of signs/symptoms with anti-CMV therapy, with the subsequent development of ocular inflammatory changes in uveovitreal tract, lens or retina, with or without associated visual changes, as documented by a non-ophthalmologist clinician.

180031 Confirmed CMV IRIS in patients without a prior history of CMV retinitis:
The development of significant ocular inflammation in the uveovitreal tract, lens, or retina attributed to CMV in the absence of ophthalmologic findings typical for acute CMV retinitis, with or without visual changes, as documented by an experienced ophthalmologist.

2. Cryptococcus neoformans

180041 Confirmed Cryptococcal IRIS in patients with a prior history of cryptococcosis:
Cryptococcal meningitis or other diagnosis of systemic cryptococcal infection [fungemia, pneumonia] by ACTG criteria and improvement of signs/symptoms with antifungal therapy, with the subsequent development of new or worsening pulmonary infiltrates, new meningeal enhancement on scan or abnormal CSF findings (low glucose, elevated WBC, or CSF CRAG with negative fungal culture), mediastinal or cervical lymphadenopathy, pleural effusion, cutaneous abscesses or cryptococcal lesions at other sites; histopathology from non-meningeal site demonstrating inflammatory changes and organisms in the absence of a positive culture or positive culture.

180042 Probable Cryptococcal IRIS in patients with a prior diagnosis of cryptococcosis:
Previous cryptococcosis diagnosis by ACTG criteria and improvement of signs/symptoms with anti-cryptococcal therapy, and the subsequent development of focal inflammatory site(s); histopathology from involved site or sterile body fluid analysis demonstrating inflammatory changes (e.g., granulomas or lymphocytes) in the absence of positive stains or cultures for any other pathogen or any non-infectious histopathologic diagnosis; and negative blood cultures for Cryptococcosis; and negative CSF CRAG if obtained or development of new onset CNS signs or symptoms with meningeal enhancement or other atypical radiographic changes with no evidence of other neurologic disease to explain the findings.
180051 Confirmed Cryptococcal IRIS in patients without a prior history of cryptococcosis: The development of meningitis with meningeal enhancement on scan with abnormal CSF findings (low glucose, elevated WBC, or positive CSF CRAG with negative or positive fungal culture), mediastinal or cervical lymphadenopathy, pleural effusion, cutaneous abscesses or cryptococcal lesions at other sites; histopathology from non-meningeal site demonstrating inflammatory changes and organisms in the absence of a positive culture or positive culture.

180052 Probable Cryptococcal IRIS in patients without a prior diagnosis of cryptococcosis: The subsequent development of focal inflammatory site(s); histopathology from involved site or sterile body fluid analysis demonstrating inflammatory changes (e.g., granulomas or lymphocytes) accompanied by evidence consistent with cryptococcosis in the absence of positive cultures for any other pathogen.

3. Mycobacterium avium complex [MAC]

180061 Confirmed MAC IRIS in patients with a prior history of disseminated MAC (dMAC): Previous disseminated MAC diagnosis by ACTG criteria and improvement of signs/symptoms with anti-MAC therapy, with the subsequent development of focal inflammatory site(s).

180062 Probable MAC IRIS in patients with a prior history of dMAC: Previous dMAC diagnosis by ACTG criteria and improvement of signs/symptoms with anti-MAC therapy, and the subsequent development of focal inflammatory site(s); histopathology from involved site demonstrating inflammatory changes (e.g., granulomas) in the absence of positive stains or cultures for any other pathogen or any non-infectious histopathologic diagnosis; and negative blood cultures for MAC.

180071 Confirmed MAC IRIS in patients without a prior history of dMAC: The development of focal inflammatory site(s); histopathology from the involved site demonstrating inflammatory changes (e.g., granulomas) accompanied by histologic or culture evidence of AFB consistent with MAC in the absence of positive cultures for any other AFB; and may have positive blood cultures for MAC.

180072 Probable MAC IRIS in patients without a prior diagnosis of MAC: The subsequent development of focal inflammatory site(s); histopathology from involved site demonstrating inflammatory changes (e.g., granulomas or lymphocytic infiltrates) and without evidence of any other specific pathogen (stains may be positive for AFB); and negative blood cultures for MAC.
4. Mycobacterium tuberculosis (TB)

180081  **Confirmed TB IRIS in patients with a prior history of TB (paradoxical TB-associated IRIS):**

There are three components to this case-definition [12]:

A) **Antecedent requirements**

i) Diagnosis of tuberculosis: previous pulmonary (smear positive or smear-negative) or extrapulmonary TB diagnosis by ACTG criteria

AND

ii) Initial response with anti-TB therapy (e.g., stabilization or improvement of signs/symptoms with appropriate anti-TB therapy prior to initiation of ART)*. For example, there has been cessation or improvement of fevers, cough, night sweats.

*NOTE: This does not apply to patients starting ART within two weeks of starting tuberculosis treatment since insufficient time may have elapsed for a clinical response to be reported.

(B) **Clinical criteria**

The onset of tuberculosis-associated IRIS manifestations should be within three months of ART initiation, reinitiation, or regimen change because of HIV treatment failure.

Of the following, at least one major criterion or two minor clinical criteria are required:

**Major criteria**

- New or enlarging lymph nodes, cold abscesses, or other focal tissue involvement, e.g., tuberculous arthritis
- New or worsening radiological features of tuberculosis (found by chest compatible radiography, abdominal ultrasonography, CT, or MRI)
- New or worsening CNS tuberculosis (meningitis or focal neurological deficit; e.g., caused by tuberculoma)
- New or worsening serositis (pleural effusion, ascites, or pericardial effusion)

**Minor criteria**

- New or worsening constitutional symptoms such as fever, night sweats, or weight loss
- New or worsening respiratory symptoms such as cough, dyspnea, or stridor
- New or worsening abdominal pain accompanied by peritonitis, hepatomegaly, splenomegaly, or abdominal adenopathy

(C) **Alternative explanations for clinical deterioration must be excluded**

- Failure of tuberculosis treatment because of tuberculosis drug resistance
- Poor adherence to tuberculosis treatment
- Another opportunistic infection or neoplasm (it is particularly important to exclude an alternative diagnosis in patients with smear-negative pulmonary tuberculosis and extrapulmonary tuberculosis where the initial tuberculosis diagnosis has not been microbiologically confirmed)
- Drug toxicity or reaction
APPENDIX 100 - Diagnoses Appendix

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180082 Probable TB IRIS in patients with a prior history of TB:
“Probable” status should be assigned for cases where criteria A and B are met (see confirmed TB IRIS with a prior history of TB definition) but an alternative diagnosis or explanation for clinical deterioration cannot be fully excluded.

180091 Confirmed TB IRIS in patients without a prior history of TB (ART “unmasking” TB-associated IRIS):
Patient is not receiving treatment for TB when ART is initiated. Active TB is diagnosed after initiation of ART and the diagnosis of TB fulfills ACTG criteria for smear-positive pulmonary TB, smear-negative pulmonary TB or extrapulmonary TB. Active TB develops within three months of starting ART and one of the following criteria is met: heightened intensity of clinical manifestations, particularly if there is evidence of a marked inflammatory component. For example, presentations may include TB lymphadenitis or TB abscesses with prominent acute inflammatory features; the development of pulmonary or extrapulmonary TB with no evidence of miliary disease accompanied by marked focal inflammation; or histopathology from involved site demonstrating inflammatory changes (e.g., granulomas, caseation) accompanied by histologic or culture evidence of AFB consistent with TB in the absence of positive cultures for any other AFB.

180092 Probable TB IRIS in patients without a prior diagnosis of TB:
Patient is not receiving treatment for TB when ART is initiated. Active TB is diagnosed after initiation of ART and the diagnosis of TB fulfills ACTG criteria for smear-positive pulmonary TB, smear-negative pulmonary TB, or extrapulmonary TB. There is heightened intensity of clinical manifestations but there is not clear evidence of a marked inflammatory component to the presentation or the subsequent development of focal inflammatory site(s) is beyond three months of ART initiation.

5. Progressive Multifocal Leukoencephalopathy (PML)

180101 Confirmed PML IRIS in patients with a prior history of PML:
Previous PML diagnosis by ACTG criteria (consistent CT or MRI scan with positive biopsy or positive CSF JC nucleic acid amplification test and the absence of any alternative diagnosis), with the subsequent development of new or worsening CT or MRI findings with contrast enhancement; brain biopsy shows focal inflammatory changes accompanied by histologic or nucleic acid amplification test evidence of PML in the absence of another diagnosis.

180102 Probable PML IRIS in patients with a prior history of PML:
Previous PML diagnosis by ACTG criteria (consistent CT or MRI scan with positive biopsy or CSF JC nucleic acid amplification test and the absence of any alternative diagnosis), with the subsequent development of new or worsening CT or MRI findings with contrast in the absence of another diagnosis.

180111 Confirmed PML IRIS in patients without a prior diagnosis of PML:
The development of new neurologic deficit(s) in patient with no previously recognized CNS infection or malignancy, accompanied by CT or MRI changes showing contrast enhancement; brain biopsy shows focal inflammatory changes accompanied by histologic or nucleic acid amplification test evidence of PML in the absence of another diagnosis.
Probable PML IRIS in patients without a prior diagnosis of PML:
The development of new neurologic deficit(s) in a patient with no previously recognized CNS infection or malignancy accompanied by CT or MRI changes showing contrast enhancement consistent with PML with no brain biopsy obtained. CSF findings cannot be attributable to another pathogen or disease process.

IRIS DIAGNOSES WITHOUT SPECIFIC CASE DEFINITIONS

PATHOGEN-DIRECTED:

Kaposi’s Sarcoma (KS)
Rapid KS progression [15] and local swelling with adjacent lymphadenopathy [20] have both been reported as a manifestation of immune reconstitution after antiretroviral therapy/regimen initiation. Whereas the latter syndrome may truly represent an IRIS syndrome, if there is no clear evidence of any characteristic inflammatory component [1, 16], then the progression could be due to a failure to reconstitute KSHV-specific immune responses. KS-IRIS is defined as a sudden or more dramatic progression of disease than expected as part of natural history that occurs within 12 weeks of initiation of ART. ART alone should be continued for four to six weeks to monitor for clinical response. If the lesions stabilize, have a regression of inflammation or lesion number or size diminishes, this will be defined as IRIS. Should disease progression continue, then this is unlikely to be IRIS and should be defined as progression of disease and managed with appropriate KSHV specific therapies. The presence of inflammation on biopsy may assist in distinguishing progression of disease vs. IRIS.

Hepatitis B and Hepatitis C
Significant increase in ALT over baseline (flares) has been documented after beginning antiretroviral therapy/regimen with HBV co-infection as well as after interruption of antiretroviral therapy/regimen (especially in patients with HBV on a 3TC- or tenofovir-containing regimen). After immune recovery HBV "flares" may occur if HBV is not being concomitantly treated, although there are other potential reasons to consider. These include:

a. Spontaneous HBeAg seroconversion
b. Treatment-induced exacerbation of the underlying disease
c. Hepatotoxic effects of treatment
d. Withdrawal of active HBV drug
e. Development of resistance and return of replication, or
f. Superimposed and unrelated acute liver disease (e.g., Hepatitis A)

Liver biopsy may be helpful in determining evidence of drug related (eosinophilic infiltrate) vs. acute viral hepatitis (hepatocyte swelling with lobular inflammation). Biopsy may also grade and stage chronic viral hepatitis. IRIS is less well-defined in HCV co-infection, and increases in liver enzymes on antiretroviral therapy/regimen may be multifactorial as well.
180158 **Herpes simplex virus (HSV)**
The development of unusual presentations of HSV following the initiation of antiretroviral therapy/regimen attributed to immune reconstitution has rarely been described. Localized HSV vesicles may occur within eight weeks of initiating ART among persons with no prior history and no new source contact to attribute it to. Symptoms and signs are frequently consistent with a primary infection. Chronic erosive or ulcerative lesions of the genitals have been described in individuals who had prior histories of genital HSV. The appearance and clinical course of the lesions attributed to immune reconstitution appeared inconsistent with these patients’ previous HSV presentations. Proctitis has also been reported [1,18,19]. Routine viral cultures may or may not yield HSV though histochemistry studies may show evidence of HSV antigens in the absence of a positive viral culture. Histopathology in some cases may demonstrate an inflammatory infiltrate with unusual prominence of plasma cells and eosinophils. Response to antivirals appears to be variable.

180168 **Pneumocystis jirovecii pneumonia (PCP)**
Despite being the most common OI with a relatively high CD4 threshold for development, few clear cases of Pneumocystis pneumonia IRIS have been documented so far (likely because steroids have been established for the use of severe PCP). Cases have been described as worsening of pneumonia and even respiratory failure, although the patients reported received suboptimal courses of steroids [20]. To entertain a diagnosis of PCP IRIS, bronchoscopy should rule out an intercurrent pulmonary process.

180178 **Syphilis**
New reactive RPR within 12 weeks of starting ART in setting of known previously treated syphilis and documented nonreactive within past two years without new attributable source. May also present serologically as less than or equal to (≤) a fourfold change in titer in someone with previous history of treated syphilis and no new attributable source. May present with systemic symptoms including arthralgia. May improve with anti-inflammatories.

180188 **Toxoplasmosis**
There is also a very small database for possible toxoplasmosis IRIS. No specific clinical pattern has been seen, and there is no clear evidence of an inflammatory component.

180198 **Varicella Zoster (VZV)**
The development of herpes zoster after initiation of antiretroviral therapy/regimen has been attributed to immune reconstitution. The incidence of zoster appears to be two- to five-fold greater in those receiving antiretroviral therapy/regimen compared to those not receiving antiretroviral therapy/regimen. Most cases occur in the first 16 weeks following initiation of antiretroviral therapy/regimen. Those with higher percentage of CD8+ lymphocytes at the time of initiation of HAART and at one month following antiretroviral therapy/regimen appeared to be at higher risk for zoster [21]. Most cases present as cutaneous dermatomal disease or mucocutaneous disease, are mild, occur without systemic symptoms and respond to antiviral therapy. Iritis and keratitis have been described rarely [1,22]. It is not clear that cutaneous zoster following initiation of antiretroviral therapy/regimen has a significant inflammatory component that differentiates this from routine VZV.
### 189008 Other viral dermatoses
Eruptive onset of new common warts, flat warts, or epidermodysplasia verruciformis-type warts, or inflammation/rapid growth of previously stable cutaneous or genital warts, have been noted during immune restoration [23-24]. In addition, eruptive onset of new molluscum contagiosum or inflammation/enlargement of pre-existing mollusca during immune restoration has been described [25].

### NON-PATHOGEN OR UNKNOWN PATHOGEN DIRECTED

#### 180208 Autoimmune disorders
Systemic lupus erythematosus, polymyositis, rheumatoid arthritis, relapsing polychondritis and Guillain-Barre have also been attributed to immune reconstitution following administration of antiretroviral therapy/regimen [1,26].

#### 180218 Follicular inflammatory eruptions
Sudden onset of follicular papulopustular inflammatory eruptions resembling acne vulgaris or acne rosacea have been reported within first 4 months of immune reconstitution [27]. Eosinophilic folliculitis, distinguished from acneiform eruptions by intense pruritus, an urticarial appearance to the papules, and histopathology showing follicular inflammation containing eosinophils, has been documented. An increased incidence of eosinophilic folliculitis has been **NOTE**d in the first six months of HAART therapy [28].

#### 180228 Graves’ Disease
New onset of clinically significant Graves’ disease (hyperthyroidism) has been reported following the initiation of antiretroviral therapy/regimen. The development of anti-thyrotropin receptor antibodies in individuals following antiretroviral therapy/regimen, not present before antiretroviral therapy/regimen initiation, has been described [1, 26].

#### 180238 Sarcoidosis
Worsening of previously-diagnosed sarcoidosis or newly-diagnosed sarcoidosis following antiretroviral therapy/regimen has been reported. Pulmonary involvement as well as extrapulmonary involvement (erythema nodosum) has been described [1, 26].
IX. INFECTIOUS DISEASES (NON-MYCOBACTERIAL)

ABDOMINAL WOUND INFECTION, Caesarean

190011 CONFIRMED PATHOGEN, specify pathogen
   All of the following:
   1. Oral temperature greater than or equal to ($\geq$) 100.4° F or 38° C in the absence of other
      source of fever
      and
   2. Pus draining/drained from wound or wound dehiscence requiring debridement
      and
   3. Positive test for specific organism

190012 NO PATHOGEN CONFIRMED (PROBABLE)
   Both of the following:
   1. At least one of the following:
      a. Erythema, edema, and tenderness
      or
      b. Health care provider diagnosis
      and
   2. No microbiologic diagnosis

ABSCESS, DENTAL

190021 CONFIRMED PATHOGEN, specify pathogen
   Both of the following:
   1. Clinical diagnosis made by an experienced practitioner
   and
   2. Positive culture for specific organism

190022 NO PATHOGEN CONFIRMED (PROBABLE)
   Both of the following:
   1. Clinical diagnosis made by an experienced practitioner
   and
   2. Negative culture or culture not done

ARTHRITE, SEPTIC ( see also Septic Arthritis)

190031 CONFIRMED PATHOGEN, specify pathogen
   Both of the following:
   1. Clinical diagnosis
   and
   2. At least one of the following:
      a. Positive Gram stain or culture of joint fluid
      or
      b. Both of the following:
         1. Characteristic inflammatory joint fluid
         and
         2. Positive blood culture
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190032  NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Clinical diagnosis  
2. Inflammatory joint fluid  
3. Negative Gram stain or culture of joint fluid and blood  
4. Specific antibacterial therapy initiated

**BACTERIAL INFECTION OF DEEP TISSUE, BODY CAVITY OR OTHER NORMALLY-STERILE SITE,** includes abscess, osteomyelitis, pyomyositis, etc.; specify site (WHO-3, if Recurrent WHO-4)

This category includes, for example, organ parenchymal, deep soft tissue (including pyomyositis) or abdominal abscesses, purulent pericarditis, and bone and joint infections.

190041  CONFIRMED PATHOGEN, specify pathogen
Demonstration of bacterial pathogen(s) in deep tissue, viscera, body cavity, or other normally-sterile site by isolation of a bacterial pathogen(s) from an aspirate or biopsy specimen

190042  NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. At least one of the following:  
   a. Evidence of an infection in a deep tissue, body cavity or other normally-sterile site demonstrated by appropriate diagnostic sampling (positive histopathology of tissue)  
   or  
   b. Demonstration of bacterial pathogen(s) by appropriate microbiological stain of aspirate or biopsy specimen  
   or  
   c. Imaging procedures documenting infection (computerized tomography, ultrasonography, or magnetic resonance imaging, radioisotope scanning, or plain radiograph)  
   and  
2. Clinical signs and symptoms compatible with the infection  
   and  
3. Appropriate treatment initiated and response demonstrated (appropriate treatment may include drainage procedures or antibacterial therapy)

190058  BACILLARY ANGIOMATOSIS, also known as CAT SCRATCH DISEASE or PELIOSIS HEPATITIS)
Appropriate clinical diagnosis confirmed by positive culture, nucleic acid amplification test, serologic or histologic tests for Bartonella henselae or related Bartonella spp.
BACTERIAL CATHETER EXIT SITE OR TUNNEL INFECTION, specify either catheter exit site or tunnel infection and specify site

190061 CONFIRMED PATHOGEN, specify pathogen
Both of the following:
1. Erythema, tenderness, induration, or purulent drainage along the subcutaneous tract or at the skin exit site
   and
2. At least one of the following:
   a. Isolation of a bacterial pathogen(s) from the exit site, tunnel, or catheter tip
   or
   b. Appropriate antibacterial therapy is initiated or recommended

190062 NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Clinical diagnosis with erythema, tenderness, induration, or purulent drainage along the subcutaneous tract or at the skin exit site
   and
2. Cultures negative or not done
   and
3. Appropriate antibacterial therapy initiated or recommended

DISSEMINATED BLASTOMYCOSIS

190071 CONFIRMED PATHOGEN, specify site
Evidence of B. dermatitidis by positive culture, positive antigen testing, or positive histopathology identifying characteristic appearance of organisms within body tissue or fluids

BRONCHIECTASIS

190082 NO PATHOGEN CONFIRMED (PROBABLE)
Focal or diffusely abnormal and chronically dilated bronchi as visualized on CT, MRI, or chest x-ray
BRONCHIOLITIS

Clinical syndrome of wheezing, cough, and hypoxia typically seen in children less than (<) two years of age, most often caused by inflammation of the bronchioles and lower respiratory tract, due to infection with respiratory syncytial virus (RSV) or, on occasion, parainfluenza virus, influenza virus, or adenovirus

190091 CONFIRMED-pathogen, specify pathogen:
Both of the following:
1. Clinical diagnosis made by experienced practitioner
   and
2. Culture or rapid diagnostic test positive for RSV or other virus

190092 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Clinical diagnosis made by experienced practitioner
   and
2. Microbiologic tests negative or not done

BRONCHITIS

190101 CONFIRMED PATHOGEND, specify pathogen:
Both of the following:
1. Clinical diagnosis of cough, often loose or with sputum production, with variable ronchi, wheezes, or crackles on lung auscultation, associated with inflammation of the bronchi
   and
2. Specific pathogen identified from respiratory tract viral or bacterial cultures or nucleic acid amplification tests or serum antibody tests

190102 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Clinical diagnosis as above
   and
2. No pathogen recovered or testing not done

CANDIDIASIS OF BRONCHI, TRACHEA OR LUNGS, specify site - bronchi, trachea, or lungs; WHO-4

190111 CONFIRMED PATHOGEN
Both of the following:
1. Characteristic white plaques in the bronchi or trachea on bronchoscopic examination
   and
2. Positive culture, KOH, or histopathology from the bronchi or trachea
190112 NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Characteristic white plaques in the bronchi or trachea on bronchoscopic examination and
2. Response to specific antifungal therapy and
3. No evidence of positive culture, KOH, or histopathology

CANDIDIASIS, ORAL/OROPHARYNGEAL, PERSISTENT OR RECURRENT, specify either oral or oropharyngeal AND pseudomembranous or erythematous

CONFIRMED PATHOGEN (WHO-3)
Both of the following:
1. One of the following case definitions:
190121 Pseudomembranous candidiasis
Clinical Descriptors: White or yellow/creamy spots or plaques that may be located in any part of the oral cavity and can usually be wiped off to reveal an erythematous surface
Patient reported symptoms: None or possible mild to moderate burning pain
Patient reported duration: Lesions/symptoms are usually intermittent, but may be long-standing

Or

190131 Erythematous candidiasis
Clinical Descriptors: Patchy erythema or red areas usually located on the palate and dorsum of the tongue, but occasionally on the buccal mucosa. At times, white spots or plaques of pseudomembranous candidiasis may also be present.
Patient reported symptoms: None or possible mild to moderate burning pain
Patient reported duration: Lesions/symptoms are usually intermittent, but may be long-standing

and
2. Positive culture, KOH, or histopathology

NO PATHOGEN CONFIRMED (PROBABLE), PERSISTENT OR RECURRENT (WHO-3)
Both of the following:
1. One of the following case definitions:
190122 Pseudomembranous candidiasis
Clinical Descriptors: White or yellow/creamy spots or plaques that may be located in any part of the oral cavity and can usually be wiped off to reveal an erythematous surface
Patient reported symptoms: None or possible mild to moderate burning pain
Patient reported duration: Lesions/symptoms are usually intermittent, but may be long-standing

Or
190132  
**Erythematous candidiasis**  
Clinical Descriptors: Patchy erythema or red areas usually located on the palate and dorsum of the tongue, but occasionally on the buccal mucosa. At times, white spots or plaques of pseudomembranous candidiasis may also be present.  
Patient reported symptoms: None or possible mild to moderate burning pain  
Patient reported duration: Lesions/symptoms are usually intermittent, but may be long-standing  

and  
2. Specific antifungal therapy initiated or recommended  

NOTE: For Probable Oral/Oropharyngeal Candidiasis, see also the Oral Disorders section.

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**CANDIDIASIS, VULVOVAGINAL**

190141  
**CONFIRMED PATHOGEN**  
Both of the following:  
1. Compatible clinical syndrome, consisting of **at least one of the following** signs or symptoms:  
   a. Vulvovaginal  
   or  
   b. Pruritus  
   or  
   c. Irritation/soreness or dyspareunia  
   or  
   d. Mucous membrane erythema  
   or  
   e. White plaques/exudates adherent to vaginal mucosa  
   or  
   f. Thick, curdy vaginal discharge  
   and  
2. Positive culture or KOH or Gram stain  

190142  
**NO PATHOGEN CONFIRMED (PROBABLE)**  
Both of the following:  
1. Compatible clinical syndrome, consisting of at least two of the following signs or symptoms:  
   a. Vulvovaginal  
   or  
   b. Pruritus  
   or  
   c. Irritation/soreness or dyspareunia  
   or  
   d. Mucous membrane erythema  
   or  
   e. White plaques/exudates adherent to vaginal mucosa  
   or  
   f. Thick, curdy vaginal discharge  
   and  
2. Specific antifungal therapy initiated or recommended
CHAGAS' DISEASE - CENTRAL NERVOUS SYSTEM INVOLVEMENT

190151  CONFIRMED PATHOGEN  
At least one of the following:
1. Direct finding of trypanosomes in cerebrospinal fluid (CSF) or brain biopsy  
or
2. CSF pleocytosis, increased protein

190152  NO PATHOGEN CONFIRMED (PROBABLE)  
All of the following:
1. Person came from endemic area  
and
2. CNS mass lesion with contrast enhancing effect that does not improve with antitoxoplasmosis treatment  
and
3. CSF pleocytosis, increased protein  
and
4. Positive serology

CHAGAS' DISEASE - MYOCARDITIS

190161  CONFIRMED PATHOGEN  
Both of the following:
1. Finding of trypanosomes nests in myocardium biopsy or in buffy coat  
and
2. EKG right bundle branch block

190162  NO PATHOGEN CONFIRMED (PROBABLE)  
All of the following:
1. Person came from endemic area  
and
2. Clinical myocarditis  
and
3. EKG with a variety of disturbances, most commonly right bundle branch block  
and
4. Positive serology
CHORIOAMNIONITIS/AMNIOTIC FLUID INFECTION, specify either chorioamnionitis or amniotic fluid infection

190171 CONFIRMED PATHOGEN, specify pathogen
Amniotic fluid with a positive Gram stain or culture

190172 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Maternal oral temperature greater than or equal to ($\geq$) 100.4º F or 38ºC not attributable to other causes
   and
2. Any two of the following:
   a. Fetal heart rate which is persistently greater than (>) 160 beats per minute (BPM)
   or
   b. Maternal heart rate which is greater than (>) 120 BPM in the absence of tocolytics or known maternal heart tachyarrhythmia
   or
   c. Uterine tenderness not associated with contractions
   or
   d. Purulent cervical discharge or amniotic fluid
   or
   e. Premature labor unresponsive to tocolytic therapy

CMV COLITIS

190181 CONFIRMED (WHO-4 if age greater than (>) 1 month)
Both of the following:
1. Persistent or chronic (greater than or equal to ($\geq$) 14 days) diarrhea (typically in small volume and associated with mucus and blood, abdominal pain, and fever)
   and
2. At least one of the following positive results:
   a. Isolation of CMV from the GI tissue
   or
   b. Detection of CMV antigen from blood or tissue
   or
   c. Isolation of CMV DNA from blood or tissue
   or
   d. Histopathology or cytopathology consistent with CMV

190182 PROBABLE (WHO-4 if age greater than (>) 1 month)
All of the following:
1. Persistent or chronic (greater than or equal to ($\geq$) 14 days) diarrhea (typically in small volume and associated with mucus and blood, abdominal pain, and fever)
   and
2. Appropriate visualization procedure (endoscopy) that reveals mucosal erythema, erosion or ulceration
   and
3. Specific antiviral therapy initiated
CMV ENCEPHALITIS

190191  CONFIRMED (WHO-4 if age greater than (>) 1 month)
Both of the following:
1. Rapidly progressive cognitive impairment, progressive change in mental status or delirium, 
or signs and symptoms of brain stem injury
   and
2. At least one of the following positive results:
   a. Isolation of CMV from CSF or brain tissue
   or
   b. Detection of CMV antigen from CSF or brain issue
   or
   c. Isolation of CMV DNA from CSF or brain tissue
   or
   d. Histopathology or cytopathology consistent with in brain tissue

CMV GASTROENTERITIS

190201  CONFIRMED (WHO-4 if age greater than (>) 1 month)
Both of the following:
1. Presence of abdominal pain
   and
2. Tissue biopsy demonstrating CMV by antigen, detection of viral nucleic acids (e.g. nucleic 
   acid amplification test) or characteristic cytopathic changes

190202  PROBABLE (WHO-4 if age greater than (>) 1 month)
All of the following:
1. Presence of abdominal pain
   and
2. Appropriate visualization procedures (endoscopy) that reveal mucosal erythema, erosion, 
or ulceration
   and
3. Anti-CMV therapy initiated or recommended

CMV MUCOCUTANEOUS ULCERS

190211  CONFIRMED
Both of the following:
1. Direct visualization of oral, vulvovaginal, or perianal ulcers
   and
2. CMV culture of lesion or histologic demonstration of typical CMV cytopathology on 
   biopsy of lesion
CMV PNEUMONITIS

190221 CONFIRMED (WHO-4 if age greater than (>) 1 month)
Both of the following:
1. Hypoxemia and infiltrates on chest x-ray or CT/MRI scan
and
2. Lung tissue biopsy demonstrating CMV by antigen, detection of viral nucleic acids (e.g., nucleic acid amplification test) or characteristic cytopathologic changes

190222 PROBABLE (WHO-4) (age greater than (>) 1 month)
All of the following:
1. Hypoxemia and infiltrates on chest x-ray or CT/MRI scan
and
2. Positive culture, detection of viral antigen, or detection of viral nucleic acids (e.g., nucleic acid amplification test) of CMV from fluid obtained by bronchoalveolar lavage
and
3. No other pathogens identified by routine testing or signs/symptoms persist or recur after treatment of co-pathogens
and
4. Specific antiviral treatment initiated or recommended

CMV PROCTITIS

190231 CONFIRMED (WHO-4 if age greater than (>) 1 month)
Both of the following:
1. Presence of rectal pain associated with tenesmus, mucus, and blood
and
2. At least one of the following positive results:
   a. Isolation of CMV from the GI tissue
   or
   b. Detection of CMV antigen from blood or tissue
   or
   c. Isolation of CMV DNA from blood or tissue
   or
   d. Histopathology or cytopathology

190232 PROBABLE (WHO-4 if age greater than (>) 1 month)
All of the following:
1. Presence of rectal pain associated with tenesmus, mucus, and blood
and
2. Appropriate visualization procedures (endoscopy) that reveal mucosal erythema, erosion, or ulceration
and
3. Specific antiviral therapy initiated
CMV RETINITIS

190241 CONFIRMED (WHO-4 if age greater than (>) 1 month)
Both of the following:
1. Typical lesions including white areas with or without hemorrhages or gray-white areas of retinal necrosis with or without hemorrhages. Lesion(s) has/have irregular, dry-appearing granular border, with little or no overlying vitreous inflammation. Must be diagnosed by an experienced ophthalmologist using indirect ophthalmoscopy.
and
2. Documented by retinal photography that can be independently verified

190242 PROBABLE (WHO-4 if age greater than (>) 1 month)
Both of the following:
1. Typical lesions including white areas with or without hemorrhages or gray-white areas of retinal necrosis with or without hemorrhages. Lesion(s) has/have irregular, dry-appearing granular border, with little or no overlying vitreous inflammation. Must be diagnosed by an experienced ophthalmologist using indirect ophthalmoscopy.
and
2. No documented retinal photographs available

COCCIDIOIDOMYCOSIS, DISSEMINATED

190251 CONFIRMED PATHOGEN (WHO-4)
Appropriate clinical syndrome with identification of the fungal organism in body tissue or fluid for C. immitis by at least one of the following:
1. Positive culture
or
2. Positive histopathology: identification of characteristic appearance of organism within body tissue or fluids

190252 NO PATHOGEN CONFIRMED (PROBABLE)
Appropriate clinical syndrome plus positive complement fixation, ELISA or immunodiffusion antibodies and specific antifungal therapy initiated

CELLULITIS
Inflammation of the skin due to infection

190261 CONFIRMED PATHOGEN, specify pathogen
By positive test for specific organism from blood or aspirate

190262 NO PATHOGEN CONFIRMED (PROBABLE)
Suspected clinically; no organism identified
CERVICITIS
Inflammation of the cervix due to infection

190271 CONFIRMED PATHOGEN, specify pathogen
By culture, nucleic acid amplification test, or other specific diagnostic test in endocervical secretions or biopsy

190272 NO PATHOGEN CONFIRMED (PROBABLE)
Diagnosed clinically, etiology unproven

CONJUNCTIVITIS
Inflammation of the conjunctiva typically caused by viral or bacterial pathogen

190281 CONFIRMED PATHOGEN, specify pathogen
Clinical diagnosis with positive cultures for bacterial or viral pathogen

190282 NO PATHOGEN CONFIRMED (PROBABLE)
Clinical diagnosis only

CROUP (LARYNGOTRACHEOBRONCHITIS), specify either Croup or Laryngotracheobronchitis
Respiratory disease characterized by a barking cough typically affecting infants and children aged three months to three years. The syndrome includes sudden onset of respiratory stridor with upper respiratory infection; inflammation of larynx, trachea, or bronchi is seen with severe illness. The most common etiology is parainfluenza virus, usually type 1 or 3; however other viruses and bacteria may be the causative agent.

190291 CONFIRMED PATHOGEN, specify pathogen
Both of the following:
1. Clinical diagnosis made by an experienced practitioner
   and
2. Positive cultures or rapid diagnostic tests for viral or bacterial pathogen

190292 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Clinical syndrome
   and
2. Negative culture or rapid diagnostic tests or tests not done

CRYPTOCOCCOSIS, OTHER ORGAN SYSTEM, e.g., skin, prostate, bone, adrenal, etc., but not pulmonary; specify organ system

190301 CONFIRMED (WHO-4)
Compatible clinical syndrome and identification of the organism C. neoformans in body tissue or fluid by at least one of the following:
1. Positive culture
   or
2. Identification of characteristic appearance of organisms in tissue or aspirate
190302 NO PATHOGEN CONFIRMED (PROBABLE) (WHO-4)
All of the following:
1. Compatible clinical syndrome
   and
2. Positive cryptococcal antigen greater than or equal to \( \geq 1:8 \) in serum or body fluid
   and
3. Specific antifungal therapy initiated or recommended

CRYPTOCOCCOSIS – PULMONARY

190311 CONFIRMED
Compatible clinical syndrome and identification of the organism C. neoformans in lung tissue or fluid by at least one of the following:
1. Positive culture
   or
2. Identification of characteristic appearance of organisms in tissue or aspirate

190312 NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Compatible clinical syndrome
   and
2. Positive cryptococcal antigen greater than or equal to \( \geq 1:8 \) in serum or body fluid
   and
3. Specific antifungal therapy initiated or recommended

CRYPTOSPORIDIOSIS

190321 CONFIRMED PATHOGEN
Both of the following:
1. At least one of the following:
   a. Diarrhea defined as three or more non-formed stools per day for three or more days
   or
   b. At least one of the following abdominal symptoms:
      1. Nausea
      or
      2. Vomiting
      or
      3. Abdominal pain
   and
2. Microscopic evidence of cryptosporidium present in stool, body fluid, or tissue specimen

There is no definition for any other level of evidence.
CYCLOSPORA GASTROENTERITIS

190331 CONFIRMED PATHOGEN
Both of the following:
1. At least one of the following:
   a. Diarrhea defined as three or more non-formed stools per day for three or more days
   or
   b. Presence of at least one of the following abdominal symptoms
      1. Nausea
      or
      2. Vomiting
      or
      3. Abdominal pain
      or
   c. Presence of at least one of the following:
      1. Biliary colic
      or
      2. Jaundice
      or
      3. Elevation in total gamma-glutamyl transpeptidase (GGTP) greater than or equal to
         \((\geq)\) 2.5 times the upper limit of normal (ULN)

   and
   2. Microscopic evidence of cyclospora present in stool, body fluid, or tissue specimen

There is no definition for any other level of evidence.

DERMATOPHYTE INFECTIONS (TINEA), specify type of infection (tinea capitus, tinea pedis, tinea cruris, tinea corporis) including disseminated
NOTE: To report fungal nail infection, use the ONYCHOMYCOSIS (FUNGAL NAIL INFECTION) code

190341 CONFIRMED PATHOGEN, specify pathogen
Diagnosed by laboratory culture or microscopy; caused by dermatophytes (Microsporum spp., Trichophyton spp.)

190342 NO PATHOGEN CONFIRMED (PROBABLE)
1. Infection suspected clinically
   and
   2. Specific tests negative or not done
DIARRHEA, ACUTE

190351 CONFIRMED PATHOGEN, specify pathogen
All of the following:
1. Clinical syndrome with acute onset of three or more unformed bowel movements (or stools) in a 24-hour period

and
2. Duration lasting greater than or equal to (≥) three days and less than or equal to (≤) 14 days

and
3. Pathogen identified, specify pathogen

190352 NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Clinical syndrome with acute onset of three or more unformed bowel movements (or stools) in a 24-hour period

and
2. Duration lasting greater than or equal to (≥) three days and less than or equal to (≤) 14 days

and
3. No pathogen identified

DIARRHEA, CHRONIC

190361 CONFIRMED PATHOGEN (WHO-3)
All of the following:
1. Clinical syndrome of three or more unformed bowel movements (or stools) in a 24-hour period.

and
2. Duration greater than or equal to (≥) 28 days if greater than or equal to (≥) 15 years of age, and greater than or equal to (≥) 14 days if age less than (<) 15 years

and
3. Pathogen identified, specify pathogen

190362 NO PATHOGEN CONFIRMED (PROBABLE) (WHO-3)
All of the following:
1. Clinical syndrome of three or more unformed bowel movements (or stools) in a 24-hour period

and
2. Duration greater than or equal to (≥) 28 days if greater than or equal to (≥) 15 years of age, and greater than or equal to (≥) 14 days if age less than (<) 15 years

and
3. Either diagnostic testing was done and no pathogen was identified or diagnostic testing was not available
DIARRHEA, SUBACUTE, PERSISTENT (AGE > 15 YEARS)

190371 CONFIRMED PATHOGEN, specify pathogen
All of the following:
1. Three (3) or more unformed bowel movements (or stools) in a 24-hour period.
   and
2. Duration greater than (>) 14 to less than (<) 28 days
   and
3. Pathogen identified, specify pathogen

190372 NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Three (3) or more unformed bowel movements (or stools) in a 24-hour period
   and
2. Duration greater than (>) 14 to less than (<) 28 days
   and
3. Either diagnostic testing was done and no pathogen was identified or diagnostic testing was not available

EMPYEMA, specify site

190381 CONFIRMED PATHOGEN, specify pathogen
Demonstration of bacterial pathogen(s) in pleural cavity by isolation of a bacterial pathogen(s) from an aspirate, chest tube, or biopsy specimen.

190382 NO PATHOGEN CONFIRMED (PROBABLE),
All of the following:
1. Evidence of an infection in a pleural cavity demonstrated by at least one of the following:
   a. Appropriate diagnostic sampling (positive histopathology of tissue)
   or
   b. Demonstration of bacterial pathogen(s) by appropriate microbiological stain of aspirate or biopsy specimen without growth on bacterial culture
   or
   c. Imaging procedures documenting likely infected pleural fluid (e.g., computerized tomography or magnetic resonance imaging)
   and
2. Clinical signs and symptoms compatible with the infection
   and
3. Appropriate treatment initiated and response demonstrated (appropriate treatment may include drainage procedures or antibacterial therapy)
ENCEPHALITIS
Bacterial, parasitic, or viral infection of the brain tissue causing change in level of consciousness, possible seizures, focal neurologic abnormalities

190391 CONFIRMED PATHOGEN, specify pathogen
1. Clinical syndrome diagnosed by an experienced practitioner
   and
2. Pathogen identified by nucleic acid amplification test, culture, or appropriate serologic or histopathologic tests

190392 NO PATHOGEN CONFIRMED (PROBABLE)
1. Clinical syndrome diagnosed by an experienced practitioner
   and
2. Pathogen not identified

ENDOCARDITIS, BACTERIAL

190401 CONFIRMED PATHOGEN, specify pathogen
At least one of the following:
1. Demonstration of a bacterial pathogen(s) by Gram stain, other histologic stain, or culture of valvular vegetation or endocardial tissue
   or
2. At least two of the following:
   a. Typical microorganisms from two separate blood cultures (viridans streptococci, Streptococcus bovis, Staphylococcus aureus, HACEK group, or enterococci (in absence of primary focus))
   or
   b. Echocardiogram positive for oscillating intracardiac or valvular mass; abscess or dehiscence of valve
   or
   c. New valvular regurgitation (not simply worsening or changing of previous murmur)
   or
3. Persistently positive blood cultures (at least two sets of blood cultures obtained on separate occasions, and at least three of the following:
   a. Predisposition from congenital heart condition or intravenous drug use
   or
   b. Temperature greater than (>) 38°C
   or
   c. Evidence of vascular phenomena (e.g., septic pulmonary emboli, major arterial emboli, mycotic aneurysm, intracranial hemorrhage, conjunctival hemorrhages, or Janeway lesions)
   or
   d. Evidence of immunologic phenomena (e.g., Roth spots, Osler’s nodes, hematuria with active urine sediment consistent with glomerulonephritis, or rheumatoid factor)
   or
4. All of the following:
   a. Predisposition from congenital heart condition or intravenous drug use
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and
b. Temperature greater than (>) 38°C
and
c. Evidence of vascular phenomena (e.g., septic pulmonary emboli, major arterial emboli, mycotic aneurysm, intracranial hemorrhage, conjunctival hemorrhages, or Janeway lesions)
and
d. Evidence of immunologic phenomena (e.g., Roth spots, Osler’s nodes, hematuria with active urine sediment consistent with glomerulonephritis, or rheumatoid factor)
and
e. One positive blood culture

ENDOMETRITIS
Infection of the uterus in a peripartum or post partum woman

190411 CONFIRMED PATHOGEN, specify pathogen
Etiology proven by positive test for specific organism in endometrial culture

190412 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Diagnosed clinically, etiology unproven, maternal postpartum fever greater than or equal to (≥) 38°C for greater than or equal to (≥) 4 hours not attributable to other causes and accompanied by uterine tenderness
and
2. Antibiotic therapy initiated

ENDOPHTHALMITIS

190421 CONFIRMED PATHOGEN, specify pathogen
Both of the following:
1. Clinically suspected infection of the eye globe
and
2. Positive culture or Gram stain on aspiration of aqueous or vitreous humor

190422 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Suspected infection of the eye globe
and
2. No positive culture or Gram stain or aspiration not done
EPIDIDYMITIS

190431 CONFIRMED PATHOGEN, specify pathogen
Both of the following:
1. Clinical diagnosis made by an experienced practitioner
and
2. Positive culture or nucleic acid amplification test for specific organism in urethral swabs

190432 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Diagnosed clinically
and
2. Tests on urethral swabs negative or not done

EPIGLOTTITIS

190441 CONFIRMED PATHOGEN, specify pathogen
Both of the following:
1. Diagnosed clinically
and
2. Etiology proven by positive culture for specific organism in blood, tissue, or direct swabs obtained at intubation

190442 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Diagnosed clinically
and
2. No pathogens on specific testing or tests not done

EPISIOTOMY INFECTION

190451 CONFIRMED PATHOGEN, specify pathogen
Both of the following:
1. Pus draining/drained from episiotomy wound or wound dehiscence (episiotomy breakdown) requiring debridement
and
2. Positive culture

190452 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Pus draining/drained from wound or wound dehiscence (episiotomy breakdown) requiring debridement
and
2. Cultures negative or not done
EPSTEIN-BARR VIRUS (EBV) INFECTION, ACUTE

190461 CONFIRMED PATHOGEN

Both of the following:
1. Clinical diagnosis of infectious mononucleosis
   and
2. Positive acute EBV-specific serology

190462 NO PATHOGEN CONFIRMED (PROBABLE)

Both of the following:
1. Clinical diagnosis of infectious mononucleosis
   and
2. Monospot or other rapid heterophile antibody test positive
   and
3. Specific EBV antibody titers not done

EPSTEIN-BARR VIRUS (EBV) INFECTION, NON-ACUTE

190471 CONFIRMED PATHOGEN

Positive tests for EBV infection by blood nucleic acid amplification tests (e.g. PCR) or EBV serology in the absence of clinical infectious mononucleosis syndrome.

ESOPHAGITIS, CANDIDIASIS

190481 CONFIRMED PATHOGEN (WHO-4)

Both of the following:
1. Compatible clinical syndrome, consisting of one or more of the following signs or symptoms:
   a. White plaques in esophagus
      or
   b. Typical filling defects on barium swallow
      or
   c. Orodynophagia (midline retrosternal discomfort with swallowing)
      and
2. Positive culture, KOH, or histopathology from esophagus

190482 NO PATHOGEN CONFIRMED (PROBABLE) (WHO-4)

Both of the following:
1. At least one of the following:
   a. Compatible clinical syndrome, consisting of two or more of the following signs or symptoms:
      1. White plaques in esophagus on endoscopy
      or
      2. Typical filling defects on barium swallow
      or
      3. Odynophagia (midline retrosternal discomfort with swallowing)
      or
   or
b. Confirmed or probable oropharyngeal candidiasis and odynophagia

and

2. Response to specific antifungal therapy for the treatment of esophagitis

ESOPHAGITIS, CMV

190491  CONFIRMED PATHOGEN (WHO-4)
Both of the following:
1. Presence of **at least one of the following** symptoms:
   a. Retrosternal pain
   
   or
   
b. Odynophagia (midline retrosternal discomfort with swallowing)
   
   and
   
2. Tissue biopsy demonstrating CMV by detection of antigen, viral nucleic acids (e.g., nucleic acid amplification test), or characteristic cytopathic changes

190492  NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Presence of **at least one of the following** symptoms:
   a. Retrosternal pain
   
   or
   
b. Odynophagia (midline retrosternal discomfort with swallowing)
   
   and
   
2. Appropriate visualization procedure (endoscopy) that reveals mucosal erythema, erosion or ulceration
   
   and
   
3. CMV is isolated from the lesion by culture alone (no biopsy tests)
   
   and
   
4. Anti-CMV therapy initiated or recommended

ESOPHAGITIS, HSV

190501  CONFIRMED PATHOGEN (WHO-4)
Both of the following:
1. Presence of **at least one of the following** symptoms:
   a. Retrosternal pain
   
   or
   
b. Odynophagia (midline retrosternal discomfort with swallowing)
   
   and
   
2. Tissue biopsy demonstrating HSV by detection of antigen, viral nucleic acids (e.g., nucleic acid amplification test), or characteristic cytopathic changes

190502  NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Presence of **at least one of the following** symptoms:
   a. Retrosternal pain
   
   or
   
b. Odynophagia (midline retrosternal discomfort with swallowing)
and
2. Appropriate visualization procedure (endoscopy) that reveals mucosal erythema, erosion, or ulceration
and
3. HSV is isolated from the lesion by culture alone (no biopsy tests)
and
4. Anti-HSV therapy initiated or recommended

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GASTROENTERITIS, ACUTE

190511 CONFIRMED PATHOGEN, specify pathogen
Both of the following:
1. Illness characterized by vomiting and diarrhea
and
2. Etiology identified by antigen testing, culture, or microscopy

190512 NO PATHOGEN CONFIRMED (PROBABLE)
1. Clinical illness
and
2. Etiology not identified or testing not done

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190522 HAIRY LEUKOPLAKIA, OHARA DEFINITION (WHO-3)

Clinical descriptors: Whitish/grey lesions on the lateral margins of the tongue. They are not removable and may exhibit vertical corrugations. Lesions range in size as they may be less than (<) one cm (centimeter), or may extend onto the ventral and dorsal surfaces of the tongue where they are usually flat. May be bilateral or unilateral.

Patient-reported symptoms: Asymptomatic.
Patient-reported duration: Lesion(s) usually longstanding.

NOTE: See also HAIRY LEUKOPLAKIA in the Oral Disease section.

HANSEN’S DISEASE/LEPROSY, specify either Hansen’s Disease or Leprosy

190531 CONFIRMED PATHOGEN
Both of the following:
1. Characteristic dermatological or neurological manifestations
and
2. Consistent histopathology:
   Multibacillary leprosy: acid fast bacilli seen on the smear
   Paucibacillary leprosy: well-formed, non-caseating granuloma and nerve involvement (AFB need not be seen)

190532 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Characteristic infiltrative skin lesion, hypoesthesia, or peripheral neuropathy
and
2. Biopsy not done
HEPATITIS A, ACUTE

170021 CONFIRMED PATHOGEN
Hepatic inflammation diagnosed within six months of having a known normal (asymptomatic) liver or chronic stable hepatitis
Hepatic inflammation is defined by at least one of the following:
1. Aminotransferase elevation of at least five times the upper limit of normal (ULN)
or
2. Seropositive for IgM antibody to HAV (Acute Hepatitis A)

NOTE: See also HEPATITIS in the Hepatobiliary section.

HEPATITIS B, ACUTE

170031 CONFIRMED PATHOGEN
Hepatic inflammation diagnosed within six months of having a known normal (asymptomatic) liver or chronic stable hepatitis
Hepatic inflammation is defined by at least one of the following:
1. Aminotransferase elevation of at least five times the upper limit of normal (ULN)
or
2. Seropositive for IgM antibody to HBc with presence of new HBsAg (acute hepatitis B)

NOTE: See also HEPATITIS in the Hepatobiliary section.

HEPATITIS B VIRUS, CHRONIC REPLICATIVE

190541 CONFIRMED
Both of the following:
1. Hepatitis B surface antigen (HbsAg) detected on two occasions at least six months apart
and
2. Hepatitis B virus DNA (HBV DNA) or a Hepatitis B e antigen (HbeAg) is detected six or more months after the initial detectable Hepatitis B surface antigen (HbsAg)

NOTE: See also HEPATITIS in the Hepatobiliary section.

HEPATITIS B VIRUS, CHRONIC NON-REPLICATIVE

190551 CONFIRMED
Both of the following:
1. Hepatitis B surface antigen (HbsAg) detected by repeat testing performed six or more months after the initial detectable HbsAg
and
2. Tests for Hepatitis B virus DNA (HBV DNA) and Hepatitis B e antigen (HbeAg) are negative six or more months after the initial HbsAg
HEPATITIS B, CHRONIC, WITH HEPATITIS D CO-INFECTION

190561 CONFIRMED PATHOGEN
Hepatitis B surface antigen (HbsAg) detected by repeat testing performed six or more months after the initial detectable HbsAg and antibody tests positive for Hepatitis D virus

HEPATITIS C, ACUTE

170041 CONFIRMED PATHOGEN
Hepatic inflammation diagnosed within six months of having a known normal (asymptomatic) liver or chronic stable hepatitis
Hepatic inflammation is defined by at least one of the following:
1. Aminotransferase elevation of at least five times ULN
or
2. Evidence of HCV viremia without HCV antibody
or
3. Documented seroconversion within six months to anti-HCV with an aminotransferase elevation above the normal level (Acute Hepatitis C)

NOTE: See also HEPATITIS in the Hepatobiliary section.

HEPATITIS C VIRUS, CHRONIC

190571 CONFIRMED PATHOGEN
Hepatitis C infection detected on two occasions at least six months apart, by positive HCV RNA assays (if HCV ELISA assays are done, positives must be confirmed by positive HCV RNA assays for identification of chronic infection)

190572 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. ALT (SGPT) results greater than (>) the upper limit of normal (ULN) on two or more occasions at least six months apart
and
2. Hepatitis C infection detected any time by HCV ELISA (confirmed by RIBA) on two occasions at least six months apart, and HCV RNA not done

HEPATITIS, ACUTE

170052 NO PATHOGEN CONFIRMED (PROBABLE)
Hepatic inflammation diagnosed within six months of having a known normal (asymptomatic) liver or chronic stable hepatitis
Hepatic inflammation is defined by at least one of the following:
1. Aminotransferase elevation of at least five times ULN
or
2. Other causes of hepatitis have been ruled out if possible where none of the following are identified:
   a. Seropositive for IgM antibody to HAV (Acute Hepatitis A)
   or
   b. IgM antibody to HBc with presence of new HBsAg (Acute Hepatitis B)
   or
   c. Evidence of HCV viremia without HCV antibody or documented seroconversion within six months to anti-HCV with an aminotransferase elevation above the normal level (Acute Hepatitis C)
   or
   d. Elevated antibody to Hepatitis E in association with travel to or residence in an endemic area (Acute Hepatitis E)

NOTE: See also HEPATITIS in the Hepatobiliary section.

ROSEOLA INFANTUM, also called HHV6 (for the specific virus) or exanthema subitum

190581 CONFIRMED PATHOGEN
   Both of the following:
   1. Clinical syndrome of roseola infantum (also known as exanthema subitum)
   and
   2. Positive viral detection studies for HHV6

190582 NO PATHOGEN CONFIRMED (PROBABLE)
   Clinical syndrome of roseola infantum (also known as exanthema subitum); virus detection studies negative or not done

HSV INFECTION, VIRAL RESISTANCE AND REFRACTORY TO THERAPY

190591 CONFIRMED PATHOGEN
   A virologically confirmed (nucleic acid amplification test or culture) HSV infection refractory to appropriate treatment for greater than (> 15 days. Use of this code must include a positive culture with in vitro sensitivity testing showing resistance to acyclovir.

HERPES SIMPLEX VIRUS (HSV), MUCOCUTANEOUS, duration less than (<) one month

190601 CONFIRMED PATHOGEN
   Both of the following:
   1. Typical (vesicular or ulcerative) HSV lesion(s) in any of the following sites: anogenital (external genitalia, cervix, vagina, perineum, perirectal, rectal), oral, perioral (including labial) or skin of the hands or feet
   and
2. Any one of the following:
   a. HSV isolated from lesion
   or
   b. HSV antigen detected by immunoassay from vesicular fluid or cells obtained from the base of a vesicle or ulcer
   or
   c. Recurrence of lesion in same general location: anogenital (external genitalia, cervix, vagina, perineum, perirectal, rectal), oral, perioral (including labial) or finger/toe with prior documented positive HSV culture or immunoassay

190602 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Clinically-apparent typical (vesicular or ulcerative) HSV lesion(s) with prodromal or concurrent symptoms of discomfort (burning, itching, pain)
   and
2. At least one of the following:
   a. Typical herpes virus inclusions or multinucleated giant cells evident in cells obtained from the base of an ulcer or vesicular fluid
   or
   b. Specific antiviral treatment initiated or recommended

HERPES SIMPLEX VIRUS (HSV), MUCOCUTANEOUS, duration greater than (>) one month

190611 CONFIRMED PATHOGEN (WHO-4)
Both of the following:
1. Typical (vesicular or ulcerative) HSV lesion(s) in any of the following sites: anogenital (external genitalia, cervix, vagina, perineum, perirectal, rectal), oral, perioral (including labial) or skin of the hands or feet
   and
2. At least one of the following:
   a. HSV isolated from lesion
   or
   b. HSV antigen detected by immunoassay from vesicular fluid or cells obtained from the base of a vesicle or ulcer
   or
   c. Recurrence of lesion in same general location: anogenital (external genitalia, cervix, vagina, perineum, perirectal, rectal), oral, perioral (including labial) or finger/toe with prior documented positive HSV culture or immunoassay

190612 NO PATHOGEN CONFIRMED (PROBABLE) (WHO-4)
Both of the following:
1. Clinically apparent typical (vesicular or ulcerative) HSV lesion(s) with prodromal or concurrent symptoms of discomfort (burning, itching, or pain)
   and
2. **At least one of the following:**
   a. Typical herpes virus inclusions or multinucleated giant cells evident in cells obtained from the base of an ulcer or vesicular fluid

   or

   b. Specific antiviral treatment initiated or recommended

### HERPES SIMPLEX VIRUS (HSV), PNEUMONITIS

190621 **CONFIRMED PATHOGEN (WHO-4)**

   **All of the following:**
   1. Hypoxemia and infiltrates on chest X-ray or CT/MRI scan
   2. Tissue biopsy or cells obtained by bronchoalveolar lavage (BAL) demonstrating HSV by culture, antigen, nucleic acid amplification (e.g., PCR), or characteristic cytopathic changes
   3. No other pathogens identified by routine testing, or signs/symptoms persist or recur after treatment of co-pathogens

190622 **NO PATHOGEN CONFIRMED (PROBABLE)**

   **All of the following:**
   1. Hypoxemia and infiltrates on chest X-ray or CT/MRI scan
   2. Microbiologic evidence (positive culture, antigen test, or nucleic acid amplification) of non-genital HSV infection elsewhere in body (e.g., pharynx, esophagus), but lung tissue or BAL fluid testing negative or not done
   3. No other pathogens identified by routine testing or signs/symptoms persist or recur after treatment of copathogens
   4. Specific antiviral treatment initiated or recommended

### HISTOPLASMOSIS, DISSEMINATED

190631 **CONFIRMED PATHOGEN (WHO-4)**

   **Both of the following:**
   1. Clinical syndrome consistent with histoplasmosis
   2. Identification of the fungal organism Histoplasma capsulatum by **at least one of the following**:
      a. Positive culture
      b. Positive histopathology: Identification of characteristic appearance of organism within body tissue or fluids
      c. Detection of positive Histoplasma antigen obtained from blood, urine, or body fluid
190632  **NO PATHOGEN CONFIRMED (PROBABLE)**
All of the following:
1. Appropriate clinical syndrome
   and
2. Plus positive complement fixation, ELISA, or immunodiffusion antibodies
   and
3. Specific antifungal therapy initiated

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**IMPETIGO**

190641  **CONFIRMED PATHOGEN, specify pathogen**
Both of the following:
1. Appropriate clinical syndrome
   and
2. Positive bacterial culture

190642  **NO PATHOGEN CONFIRMED (PROBABLE)**
Both of the following:
1. Appropriate clinical syndrome
   and
2. Bacterial culture negative or not obtained

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**INFLUENZA**

190651  **CONFIRMED PATHOGEN, specify influenza virus type if known**
Both of the following:
1. Influenza-like illness as defined by the sudden onset of fever and cough or sore throat in the absence of other diagnoses
   and
2. Positive culture, nucleic acid amplification test, or rapid antigen test for influenza virus

If available, specify type (influenza A, B, C) and subtype (e.g., pandemic or seasonal H1N1, H3N2, etc.)

190652  **NO PATHOGEN CONFIRMED (PROBABLE)**
Both of the following:
1. Influenza-like illness as defined by the sudden onset of fever and cough or sore throat in the absence of other diagnoses
   and
2. Evidence of regional endemic/epidemic area or travel to endemic/epidemic area
ISOSPORAISIS

190661 CONFIRMED PATHOGEN (WHO-4)
Both of the following:
1. At least one of the following:
   a. Diarrhea defined as three or more non-formed stools per day for three or more days
   or
   b. Presence of at least one of the following abdominal symptoms
      1. Nausea
      or
      2. Vomiting
      or
      3. Abdominal pain
   and
   2. Microscopic evidence of Isospora present in stool, body fluid, or tissue specimen

There is no definition for any other level of evidence.

LEISHMANIASIS, CUTANEOUS OR MUCOSAL, specify site

190671 CONFIRMED PATHOGEN
Both of the following:
1. Compatible clinical syndrome
   and
2. Histologic evidence of disease from an aspirate or biopsy

190672 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Compatible clinical syndrome
   and
2. Specific treatment initiated or recommended

LEISHMANIASIS, VISCERAL OR DISSEMINATED, specify site

190681 CONFIRMED PATHOGEN (WHO-4)
Both of the following:
1. Compatible clinical syndrome
   and
2. Histologic evidence of disease from an aspirate or biopsy

190682 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Compatible clinical syndrome
   and
2. Specific treatment initiated or recommended
LYMPHOID INTERSTITIAL PNEUMONITIS (LIP)

190691 CONFIRMED PATHOGEN (WHO-3)
All of the following:
1. Characteristic clinical course (ranges from asymptomatic to chronic bronchitis or wheezing, without response to antibiotic treatment)
   and
2. Compatible chest x-ray (chronic bilateral reticulonodular interstitial infiltrates)
   and
3. Lymphocytic interstitial infiltration on lung biopsy

190692 NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Characteristic clinical
   and
2. Radiologic course as above
   and
3. Lung biopsy not done

LYME DISEASE, EARLY

190702 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Clinical diagnosis is sufficient if characteristic erythema migrans (erythema chronicum migrans) lesions are diagnosed by an experienced practitioner.
   and
2. There is a single lesion.

LYME DISEASE, EARLY DISSEMINATED

190712 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Clinical diagnosis is sufficient if characteristic erythema migrans (erythema chronicum migrans) lesions are diagnosed by an experienced practitioner.
   and
2. There are multiple lesions.
LYME DISEASE, LATE (OR SUSPECTED EARLY DISSEMINATED DISEASE WITHOUT ERYTHEMA MIGRANS LESIONS)

190721  CONFIRMED PATHOGEN
All of the following:
1. One of the following:
   a. Early disseminated disease without erythema migrans lesions (e.g., isolated facial nerve palsy, or heart block)
   or
   b. Late Lyme disease (e.g., Lyme arthritis)

2. Appropriate clinical diagnosis

3. Serologic (2-step EIA and immunoblot) diagnosis

LYMPHADENITIS

190731  CONFIRMED PATHOGEN, specify pathogen
Both of the following:
1. Clinical syndrome of acute swelling of lymph gland(s), with tenderness and other signs of inflammation

2. Positive culture for specific organism from node biopsy or aspirate, or blood

190732  NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Clinical syndrome of acute swelling of lymph gland(s), with tenderness and other signs of inflammation

2. Cultures for specific organism from node biopsy or aspirate not done or negative

LYMPHADENOPATHY, PERSISTENT GENERALIZED (WHO-1)

190742  NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Clinical syndrome of painless swollen lymph nodes, each greater than (>) 0.5 mm in size

2. Present at greater than (>) two sites

3. Lasting for greater than (>) three months
MALARIA

190751 CONFIRMED PATHOGEN
Both of the following:
1. Appropriate clinical syndrome
   and
2. Identification of Plasmodium spp. on a smear of peripheral blood by microscopy, rapid antigen test, or nucleic acid amplification test

190752 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Appropriate clinical syndrome
   and
2. Specific treatment initiated or recommended

MASTITIS, POSTPARTUM

190761 CONFIRMED PATHOGEN, specify pathogen
Appropriate clinical syndrome in a postpartum woman, as defined by both of the following:
1. Positive bacterial culture from breast or expressed secretions
   and
2. Any two of the following:
   a. Unilateral breast (not nipple) pain
   or
   b. Erythema and induration in one area of the breast
   or
   c. Fluctuance of one area of the breast
   or
   d. Temperature greater than (> 38.0°C

190762 NO PATHOGEN CONFIRMED (PROBABLE)
Appropriate clinical syndrome in a postpartum woman, as defined by:
Any two of the following, but bacterial cultures of breast/breast secretions negative or not done:
1. Unilateral breast (not nipple) pain
   or
2. Erythema and induration in one area of the breast
   or
3. Fluctuance of one area of the breast
   or
4. Temperature greater than (> 38.0°C
MASTOIDITIS

190771 CONFIRMED PATHOGEN, specify pathogen
All of the following:
1. Appropriate clinical syndrome
   and
2. Positive compatible radiography (plain films, CT scan, or MRI scan)
   and
3. Positive bacterial culture in surgical specimen or mastoid aspirate

190772 NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Appropriate clinical syndrome
   and
2. Positive compatible radiography (plain films, CT scan, or MRI scan)
   and
3. Bacterial cultures negative or not done

MEASLES - also called RUBEOLA,
Highly-contagious viral disease that causes fever, cough, catarrh, and conjunctivitis followed by characteristic rash; complications include pneumonia, encephalitis, and death

190781 CONFIRMED PATHOGEN
Both of the following:
1. Appropriate clinical syndrome
   and
2. Positive IgM serology, viral culture, or nucleic acid amplification test

190782 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Appropriate clinical syndrome
   and
2. Virus-specific serology or culture not done
MENINGITIS, non-AIDS defining if cryptococcal or mycobacterial meningitis, or histoplasmosis or coccidioidomycosis; also see codes specific for each of these pathogens

190791  CONFIRMED PATHOGEN (WHO-3), specify pathogen
All of the following:
1. Appropriate clinical syndrome
   and
2. Abnormal cerebrospinal fluid (CSF), with identification of organism in CSF or blood by at least one of the following:
   a. Culture
   or
   b. Antigen test
   or
   c. Nucleic acid amplification test (e.g., PCR)
   or
   d. For syphilis, a positive CSF VDRL

Specify pathogen; examples include, but are not limited to, Streptococcus pneumoniae, Haemophilus influenzae, Neisseria meningitidis, Candida albicans, enterovirus, Treponema pallidum (neurposyphilis), or certain parasites.

190792  NO PATHOGEN CONFIRMED (PROBABLE) (WHO-3)
All of the following:
1. Appropriate clinical syndrome
   and
2. Abnormal cerebrospinal fluid (CSF)
   and
3. No positive tests for specific pathogen from CSF or blood

MICROSPORIDIOSIS

190801  CONFIRMED PATHOGEN
Both of the following:
1. At least one of the following:
   a. Diarrhea defined as three or more non-formed stools per day for three or more days
   or
   b. Presence of at least one of the following abdominal symptoms:
      1. Nausea
      or
      2. Vomiting
      or
      3. Abdominal pain
   and
2. Microscopic evidence of microsporidia present in stool, body fluid, or tissue specimen

There are no criteria for any other level of evidence.
MOLLUSCUM CONTAGIOSUM, LIMITED

190812 NO PATHOGEN CONFIRMED (PROBABLE)
Clinical diagnosis of characteristic skin lesions of small flesh-colored, pearly or pink, dome-shaped or umbilicated growths

MOLLUSCUM CONTAGIOSUM, EXTENSIVE (WHO-2)

190822 NO PATHOGEN CONFIRMED (PROBABLE)
Clinical diagnosis of characteristic skin lesions of small flesh-colored, pearly or pink, dome-shaped or umbilicated growths, may be inflamed or red, may cover more than 5% (percent) of body area or be disfiguring

MYOSITIS/PYOMYOSITIS, specify either myositis or pyomyositis

190831 CONFIRMED PATHOGEN, specify pathogen
Both of the following:
  1. At least one of the following:
     a. Appropriate clinical diagnosis including elevated serum CK level
     or
     b. Radiologic imaging findings of pyomyositis
     and
  2. Positive blood or surgical specimen culture or aspirate culture

190832 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
  1. Appropriate clinical diagnosis including elevated serum CK
  and
  2. Negative blood or surgical specimen cultures or cultures not done; with or without positive imaging

OMPHALITIS

190841 CONFIRMED PATHOGEN, specify pathogen
Both of the following:
  1. Clinical diagnosis of neonatal umbilical infection with purulent umbilical discharge and periumbilical cellulitis
  and
  2. Pathogen identified

190842 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
  1. Clinical diagnosis of neonatal umbilical infection with purulent umbilical discharge and periumbilical cellulitis
  and
  2. No pathogen identified; testing negative or not done
ONYCHOMYCOSIS, also called FUNGAL NAIL INFECTION (NOTE: For Tinea, seeDERMATOPHYTE INFECTIONS)

190851 CONFIRMED PATHOGEN (WHO-2), specify pathogen
Both of the following:
1. Appropriate clinical diagnosis
and
2. Positive fungal culture of the nail or nail plate material

190852 NO PATHOGEN CONFIRMED (PROBABLE) (WHO-2)
Both of the following:
1. Appropriate clinical diagnosis
and
2. Cultures of nail or nail plate negative or not done

OSTEOMYELITIS, specify bone(s)

190861 CONFIRMED PATHOGEN, specify pathogen
Both of the following:
1. Appropriate clinical diagnosis
and
2. Positive cultures of blood or bone tissue, with or without positive bone histology

190862 NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Appropriate clinical diagnosis
and
2. Supportive radiologic imaging
and
3. Negative cultures or histology, or cultures/surgery not done

OTITIS EXTERNA

190871 CONFIRMED PATHOGEN, specify pathogen
Both of the following:
1. Appropriate clinical syndrome
and
2. Positive Gram stain or culture for specific organism

OTITIS MEDIA, ACUTE

190881 CONFIRMED PATHOGEN, specify pathogen
Both of the following:
1. Appropriate clinical syndrome
and
2. Confirmed by tympanocentesis yielding positive Gram stain or culture for specific organism
<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
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<td>190882</td>
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<td>All of the following:</td>
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<tr>
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<td>1. Appropriate clinical syndrome</td>
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<td>and</td>
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<td>2. Abnormal pneumatic otoscopy</td>
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<td>and</td>
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<td>3. Tympanocentesis cultures negative or not done</td>
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**OTITIS MEDIA, WITH EFFUSION**

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<th>Description</th>
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<tr>
<td>190892</td>
<td>NO PATHOGEN CONFIRMED (PROBABLE)</td>
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<tr>
<td></td>
<td>Clinical syndrome of persistent effusion (present for greater than or equal to ((\geq) three months) diagnosed by physical exam with pneumatic otoscopy or tympanometry</td>
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**OTITIS MEDIA, CHRONIC SUPPURATIVE**

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<td>NO PATHOGEN CONFIRMED (PROBABLE)</td>
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<tr>
<td></td>
<td>Clinical syndrome of otorrhea for greater than or equal to ((\geq) three months)</td>
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**PAPILLOMATOSIS, LARYNGEAL**

<table>
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<th>Description</th>
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<td>Both of the following:</td>
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<tr>
<td></td>
<td>1. Appropriate clinical diagnosis</td>
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<td>and</td>
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<td>2. Confirmed by characteristic findings on laryngoscopy, cytology, or histopathology</td>
</tr>
</tbody>
</table>

**PAPILLOMATOSIS, ANOGENITAL, also known as CONDYLOMA ACUMINATA, GENITAL WARTS**

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
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</thead>
<tbody>
<tr>
<td>190921</td>
<td>CONFIRMED PATHOGEN</td>
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<tr>
<td></td>
<td>All of the following:</td>
</tr>
<tr>
<td></td>
<td>1. Appropriate clinical diagnosis</td>
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<td></td>
<td>and</td>
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<td></td>
<td>2. Confirmed by characteristic findings of anal or genital warts on exam</td>
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<td></td>
<td>and</td>
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<td>3. Confirmed by positive cytology, histopathology, or nucleic acid amplification tests for papillomavirus</td>
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<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>190922</td>
<td>NO PATHOGEN CONFIRMED (PROBABLE)</td>
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<tr>
<td></td>
<td>Both of the following:</td>
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<tr>
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<td>1. Genital or anal warts on exam</td>
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<tr>
<td></td>
<td>and</td>
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<tr>
<td></td>
<td>2. Cytology, histopathology, or nucleic acid amplification tests negative or not done</td>
</tr>
</tbody>
</table>

**PARACOCCIDIOIDOMYCOSIS**
CONFIRMED PATHOGEN
All of the following:
1. Appropriate clinical syndrome
   and
2. Identification of *P. brasiliensis* by **at least one of the following:**
   a. Positive culture from sputum, bronchoalveolar lavage, cerebrospinal fluid lymph nodes, lung tissue, skin, or any other tissue
   or
   b. Positive histopathology: observation of the characteristic "pilot wheel" shape of *P. brasiliensis*

NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Appropriate clinical syndrome
   and
2. Positive complement fixation, ELISA, or immunodiffusion antibodies
   and
3. Specific antifungal therapy initiated

PAROTITIS, ACUTE (includes Mumps And Non-Mumps Parotitis), specify either mumps or non-mumps parotitis if pathogen known

CONFIRMED PATHOGEN, specify pathogen
Both of the following:
1. Unilateral or bilateral swelling of the parotid gland with loss of the angle of the mandible
   and
2. Causative agent identified by viral or bacterial culture, nucleic acid amplification test, IgM mumps serology, or histology

NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Unilateral or bilateral swelling of the parotid gland with loss of the angle of the mandible
   and
2. No causative agent identified or tests not done

PAROTITIS, CHRONIC (WHO-2, CDC A)

NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Clinical diagnosis characterized by asymptomatic bilateral swelling that may spontaneously resolve and recur, usually painless
   and
2. No other known cause
PARVOVIRUS B19 INFECTION

190961  CONFIRMED PATHOGEN
Both of the following:
1. Appropriate clinical diagnosis characterized by rash of “fifth disease” (also known as
erythema infectiosum), aplastic crisis, or other characteristic syndrome (e.g., hydrops
fetalis)
   and
2. Positive serologic or nucleic acid amplification tests (e.g., PCR) for parvovirus B19

PELVIC INFLAMMATORY DISEASE (PID), includes SALPINGITIS, TUBO-OVARIAN
ABSCESS; specify which form

150111  CONFIRMED PATHOGEN
Both of the following:
1. Clinical diagnosis made by an experienced practitioner
   and
2. Positive culture for specific organism (e.g., Chlamydia or Neisseria gonorrhoeae) from
material obtained by laparoscopy or cul de sac aspiration, or cervical/vaginal diagnostic
test in a woman with appropriate symptoms

150112  NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Clinical diagnosis made by an experienced practitioner
   and
2. Negative cultures in a woman with appropriate symptoms

NOTE: See also PELVIC INFLAMMATORY DISEASE in Genitourinary/Sexually Transmitted
Infections section.

PENICILLIUM MARNEFFEI, DISSEMINATED

190971  CONFIRMED PATHOGEN
Both of the following:
1. Appropriate clinical syndrome
   and
2. At least one of the following:
   a. Isolation of Penicillium marneffei from blood, bone marrow, tissue, or other normally-
sterile body fluids
   or
   b. Positive histology (elongated yeast-like organism with clear central septum in
Wright's-stained skin biopsy, touch smear, or scraping of a skin lesion)
190972  NO PATHOGEN CONFIRMED (PROBABLE)  
All of the following:  
1. Appropriate clinical syndrome for penicilliosis  
   and  
2. Cultures or histology negative or not done  
   and  
3. Appropriate antibiotic therapy recommended or initiated  

PERITONITIS  

190981  CONFERMED PATHOGEN, specify pathogen  
Both of the following:  
1. Appropriate clinical diagnosis of acute illness  
   and  
2. Positive cultures from peritoneal fluid or ascites  

190982  NO PATHOGEN CONFIRMED (PROBABLE)  
Both of the following:  
1. Appropriate clinical diagnosis of acute illness  
   and  
2. Cultures from peritoneal fluid or ascites negative or not done  

PERTUSSIS  

190991  CONFERMED PATHOGEN  
Both of the following:  
1. Appropriate clinical diagnosis of chronic cough syndrome  
   and  
2. Positive culture, direct fluorescent antibody test, or nucleic acid amplification test (e.g.,  
   PCR) for Bordetella pertussis or Bordetella parapertussis  

190992  NO PATHOGEN CONFIRMED (PROBABLE)  
Both of the following:  
1. Appropriate clinical diagnosis of chronic cough syndrome  
   and  
2. Culture, direct fluorescent antibody test, or nucleic acid amplification test negative or not  
   done  

PHARYNGITIS  

191001  CONFERMED PATHOGEN, specify pathogen  
Both of the following:  
1. Sore throat, inflamed pharynx on clinical exam  
   and  
2. Positive throat culture or other antigen detection test
NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Sore throat and inflamed pharynx on exam
and
2. Culture or specific identification tests negative or not done

PROGRESSIVE MULTIFOCAL LEUKOENCEPHALOPATHY (PML)

CONFIRMED PATHOGEN (WHO-4)
All of the following:
1. Appropriate clinical diagnosis of subacute or chronic progressive neurologic illness characterized by hemiparesis, aphasia, hemianopsia, ataxia, and other focal deficits
and
2. Compatible white matter changes on CAT or MRI scans
and
3. At least one of the following:
   a. Positive histopathology or in situ hybridization for JC polyomavirus from a brain biopsy
   or
   b. Positive nucleic acid amplification test (e.g., PCR) of cerebrospinal fluid (CSF) for JC polyomavirus

NO PATHOGEN CONFIRMED (PROBABLE) (WHO-4)
Both of the following:
1. Appropriate clinical and radiologic diagnosis as defined above
and
2. Brain biopsy or CSF nucleic acid amplification tests negative or not done

PNEUMOCYSTIS JIROVECII PNEUMONIA (PCP), formerly known as Pneumocystis carinii pneumonia

CONFIRMED PATHOGEN
Both of the following:
1. A history (within the past three months) of shortness of breath, dyspnea on exertion, cough, or fever
and
2. Histological, cytological, fluorescent antibody test or nucleic acid amplification test evidence of Pneumocystis jirovecii on bronchoalveolar lavage, lung biopsy, or sputum specimen

NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. A history (within the past three months) of shortness of breath, dyspnea on exertion, cough, or fever
and
2. Abnormal chest X-ray (or CT scan) or hypoxemic arterial blood gas PaO2 less than (<) 80 mm Hg or (A-a) DO2 mm Hg greater than (>) 15, on room air
and
3. Specific anti-Pneumocystis therapy initiated
PNEUMOCYSTIS JIROVECII, EXTRAPULMONARY, specify site

191031 CONFIRMED PATHOGEN
One of the following:
1. Histological, cytological, fluorescent antibody test or nucleic acid amplification test evidence of Pneumocystis jirovecii in an aspirate or biopsy of a normally-sterile body site other than the lung

or
2. For ophthalmologic disease only:
   a. Characteristic lesions of Pneumocystis choroiditis according to an experienced ophthalmologist
   
   and
   b. Improvement with systemic anti-Pneumocystosis therapy

There is no definition for any other level of evidence.

PNEUMONIA, BACTERIAL SINGLE EPISODE within six months

191041 CONFIRMED PATHOGEN (WHO-3), specify pathogen
Both of the following in a patient with no previous episode of pneumonia in past six months:
1. Chest radiographic examination shows new or progressive infiltrate, consolidation, or cavitation

and
2. At least one of the following:
   a. Bacterial organism(s) cultured from blood with no alternative site of infection
   or
   b. Isolation of a bacterial pathogen(s) from a culture specimen obtained by transtracheal aspirate, protected bronchial brushing, or biopsy
   or
   c. Histopathologic evidence of pneumonia with bacterial organism(s) demonstrated by Gram stain or culture of tissue specimen or positive Quellung test for Streptococcus pneumoniae
   or
   d. Demonstration of a predominant bacterial organism by positive culture or Gram stain of an adequate sputum specimen (fewer than 10 epithelial cells and greater than (> 25 PMNs per high power field)
   or
   e. Urine antigen detection method positive for Legionella or Streptococcus pneumoniae, or appropriately-collected (acute and convalescent) serologic tests positive for Legionella, Chlamydia, or Mycoplasma
191042 NO PATHOGEN CONFIRMED (PROBABLE), (WHO-3),
single episode within six months

All of the following in a patient with no previous episode of pneumonia in past six months:
1. Chest radiographic examination shows new or progressive infiltrate, consolidation, or cavitation

   and

2. At least one of the following:
   a. Fever or cough
   or
   b. New onset of purulent sputum or change in character of sputum

   and

3. Appropriate antibacterial therapy initiated or recommended

PNEUMONIA, BACTERIAL RECURRENT EPISODE within six months

191051 CONFIRMED PATHOGEN (WHO-4), specify pathogen

Both of the following in a patient with one or more episodes (in addition to current episode) of pneumonia in past six months:
1. Chest radiographic examination shows new or progressive infiltrate, consolidation, or cavitation

   and

2. At least one of the following:
   a. Bacterial organism(s) cultured from blood with no alternative site of infection
   or
   b. Isolation of a bacterial pathogen(s) from a culture specimen obtained by transtracheal aspirate, protected bronchial brushing, or biopsy
   or
   c. Histopathologic evidence of pneumonia with bacterial organism(s) demonstrated by Gram stain or culture of tissue specimen or positive Quellung test for Streptococcus pneumoniae
   or
   d. Demonstration of a predominant bacterial organism by positive culture or Gram stain of an adequate sputum specimen (fewer than 10 epithelial cells and greater than (> ) 25 PMNs per high power field)
   or
   e. Urine antigen detection method positive for Legionella or Streptococcus pneumoniae, or appropriately collected (acute and convalescent) serologic tests positive for Legionella, Chlamydia, or Mycoplasma
191052  NO PATHOGEN CONFIRMED (PROBABLE) (WHO-4)

All of the following in a patient with one or more episodes (in addition to current episode) of pneumonia in past six months:

1. Chest radiographic examination shows new or progressive infiltrate, consolidation, or cavitation.

and

2. At least one of the following:
   a. Fever or cough
   or
   b. New onset of purulent sputum or change in character of sputum

and

3. Appropriate antibacterial therapy initiated or recommended

PNEUMONIA, NON-BACTERIAL

191062  NO PATHOGEN CONFIRMED (PROBABLE)

All of the following:

1. Compatible clinical syndrome of pneumonia (e.g., productive cough and fever)

and

2. Radiologic evidence of pulmonary infiltrate

and

3. Antibacterial therapy not initiated

NOTE: This is not to be used for diagnoses of bacterial pneumonia, or to fungal pneumonia caused by a mycosis for which exist codes elsewhere. Refer to the bacterial pneumonia criteria for the Bacterial Pneumonia diagnoses, or to the appropriate fungal diagnosis code. This diagnosis would apply to viral pneumonia of known cause, other than measles or CMV, or HSV.

PROCTITIS, NON-CMV, see CMV-specific code

191071  CONFIRMED PATHGEN, specify pathogen.

All of the following:

1. Appropriate clinical diagnosis of rectal pain

and

2. Associated with tenesmus, mucus, and blood

and

3. Positive diagnostic test for specific organism in anal lesions other than CMV, which is coded with CMV Proctitis code

PROSTATITIS

191081  CONFIRMED PATHGEN, specify pathogen

Both of the following:

1. Appropriate clinical diagnosis

and

2. Positive diagnostic test for specific organism in urine or prostatic secretions
PYELONEPHRITIS
NOTE: In children less than (<) five years of age, it may be difficult to distinguish pyelonephritis and lower urinary tract infection.

191091 CONFIRMED PATHOGEN, specify pathogen
Both of the following:
1. Clinical signs and symptoms of upper urinary tract disease
   and
2. Positive culture for specific organism in urine or blood

191092 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Clinical signs and symptoms of upper urinary tract disease
   and
2. Negative culture for specific organism in urine or blood

RUBELLA, CONGENITAL

191101 CONFIRMED PATHOGEN (WHO criteria)
Both of the following:
1. One of the following:
   a. Appropriate clinical syndrome in an infant less than (<) 12 months of age. For example, a combination of two or more of the following:
      1. Cataracts
      or
      2. Congenital glaucoma
      or
      3. Congenital heart disease
      or
      4. Hearing impairment
      or
      5. Pigmentary retinopathy
   or
   b. Both of the following:
      1. History of one of the following in a child:
         a. Purpura
         or
         b. Splenomegaly
         or
         c. Microcephaly
         or
         d. Mental retardation
         or
         e. Meningoencephalitis
         or
         f. Radiolucent bone disease
         or
         g. Jaundice with onset within 24-hours after birth
2. One of the following in a child:
   a. Cataracts
   or
   b. Congenital glaucoma
   or
   c. Congenital heart disease
   or
   d. Hearing impairment
   or
   e. Pigmentary retinopathy

and

2. Positive viral culture or nucleic acid amplification test for rubella virus or positive IgM antibody if IgM performed at less than (<) three months of age

RUBELLA, POSTNATAL

191111 CONFIRMED PATHOGEN
Both of the following:
1. Appropriate clinical syndrome of rash, fever, and lymphadenopathy
and
2. Positive IgM serology, or by virus isolation or nucleic acid amplification

SEPSIS, BACTERIAL - NOT CATHETER-ASSOCIATED

191121 CONFIRMED PATHOGEN, specify pathogen
Both of the following:
1. Systemic inflammatory response syndrome criteria as defined by two or more of the following:
   a. All of the following:
      1. Temperature greater than (>) 38°C or less than (<) 36°C
      and
      2. Heart rate greater than (>) 90 beats per minute (BPM)
      and
      3. Respiratory rate greater than (>) 20 breaths per minute
   or
   b. PaCO2 (<) 32 mm Hg; WBC greater than (>) 12,000 cells/mm³ (cubic millimeters) or less than (<) 4000 cells/mm³
   or
   c. Greater than (>) 10% (percent) immature (band) forms
and
2. Bacterial infection that is identified by culture, Gram stain, serology, nucleic acid amplification test, or antigen testing of blood or normally-sterile body fluid
191122

**NO PATHOGEN CONFIRMED (PROBABLE)**

**Both of the following:**
- Systemic inflammatory response syndrome (as defined above) with suspected infection
  and
- Negative culture, Gram stain, serology, nucleic acid amplification test, or antigen testing

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**SEPSIS, BACTERIAL-CATHETER-ASSOCIATED**

191131

**CONFIRMED PATHOGEN, specify pathogen**

**All of the following:**
- Systemic inflammatory response syndrome criteria as defined by two or more of the following:
  - **All of the following:**
    - Temperature greater than (> ) 38°C or less than (< ) 36°C
    and
    - Heart rate greater than (> ) 90 beats per minute (BPM)
    and
    - Respiratory rate greater than (> ) 20 breaths per minute
  or
  - Breaths/min, or PaCO2 less than (< ) 32 mm Hg; WBC greater than (> ) 12,000 cells/mm³ (cubic millimeters) or less than (< ) 4000 cells/mm³
  or
  - Greater than (> ) 10% (percent) immature (band) forms
  and
  - Bacterial infection that is identified by culture, Gram stain, serology, nucleic acid amplification test, or antigen testing of blood
  and
  - With an indwelling catheter in place

191132

**NO PATHOGEN CONFIRMED (PROBABLE)**

**All of the following:**
- Systemic inflammatory response syndrome (as defined above) with suspected infection
  and
- With an indwelling catheter in place
  and
- Negative culture, Gram stain, serology, nucleic acid amplification test, or antigen testing.

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**SALMONELLA SEPSIS, non-typhoid; includes nontyphoid enteric fever**

191141

**CONFIRMED PATHOGEN**

**Both of the following:**
- Appropriate clinical syndrome
  and
- Positive blood culture
SEPTIC ARTHRITIS (see also ARTHRITIS, SEPTIC)

190031 CONFIRMED PATHOGEN, specify pathogen
Appropriate clinical diagnosis confirmed by one of the following:
1. Positive Gram stain or culture of joint fluid
   or
2. Characteristic inflammatory joint fluid AND positive blood culture

190032 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Appropriate clinical diagnosis
2. Inflammatory joint fluid with both of the following:
   a. Negative Gram stain or culture of joint fluid and blood
   b. Specific antibacterial therapy initiated

SINUSITIS, ACUTE

191151 CONFIRMED PATHOGEN, specify pathogen
Both of the following:
1. Appropriate clinical findings for illness with duration less than (<) three months, with compatible imaging test results, if done
2. Specific identification of organism in sinus aspirate cultures or blood cultures

191152 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Appropriate clinical findings as above
2. Cultures negative or no testing performed

SINUSITIS, CHRONIC

191161 CONFIRMED PATHOGEN, specify pathogen
Both of the following:
1. Appropriate clinical findings for illness with duration greater than (>) three months, with compatible imaging test results, if done
2. Specific identification of organism in sinus aspirate cultures or blood cultures
191162 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Appropriate clinical findings for illness with duration greater than (>3) three months, with compatible imaging test results, if done
   and
2. Cultures negative or no testing performed

TONSILLITIS

191171 CONFIRMED PATHOGEN, specify pathogen
Both of the following:
1. Sore throat, inflamed tonsils on clinical exam
   and
2. Positive throat culture or other antigen detection test

191172 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Sore throat and inflamed tonsils on exam
   and
2. Culture or specific identification tests negative or not done.

TOXOPLASMA ENCEPHALITIS

191181 CONFIRMED PATHOGEN (WHO-4)
At least one of the following:
1. Histologic or nucleic acid amplification test evidence of Toxoplasma gondii in tissue obtained by brain biopsy or autopsy
   or
2. All of the following:
   a. Compatible clinical syndrome consisting of headache, seizure, neurologic deficits, or fever
      and
   b. Presence of characteristic mass lesion(s) on brain imaging study (CAT or MRI)
      and
   c. Response after a minimum of two weeks of antitoxoplasmosis therapy with documented clinical or radiographic improvement
      and
   d. Positive serologic test for Toxoplasma gondii
191182 NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Compatible clinical syndrome consisting of headache, seizure, neurologic deficits, or fever
   and
2. Presence of characteristic mass lesion(s) on brain imaging study (CT or MRI)
   and
3. Response after a minimum of two weeks of antitoxoplasmosis therapy with documented clinical or radiographic improvement

TOXOPLASMOsis, CONGENITAL

191191 CONFIRMED PATHOGEN
Both of the following:
1. Appropriate clinical syndrome in an infant
   and
2. Positive serology, ophthalmologic exam, or histopathology indicative of Toxoplasma gondii

191192 NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Appropriate clinical syndrome
   and
2. No documented positive serology, ophthalmologic exam, or histopathology indicative of Toxoplasma gondii
   and
3. No evidence of CMV or rubella infection

NOTE: See also TOXOPLASMOsis, CONGENITAL in the Neonatal section.

TRACHEITIS

191201 CONFIRMED PATHOGEN, specify pathogen
All of the following:
1. Appropriate clinical syndrome
   and
2. Endoscopy
   and
3. Etiology proven by positive culture of blood or tracheal culture

191202 NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Appropriate clinical syndrome
   and
2. Endoscopy
   and
3. Cultures of blood or trachea negative or not done
URINARY TRACT INFECTION
(NOTE that in children < five years of age, it may be difficult to distinguish pyelonephritis and lower urinary tract infection)

191211 CONFIRMED PATHOGEN, specify pathogen
Both of the following:
1. Appropriate clinical syndrome WITHOUT signs and symptoms of upper urinary tract disease.
2. Positive culture for specific organism in urine obtained by suprapubic tap or catheterization of bladder (for children less than five years old) or clean catch specimen (patients over five years old)

VAGINOSIS, BACTERIAL

191222 NO PATHOGEN CONFIRMED (PROBABLE)
A clinical syndrome defined by three of the following clinical criteria:
1. Homogeneous, white, non-inflammatory discharge that adheres
2. Clue cells on microscopic exam
3. Ph of vaginal fluid greater than (> ) 4.5
4. Fishy odor or vaginal discharge before or after addition of 10% (percent) KOH

VULVOVAGINITIS, TRICHOMONAS

191231 CONFIRMED
Both of the following:
1. Appropriate clinical syndrome
2. Confirmed by positive nucleic acid amplification test (e.g., PCR) or wet mount exam for Trichomonas vaginalis

191232 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Appropriate clinical syndrome
2. Specific tests negative or not done

NOTE: See code for VULVOVAGINITIS, CANDIDA under Candida
VARICELLA-ZOSTER VIRUS (VZV), CHICKENPOX, UNCOMPlicated, specify chickenpox, uncomplicated or varicella zoster virus primary disease, uncomplicated

191242 NO PATHOGEN CONFIRMED (PROBABLE)
Clinical diagnosis of uncomplicated chickenpox (VZV primary disease)

VARICELLA-ZOSTER VIRUS (VZV), CHICKENPOX, COMPLICATED (DISSEMINATED), specify chickenpox or varicella zoster virus primary disease and either complicated or disseminated in the description

191252 NO PATHOGEN CONFIRMED (PROBABLE)
Clinical diagnosis of complicated chickenpox with disseminated disease, including VZV pneumonia, encephalitis, or hepatitis

VARICELLA-ZOSTER VIRUS (VZV) - ZOSTER (SHINGLES, HERPES ZOSTER), UNCOMPlicated, specify zoster, herpes zoster, or shingles

191261 CONFIRMED PATHOGEN
All of the following:
1. Appropriate clinical diagnosis of rash typical of herpes zoster or shingles, first episode
   and
2. Without complications
   and
3. Proven by virus isolation, antigen detection, or nucleic acid amplification test from skin lesions

191262 NO PATHOGEN CONFIRMED (PROBABLE) (WHO-2)
All of the following:
1. Appropriate clinical diagnosis of rash typical of herpes zoster or shingles, first episode
   and
2. Without complications
   and
3. Diagnostic culture, nucleic acid amplification test, or antigen tests negative or not done

VARICELLA-ZOSTER VIRUS (VZV) - ZOSTER, (SHINGLES, HERPES ZOSTER), DISSEMINATED, specify zoster, herpes zoster, or shingles and disseminated in the description

191271 CONFIRMED PATHOGEN
All of the following:
1. Appropriate clinical diagnosis of rash typical of herpes zoster or shingles
   and
2. Signs of dissemination present, such as visceral involvement or cutaneous involvement with greater than (> ) two distinct dermatomes
   and
3. Proven by virus isolation, antigen detection or nucleic acid amplification test from skin lesions
CANDIDIASIS, OTHER, specify diagnosis
This includes disseminated candidemia, chronic cutaneous candidiasis, and invasive candidiasis. When reporting other candidiasis, use the following guidelines for confirmed and probable diagnoses:

199001 Confirmed diagnosis criteria may include:
1. A compatible clinical syndrome plus any diagnostic evidence that supports the diagnosis
or
2. There may be the diagnostic test confirmation of the diagnosis without a clinical syndrome. Examples of diagnostic tests are: Microbiologic, cytologic, or histologic evidence that supports the diagnosis or any currently-approved diagnostic test.

199002 Probable diagnosis criteria may include:
1. A compatible clinical syndrome
or
2. One or more test results from procedures generally accepted as supportive of the diagnosis in standard medical practice
or
3. Initiation or recommendation of specific therapy when appropriate

CMV SYNDROMES, OTHER, specify

199011 CONFIRMED (WHO-4 if Age greater than (>) 1 month) (this includes, but is not limited to, the following)
Hepatitis or cholangitis:
1. ALT or alkaline phosphatase significantly elevated above the study participant’s baseline values
and
2. Tissue biopsy demonstrating CMV by antigen, detection of viral nucleic acids (e.g., nucleic acid amplification test), or characteristic cytopathic changes

Radiculomyelopathy
Clinical presentation compatible with CMV end-organ disease including all of the following:
1. Decreased lower extremity strength and reflexes or syndrome consistent with a cord lesion present subacutely (over days to weeks)
and
2. Myelogram or MRI reveals no mass lesions but lower spinal nerve roots thickened
and
3. CMV positive culture in CSF or detection of CMV viral nucleic acids (e.g., nucleic acid amplification test) in CSF
CONNECTIVE TISSUE DISORDER, OTHER, specify diagnosis
Various syndromes including but not limited to:
  Systemic lupus erythematosus (SLE)
  Vasculitides, non-specific
  Sjögren’s syndrome
  Rheumatoid arthritis (also coded under ARTHRITIS)
  Connective tissue diseases not specified

When reporting other connective tissue disorders, use the following guidelines for confirmed and probable diagnoses:

199021 Confirmed diagnosis criteria may include:
  1. A compatible clinical syndrome plus any diagnostic evidence that supports the diagnosis or
  2. There may be the diagnostic test confirmation of the diagnosis without a clinical syndrome. Examples of diagnostic tests are: Microbiologic; cytologic, or histologic evidence that supports the diagnosis or any currently-approved diagnostic test.

199022 Probable diagnosis criteria may include:
  1. A compatible clinical syndrome or
  2. One or more test results from procedures generally accepted as supportive of the diagnosis in standard medical practice or
  3. Initiation or recommendation of specific therapy when appropriate

FUNGAL INFECTION, OTHER, specify diagnosis (other than Candida, cryptococcosis, histoplasmosis, coccidioidomycosis, paracoccidioidomycosis, blastomycosis, for which codes exist elsewhere)

199031 CONFIRMED PATHOGEN, specify pathogen
  All of the following:
  1. Compatible clinical syndrome and
  2. Histologic evidence of invasive disease and
  3. Positive culture or smear from a sterile tissue site, specify pathogen

199032 NO PATHOGEN CONFIRMED (PROBABLE)
  All of the following:
  1. Compatible clinical syndrome and
  2. Positive culture or smear from a non-sterile site and
  3. Specific antifungal treatment initiated or recommended

TOXOPLASMOSIS, OTHER, postnatal, non-encephalitic
CONFIRMED PATHOGEN

At least one of the following:

1. Histologic evidence of Toxoplasma gondii present in tissue or body fluid obtained by biopsy or aspirate (e.g., from lymph node, lung, or bronchoalveolar lavage)

2. Characteristic post-natal ophthalmologic findings diagnosed by an experienced ophthalmologist in an older child or adult without history of congenital toxoplasmosis
X. METABOLIC/ENDOCRINE DISORDERS

ADDISON’S DISEASE, specify primary or secondary (Also known as chronic adrenal insufficiency, hypocortisolism, or hypocorticism)
A rare endocrine disorder in which the adrenal glands do not produce enough steroid hormones (glucocorticoids and often mineralocorticoids)

200011 CONFIRMED
Identified by an abnormal cosyntropin test (i.e., ACTH is administered and there is a subnormal increase in plasma cortisol between baseline and 60 minutes post challenge)

200012 PROBABLE
Low serum cortisol levels, particularly in the early morning, accompanied by high plasma levels of ACTH

CUSHING’S SYNDROME, also called hyperadrenocorticism or hypercorticism
An endocrine disorder caused by high levels of cortisol (hypercortisolism) in the blood. This can be caused by taking glucocorticoid drugs, by adrenal tumors that produce cortisol, or, in the case of Cushing disease, by a pituitary tumor that produces large amounts of ACTH, which in turn elevates cortisol.

200021 CONFIRMED
At least one of the following:
1. An abnormal 24-hour urinary measurement of free cortisol
   or
2. An abnormal dexamethasone suppression test (administration of dexamethasone followed by determination of cortisol and ACTH levels)

200022 PROBABLE
Both of the following:
1. Compatible clinical signs such as: rapid weight gain, particularly of the trunk and face with sparing of the limbs (central obesity), growth of fat pads along the collar bone and on the back of the neck (buffalo hump), and a round or “moon” face, thinning weakening of the skin, easy bruising, and, in children, decreased growth rate, hypertension, and glucose intolerance
   and
2. No urinary cortisol or dexamethasone suppression testing performed
DIABETES, TYPE 1
Type 1 diabetes (formerly known as "childhood," "juvenile," or "insulin-dependent" diabetes) occurs in both children and adults. Insulin treatment is mandatory for survival.

200031 CONFIRMED
According to guidelines set forth by the American Diabetes Association, Standards of Medical Care in Diabetes (2010), diabetes (in the non-pregnant state) is present if:
1. The fasting plasma glucose (FPG) is greater than or equal to ($\geq$) 126 mg/dL (milligrams/deciliter) or 7.0 mmol/L (millimoles/liter). Fasting is defined as no caloric intake for at least eight hours.

or
2. A 2-hour plasma glucose is greater than or equal to ($\geq$) 200 mg/dL (milligrams/deciliter) or 11.1 mmol/L (millimoles/liter) during an oral glucose tolerance test (OGTT). The test should be performed as described by the World Health Organization using a glucose load containing the equivalent of 75 grams of anhydrous glucose dissolved in water.

or
3. A casual (random) blood glucose concentration is greater than or equal to ($\geq$) 200 mg/dL (milligrams/deciliter) or 11.1 mmol/L (millimoles/liter) in a person with symptoms of hyperglycemia or hyperglycemic crisis.

or
4. Glycosylated hemoglobin (A1c) of greater than or equal to ($\geq$) 6.5% (percent). The test should be performed in a laboratory using a method that is National Glycohemoglobin Standardized Program (NGSP) certified and standardized to the Diabetes Control and Complications Trial (DCCT) assay.

DIABETES, TYPE 2
Type 2 diabetes accounts for approximately 90–95% (percent) of those with diabetes, and has previously been referred to as non-insulin-dependent diabetes, type II diabetes, or adult-onset diabetes.

200041 CONFIRMED
According to guidelines set forth by the American Diabetes Association, Standards of Medical Care in Diabetes (2010), diabetes (in the non-pregnant state) is present if:
1. The fasting plasma glucose (FPG) is greater than or equal to ($\geq$) 126 mg/dL (milligrams/deciliter) or 7.0 mmol/L (millimoles/liter). Fasting is defined as no caloric intake for at least eight hours.

or
2. A 2-hour plasma glucose is greater than or equal to ($\geq$) 200 mg/dL (milligrams/deciliter) or 11.1 mmol/L (millimoles/liter) during an oral glucose tolerance test (OGTT). The test should be performed as described by the World Health Organization using a glucose load containing the equivalent of 75 g anhydrous glucose dissolved in water.

or
3. A casual (random) blood glucose concentration is greater than or equal to ($\geq$) 200 mg/dL (milligrams/deciliter) or 11.1 mmol/L (millimoles/liter) in a person with symptoms of hyperglycemia or hyperglycemic crisis.

or
4. Glycosylated hemoglobin (A1c) of greater than or equal to ($\geq$) 6.5% (percent). The test should be performed in a laboratory using a method that is National Glycohemoglobin
DIABETES, PRE-GESTATIONAL
Refers to diabetes (Type 1 or 2) that is pre-existing when a woman becomes pregnant

200051 CONFIRMED
According to guidelines set forth by the American Diabetes Association, Standards of Medical Care in Diabetes (2010), diabetes (in the non-pregnant state) is present if:
1. The fasting plasma glucose (FPG) is greater than or equal to (≥) 126 mg/dL (milligrams/deciliter) or 7.0 mmol/L (millimoles/liter). Fasting is defined as no caloric intake for at least eight hours.

or
2. A 2-hour plasma glucose is greater than or equal to (≥) 200 mg/dL (milligrams/deciliter) or 11.1 mmol/L (millimoles/liter) during an oral glucose tolerance test (OGTT). The test should be performed as described by the World Health Organization using a glucose load containing the equivalent of 75 g anhydrous glucose dissolved in water.

or
3. A casual (random) blood glucose concentration is greater than or equal to (≥) 200 mg/dL (milligrams/deciliter) or 11.1 mmol/L (millimoles/liter) in a person with symptoms of hyperglycemia or hyperglycemic crisis.

or
4. Glycosylated hemoglobin (A1c) of greater than or equal to (≥) 6.5 % (percent). The test should be performed in a laboratory using a method that is National Glycohemoglobin Standardized Program (NGSP) certified and standardized to the Diabetes Control and Complications Trial (DCCT) assay.

IMPAIRED GLUCOSE TOLERANCE (IGT)

200061 CONFIRMED
The American Diabetes Association recognizes an intermediate group of subjects whose glucose levels, although not meeting criteria for diabetes, are nevertheless too high to be considered normal.

This group is defined as at least one of the following:
1. Impaired fasting glucose (IFG): defined as having a fasting plasma glucose (FPG) levels greater than or equal to (≥) 100 mg/dL (milligrams/deciliter), but less than (<) 126 mg/dL (milligrams/deciliter).

or
2. Impaired glucose tolerance (IGT) defined as having a 2-hour plasma glucose values during an oral glucose tolerance test (OGTT) of greater than or equal to (≥) 140 mg/dL (milligrams/deciliter), but less than (<) 200 mg/dL (milligrams/deciliter).
IMPAIRED FASTING GLUCOSE (IFG)

200071 CONFIRMED
The American Diabetes Association recognizes an intermediate group of subjects whose glucose levels, although not meeting criteria for diabetes, are nevertheless too high to be considered normal.

This group is defined as at least one of the following:
1. Impaired fasting glucose (IFG): defined as having a fasting plasma glucose (FPG) levels greater than or equal to ($\geq$) 100 mg/dL (milligrams/deciliter), but less than (<) 126 mg/dL.
   or
2. Impaired glucose tolerance (IGT) defined as having a 2-hour plasma glucose values during an oral glucose tolerance test (OGTT) of greater than or equal to ($\geq$) 140 mg/dL (milligrams/deciliter), but less than (<) 200 mg/dL.

GESTATIONAL DIABETES (GDM)
Defined as any degree of glucose intolerance with initial onset during pregnancy. The definition applies regardless of whether insulin or only diet modification is used for treatment, or whether the condition persists after pregnancy. It does not exclude the possibility that unrecognized glucose intolerance may have antedated or begun concomitantly with the pregnancy.

200081 CONFIRMED
According to guidelines set forth by the American Diabetes Association, GDM is present if two or more of the following venous plasma concentrations during an oral glucose tolerance test (OGTT) (100 milligram load) are exceeded:
1. Fasting greater than or equal to ($\geq$) 95 mg/dL (milligrams/deciliter)
2. 1-hour greater than or equal to ($\geq$) 180 mg/dL
3. 2-hour greater than or equal to ($\geq$) 155 mg/dL
4. 3-hour greater than or equal to ($\geq$) 140 mg/dL

DIABETES INSIPIDUS (DI)
A condition occurring in both children and adults that is characterized by excessive thirst and excretion of large amounts of severely diluted urine, with reduction of fluid intake having no effect on the latter. There are several different types of DI, each with a different cause. The most common type is neurogenic or central DI, caused by a deficiency of arginine vasopressin (AVP), also known as anti-diuretic hormone (ADH). The other type of DI is nephrogenic diabetes insipidus, which is caused by an insensitivity of the kidneys to ADH and which can also be a side artifact of certain medications.

Diagnosis is usually made with a water deprivation test, in which a person is not allowed to drink; serial samples are obtained for measurements of urine osmolality and serum osmolality and serum sodium. Weight is also monitored. Diagnosis is made if the patient becomes hyperosmolar while still having a dilute urine.
CENTRAL DIABETES INSIPIDUS (DI)

200091  CONFINED
   o  All of the following:
      •  Hyperosmolarity with dilute urine
        and
      2.  Decreased level of vasopressin
        and
      3.  Response to desmopressin with decreased urine output and increased urine osmolality

NEPHROGENIC DIABETES INSIPIDUS (DI)

200101  CONFINED
   All of the following:
   1.  Hyperosmolarity with dilute urine
        and
   2.  Increased level of vasopressin
        and
   3.  No response to desmopressin. No decrease in urine output.

DIABETIC KETOACIDOSIS (DKA)

200111  CONFINED
   Both of the following:
   1.  Compatible clinical symptoms including vomiting, dehydration, deep gasping breathing, confusion, and occasionally coma in an individual with hyperglycemia
        and
   2.  Blood or urine test results that demonstrate ketoacidosis

FAILURE TO THRIVE (FTT)
Denotes the presence of poor weight gain (with or without poor linear growth) over an extended period of time in infancy or childhood.

200121  CONFINED
   FTT can be defined as at least one of the following:
   1.  Weights consistently less than (<) 3rd - 5th percentiles for age
        or
   2.  A persistent decrease in weight of more than 10% (percent) of baseline
        or
   3.  A decrease two major growth percentile lines over time
GROWTH HORMONE DEFICIENCY (GHD)

200131 CONFIRMED
All of the following:
Children:
1. Short stature
   and
2. Decreased levels of growth hormone

Adults
1. Diminished lean body mass
   and
2. Poor bone density
   and
3. Loss of strength
   and
4. Decreased growth hormone

200132 PROBABLE
Suspected clinical syndrome as described above, no measurement of growth hormone

HYPERTHYROIDISM
A condition of both children and adults in which the thyroid gland overproduces the thyroid hormones, thyroxine (T4), and tri-iodothyronine (T3). It is most often due to Graves’ disease, a condition in which antibodies to the thyroid-stimulating hormone receptor overstimulate the gland. Thyroid hormones are critical to normal cellular functions.

In excess, they both overstimulate metabolism and exacerbate the effects of the sympathetic nervous system, causing "speeding up" of various bodily systems.

200141 CONFIRMED
Both of the following:
1. Clinical syndrome of fast heart beat, palpitations, tremors, anxiety, difficulty concentrating leading to poor school performance (if in school), diarrhea, and weight loss
   and
2. Increased levels of thyroxine (T4) and tri-iodothyronine (T3)

200142 PROBABLE
Clinical syndrome of fast heart beat, palpitations, tremors, anxiety, difficulty concentrating leading to poor school performance (if in school), diarrhea, and weight loss. Laboratory testing not done.
HYPOTHYROIDISM
A condition that can occur at all ages in which the thyroid gland underproduces tri-iodothyronine (T3) and thyroxine (T4). In infants, it is most likely due to aplasia, hypoplasia, or ectopia whereas, in older children and adults, due to autoimmune destruction. Thyroid hormones function as stimuli to metabolism and are critical to normal cellular functions. Affected infants most notably are at risk for mental retardation if not properly diagnosed and treated. In childhood, the most common manifestations are goiter, slow height velocity, and mild excess rate of weight gain. Children and adults also are at risk for fatigue, cold intolerance, dry skin, constipation, bradycardia, and, in females, changes in menstrual pattern.

200151 CONFIRMED
Both of the following:
1. At least one of the following:
   Childhood
   a. Goiter
   or
   b. Slow height velocity
   or
   c. Mild excess rate of weight gain
   or
   d. Fatigue
   or
   e. Cold intolerance
   or
   f. Dry skin
   or
   g. Constipation
   or
   h. Bradycardia

   Adults
   a. Goiter
   or
   b. Fatigue
   or
   c. Cold intolerance
   or
   d. Dry skin
   or
   e. Constipation
   or
   f. Bradycardia
   or
   g. Weight gain
   or
   h. Changes in menstrual pattern for females

and
2. Decreased levels thyroxine (T4) and tri-iodothyronine (T3) or increased TSH

200152 PROBABLE
1. At least one of the following:
   Childhood
   a. Goiter
      or
   b. Glow height velocity
      or
   c. Mild excess rate of weight gain
      or
   d. Fatigue
      or
   e. Cold intolerance
      or
   f. Dry skin
      or
   g. Constipation
      or
   h. Bradycardia

   Adults
   a. Goiter
      or
   b. Fatigue
      or
   c. Cold intolerance
      or
   d. Dry skin
      or
   e. Constipation
      or
   f. Bradycardia
      or
   g. Weight gain
      or
   h. Changes in menstrual pattern for females

HYPERCHOLESTEROLEMIA

200161 CONFIRMED
Abnormally elevated levels of total and LDL cholesterol

NOTE: When measuring serum lipid profiles, care must be taken to be sure the patient has fasted a minimum of 12 hours at the time of the measure and that age-appropriate normal reference values are used for comparison.
HYPERTRIGLYCERIDEMIA

200171 CONFIRMED
Abnormally elevated levels of triglycerides.

NOTE: When measuring serum lipid profiles, care must be taken to be sure the patient has fasted a minimum of 12 hours at the time of the measure and that age-appropriate normal reference values are used for comparison.

HYPOGONADISM, specify gender

200181 CONFIRMED
Reduced gonadal (ovaries or testes) function, i.e., reduced or absent production of sex hormones and gametes (eggs or sperm) in response to cyclical production of hypothalamic gonadotropin-releasing hormone (GnRH) and, subsequently, pituitary luteinizing hormone (LH) and follicle-stimulating hormone (FSH).

LACTIC ACIDOSIS, ASYMPTOMATIC

200191 CONFIRMED
The persistent occurrence of a lactate level greater than (> 1.5 times normal on at least two separate blood draws.

LACTIC ACIDOSIS, SYMPTOMATIC

200201 CONFIRMED
Both of the following:
1. Potentially life-threatening condition; elevated plasma lactate level and pH less than (<) 7.35 with an increased anion gap
   and
2. Symptoms which may include fatigue, abdominal pain, gastric discomfort, or increased liver function tests

LIPODYSTROPHY

This syndrome is identified by abnormal fat distribution which manifests as fat accumulation (lipoaccumulation) or fat loss (lipoatrophy). Lipodystrophy can be caused by genetic syndromes or by certain medications used to treat HIV-infected patients.

FAT ACCUMULATION (LIPOOACCUMULATION), specify site and either fat accumulation or lipoaccumulation

200211 CONFIRMED - refer to the body area being reported for specific criteria

200212 PROBABLE - refer to the body area being reported for specific criteria

Symptoms due to fat accumulation in various places occur following the initiation or change of antiretroviral therapy. Lipocaccumulation may manifest as increasing abdominal girth with an increasing belt or waist size which may be accompanied by complaints of bloating or distension; fat
accumulation in the back of the neck or increasing neck size; increasing breast size due to fatty tissue infiltration which may be accompanied by complaints of breast pain; and other new fat accumulations either circumscribed (lipomas) or general such as increase in chest size in absence of breast enlargement. In reporting fat accumulation, the body area involved needs to be specified. In males, unilateral or bilateral breast enlargement may occur, occasionally with nodular lesions. This syndrome should be distinguished from gynecomastia, which is an increase in breast size due to increased breast tissue, which is not the same as fat accumulation of the breast.

BODY AREA(S) INVOLVED:
1) ABDOMINAL OR TRUNCAL OBESITY
Criteria for reporting this body site:
CONFIRMED
Cross-sectional: Self-reported increase of abdominal girth; increasing belt or waist size (may be accompanied by complaint of bloating, distention) plus waist-to-hip ratio (WHR) greater than (> ) 0.95 (Males) or 0.85 (Females)
Longitudinal: Measured increase in waist circumference of 2.5 cm (centimeter) or 1” (inch), or 5% (percent) increase in WHR, sagittal diameter, or abdominal fat (by paired MRI, DEXA, or CT measurements obtained under identical, controlled conditions) in the past 12 months

PROBABLE
Self-report of increasing abdominal girth; increasing belt or waist size (may be accompanied by complaints of bloating, distention)

2) DORSOCERVICAL FAT PAD ENLARGEMENT
Criteria for reporting this body site:
CONFIRMED
Cross-sectional: Physical findings consistent with accumulations of fat in dorsocervical area
Longitudinal: Measured increase in neck circumference of 1.5 cm (centimeter) or .5” (inch) in the past 12 months

PROBABLE
Self-report of increasing size of dorsocervical region; may be accompanied by increasing shirt neck size or inability to button shirts

3) BREAST ENLARGEMENT (BOTH GENDERS), specify gender
Criteria for reporting this body site:
CONFIRMED
Cross-sectional: Self-reported increase in bra size or shirt/blouse size to accommodate increasing breast size plus physical findings consistent with enlarged breasts due to increase in fat deposition (NOTE: gynecomastia is an increase in breast tissue, a distinct syndrome and finding)
Longitudinal: Measured increase in chest circumference of 5% (percent) in past 12 months
PROBABLE
Self-report of increasing bra size or shirt/blouse size to accommodate increasing breast size; may be accompanied by complaints of breast pain

FAT LOSS (LIPOATROPHY), specify site and either fat loss or lipoatrophy

NOTE: If multiple sites are affected, e.g., face, extremities and buttocks, specify generalized or disseminated.

200221 CONFIRMED - refer to the body area being reported for specific criteria

BODY AREA(S) INVOLVED:
1) FACE
Criteria for reporting this body site:
CONFIRMED
Study participant may report “sunken cheeks” or “drawn face” or indicate that family members or friends have noticed such changes since initiation or change of antiretroviral therapy, although it may be induced by HIV itself. The loss of facial tissue should be just proximal to the nasolabial fold (the area of the buccal fat pad, the largest fat deposit in the face). Facial lipoatrophy is noted on exam.

2) EXTREMITIES
Criteria for reporting this body site:
CONFIRMED
Study participant reports that pants/slacks are progressively fitting more loosely through the thighs, new onset of looseness of watch or wristbands, and awareness the extremities appear thinner since the initiation or change of antiretroviral therapy. The relationship is strengthened by reporting of awareness that veins in the extremities appear more prominent.
On exam, extremities appear thin and veins prominent.

3) BUTTOCKS
Criteria for reporting this body site:
CONFIRMED
Self-reported change in the buttocks in which there is a perception of loss of volume in the subgluteal region. Loss of firmness is by itself not diagnostic as it could be because of muscle atrophy.
Lipoatrophy noted in buttocks on exam.

PRECOCIOUS PUBERTY
The unusually early onset of puberty, the process of sexual maturation triggered by the brain or exogenous hormones, which usually begins in late childhood and ultimately results in reproductive maturity and completion of linear growth

200231 CONFIRMED
In general, precocious puberty can be defined by:
In girls: Breast development (Tanner stage 2 or greater) before eight years of age
In boys: Testicular enlargement (greater than (>)) three cc (cubic centimeters)) before nine years of age
MODERATE ACUTE MALNUTRITION (MAM)

200241 CONFIRMED
At least one of the following:
1. Weight-for-height measurement of less than (<) two standard deviations (SD) below the mean (from NCHS/WHO reference curves as appropriate)
   or
2. Mid-Upper-Arm Circumference (MUAC) less than (<) 125 mm (millimeters) in children age one - five years

SEVERE ACUTE MALNUTRITION (SAM)

200251 CONFIRMED
At least one of the following:
1. Weight-for-height measurement less than (<) three standard deviations (SD) below the mean (from NCHS/WHO reference curves as appropriate)
   or
2. The presence of bilateral pitting oedema of nutritional origin
   or
3. A Mid-Upper-Arm Circumference (MUAC) less than (<) 115 mm (millimeters) in children age one - five years

PRECOCIOUS ADRENARCHE

200261 CONFIRMED
Both of the following:
1. In girls: The development of pubic hair before the age of eight years with or without axillary hair and apocrine odor
   In boys: The development of pubic hair before the age of nine years with or without axillary hair and apocrine odor
   and
2. No other signs of sexual development

PRECOCIOUS THELARCHE

200271 CONFIRMED
Both of the following:
1. Breast development in girls younger than eight years of age
   and
2. No other signs of sexual development
PAROTID ENLARGEMENT

200282 PROBABLE
Clinical descriptors: enlarged parotid glands, usually bilateral
Patient-reported symptoms: Usually asymptomatic enlargement; may report xerostomia (perception of dry mouth)
Patient-reported duration: Usually long-standing

NOTE: See also PAROTID ENLARGEMENT in the ORAL DISORDERS section.

SALIVARY HYPOFUNCTION (HYPOSECRETION), specify either salivary hypofunction or salivary hyposcretion

200291 CONFIRMED
Defined as unstimulated whole salivary flow rate less than (<) 2.5 mL (milliliters) per five minutes (0.5mL/min)

ADRENAL INSUFFICIENCY, OTHER

209001 CONFIRMED diagnosis criteria may include:
1. A compatible clinical syndrome plus any diagnostic evidence that supports the diagnosis
   or
2. There may be the diagnostic test confirmation of the diagnosis without a clinical syndrome. Examples of diagnostic tests are: Microbiologic; cytologic, or histologic evidence that supports the diagnosis or any currently-approved diagnostic test.

209002 Probable diagnosis criteria may include:
1. A compatible clinical syndrome
   or
2. One or more test results from procedures generally accepted as supportive of the diagnosis in standard medical practice
   or
3. Initiation or recommendation of specific therapy when appropriate

NEW FAT ACCUMULATION, OTHER, must specify location

209011 CONFIRMED
Cross-sectional: Self-report of new fat accumulation plus physical findings consistent with lipoma(s) or lipomatosis (multiple fat accumulations or one greater than (> two cm (centimeter))

209012 PROBABLE
Self-report of new regional circumscribed accumulation of fat; increase in neck size in absence of dorsocervical fat pad enlargement; increase in chest size in absence of breast enlargement
XI. MITOCHONDRIAL DISORDERS

Mitochondrial disorders are a heterogeneous group of conditions associated with failure of the mitochondria. These organelles are specialized compartments present in every cell of the body except red blood cells. Mitochondrial disorders present complex diagnostic problems whose presentations are variable and nonspecific. The same phenotypic disorder may have more than one genetic causative abnormality; conversely the same genetic abnormality may be associated with more than one phenotype. Adding to difficulties in clinical recognition, there is no single biologic marker that can serve as the gold standard for diagnosis. Inherited mitochondrial diseases primarily affect children but adult onset may occur. Mitochondrial abnormalities have been proposed to be a consequence of exposure to nonnucleoside reverse transcriptase inhibitor (NRTI) exposure in HIV-infected children and adults.

Please note that great care is required when applying one of the following diagnoses due to the overlapping of symptoms among these disorders. Syndromes listed below are grouped according to the primary organ system involved with other associated findings also listed.

BARTH SYNDROME

210011 CONFINED
Cardiomyopathy, mitochondrial myopathy, short stature, and cyclic neutropenia

ENCEPHALOMYOPATHY

210021 CONFINED
Isolated or combined symptoms and signs of encephalopathy, seizures, hypotonia, or ophthalmopathy

GROWTH RETARDATION, AMINO ACIDURIA, CHOLESTASIS, IRON OVERLOAD, LACTIC ACIDOSIS, AND EARLY DEATH (GRACILE)

210031 CONFINED
Presents in infancy with severe growth retardation, lactic acidosis, hepatic hemosiderosis, increased ferritin, and death within the first few months of life

KEARNS-SAYRE SYNDROME

210041 CONFINED
Development of paresis of extraocular muscles and bilateral ptosis along with pigmentary retinopathy. Onset is usually before age 20 years and can be associated with short stature, cerebellar ataxia, cardiac conduction defects, or cognitive deficits/mental retardation.
LEBER HEREDITARY OPTIC NEUROPATHY (LHON)

210051  CONFIRMED
Delayed bilateral loss of vision which could lead to total blindness due to degeneration of the optic nerve. Early signs include localized collection of distended blood capillary vessels around the start of the optic nerve. May also be associated with mild dementia, ataxia, spasticity, peripheral neuropathy, and heart conduction defects.

LEIGH SYNDROME (SUBACUTE NECROTIZING ENCEPHALOMYELOPATHY)

210061  CONFIRMED
Usually presents in infancy or early childhood and is characterized by developmental delay or psychomotor regression, ataxia, dystonia, external ophthalmoplegia, seizures, lactic acidosis, vomiting, and weakness. Erratic breathing patterns and respiratory failure are common. Brain MRI shows abnormal white matter signal in the putamen, basal ganglia, and brain stem on T2 and FLAIR sequence images consistent with necrotizing lesions.

LETHAL INFANTILE MITOCHONDRIAL DISEASE (CONGENITAL MYOPATHY)

210071  CONFIRMED
Presents in infancy with marked hypotonia, respiratory muscle weakness, and feeding difficulty. Can be associated with renal dysfunction (Fanconi syndrome).

MATERNALLY INHERITED DEAFNESS AND DIABETES (MIDD)

210081  CONFIRMED
Defective insulin secretion, sensorineural hearing loss with onset in adulthood. May also demonstrate macular retinal dystrophy, myopathy, cardiac disorders, gestational diabetes, renal disease, particularly focal segmental glomerular sclerosis, short stature, and gastrointestinal disease.

MITOCHONDRIAL ENCEPHALOPATHY WITH LACTIC ACIDOSIS AND STROKE-LIKE EPISODES (MELAS)

210091  CONFIRMED
Usually presents in childhood after normal infant development. Relapsing/remitting course of stroke-like episodes that result in hemiparesis, hemianopia, or cortical blindness and dementia. Other symptoms include focal or generalized seizures, recurrent migraine-like headaches, vomiting, short stature, hearing loss, and muscle weakness.

MITOCHONDRIAL NEUROGASTROINTESTINAL ENCEPHALOPATHY (MNGIE)

210101  CONFIRMED
Severe gastrointestinal dysmotility and cachexia, ptosis, ophthalmoplegia or ophthalmoparesis without diplopia, symmetric polyneuropathy, and asymptomatic leukoencephalopathy. Symptoms of the neuropathy include paresthesia, pain, and distal weakness. Onset is usually before age 20 years.
MYOClonic epilepsy with ragged red fibers (merrF)

210111 CONFIRMED
Development of myoclonus is typically the first symptom, and is associated with generalized epilepsy, ataxia, myopathy. Additional features can include dementia, optic atrophy, bilateral deafness, peripheral neuropathy, spasticity, lipomatosis, or cardiomyopathy with wolff-Parkinson-white syndrome. Presentation in childhood after a normal early development is typical.

myopathy, infantile

210121 CONFIRMED
Following normal early development until one year of age, weakness appears and worsens rapidly, causing respiratory failure and death typically within a few years.

myopathy, infantile /hepatopathy

210131 CONFIRMED
Enlarged liver and intractable liver function, myopathy, and severe lactic acidosis. Death within the first year.

myopathy, isolated with or without exercise intolerance or myalgia

210141 CONFIRMED
Proximal myopathy which may be associated with fatigue and exercise induced myalgia and myoglobinuria.

neuropathy, ataxia and retinitis pigmentosa (narp)

210151 CONFIRMED
Developmental delay, sensory polyneuropathy, ataxia, pigmentary retinopathy, muscle weakness, epilepsy, and dementia. Onset is usually late childhood or early adult.

ophthalmoplegia, chronic progressive external

210161 CONFIRMED
Development of paresis of extraocular muscles along with bilateral ptosis which generally begins in early adulthood and is slowly progressive. May be associated with mild proximal myopathy.

pearson syndrome

210171 CONFIRMED
Severe anemia, ring sideroblasts in the bone marrow, neutropenia, thrombocytopenia, and exocrine pancreatic insufficiency.
XII. MYCOBACTERIAL INFECTIONS

MYCOBACTERIUM TUBERCULOSIS LATENT

220011  CONFIRMED PATHOGEN
Both of the following:
1.  At least one of the following:
   a.  Positive tuberculin skin test (TST) defined by greater than or equal to (≥) five mm induration for HIV-infected persons, or greater than or equal to (≥)10 mm induration for HIV-uninfected persons
   or
   b.  Positive interferon gamma release assay (IGRA) test

and
2.  No clinical, bacteriologic, or radiographic evidence of active tuberculosis

MYCOBACTERIUM TUBERCULOSIS PULMONARY

220021  CONFIRMED PATHOGEN
All of the following:
1.  Compatible clinical symptoms (e.g., cough-no duration, hemoptysis, shortness of breath, chest pain, weight loss, fever, or night sweats) with or without abnormal chest x-ray, chest CT scan or other chest imaging (e.g., hilar lymphadenopathy, paratracheal lymphadenopathy, alveolar consolidation, miliary pattern, lung parenchymal breakdown/cavitation, or Ghon focus)

and
2.  Sputum, bronchial alveolar lavage, pleural fluid, pleural tissue, lung tissue culture or gastric acid lavage or nucleic acid amplification test positive for Mycobacterium tuberculosis or nucleic acid amplification test positive for MTB Complex.

220022  NO PATHOGEN CONFIRMED (PROBABLE)
AGE < 2 YEARS
All of the following:
1.  At least one of the following:
   a.  Non-specific signs and symptoms are present, such as productive or non-productive chronic cough, hemoptysis, fever, night sweats, or anorexia or weight loss (child with weight less than (<) 3rd percentile for age or a decrease in weight that has crossed two major growth percentiles since the last documented weight)
   or
   b.  abnormal chest x-ray (e.g., hilar adenopathy, paratracheal lymphadenopathy, alveolar consolidation, miliary pattern, parenchymal breakdown/cavitation, or Ghon focus).

and
2.  History of contact with an individual with known or suspected tuberculosis (TB)

and
3. **At least one of the following:**
   a. No response achieved with standard broad spectrum antibiotic treatment
   or
   b. Specific antituberculogous therapy initiated

**AGE ≥ 2 YEARS**

**All of the following:**

1. Compatible clinical symptoms (e.g., cough, hemoptysis, shortness of breath, chest pain, weight loss, fever, or night sweats of greater than or equal to \((\geq)\) two weeks duration)

   and

2. **At least one of the following:**
   a. Positive sputum smear for acid fast bacilli (AFB)
   or
   b. Either abnormal chest x-ray, chest CT scan or other chest imaging (e.g., hilar lymphadenopathy, paratracheal lymphadenopathy, alveolar consolidation, miliary pattern, parenchymal breakdown/cavitation, or Ghon focus)
   or
   c. Evidence of granulomata with organisms positive for AFB or caseating granulomata on lung tissue biopsy or at autopsy
   or
   d. Positive tuberculin skin test (TST) \((\geq)\) five mm) or interferon gamma release assay (IGRA)

   and

3. Without concurrent illness that would explain the findings

**MYCOBACTERIUM TUBERCULOSIS EXTRAPULMONARY**

This definition refers to extrapulmonary tuberculosis where a specific site or sites of organ involvement can be identified and the site or sites must be specified: abdominal, bone/joint, extrathoracic lymphadenitis, pericarditis, etc. Miliary/disseminated tuberculosis where there is diffuse involvement of organ sites is included in this definition set and should be specified. There is no age distinction for this diagnosis.

**220031 CONFIRMED PATHOGEN**

**Both of the following:**

1. Systemic illness usually with prolonged fever, night sweats or weight loss plus clinical features of organs involved, such as sterile pyuria, pericarditis, ascites, pleuraleffusion, meningitis, arthritis, orchitis, pericarditis, enteritis, extrathoracic lymphadenitis, or osteitis.

   and

2. Positive culture for Mycobacterium tuberculosis or nucleic acid amplification test positive for Mycobacterium tuberculosis or Mycobacterium tuberculosis complex.
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APPENDIX 100 - Diagnoses Appendix

220032 NO PATHOGEN CONFIRMED (PROBABLE)

Both of the following:
1. Systemic illness usually with prolonged fever, night sweats or weight loss plus clinical features of organs involved, such as sterile pyuria, pericarditis, ascites, pleural effusion, meningitis, arthritis, orchitis, pericarditis, enteritis, extrathoracic lymphadenitis or osteitis or evidence of granulomata with organisms positive for AFB or caseating granulomata on tissue biopsy from relevant site
2. Specific antituberculous therapy initiated or recommended

---

MONORESISTANT MYCOBACTERIUM TUBERCULOSIS

220041 CONFIRMED PATHOGEN

Both of the following:
1. Confirmed tuberculosis diagnosis (refer to CONFIRMED PATHOGEN MYCOBACTERIUM TUBERCULOSIS in relevant section)
2. M. tuberculosis isolate resistant to isoniazid or rifampin

---

MULTIDRUG RESISTANT MYCOBACTERIUM TUBERCULOSIS (MDR-TB)

220051 CONFIRMED PATHOGEN

Both of the following:
1. Confirmed tuberculosis diagnosis (refer to CONFIRMED PATHOGEN MYCOBACTERIUM TUBERCULOSIS in relevant section)
2. M. tuberculosis isolate resistant to isoniazid and rifampin

---

EXTENSIVELY DRUG RESISTANT TUBERCULOSIS (XDR-TB)

220061 CONFIRMED PATHOGEN

Both of the following:
1. Confirmed tuberculosis diagnosis (Refer to CONFIRMED PATHOGEN MYCOBACTERIUM TUBERCULOSIS in relevant section.)
2. M. tuberculosis isolate resistant to isoniazid, rifampin, any fluoroquinolone drug and at least one of the three second-line injectable drugs such as amikacin, kanamycin, or capreomycin.
MYCOBACTERIUM AVIUM COMPLEX (MAC)

220071 CONFIRMED PATHOGEN
At least one of the following:
1. Positive cultures or nucleic acid amplification test for MAC from a normally sterile site (blood, bone marrow, lymph node, liver, cerebrospinal fluid or other normally sterile body fluid, tissue or organ)

Or
2. Positive culture or nucleic acid amplification test for MAC from bronchopulmonary, gastrointestinal, skin surface or other non-sterile site(s) as the only pathogen, and histopathologic confirmation of AFB in tissue specimen(s) from which MAC was identified

220072 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. A clinical syndrome compatible with MAC, consisting of at least one of the following:
   a. Persistent fever greater than or equal to (≥) 38°C (centigrade) for more than one week
   Or
   b. Night sweats
   Or
   c. Diarrhea
   Or
   d. Weight loss or wasting
   Or
   e. Radiographically documented pulmonary infiltrates
   Or
   f. Hepatomegaly
   Or
   g. Splenomegaly
   Or
   h. Anemia
   Or
   i. Elevated alkaline phosphatase
   Or
   j. Cultures negative, not done, or repeatedly positive cultures or nucleic acid amplification test for MAC from the same non-sterile body site (e.g., bronchopulmonary, gastrointestinal, skin surface or other non-sterile sites) without histopathologic confirmation of AFB in tissue

And
2. Specific treatment initiated or recommended for MAC
MYCOBACTERIUM BOVIS

220081 CONFIRMED PATHOGEN
At least one of the following:
1. Positive cultures or nucleic acid amplification test for M. bovis from a normally-sterile site (blood, bone marrow, lymph node, liver, cerebrospinal fluid or other normally sterile body fluid, tissue, or organ)
   or
2. Positive cultures or nucleic acid amplification test for M. bovis from bronchopulmonary, gastrointestinal, skin surface, or other non-sterile site(s) as the only pathogen, and histopathologic confirmation of AFB in tissue specimen(s) from which M. bovis was identified

220082 NO PATHOGEN CONFIRMED (PROBABLE)
Both of the following:
1. Repeatedly positive cultures or nucleic acid amplification test for M. bovis from the same non-sterile body site (e.g., bronchopulmonary, gastrointestinal, skin surface or other non-sterile sites) without histopathologic confirmation of AFB in tissue
   and
2. Specific treatment initiated or recommended for M. bovis

MYCOBACTERIA OTHER THAN TB, MAC, OR M. BOVIS

220091 CONFIRMED PATHOGEN, specify mycobacterial species: M. kansasii, M. cheloneae, M. abscessus, M. haemophilum, M. genovensii, etc.
At least one of the following:
1. Positive cultures or nucleic acid amplification test for other mycobacteria, including, but not limited to M. kansasii, M. cheloneae, M. abscessus, M. haemophilum, M. genovensii, from a normally-sterile site (blood, bone marrow, lymph node, liver, cerebrospinal fluid, or other normally-sterile body fluid, tissue or organ)
   or
2. Positive cultures or nucleic acid amplification test for other mycobacteria, including, but not limited to those species listed above from other non-sterile sites (e.g., bronchopulmonary, gastrointestinal, skin surface or other non-sterile sites) from which the species was identified.

220092 PROBABLE, specify mycobacterial species
Both of the following:
1. Repeatedly positive cultures or nucleic acid amplification test for other mycobacteria, including, but not limited to M. kansasii, M. cheloneae, M. abscessus, M. haemophilum, M. genovensii, from the same non-sterile body site (e.g., bronchopulmonary, gastrointestinal, skin surface or other non-sterile sites)
   and
2. Specific treatment initiated for other mycobacteria
XIII. NEONATAL DISORDERS

CYSTIC FIBROSIS

120071 CONFIRMED
Systemic inherited disease of the exocrine glands that affects the lung and digestive systems. Confirmed by pilocarpine iontophoresis of sweat greater than or equal to ($\geq$) 60 mEq/L.

120072 PROBABLE
Both of the following:
1. Compatible syndrome
   and
2. Pilocarpine iontophoresis of sweat test less than (<) 60 mEq/L (milliequivalents/liter)

NOTE: See also CYSTIC FIBROSIS in the Congenital/Birth Defects section.

CYTOMEGALOVIRUS DISEASE, CONGENITAL

230011 CONFIRMED
1. At least one of the following:
   a. Hepatitis
      or
   b. Jaundice
      or
   c. Hepatosplenomegaly
      or
   d. Thrombocytopenia
      or
   e. Petechiae
      or
   f. Intrauterine growth retardation
      or
   g. Retinitis
      or
   h. Periventricular calcifications
      or
   i. Sensorineural hearing loss,

   and

2. At least one of the following:
   a. Positive urine, blood, or saliva CMV culture
      or
   b. Positive blood nucleic acid amplification test
      or
   c. Antigen test
ABO HEMOLYTIC DISEASE OF THE NEWBORN

160111  CONFIRMED
Mediated by maternal antibody as identified by a positive direct Coombs test

NOTE: See also ABO HEMOLYTIC DISEASE OF THE NEWBORN in the Hematologic section.

RH HEMOLYTIC DISEASE OF THE NEWBORN

160121  CONFIRMED
Mediated by maternal antibody; Coomb's positive.

NOTE: See also HEMOLYTIC DISEASE OF THE NEWBORN in the Hematologic section.

HEPATITIS, NEONATAL

170072  NO PATHOGEN CONFIRMED (PROBABLE)
Characterized by elevated transaminases 1.5 times ULN with or without clinical findings such as jaundice, hepatomegaly, and hepatic failure

NOTE: See also HEPATITIS, NEONATAL in the Hepatobiliary section.

HERPES SIMPLEX VIRUS (HSV), NEONATAL, Skin, Eye, Mouth Disease

230021  CONFIRMED
All of the following:
1. HSV detected from skin lesions, nasopharynx, or conjunctiva
   and
2. Location limited to mucosal surfaces, skin, or eyes
   and
3. No evidence of CNS infection (normal CSF parameters and negative CSF HSV DNA nucleic acid amplification test) or systemic disease (normal LFTs; no thrombocytopenia or pneumonia)

HERPES SIMPLEX VIRUS (HSV), NEONATAL, Disseminated Disease

230031  CONFIRMED
Both of the following:
1. HSV detected from blood, skin, or CSF
   and
2. Evidence of hepatitis, pneumonitis, or thrombocytopenia
HERPES SIMPLEX VIRUS (HSV), NEONATAL, CNS Disease

230041  CONFIRMED
HSV detected from CSF by culture or nucleic acid amplification test

INTRAVENTRICULAR HEMORRHAGE, GRADE 3, NEONATAL

230051  CONFIRMED
Radiologic diagnosis of hemorrhage into the germinal matrix tissues of the developing brain with possible rupture into the ventricular system and parenchyma

INTRAVENTRICULAR HEMORRHAGE, GRADE 4, NEONATAL

230061  CONFIRMED
Radiologic diagnosis of hemorrhage into the germinal matrix tissues of the developing brain with possible rupture into the ventricular system and parenchyma

KERNICTERUS

230071  CONFIRMED
All of the following:
1. Clinical neonatal syndrome
2. Severe indirect hyperbilirubinemia (greater than (>) 20 mg/dL (milligrams/deciliter))
3. Associated with CNS symptoms including bulging fontanel, opisthotonic posturing, fever, hypertonicity, paralysis, or seizures

MECONIUM ASPIRATION SYNDROME

230081  CONFIRMED
Aspiration of meconium mixed with amniotic fluid in utero or during delivery causing a partial or complete blockage of the airways associated with poor gas exchange in the lungs and chemical pneumonitis
NECROTIZING ENTEROCOLITIS, NEONATAL

230091 CONFIRMED
Both of the following:
1. Inflammation causing destruction of part of the bowel, may involve only the innermost lining or the entire thickness of the bowel, variable amounts of the bowel
   and
2. Confirmed by either surgery or radiographic study

230092 PROBABLE
1. Inflammation causing destruction of part of the bowel, may involve only the innermost lining or the entire thickness of the bowel, variable amounts of the bowel.
   and
2. Radiographic study non-diagnostic

OPHTHALMIA NEONATORUM

230101 CONFIRMED, specify pathogen
Clinical conjunctivitis and pathogen identified

230102 PROBABLE
Clinical conjunctivitis and no pathogen identified

RESPIRATORY DISTRESS SYNDROME, NEWBORN

230111 CONFIRMED
Clinical presentation of respiratory distress in a premature infant assumed to be due to surfactant deficiency; excludes other etiologies such as sepsis

RUBELLA, CONGENITAL

191101 CONFIRMED PATHOGEN (WHO criteria)
Both of the following:
1. One of the following:
   a. Appropriate clinical syndrome in an infant less than (<) 12 months of age.
      For example, a combination of two or more of the following:
      1. Cataracts
      or
      2. Congenital glaucoma
      or
      3. Congenital heart disease
      or
      4. Hearing impairment
      or
      5. Pigmentary retinopathy
      or
b. Both of the following:
   1. History of one of the following in a child:
      a. Purpura
      or
      b. Splenomegaly
      or
      c. Microcephaly
      or
      d. Mental retardation
      or
      e. Meningoencephalitis
      or
      f. Radiolucent bone disease
      or
      g. Jaundice with onset within 24-hours after birth
      and
   2. One of the following in a child:
      a. Cataracts
      or
      b. Congenital glaucoma
      or
      c. Congenital heart disease
      or
      d. Hearing impairment
      or
      e. Pigmentary retinopathy
      and
   2. Positive viral culture or nucleic acid amplification test for rubella virus or positive IgM antibody if IgM performed at less than (<) three months of age

NOTE: See also RUBELLA, CONGENITAL in the Infectious Diseases (non-mycobacterial) section.

SEPSIS, NEONATAL

230131 CONFIRMED, specify pathogen
Both of the following:
1. Clinical or laboratory findings indicating the presence of disseminated infection in the infant (zero to six weeks of age)
   and
2. Pathogen identified

230132 PROBABLE
Both of the following:
1. Clinical or laboratory findings indicating the presence of disseminated infection in the infant (zero to six weeks of age)
   and
2. No pathogen identified
SYMPHILIS, CONGENITAL

150121 CONFIRMED PATHOGEN
Diagnosis defined by having at least one of the following:
1. Demonstration of Treponema pallidum by dark field microscopy or specific fluorescent antibody stains from lesions of a newborn infant or from placenta, umbilical cord, or fetal autopsy

or
2. Newborn infant with a nontreponemal test titer four-fold greater than (> the mother’s titer using the same test preferably in the same laboratory

or
3. Newborn infant who has a reactive treponemal test for syphilis and at least one of the following:
   a. Abnormal physical examination (including, but not limited to, hepatosplenomegaly, rash, or snuffles)
   or
   b. Abnormal long bone radiographs with characteristic epiphyseal and metaphyseal changes
   or
   c. Positive CSF VDRL
   or
   d. Abnormal CSF cell count or protein in the absence of other explanations
   or
   e. Reactive IgM EIA or IgM 19S-FTA-ABS test

or
4. A child greater than (> two years of age with a reactive nontreponemal test for syphilis and stigmata of untreated congenital syphilis (e.g., interstitial keratitis, eighth cranial nerve deafness, anterior bowing of shins, frontal bossing, mulberry molars, peg-shaped notched central incisors (Hutchinson teeth), saddle nose, rhagades, or symmetric, painless knee swelling (Clutton joints)

150122 NO PATHOGEN CONFIRMED (PROBABLE)
Diagnosis defined by having at least one of the following:
1. A stillbirth at greater than (> 20 weeks gestation or greater than (> 500 grams fetal weight to a woman with untreated or inadequately treated syphilis, i.e., any non-penicillin regimen during pregnancy or a penicillin regimen administered less than (<) 30 days before delivery

or
2. A newborn infant born to a mother with untreated or inadequately treated syphilis, i.e., any non-penicillin regimen during pregnancy or a penicillin regimen administered less than (<) 30 days before delivery; serologic tests negative or not done, but penicillin therapy initiated

NOTE: See also SYMPHILIS, CONGENITAL in the Genitourinary/Sexually Transmitted Diseases section.
TRANSIENT TACHYPNEA, NEWBORN

230141 CONFIRMED
Signs of respiratory distress (e.g., tachypnea, nasal flaring, grunting, retractions, or cyanosis in extreme cases) become evident shortly after birth which is transient, with resolution occurring usually by age 72 hours. Excludes other etiologies such as sepsis.

TOXOPLASMOSIS, CONGENITAL

191191 CONFIRMED PATHOGEN
Both of the following:
1. Appropriate clinical syndrome in an infant
   and
2. Positive serology, ophthalmologic exam, or histopathology indicative of Toxoplasma gondii

191192 NO PATHOGEN CONFIRMED (PROBABLE)
All of the following:
1. Appropriate clinical syndrome
   and
2. No documented positive serology, ophthalmologic exam, or histopathology indicative of Toxoplasma gondii
   and
3. No evidence of CMV or rubella infection

NOTE: See also TOXOPLASMOSIS, CONGENITAL in the Infectious Diseases (non-mycobacterial section.

OTHER CONGENITAL INFECTIONS, specify

239001 CONFIRMED, specify pathogen
Pathogen identified

239002 PROBABLE
No pathogen identified; compatible clinical syndrome
XIV. NEOPLASTIC DISEASES

ANORECTAL CANCER, INCLUDES ANAL
Location: Contiguous or overlapping sites of rectum, rectosigmoid junction, and anus with unknown point of origin, anal canal, or sphincter
Specify: Adenocarcinoma or squamous cell carcinoma

240011 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240012 PROBABLE
1. One of the following:
   a. Characteristic mass on examination
   or
   b. Compatible radiography
   and
   2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

ANAL ATYPIA
Location: Perianal
Specify: Squamous or glandular cells

240021 CONFIRMED
One of the following:
1. Diagnostic cytology results showing atypical cells of uncertain significance
   or
2. PAP smear results showing atypical cells of uncertain significance

ANAL DYSPLASIA/INTRAEPITHELIAL NEOPLASIA
Location: Perianal
Specify: Low-grade squamous intraepithelial lesion (LGSIL), high-grade squamous intraepithelial lesion (HGSIL), or carcinoma in situ (CIS)

240031 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology.

240032 PROBABLE
One of the following:
1. Diagnostic cytology
   or
2. PAP smear results
BILIARY TRACT CANCER
Location: All sites of gall bladder, intrahepatic or extrahepatic bile ducts

240041 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240042 PROBABLE
1. One of the following:
   a. Endoscopy
   or
   b. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

BLADDER CANCER
Location: All parts of the bladder, bladder neck, ureteric orifice, and urachus

240051 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240052 PROBABLE
1. One of the following:
   a. Compatible endoscopy
   or
   b. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

BONE AND JOINT CANCER
Location: All bones and articular cartilage

240061 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240062 PROBABLE
Both of the following:
1. One of the following:
   a. Characteristic mass on examination
   or
   b. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis
BRAIN AND NERVOUS SYSTEM CANCER, excluding non-Hodgkin’s Lymphoma, Primary CNS
Location: All parts of the brain or nervous system (primary)

240071 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240072 PROBABLE
Both of the following:
1. Compatible radiography
and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

NOTE: See NON-HODGKIN’S LYMPHOMA or PRIMARY CNS criteria in this section.

BREAST CANCER
Location: All parts of the breast, nipple, and areola
Specify: Intraductal/ductal insitu (DIS), infiltrating/invasive ductal, lobular in situ (LCIS)/infiltrating lobular, other, or unspecified

240081 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240082 PROBABLE
Both of the following:
1. One of the following:
   a. Characteristic mass on examination
      or
   b. Compatible radiography
      and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

CERVICAL CANCER, INVASIVE
Location: Endocervix, exocervix, cervix uteri, or any unspecified site of cervix
Specify: Adenocarcinoma or squamous cell carcinoma

240091 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240092 PROBABLE
1. One of the following:
   a. Characteristic mass on examination
   or
   b. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

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CERVICAL ATYPIA
Location: Endocervix, exocervix, cervix uteri, or any unspecified site of cervix
Specify: Squamous or glandular cells

240101 CONFIRMED
Diagnostic cytology or PAP smear results showing atypical cells of uncertain significance

CERVICAL DYSPLASIA/INTRAEPITHELIAL NEOPLASIA
Location: Endocervix, exocervix, cervix uteri, or any unspecified site of cervix
Specify: Low-grade squamous intraepithelial lesion (LGSIL), high-grade squamous intraepithelial lesion (HGSIL), or carcinoma in situ (CIS)

240111 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240112 PROBABLE
One of the following:
1. Diagnostic cytology
or
2. PAP smear results

COLON CANCER
Location: All parts of the large bowel or colon
Specify: Distal, proximal, or unspecified

240121 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240122 PROBABLE
1. One of the following:
   a. Compatible endoscopy
   or
   b. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

ESOPHAGUS CANCER
Location: All parts of the esophagus

240131 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology
240132 PROBABLE
1. One of the following:
   a. Compatible endoscopy
   or
   b. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

EYE CANCER
Location: All parts of the eye
Specify: Conjunctiva, other or unspecified

240141 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240142 PROBABLE
1. One of the following:
   a. Ophthalmologic examination
   or
   b. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

HODGKIN'S DISEASE
Location: All parts of the lymphatic system, spleen, bone marrow, and other organs (liver, lung, brain, etc.)
Specify: Nodular sclerosis, mixed cellularity, lymphocyte-rich, and lymphocyte-depletion

240151 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240152 PROBABLE
1. One of the following:
   a. Characteristic mass on examination
   or
   b. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

KAPOSI'S SARCOMA
Location: Skin, mucus membranes, or organs
Specify: Mucocutaneous, visceral, or unspecified

240161 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology
240162 PROBABLE
Mucocutaneous: Physical examination by experienced provider
Visceral: 1. One of the following:
   a. Compatible endoscopy
   or
   b. Compatible radiography.
   and
   2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

KIDNEY CANCER
Location: Any part of the kidney or renal pelvis

240171 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240172 PROBABLE
Both of the following:
1. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

LARYNX CANCER
Location: Any part of the larynx including glottis, subglottis, or laryngeal cartilage

240181 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240182 PROBABLE
1. One of the following:
   a. Compatible endoscopy
   or
   b. Compatible radiography
   and
   2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

LEUKEMIA
Location: Blood and bone marrow
Specify: Acute lymphocytic, acute monocytic, acute myeloid, chronic lymphocytic, chronic myeloid,
other lymphocytic, other myeloid/monocytic, or other

240191 CONFIRMED
Diagnostic histopathology on blood smear or bone marrow biopsy
**LIVER CANCER**
Location: Any part of the liver

**240201 CONFIRMED**
Diagnostic histopathology on biopsy or surgical pathology

**240202 PROBABLE**
*Both of the following:*
1. Compatible radiography
   *and*
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

**LUNG CANCER**
Location: Any part of the lung or bronchus
Specify: Adenocarcinoma, large cell carcinoma, small cell carcinoma, or other non-small cell carcinoma

**240211 CONFIRMED**
Diagnostic histopathology on biopsy or surgical pathology

**240212 PROBABLE**
1. *One of the following:*
   a. Compatible endoscopy
   *or*
   b. Compatible radiography
   *and*
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

**MULTIPLE MYELOMA**
Location: blood and bone marrow

**240221 CONFIRMED**
Diagnostic histopathology on blood smear or bone marrow biopsy

**NASAL CAVITIES, MIDDLE EAR, AND SINUSES (NOT PYRIFORM) CANCER**
Location: Nasal cavity, auditory tube, middle ear, mastoid air cells, maxillary sinus, ethmoidal sinus, frontal sinus, sphenoidal sinus, accessory sinus and any other unspecified part of these areas, except pyriform sinus. Specify location.

**240231 CONFIRMED**
Diagnostic histopathology on biopsy or surgical pathology
240232 PROBABLE
1. One of the following:
   a. Characteristic mass on examination
   or
   b. Compatible radiography
   or
   c. Compatible endoscopy
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

NON-HODGKIN’S LYMPHOMA, NON-PRIMARY CNS
Location: All parts of the lymphatic system, spleen, bone marrow, and other organs (liver, lung), except primary CNS disease.
Specify: Small cleaved cell diffuse (Burkitts), mixed small cleaved and large cell diffuse, large cell diffuse, small cleaved cell follicular, mixed small cleaved and large cell follicular, large cell follicular, undifferentiated (diffuse), immunoblastic or unspecified

240241 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240242 PROBABLE
1. One of the following:
   a. Characteristic mass on examination
   or
   b. Compatible radiography
   or
   c. Compatible endoscopy
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

NOTE: See also ORAL NON-HODGKIN’S LYMPHOMA in the Oral Disorders section.

NON-HODGKIN’S LYMPHOMA, PRIMARY CNS
Location: Only primary CNS disease

240251 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology
240252  **PROBABLE**

All of the following:

1. Neurologic signs with CD4 lymphocyte count less than (<) 100 cells/mm\(^3\)  
2. Mass lesion(s) on head CT/MRI scan  
3. Failure of clinical response to antitoxoplasmosis chemotherapy or other anti-infective chemotherapy (e.g. tuberculosis, cryptococcosis)  
4. Lesion(s) becomes markedly reduced or disappears following high-dose glucocorticoid or radiation therapy

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**ORAL CAVITY AND PHARYNX CANCER**

Location: All parts of lip, all parts of tongue, junctional zone, parotid gland, all types of salivary glands, all parts of gums, all parts of mouth, all parts of palate, uvula, all parts of nasopharynx, postcrioid region, pyriform sinus, all parts of hypopharynx, all parts of oropharynx, all parts of tonsil, vallecula, and Waldeyer’s ring

Specify: Anterior epiglottis, base of tongue, junctional region, lateral wall of oropharynx, posterior wall of oropharynx, tonsil, vallecula, Waldeyer’s ring, other site, or unspecified

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240261  **CONFIRMED**

Diagnostic histopathology on biopsy or surgical pathology

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240262  **PROBABLE**

1. One of the following:
   a. Characteristic mass on examination  
   or  
   b. Compatible radiography  
   or  
   c. Compatible endoscopy  
   and  
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis.

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**OVARIAN CANCER**

Location: Any part of the ovary

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240271  **CONFIRMED**

Diagnostic histopathology on biopsy or surgical pathology.
240272  PROBABLE
1. One of the following:
   a. Compatible endoscopy
   or
   b. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

PANCREAS CANCER
Location: Any part of the pancreas including head, body, tail, pancreatic duct, or islets of Langerhans

240281  CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240282  PROBABLE
1. One of the following:
   a. Compatible endoscopy
   or
   b. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

PENIS CANCER
Location: Any part of the penis including prepuce, glans, body, or unspecified

240291  CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240292  PROBABLE
1. One of the following:
   a. Characteristic mass on examination
   or
   b. Compatible radiography
   or
   c. Compatible endoscopy
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

PERITONEUM AND RETROPERITONEUM CANCER,
Location: Any part of the peritoneum or retroperitoneum not part of a retroperitoneal organ on this list (e.g. pancreas or kidney)
Specify: Either peritoneum or retroperitoneum
Diagnostic histopathology on biopsy or surgical pathology

Both of the following:
1. Compatible endoscopy or compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

PLEURA CANCER
Location: Any part of the parietal or visceral pleura

Diagnostic histopathology on biopsy or surgical pathology

Both of the following:
1. Compatible endoscopy or compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

PROSTATE CANCER
Location: Any part of the prostate

Diagnostic histopathology on biopsy or surgical pathology.

1. One of the following:
   a. Characteristic mass on examination
   or
   b. Compatible radiography
   and
   2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

SKIN CANCER
Location: Skin of the lip, eye lid, ear, face, scalp, neck, trunk, upper limb, lower limb, other or unspecified site

Specify: Melanoma, non-melanoma (specify the type of non-melanoma cell - squamous cell, basal cell, Merkel cell, or other non-melanoma)

Diagnostic histopathology on biopsy or surgical pathology
240332 PROBABLE
Characteristic lesion on examination by a dermatologist highly suggestive of this diagnosis

SMALL INTESTINE CANCER
Location: Duodenum, jejunum, ileum, Meckel’s diverticulum, other, or unspecified small bowel

240341 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240342 PROBABLE
1. One of the following:
   a. Compatible endoscopy
   or
   b. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

240351 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240352 PROBABLE
1. One of the following:
   a. Characteristic mass on examination
   or
   b. Compatible radiography
   or
   c. Compatible endoscopy
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

240361 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology
240362 PROBABLE
1. One of the following:
   a. Compatible endoscopy
   or
   b. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

TESTICULAR CANCER
Location: All parts of the testis

240371 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240372 PROBABLE
1. One of the following:
   a. Characteristic mass on examination
   or
   b. Compatible radiography
   or
   c. Compatible endoscopy
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

THYROID CANCER
Location: All parts of the thyroid

240381 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240382 PROBABLE
1. One of the following:
   a. Characteristic mass on examination
   or
   b. Compatible radiography
   or
   c. Compatible endoscopy
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

TRACHEA, MEDIASTINUM, AND OTHER RESPIRATORY ORGANS CANCER
Location: Trachea, upper respiratory tract, all parts of mediastinum, other or ill-defined parts of the respiratory system
Specify: Site

240391 CONFIRMED
240392 PROBABLE
1. **One of the following:**
   a. Compatible endoscopy
   **or**
   b. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

**UTERUS (NOT SPECIFIED) OR CORPUS CANCER,**
Location: Corpus uteri, isthmus, or uterus not specified
Specify: Uterus or corpus

240401 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240402 PROBABLE
1. **One of the following:**
   a. Compatible endoscopy
   **or**
   b. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

**VAGINAL CANCER**
Location: All parts of the vagina

240411 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240412 PROBABLE
1. **One of the following:**
   a. Characteristic mass on examination
   **or**
   b. Compatible radiography
   **or**
   c. Compatible endoscopy
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

**VAGINAL ATYPIA**
Location: Vagina
Specify: Squamous or glandular cells

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240421 CONFIRMED
One of the following:
1. Diagnostic cytology results showing atypical cells of uncertain significance
   or
2. PAP smear results showing atypical cells of uncertain significance

VAGINAL DYSPLASIA/INTRAEPITHELIAL NEOPLASIA
Location: Vagina
Specify: Low-grade squamous intraepithelial lesion (LGSIL), high-grade squamous intraepithelial lesion (HGSIL), or carcinoma in situ (CIS)

240431 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240432 PROBABLE
One of the following:
1. Diagnostic cytology
   or
2. PAP smear results

VULVA CANCER
Location: All parts of the vulva including labia major, labia minor, and clitoris

240441 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240442 PROBABLE
1. One of the following:
   a. Characteristic mass on examination
      or
   b. Compatible radiography
      or
   c. Compatible endoscopy
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

VULVAR ATYPIA
Location: Vulva
Specify: Squamous or glandular cells

240451 CONFIRMED
One of the following:
1. Diagnostic cytology results showing atypical cells of uncertain significance
   or
2. PAP smear results showing atypical cells of uncertain significance
VULVAR DYSPLASIA/INTRAEPITHELIAL NEOPLASIA
Location: Vulva
Specify: Low-grade squamous intraepithelial lesion (LGSIL), high-grade squamous intraepithelial lesion (HGSIL), or carcinoma in situ (CIS)

240461 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

240462 PROBABLE
One of the following:
1. Diagnostic cytology
   or
2. PAP smear results

OTHER CANCERS

OTHER ENDOCRINE ORGAN CANCERS
Location: Thymus, adrenal gland, parathyroid gland, pituitary gland and craniopharyngeal duct, pineal gland, carotid body, aortic body, other or unspecified
Specify: Site

249001 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

249002 PROBABLE
Both of the following:
1. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

OTHER FEMALE GENITAL ORGANS CANCER, specify site
Location: Fallopian tube, broad ligament, parametrium, round ligament, uterine adnexa, other, and unspecified
Specify: Site

249011 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology
249012 PROBABLE
1. One of the following:
   a. Characteristic mass on examination
   or
   b. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

OTHER LYMPHATIC AND HEMATOPOIETIC TISSUE CANCERS
Location: Lymph nodes or spleen
Specify: Lymphatic or hematopoietic tissue

249021 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

249022 PROBABLE
1. One of the following:
   a. Endoscopy
   or
   b. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

OTHER MALE GENITAL ORGANS CANCER
Location: Epididymis, spermatic cord, scrotum, other, or unspecified
Specify: Site

249031 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

249032 PROBABLE
1. One of the following:
   a. Characteristic mass on examination
   or
   b. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis
OTHER URINARY CANCER, specify location
Location: Urethra, paraurethral glands, ureter, other, or unspecified
Specify: Location

249041 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

249042 PROBABLE
1. One of the following:
   a. Endoscopy
   or
   b. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis

OTHER CANCER, specify
Location: Any cancer with unknown primary location or other primary location not otherwise listed in this document
Specify: Adenocarcinoma of unknown primary, adenocarcinoma of other primary (specify the primary site), other cancer of unknown primary, other cancer of other primary (specify the primary site)

249051 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

249052 PROBABLE
1. One of the following:
   a. Characteristic mass on examination
   or
   b. Compatible radiography
   and
2. Cytology of aspirate or body fluid highly suggestive of this diagnosis
XV. NEUROLOGICAL DISORDERS

ATAXIA

250011 CONFIRMED
Abnormalities of muscle balance or inability to finely coordinate movements

CNS MASS LESION, OTHER THAN CNS TOXOPLASMOSIS, NON-HODGKIN LYMPHOMA, PRIMARY CNS

250021 CONFIRMED
1. Presence of mass lesion(s) on brain imaging study (e.g., CT or MRI)
   and
2. At least one of the following:
   a. Study participant does not meet criteria for other diagnosis with CNS mass lesion (e.g., CNS toxoplasmosis, CNS lymphoma)
   or
   b. Diagnostic testing not available for diagnosis of a specific CNS mass lesion

CRANIAL NERVE ABNORMALITY, specify cranial nerve.

250031 CONFIRMED
Cranial nerve palsy or paralysis on clinical exam. Excludes Bell’s Palsy.

NOTE: See also PALSY, BELL’S in this section.

ENCEPHALOMYELITIS, ACUTE DISSEMINATED (ADEM)

250041 CONFIRMED
An acute, multifocal encephalomyelitis that follows vaccination or (often viral) infections, with white matter abnormalities on MRI

250042 PROBABLE.
Acute ADEM and MRI either negative or not done

ENCEPHALOPATHY, PROGRESSIVE
Progressive encephalopathy needs to be distinguished from mental retardation secondary to other causes, such as maternal drug addiction and prematurity, which can be determined only by longitudinal assessment. AN ETIOLOGY SHOULD BE SPECIFIED, IF IDENTIFIED.

250051 CONFIRMED
All of the following:
1. Impaired brain growth
   and
2. Progressive motor dysfunction
   and
APPENDIX 100 - Diagnoses Appendix

3. Loss or plateau of developmental milestones

250052 PROBABLE
One or two of the following, but not all three:
1. Impaired brain growth
   or
2. Progressive motor dysfunction
   or
3. Loss or plateau of developmental milestones

FAILURE OF BRAIN GROWTH

250062 PROBABLE
Impaired brain growth in childhood, as documented by either absence of normal rate of head growth in children or progressive loss of cerebral parenchymal volume by CT or MRI.

HIV-ASSOCIATED NEUROLOGICAL DISORDERS (HAND)

HIV-ASSOCIATED ASYMPTOMATIC NEUROCOGNITIVE IMPAIRMENT

250071 CONFIRMED
Impairment diagnosed by neurocognitive testing without impact on activities of daily living (ADLs).

HIV-ASSOCIATED MILD NEUROCOGNITIVE DISORDER

250081 CONFIRMED
Impairment diagnosed by neurocognitive testing with impact on activities of daily living (ADLs) present.

HIV-ASSOCIATED DEMENTIA

250091 CONFIRMED
All of the following:
1. Acquired cognitive/motor dysfunction for at least one month causing impairment of work or activities of daily living (verifiable by report of a key informant), not attributable solely to severe systemic illness, medication adverse effects, active psychiatric disorders, active alcohol or substance use or substance withdrawal.
   and
2. Abnormalities from at least two of the following categories:
   a. Motor abnormality: For example, slowed rapid movements, release signs, abnormal gait, limb incoordination, diffuse hyperreflexia, hypertonia, or weakness.
   b. Behavioral abnormality: For example, change in personality with apathy, inertia, irritability, or emotional lability or new onset of impaired judgment characterized by socially inappropriate behavior or disinhibition.
   c. Cognitive abnormality (two or more): memory, judgment, flexibility, visual, constructional difficulties, reaction time, speed of mental processing, attention or
APPENDIX 100 - Diagnoses Appendix

concentration as determined by appropriate neuropsychological instruments, with
interpretation of abnormality or decline by a neurologist/neuropsychologist.

and

3. No other etiology is identified by CNS imaging, or culture, serology, or nucleic acid
amplification test testing of CSF.

250092  PROBABLE
All of the following:
1. Acquired cognitive/motor dysfunction for at least one month causing impairment of work
or activities of daily living (verifiable by report of a key informant), not attributable solely
to severe systemic illness or medication adverse effects.

and

2. Abnormalities from at least two of the following categories:
   a. Motor abnormality: For example, slowed rapid movements, release signs, abnormal
gait, limb incoordination, diffuse hyperreflexia, hypertonia, or weakness.
   b. Behavioral abnormality: For example, change in personality with apathy, inertia,
irritability, or emotional lability or new onset of impaired judgment characterized by
socially inappropriate behavior or disinhibition.
   c. Cognitive abnormality (two or more): memory, judgment, flexibility, visual,
constructional difficulties, reaction time, speed of mental processing, attention or
concentration as determined by appropriate neuropsychological instruments, with
interpretation of abnormality or decline by a neurologist/neuropsychologist.

and

3. Tests for other possible etiology (active CNS opportunistic infections or malignancy,
active psychiatric disorders, active alcohol or substance abuse or substance withdrawal)
are not completed, results are not available or results do not exclude other CNS processes.

HEARING LOSS, specify type

250101  CONFIRMED
Hearing loss sufficient to present clinically, regardless of etiology and site (i.e., conductive,
sensorineural, etc) and by audiometric testing

250102  PROBABLE
Hearing loss sufficient to present clinically, regardless of etiology and site (i.e., conductive,
sensorineural, etc) but not by audiometric testing

INFANTILE SPASMS (West Syndrome)

250111  CONFIRMED
Syndrome characterized by the triad of infantile spasms (generalized seizures),
hypsarrhythmia, and arrest of psychomotor development at seizure onset
MACROCEPHALY

250122 PROBABLE
Head circumference that measures greater than or equal to (≥)2 SD above the mean (+2 SD) for age and sex.

MICROCEPHALY

250132 PROBABLE
Head circumference that measures less than (<)2 SD below the mean (<−2 SD) for age and sex.

MIGRAINE

250141 CONFIRMED
Clinical syndrome of recurrent episodes of severe cephalgia, often associated with nausea, vomiting, or aura

MOTOR DEVELOPMENTAL DELAY

250152 PROBABLE
Both of the following:
1. One or two of the following, but not all three:
   a. impaired brain growth
   or
   b. progressive motor dysfunction
   or
   c. loss or plateau of developmental milestones
   and
2. Study participant does not have abnormalities of reflexes, tone or muscle bulk, cognitive development is reasonable but motor landmarks are delayed, for example, late walking without weakness, cerebral palsy (CP), etc.

MOTOR NEURON DISEASE

250161 CONFIRMED
Clinical disorder that affects the anterior horn cells leading to muscle weakness and atrophy

MULTIPLE SCLEROSIS

250171 CONFIRMED
A disease characterized by CNS demyelination, with multiple lesions occurring at different times and spatial locations. Must be diagnosed by a clinician experienced with making the diagnosis of multiple sclerosis.

MUSCULAR DYSTROPHY

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V.1.3 November, 2013
A group of diseases characterized by progressive muscle weakness and loss of muscle tissue including but not limited to Becker Syndrome, Duchenne’s muscular dystrophy and limbgirdle muscle dystrophy. Must be diagnosed by a clinician experienced with making the diagnosis of muscular dystrophy.

**MYELOPATHY, HIV-ASSOCIATED (VACUOLAR)**

- **250191 CONFIRMED**
  Vacuolation of white matter in the spinal cord, which is most pronounced in the lateral and posterior columns of the thoracic cord at autopsy or biopsy

- **250192 PROBABLE**
  Lower extremity weakness and spasticity with hyperreflexia. CSF profile is typically characterized by few white blood cells, normal glucose and normal or elevated protein. Myelopathy due to CMV, VZV, and HTLV-1 have been excluded.

**MYELOPATHY, NON-HIV-ASSOCIATED**

- **250201 CONFIRMED**
  A disorder of the spinal cord, specify cause (e.g., CMV, VZV, HTLV-1, other)

- **250202 PROBABLE**
  A disorder of the spinal cord of unknown cause that is not consistent with HIV-associated vacular myelopathy.

**MYASTHENIA**, includes myasthenia gravis and related syndromes

- **250211 CONFIRMED**
  Clinical syndrome with abnormalities of the myoneural junction. (e.g., myasthenia gravis or myathenic syndromes). Must be diagnosed by an experienced clinician.

**MYOPATHY, HIV-ASSOCIATED**

- **250221 CONFIRMED**
  All of the following:
  1. Symptoms of weakness in proximal muscles (thighs, shoulders, or upper arms) with evidence of proximal weakness on physical examination
  2. CPK elevated to greater than (>) twice normal (no EMG, physical trauma, or IM injection within two weeks)
  3. ZDV muscle toxicity excluded by one of the following:
     a. No history of ZDV in the immediately preceding three months
    or
APPENDIX 100 - Diagnoses Appendix

b. Drug holiday from ZDV for at least one month with no improvement in signs, symptoms, or CPK elevation

and

4. Neurodiagnostic confirmation by at least one of the following:
   a. EMG documenting myopathic features
   or
   b. Muscle biopsy documenting myofiber degeneration or inflammation

and

5. Other causes of myopathy have been excluded

250222 PROBABLE
All of the following:
1. Symptoms of weakness in proximal muscles (thighs, shoulders, or upper arms) with evidence of proximal weakness on physical examination

and

2. CPK elevated to greater than (> ) two times normal (no EMG, physical trauma, or IM injection within two weeks)

and

3. ZDV muscle toxicity excluded by one of the following:
   a. No history of ZDV in the immediately preceding three months
   or
   b. Drug holiday from ZDV for at least one month with no improvement in signs, symptoms, or CPK elevation

and

4. Other causes of myopathy have been excluded

MYOPATHY, NON-HIV ASSOCIATED, specify type

250231 CONFIRMED
A disorder of the muscle that is not consistent with HIV-associated myopathy. Confirmed with appropriate laboratory testing; specify type.

250232 PROBABLE
A disorder of the muscle that is not consistent with HIV-associated myopathy, appropriate laboratory testing negative or not done, specify type.

Palsy, Bell’s

250241 CONFIRMED
Idiopathic paralysis of cranial nerve VII.

Palsy, Cerebral

250251 CONFIRMED
A form of a static encephalopathy, characterized by motor difficulties (hypertonia/spasticity, hypotonia, dystonia, or choreoathetosis).
PERIPHERAL NEUROPATHY

250261 CONFIRMED
1. Both of the following:
   a. Symmetrical, chronic pain (greater than (> three days), burning, or dysesthesias affecting both feet
   and
   b. Absent or diminished (compared to knee) ankle reflexes

or

2. Both of the following:
   a. At least one abnormal sensory sign: elevated vibratory thresholds, stocking distribution, loss of pinprick or temperature, or cutaneous allodynia
   and
   b. Proven by laboratory studies, EMG/NCV, or nerve biopsy

NOTE: Specify possible contributing factors including: diabetes mellitus, B12 deficiency, medication toxicity (ddl, ddC, or d4T, other)

250262 PROBABLE
At least one abnormal sensory sign:
1. Elevated vibratory thresholds
or
2. Stocking distribution
   Or
3. Loss of pinprick or temperature
   or
4. Cutaneous allodynia in the absence laboratory studies
   or
5. EMG/NCV or nerve biopsy

NOTE: Specify possible contributing factors including: diabetes mellitus, B12 deficiency, medication toxicity (ddl, ddC, or d4T, other)

POST-HERPETIC NEURALGIA

250271 CONFIRMED
A painful dermatomal syndrome following herpes zoster infection related to previous nerve inflammation

PSEUDOTUMOR CEREBRI

250281 CONFIRMED
A clinical syndrome of increased intracranial pressure as documented on lumbar puncture without an identifiable cause such as a mass
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250282 PROBABLE
A clinical syndrome of increase intracranial pressure as documented on lumbar puncture and no radiologic testing done

SEIZURE, FEBRILE, SIMPLE

250291 CONFIRMED
Syndrome of childhood, consisting of a seizure event associated with a febrile episode
Seizures are brief (less than \(<\) 10 - 15 minutes), generalized convulsions, with fever, not recurrent within the same 24-hours, in a child without preexisting neurological or developmental abnormalities.
First occurrence is generally between ages three months and three years. Subsequent occurrences may continue to age six years.

SEIZURE, FEBRILE, COMPLEX

250301 CONFIRMED
Syndrome of childhood, consisting of a seizure event associated with a febrile episode.
Seizures have **at least one of the following** complex characteristics:
1. Focal
   *or*
2. Prolonged (greater than \(>\) 15 min)
   *or*
3. Multiple in same 24-hours
   *or*
4. Preexisting neurological or developmental abnormalities

SEIZURE, NEONATAL

250311 CONFIRMED
Seizures occurring up to one month of age

SEIZURE, SINGLE UNPROVOKED

250321 CONFIRMED
Isolated seizure of any type without identifiable provoking condition. Approximately 40% (percent) of children with a single unprovoked seizure have a second seizure and meet criteria for diagnosis of epilepsy.

SEIZURE DISORDER/ EPILEPSY

250331 CONFIRMED
A syndrome of recurrent, unprovoked seizures. Febrile seizures, neonatal seizures, single isolated seizure, and other provoked seizure are excluded from this diagnosis.

SEIZURE DISORDER (not Epilepsy), specify disorder
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250341 CONFIRMED
A syndrome of recurrent provoked seizures caused by an acute systemic condition such as: toxin exposure, mass lesion, CVA, hyponatremia, hypoglycemia, intoxication, drug withdrawal, acute trauma, or immediately after neurological injury or as a response to syncope.

SLEEP APNEA

250351 CONFIRMED
A disruption of normal sleep architecture as a result of recurrent apnea, which is usually obstructive with positive sleep plethysmography

250352 PROBABLE
A disruption of normal sleep architecture as a result of recurrent apnea, which is usually obstructive without positive sleep plethysmography

SPINOCEREBELLAR DISEASE, specify disease (if identified)

250361 CONFIRMED
Degenerative diseases involving the cerebellum, brainstem and spinal cord, specify if identified. Diagnosis made by an experienced clinician.

STROKE, HEMORRHAGIC

110261 CONFIRMED
1. Both of the following:
   a. Demonstrable lesion compatible with an acute hemorrhagic stroke on a CT or MRI by at least one of the following:
      1. Blood in subarachnoid space or intraparenchymal hemorrhage by CT scan.
         (Intraparenchymal blood must be dense and not mottled-mixed hyperdensity and hypodensity.)
       or
      2. Bloody spinal fluid by lumbar puncture. (Bloody CSF means greater than (> 100 cells/mm³ (cubic millimeter.) The LP is thought to be non-traumatic and counts in the last tube are similar to those in the first tube (no clearing) or xanthochromia when the specimen is spun down.)
       or
      3. Surgical evidence of hemorrhage as cause of clinical syndrome
   and
   b. Acute onset with a clinically compatible course, including unequivocal objective findings of a localizing neurologic deficit
     or
   2. Stroke diagnosed as cause of death at autopsy
110262 PROBABLE
1. Demonstrable lesion compatible with an acute hemorrhagic stroke on a CT or MRI by at least one of the following:
   a. Blood in subarachnoid space or intraparenchymal hemorrhage by CT scan.
      (Intraparenchymal blood must be dense and not mottled-mixed hyperdensity and hypodensity.)
   or
   b. Bloody spinal fluid by lumbar puncture. (Bloody CSF means greater than (>) 100 cells/mm³ (cubic millimeter.) The LP is thought to be non-traumatic and counts in the last tube are similar to those in the first tube (no clearing) or xanthochromia when the specimen is spun down.)
   or
   c. Surgical evidence of hemorrhage as cause of clinical syndrome

and
2. At least one of the following:
   a. Positive lumbar puncture compatible with subarachnoid hemorrhage
   or
   b. Death certificate or death NOTE from medical record listing stroke as cause of death

NOTE: See also STROKE, HEMORRHAGIC in the Cardiovascular Disease section.

STROKE, ISCHEMIC INFARCTION

110271 CONFIRMED
1. Both of the following:
   a. Demonstrable lesion compatible with an acute stroke with ischemic infarction on a CT or MR) by at least one of the following:
      1. Focal brain deficit without CT or LP evidence of blood, except mottled cerebral pattern. Either decreased density by CT in a compatible location or a negative CT or none done.
   or
   2. Surgical evidence of ischemic infarction
   and
   b. Acute onset with a clinically compatible course including unequivocal objective findings of a localizing neurologic deficit
   or
   2. Stroke diagnosed as cause of death at autopsy

110272 PROBABLE
1. Demonstrable lesion compatible with an acute stroke with ischemic infarction on a CT or MRI by at least one of the following:
   a. Focal brain deficit without CT or LP evidence of blood, except mottled cerebral pattern. Either decreased density by CT in a compatible location or a negative CT or none done.
   or
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b. Surgical evidence of ischemic infarction

and

2. At least one of the following:
   a. Positive lumbar puncture compatible with subarachnoid hemorrhage
   or
   b. Death certificate or death **NOTE** from medical record listing stroke as cause of death

**NOTE**: See also STROKE, ISCHEMIC INFARCTION in the Cardiovascular Disease section.

STROKE, UNKNOWN TYPE

110281 CONFIRMED
1. Both of the following:
   a. Demonstrable lesion compatible with an acute stroke on a CT or MRI with inadequate information to categorize as hemorrhagic or ischemic infarction
   and
   b. Acute onset with a clinically compatible course, including unequivocal objective findings of a localizing neurologic deficit
   or
2. Stroke diagnosed as cause of death at autopsy

**NOTE**: See also STROKE, UNKNOWN TYPE in the Cardiovascular Disease section.

ENCEPHALOPATHY, OTHER, specify etiology if known.

259001 CONFIRMED
Changes in mental status or brain function caused by inborn error of metabolism, electrolyte imbalance, mitochondrial dysfunction, or neurometabolic disease affecting the CNS or toxic substances. Excludes HAD (HIV-Associated Dementia), HAND (HIV-Associated Neurological Disorders), and Toxoplasmosis.

NEUROLOGIC DISEASE, OTHER, specify disease; excludes known conditions listed elsewhere in this document

259011 CONFIRMED
Could include gait/balance disorder, hypotonia, hypertonia, or involuntary movement disorder.
FUNGAL INFECTIONS

ORAL/OROPHARYNGEAL PSEUDOMEMBRANOUS OR ERYTHEMATOUS CANDIDIASIS, Specify oral or oropharyngeal and pseudomembranous or erythematous

PROBABLE
Both of the following:
1. One of the following case definitions:

   190122 Pseudomembranous candidiasis
   Clinical Descriptors: White or yellow/creamy spots or plaques that may be located in any part of the oral cavity and can usually be wiped off to reveal an erythematous surface
   Patient reported symptoms: None or possible mild to moderate burning pain
   Patient reported duration: Lesions/symptoms are usually intermittent, but may be long-standing

   190132 Erythematous candidiasis
   Clinical Descriptors: Patchy erythema or red areas usually located on the palate and dorsum of the tongue, but occasionally on the buccal mucosa. At times, white spots or plaques of pseudomembranous candidiasis may also be present.
   Patient reported symptoms: None or possible mild to moderate burning pain
   Patient reported duration: Lesions/symptoms are usually intermittent, but may be long-standing

   and

2. Specific antifungal therapy initiated or recommended

NOTE: See also ORAL/OROPHARYNGEAL CANDIDIASIS, PROBABLE in the Infectious Diseases (non-mycobacterial) section.

ANGULAR CHEILITIS

260012 PROBABLE
   Clinical descriptors: Red or white fissures or linear ulcers located at the lip commissures or corners of the mouth.
   Patient-reported symptoms: None or possible mild pain when opening mouth.
   Patient-reported duration: Lesions/symptoms usually intermittent, but may be long-standing.

VIRAL INFECTIONS

190522 HAIRY LEUKOPLAKIA, OHARA DEFINITION (WHO-3)

Clinical descriptors: Whitish/grey lesions on the lateral margins of the tongue. They are not removable and may exhibit vertical corrugations. Lesions range in size as they may be less than (<) one cm (centimeter), or may extend onto the ventral and dorsal surfaces of the tongue where they are usually flat. May be bilateral or unilateral.
APPENDIX 100 - Diagnoses Appendix

Patient-reported symptoms: Asymptomatic.
Patient-reported duration: Lesion(s) usually long-standing.

PRIMARY HERPETIC GINGIVOSTOMATITIS

260022 PROBABLE
Clinical descriptors: Vesicular eruption that may occur anywhere on oral mucosa, gingiva, vermillion, and peri-oral skin. The vesicles rupture within one day and are replaced by small ulcers that eventually coalesce to form larger map-like shallow ulcerations. The lesions are accompanied by fever and cervical lymphadenopathy.
Patient-reported symptoms: Arthralgia, malaise, moderate to severe pain in mouth and throat
Patient-reported duration: Lesion(s) usually present for at most 10-14 days.

HERPES LABIALIS, specify site, oral

260032 PROBABLE
Clinical descriptors: Single or multiple vesicles or ulcers with crusting on vermillion portion of lips and adjacent facial skin.
Patient-reported symptoms: Usually mild to moderate pain.
Patient-reported duration: Lesion(s) usually present for at most 10-14 days. Prior history of (or recurrent) lesion(s).

RECURRENT INTRA-ORAL HERPES SIMPLEX

260042 PROBABLE
Clinical descriptors: Solitary, or cluster of multiple or confluent ulcers that may be noted together with vesicles on keratinized mucosa, including hard palate, attached gingiva, and dorsum of tongue. Exceptionally, non-keratinized tissue may be involved. Round to slightly irregular (map-like) margins with minimal to no erythematous halos are present. The base of the ulcers is usually pink.
Patient-reported symptoms: Usually mild to moderate pain.
Patient-reported duration: Lesion(s) usually present for at most 10-14 days. Prior history of (or recurrent) lesion(s).

ORAL WARTS

260052 PROBABLE
Clinical descriptors: Mucosal color or white, solitary or multiple (often clustered) raised lesions that range in texture as they may be smooth, spiky, or cauliflower-like, and located in any part of the oral cavity.
Patient-reported symptoms: Usually asymptomatic. NOTE: Warts on the buccal or labial mucosa or tongue may get traumatized by biting, and may be painful.
Patient-reported duration: Lesion(s) usually long-standing.
IDIOPATHIC CONDITIONS

RECURRENT APHTHOUS STOMATITIS

260062 PROBABLE
Clinical descriptors: Single or multiple, white/yellow well circumscribed, painful ulcer(s) on non-keratinized tissue. A red halo is usually present around each ulcer. Minor aphthous ulcers may be 0.2 to 0.5 cm (centimeter) in diameter, while major aphthous ulcers are greater than (> 0.5 cm (centimeter) (may be as large as two cm (centimeter) in diameter).
Patient-reported symptoms: Moderate to severe pain, especially upon eating.
Patient-reported duration: Each minor ulcer usually lasts 7-10 days, while major aphthous ulcers may last for weeks. Patient reports a long-term history of recurrent ulcers.

ULCERATIONS NOS (NOT OTHERWISE SPECIFIED) / NECROTIZING ULCERATIVE STOMATITIS, specify ulcerations or necrotizing ulcerative stomatitis

260072 PROBABLE
Clinical descriptors: Large (greater than (> 0.5 cm (centimeter) and sometimes up to three cm (centimeter)) ulceration(s) with white/yellow necrotic base that may be located on either keratinized or non-keratinized mucosa. (NOTE: clinical appearance is similar to that of major aphthous ulcer, but there is no history of recurrent lesions). Necrotizing ulcerative stomatitis presents as localized, painful ulceronecrotic lesions of the oral mucosa that exposes underlying bone or penetrates or extends into contiguous tissues. These lesions may extend from areas of necrotizing periodontitis.
Patient-reported symptoms: Severe pain may be a prominent feature.
Patient-reported duration: Sudden onset, but may be long-standing or recurrent.

LINEAR GINGIVAL ERYTHEMA (LGE)

260082 PROBABLE
Clinical descriptors: Band shaped or punctate erythema characterized by distinctive erythema of the free and attached gingiva and alveolar mucosa. Erythema is disproportionately intense in relation to the plaque accumulation, which may be minimal.
Patient-reported symptoms: May bleed upon tooth brushing, or spontaneously in severe cases
Patient-reported duration: May be long standing. Lineal gingival erythema is resistant to local treatment measures.
APPENDIX 100 - Diagnoses Appendix

BACTERIAL INFECTION

NECROTIZING ULCERATIVE GINGIVITIS OR PERIODONTITIS

260092 PROBABLE
Clinical descriptors: Destruction of one or more interdental gingival papillae. In the acute stage of the process ulceration, necrosis, and sloughing may be seen with ready hemorrhage and characteristic fetid odor. In the case of Necrotizing Ulcerative Periodontitis, the condition is characterized by soft tissue loss as a result of ulceration or necrosis. Exposure, destruction or sequestration of alveolar bone may be seen, and the teeth may become loosened.
Patient-reported symptoms: Moderate to severe pain may be a prominent feature.
Patient-reported duration: Usually sudden onset and rapidly worsening.

NEOPLASMS

ORAL KAPOSI SARCOMA

260102 PROBABLE
Clinical descriptors: Early lesions are typically flat (or macular) with color ranging from red to purple. At a later stage lesions become nodular, raised and ulcerated. Predominantly seen on the palate or gingiva.
Patient-reported symptoms: At early stage, lesions are asymptomatic. Mild to moderate pain may develop as lesions become nodular and ulcerated. Local trauma to more advanced lesions may induce bleeding.
Patient-reported duration: Nodular lesions are long-standing

ORAL NON-HODGKIN’S LYMPHOMA

260112 PROBABLE
Clinical descriptors: A firm elastic, often somewhat reddish swelling, with or without ulceration. The gingiva, palatal mucosa, and fauces are sites of predilection. (The fauces are the two pillars of mucous membrane, the palatoglossal arch on the anterior and the palatopharyngeal arch on the posterior, surrounding the palatine tonsils).
Patient-reported symptoms: At early stage, lesions are usually asymptomatic. Moderate to severe pain may develop as lesions become ulcerated.
Patient-reported duration: Ulcerated lesions and swellings are long-standing.

SALIVARY GLAND DISEASE

PAROTID ENLARGEMENT

200282 PROBABLE
Clinical descriptors: Enlargement of the parotid glands that may result in facial disfigurement, usually bilateral.
Patient-reported symptoms: Usually asymptomatic enlargement. Patient may report xerostomia (perception of dry mouth).
Patient-reported duration: Usually long-standing.
## XVII. PERINATAL/PREGNANCY

### ABORTION, SPONTANEOUS/MISCARRIAGE

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
<th>Details</th>
</tr>
</thead>
<tbody>
<tr>
<td>270011</td>
<td>CONFIRMED</td>
<td>Loss of a pregnancy at less than (&lt;) 20 weeks gestation either spontaneously or through medical or surgical procedure after documentation of no fetal heart activity</td>
</tr>
</tbody>
</table>

### ABORTION, THERAPEUTIC, elective/induced

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
<th>Details</th>
</tr>
</thead>
<tbody>
<tr>
<td>270021</td>
<td>CONFIRMED</td>
<td>Termination of pregnancy prior to viability utilizing a medical or surgical procedure.</td>
</tr>
</tbody>
</table>

### BLEEDING, VAGINAL < 28 WEEKS

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
<th>Details</th>
</tr>
</thead>
<tbody>
<tr>
<td>270031</td>
<td>CONFIRMED</td>
<td>Any vaginal bleeding occurring during pregnancy prior to 28 weeks gestation and prior to the onset of labor</td>
</tr>
</tbody>
</table>

### BLEEDING, VAGINAL ≥ 28 WEEKS

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
<th>Details</th>
</tr>
</thead>
<tbody>
<tr>
<td>270041</td>
<td>CONFIRMED</td>
<td>Any vaginal bleeding occurring during pregnancy at or after 28 weeks gestation and prior to the onset of labor</td>
</tr>
</tbody>
</table>

### CORD PROLAPSE

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
<th>Details</th>
</tr>
</thead>
<tbody>
<tr>
<td>270051</td>
<td>CONFIRMED</td>
<td>Documentation of protrusion of the umbilical cord through the cervical os</td>
</tr>
</tbody>
</table>

### FEBRILE MORBIDITY, specify intrapartum or postpartum

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
<th>Details</th>
</tr>
</thead>
<tbody>
<tr>
<td>270061</td>
<td>CONFIRMED INTRAPARTUM</td>
<td>One of the following:</td>
</tr>
<tr>
<td></td>
<td></td>
<td>1. Oral, aural/tympanic or forehead temperature greater than or equal to (≥) 100.4º F or 38º C or</td>
</tr>
<tr>
<td></td>
<td></td>
<td>2. Rectal temperature greater than or equal to (≥) 100ºF or 38.3ºC</td>
</tr>
<tr>
<td></td>
<td>POSTPARTUM</td>
<td>Oral, aural/tympanic, or forehead temperature greater than or equal to (≥) 100.4º F or 38ºC on any two occasions 4 hours apart from greater than (&gt; ) 24-hours post delivery through 10 days postpartum.</td>
</tr>
</tbody>
</table>
ECLAMPSIA

270071 CONFIRMED
1. Seizure during pregnancy in the absence of any underlying known etiology or without any known reason for seizure
   and
2. No suspicion of epilepsy or trauma

PRE-ECLAMPSIA

270081 CONFIRMED
1. Both of the following:
   a. Must occur after 20 weeks of gestation
   and
   b. Blood pressure persistently greater than or equal to ($\geq$) 140/90 mm Hg.
   and
2. At least one of the following:
   a. Proteinuria of greater than or equal to ($\geq$) 1+ by dipstick, on two separate occasions
   or
   b. Greater than or equal to ($\geq$) 300mg protein in 24-hour collection

HELLP SYNDROME (hemolysis, elevated liver enzymes, low platelets)

270091 CONFIRMED
This diagnosis should be reviewed by an obstetrician for confirmation before being reported.
All of the following:
1. The diagnosis must be made after 20 weeks gestation
   and
2. One of the following:
   a. For women with no hypertension or proteinuria before 20 weeks gestation: Pregnancy associated hypertension consisting of a diastolic blood pressure of 90 mm Hg or greater on two occasions, 4 hours to one week (or 168 hours) apart
   or
   b. For women with hypertension but no proteinuria before 20 weeks gestation: no hypertension requirement
   or
   c. For women with proteinuria but no hypertension before 20 weeks gestation: no hypertension requirement
   and
3. All of the following:
   a. Thrombocytopenia: at least one platelet count less than (<) 100,000/ mm$^3$ (cubic millimeter) or less than (<) 100,000 x $10^9$/L (liter; SI units)
   and
   b. AST/SGOT greater than (>) 70 U per liter (U/L) above the upper limit of normal (ULN)
c. Hemolysis: LDH greater than or equal to ($\geq$) 600 U/L (units/liter) or total bilirubin concentration greater than or equal to ($\geq$) two milligrams per deciliter (mg/dL) or greater than or equal to ($\geq$) 34.2 µmol/L (micromoles/Liter; SI units) or a peripheral smear with nucleated RBCs or schistocytes

**HYPERTENSION, CHRONIC, IN PREGNANCY**

110141 CONFIRMED

At least one of the following:

1. Blood pressure persistently greater than or equal to ($\geq$) 140/90 mm Hg that began prior to pregnancy or in the first 20 weeks of pregnancy

   or

2. On anti-hypertension medication at the onset of pregnancy

**NOTE:** See also CHRONIC HYPERTENSION IN PREGNANCY in the Cardiovascular Disease section.

**HYPERTENSION, PREGNANCY INDUCED**

110151 CONFIRMED

1. Blood pressure persistently greater than or equal to ($\geq$) 140/90 mm Hg WITHOUT proteinuria

   and

2. Onset after first 20 weeks gestation with no hypertension prior to pregnancy

**NOTE:** See also PREGNANCY INDUCED HYPERTENSION in the Cardiovascular Disease section.

**HEMATOMA, VAGINAL OR VULVAR,** specify site, either vaginal or vulvar

270101 CONFIRMED

Documentation of a collection of blood

**HEMORRHAGE WITH HEMODYNAMIC INSTABILITY INTRAPARTUM**

270111 CONFIRMED

1. Bleeding and hypotension (blood pressure less than ($<$) 90/60 mm Hg)

   or

2. Maternal heart rate greater than ($>$) 120 beats per minute (BPM) and required treatment with fluid/volume expanders
HEMORRHAGE REQUIRING SURGICAL PROCEDURE INTRAPARTUM

270121 CONFIRMED
Bleeding that necessitates surgical intervention, such as dilation and curettage, hysterectomy or uterine artery ligation or embolization

HEMORRHAGE REQUIRING TRANSFUSION, INTRAPARTUM

270131 CONFIRMED
1. Bleeding with estimated maternal blood loss of
   a. Greater than (>) 750 mL in vaginal delivery
      or
   b. Greater than (>) 1200 mL in caesarean delivery
      and
2. Necessitates transfusion intrapartum

HEMORRHAGE WITH HEMODYNAMIC INSTABILITY POSTPARTUM

270141 CONFIRMED
All of the following:
1. Postpartum maternal hemorrhage with estimated maternal blood loss of greater than (>) 750 milliliters (mL) in vaginal delivery or greater than (>) 1200 milliliters (mL) in caesarean delivery
   and
2. Hemodynamic instability
   and
3. Blood pressure (BP) less than (<) 90/60 mm Hg or heart rate (HR) greater than (>) 120 beats per minute (BPM)
   and
4. Treated with fluid/volume expanders

HEMORRHAGE REQUIRING SURGICAL PROCEDURE POSTPARTUM

270151 CONFIRMED
1. Bleeding with estimated maternal blood loss of greater than (>) 750 mL (milliliters) in vaginal delivery or greater than (>) 1200 mL in Caesarean delivery
   and
2. Requires additional surgery such as dilation and curettage, hysterectomy or uterine artery ligation or embolization to control bleeding

Examples include retained placenta requiring curettage, placenta accreta requiring hysterectomy, and vaginal lacerations requiring repair in an operating room
HEMORRHAGE REQUIRING TRANSFUSION, POSTPARTUM

270161 CONFIRMED
1. Bleeding with estimated maternal blood loss of greater than (> 750 mL (milliliters) in vaginal delivery or greater than (> 1200 mL in cesarean delivery

   and

2. Necessitates transfusion to maintain hemodynamic stability as defined by at least one of the following:
   a. To correct blood pressure (BP) less than (<) 90/60 mm Hg or heart rate (HR) greater than (> 120 beats per minute (BPM)
   
   or
   b. To maintain hematocrit greater than (> 20 percent

INCOMPETENT CERVIX

270171 CONFIRMED
History consistent with incompetent cervix or current exam by physical diagnosis or imaging study as determined by experienced clinician

INCOMPETENT CERVIX, PROPHYLACTIC CERCLAGE

270181 CONFIRMED
History consistent with incompetent cervix, resulting in prophylactic cerclage placement

INCOMPETENT CERVIX, EMERGENCY CERCLAGE

270191 CONFIRMED
History consistent with incompetent cervix, resulting in emergency cerclage placement

INTRAUTERINE FETAL DEMISE, specify gestational age at diagnosis of fetal death

270201 CONFIRMED
Intrauterine death or stillbirth at greater than or equal to (≥) 20 weeks gestational age

INTRAUTERINE GROWTH RESTRICTION (IUGR) FETAL

270211 CONFIRMED
Based on ultrasound with estimated fetal weight less than or equal to (≤) 10th percentile for gestational age
INTRAUTERINE GROWTH RESTRICTION (IUGR) FETAL, SEVERE

270221 CONFIRMED
Based on ultrasound with estimated fetal weight less than or equal to ($\leq$) 3rd percentile for gestational age

SIGNIFICANT GROWTH LAG

270231 CONFIRMED
At least one of the following:
1. Fetal growth lagging requiring treatment that does not meet the requirements for intrauterine growth restriction (IUGR)
   or
2. Necessitates delivery

OLIGOXYDRAMNIOIS

270241 CONFIRMED
At least one of the following:
1. Amniotic fluid index (AFI) less than (<) five cm (centimeter) or largest vertical pocket less than (<) two cm (centimeter)
   or
2. Diagnosis by ultrasound without AFI information

POLYHYDRAMNIOIS

270251 CONFIRMED
At least one of the following:
1. Amniotic Fluid Index (AFI) greater than or equal to ($\geq$) 25 cm (centimeter) or maximum vertical pocket less than (<) eight cm (centimeters)
   or
2. Diagnosis by ultrasound without AFI information

PLACENTAE, ABRUPTIO

270261 CONFIRMED
At least one of the following:
1. Examination of the placenta at delivery reveals retroplacental clot
   or
2. Clinical diagnosis in study participant with at least two of the following:
   a. Vaginal bleeding
      or
   b. Uterine tenderness without other evidence of chorioamnionitis
      or
c. Hypercontractility

or
d. Hypertonus

PLACENTA ACCRETA (Total or partial)

270271 CONFIRMED
Documentation of placental villi invasion of the myometrium at the site of implantation and leading to obliteration of the normal cleavage plane

PLACENTA INCRETA

270281 CONFIRMED
Documentation of abnormal placental implantation with the villi extending into the myometrium

PLACENTA PERCRETA

270291 CONFIRMED
Documentation of invasion of villi through the full thickness of the myometrium

PLACENTA PREVIA

270301 CONFIRMED
Documentation that the placenta overlies the cervical os by one of the following:
1. By ultrasound

or

2. At delivery

or

3. At time of Caesarean section.

PREGNANCY, ECTOPIC

270311 CONFIRMED
Implantation of the fertilized ovum outside the uterine cavity

PREGNANCY, INTRAUTERINE

270321 CONFIRMED
Implantation of the fertilized ovum inside the uterine cavity

PREGNANCY, POST-TERM

270331 CONFIRMED
Pregnancy at greater than or equal to (≥) 42-weeks gestation
PRETERM LABOR

270341 CONFIRMED
Uterine contractions after 20 weeks and before 37 weeks necessitating tocolytic therapy or resulting in delivery

PREMATURE RUPTURE OF MEMBRANES, PRETERM

270351 CONFIRMED
Both of the following:
1. Spontaneous rupture of membranes less than (<) 37 weeks and
2. Must be documented by one of the following:
   a. Visualizing a pool of amniotic fluid in the vagina or
   b. Gross leakage of amniotic fluid from the vagina or
   c. Positive peri-pad test after installation of indigo carmine dye or
   d. Elevated pH in addition to at least one other listed criteria or
   e. Ferning of dried fluid on a microscope slide or
   f. History consistent with premature rupture of membranes or
   g. Decreased amniotic fluid volume on ultrasound with no other explanation for the oligohydramnios

270352 PROBABLE
Clinically suspected but not confirmed

PREMATURE RUPTURE OF MEMBRANES, TERM

270361 CONFIRMED
Both of the following:
1. Spontaneous rupture of membranes greater than or equal to (≥) 37 weeks and
2. Must be documented by one of the following:
   a. Visualizing a pool of amniotic fluid in the vagina or
   b. Gross leakage of amniotic fluid from the vagina or
   c. Positive peri-pad test after installation of indigo carmine dye or
d. Elevated pH in addition to at least one other listed criteria

or

e. Ferning of dried fluid on a microscope slide

or

f. History consistent with premature rupture of membranes

or

g. Decreased amniotic fluid volume on ultrasound with no other explanation for the oligohydramnios

270362 PROBABLE
Clinically suspected but not confirmed

PRETERM DELIVERY

270371 CONFIRMED
Delivery before 37 completed weeks gestation

UTERINE ATONY

270381 CONFIRMED
Failure of the uterus to contract postpartum, requiring intervention

UTERINE INVERSION

270391 CONFIRMED
Clinical diagnosis by experienced provider

UTERINE RUPTURE

270401 CONFIRMED
Spontaneous rupture of pregnant uterus resulting in fetal distress, maternal hemorrhage, or extrusion of all or part of the fetus

UTERINE SCAR DEHISCENCE

270411 CONFIRMED
Separation of scar from prior uterine surgery without meeting any of the criteria for uterine rupture

GROUP B STREPTOCOCCAL INFECTION, ASYMPTOMATIC, specify during pregnancy or not pregnant

270421 CONFIRMED
Culture positive at relevant site for Group B streptococci without any symptoms (“carrier”)
GROUP B STREPTOCOCCAL INFECTION, SYMPTOMATIC, specify during pregnancy or not pregnant

270431 CONFIRMED
Both of the following:
1. Culture positive at relevant site for Group B streptococci
   and
2. Patient symptomatic (e.g., fever, malaise, uterine inflammation in women; shock, meningitis, bacteremia in infants)

CHOLESTASIS OF PREGNANCY

270442 PROBABLE
Intense itching typically occurring in late pregnancy and most likely involving hands and feet; thought to be due to blockage of the flow of bile

PRURITIC URTICARIAL PAPULES AND PLAQUES OF PREGNANCY (PUPPS)

270452 PROBABLE
Most common rash of pregnancy. Typical appearance is small red wheals or vesicles which start in the stretch marks and may spread over thighs, buttocks, breast, and arms. This rash is very pruritic but harmless to mother and baby. Clinical diagnosis only.

UTERINE LEIOMYOMA

270461 CONFIRMED
Diagnostic histopathology on biopsy or surgical pathology

270462 PROBABLE
Clinical findings on physical exam, visualization at surgery, or imaging studies consistent with leiomyoma

OTHER PREGNANCY RELATED DIAGNOSIS, NOT LISTED IN APPENDIX, specify diagnosis
When reporting other pregnancy-related diagnosis, use the following general guidelines for confirmed and probable diagnoses:

279001 Confirmed diagnosis criteria may include:
1. A compatible clinical syndrome plus any diagnostic evidence that supports the diagnosis
   or
2. There may be the diagnostic test confirmation of the diagnosis without a clinical syndrome. Examples of diagnostic tests are: Microbiologic; cytologic, or histologic evidence that supports the diagnosis or any currently-approved diagnostic test.
Probable diagnosis criteria may include:
1. A compatible clinical syndrome
   or
2. One or more test results from procedures generally accepted as supportive of the diagnosis in standard medical practice
   or
3. Initiation or recommendation of specific therapy when appropriate
XVIII. PSYCHOLOGICAL SYMPTOMS AND DISORDERS

NOTE: There is no distinction between confirmed and probable levels of evidence for this section of the document.

PSYCHOLOGICAL SYMPTOMS
The following are symptoms that may be encountered as a feature of disease, adjustment to diagnosis, or side effects of medication. Generally speaking, symptoms are isolated experiences and do not cause significant impairment. If there are multiple symptoms or if symptoms are causing significant impairment, consideration should be given to reviewing possible psychiatric/psychological diagnoses found below under DISORDERS.

280018 ANXIETY
Anxiety is defined as persistent and excessive anxiety and worry for at least 10 out of 14 consecutive days. If significant impairment is noted, may meet criteria for Anxiety Disorder (see below).

280028 DEPRESSED MOOD
Depressed mood is defined a low or sad mood for most of the day for at least 10 of past 14 days. If accompanied by other symptoms and causes impairment may meet criteria for a Mood Disorder (see below).

280038 DELUSIONS
Delusions are defined as erroneous beliefs that usually involve a misinterpretation of perceptions or experiences. Persecutory, religious, grandiose or delusions of reference (belief that certain gestures, messages are intended as a special meaning for them individually).

280048 DREAM ABNORMALITY
Dream abnormality is defined as new onset of abnormal dreams which are described by the study participant as vivid, bizarre, or frightening and which have been present for at least 4 days out of seven consecutive days.

280058 DROWSINESS
Drowsiness is defined as new or worsening pathologic increase in absolute sleep hours by 25% (percent) for more than seven days.

280068 HALLUCINATIONS
Hallucinations are defined as presence of false visual, auditory, tactile, olfactory, or gustatory perceptions that have no basis in external stimulation.
280078  **HYPOMANIA/MANIA**

**All of the following:**

1. A hypomanic episode is defined as a distinct period of abnormal and persistently elevated, expansive mood lasting at least three out of 4 consecutive days. A manic episode is defined as a distinct period of abnormal and persistently elevated, expansive mood lasting at least six out of seven consecutive days.

2. Three or more of the following:
   a. Inflated self-esteem
   or
   b. decreased need for sleep (feels rested after only three hours of sleep)
   or
   c. pressure to keep talking
   or
   d. flight of ideas or racing thoughts
   or
   e. distractibility
   or
   f. increase in goal-directed activity
   or
   g. excessive and risky pleasure-seeking (e.g., unrestrained buying sprees, sexual indiscretions, foolish investments, etc.).

3. If a hypomanic/manic episode is part of a more pervasive mood disorder, consider BIPOLAR DISORDER (see below).

280088  **INSOMNIA**

Insomnia is characterized by difficulty in falling asleep or in staying asleep or by disturbed sleep patterns resulting in insufficient sleep for more than 10 of nights the past 14 nights.

280098  **PSYCHOSIS**

Psychosis is a condition of significantly disturbed mental status characterized by hallucinations, delusions or other impairments in reality testing. May be due to effects of substances, medical illness, or primary psychiatric disorder (see below).

**PSYCHOLOGICAL/PSYCHIATRIC DISORDERS**

Specific Diagnoses are categorized by primary symptoms. In general the required symptoms must be at a level that causes a clinically significant impairment or distress. This is not a complete list of psychiatric disorders, but these are the most common mental disorders seen in primary care settings. More complete information about these disorders and other disorders not included may be found in DSM-IV.
280108  ADJUSTMENT DISORDERS, specify category (see below):

All of the following:
1. Characterized by the development of clinically significant emotional or behavioral symptoms in response to an identifiable psychosocial stressor.
   and
2. The symptoms develop within three months of the onset of the stressor
   and
3. One of the following:
   a. Are in excess of what would be considered based on the nature of the stress or
   b. By significant impairment in functioning.

The diagnosis should not be used if the symptoms meet criteria for another mental disorder such as Major Depressive Disorder, Anxiety Disorder, etc. or are exacerbations of existing disorders.

Categories for adjustment disorders include:
- Adjustment Disorder with Depressed Mood
- Adjustment Disorder with Anxiety
- Adjustment Disorder with Anxiety and Depressed Mood
- Adjustment Disorder with Disturbance of Conduct
- Adjustment Disorder with Mixed Disturbance of Emotions and Conduct

ANXIETY DISORDERS

280118  GENERALIZED ANXIETY DISORDER
Characterized by excessive worry and anxiety for more days than not for at least six months duration. The anxiety is hard to control.
Anxiety associated with three of six symptoms (only one required for children less than (<) 18 years):
1. Restlessness, feeling on edge or
2. Easily fatigued or
3. Difficulty with concentration or
4. Irritability or
5. Muscle tension or
6. Sleep disturbance (difficulty falling asleep, staying asleep, or restless sleep)
**280128 PANIC DISORDER**
Characterized by recurrent, spontaneous episodes of panic that are associated with physiological and psychological symptoms. Panic attacks are discrete periods of intense fear or discomfort characterized by **four or more symptoms:**
1. Palpitations, pounding heart, or tachycardia  
   or
2. Sweating  
   or
3. Trembling or shaking  
   or
4. Feelings of dyspnea or smothering  
   or
5. Feeling of choking  
   or
6. Chest pain  
   or
7. Nausea, abdominal discomfort  
   or
8. Feeling dizzy, unsteady, lightheaded, or faint  
   or
9. Derealization or depersonalization  
   or
10. Fear of losing control or going crazy  
   or
11. Fear of dying  
   or
12. Paresthesias  
   or
13. Chills or hot flashes

**280138 POST-TRAUMATIC STRESS DISORDER**
Characterized by a pattern of arousal and avoidance following exposure to a traumatic event in which the person experienced or witnessed an event that involved actual or perceived serious injury or death. The response involves fear, helplessness or horror (or agitation in children). **All of the following:**
1. The trauma is re-experienced in one or more ways:  
   a. Recurrent and intrusive recollections (repetitive play in children)  
   b. Recurrent dreams of the event  
   c. Acting or feeling as if the event is recurring (flashbacks or other experiences)  
   d. Intense distress to cues that symbolize the event  
   e. Physiologic reactions following exposure to cues
   and
2. Avoidance of stimuli as indicated by three or more of the following:
   a. Efforts to avoid thoughts, feelings, or conversations about trauma
   b. Efforts to avoid activities that arouse recollections
   c. Inability to recall important aspects of the trauma
   d. Diminished interest in significant activities
   e. Feelings of detachment or estrangement
   f. Restricted range of feelings/affect
   g. Sense of foreshortened future
   
\textit{and}

3. Persistent symptoms of arousal manifested by two or more of the following:
   a. Sleep difficulties
   b. Outbursts of anger
   c. Difficulty concentrating
   d. Hypervigilance (“on-edge” or hyper-alert)
   e. Exaggerated startle response

\textbf{280148 ACUTE STRESS DISORDER}

\textbf{Both of the following:}

1. Characterized by a pattern of anxiety, dissociation, and other symptoms occurring within a month of an extreme traumatic event.
   a. Either during or after the event the person experiences three or more of the following dissociative symptoms:
      1. Subjective sense of numbing or detachment
      2. Reduction in awareness of surroundings
      3. Serialization
      4. Depersonalization
      5. Dissociative amnesia
   \textit{and}

   b. The event is re-experienced by \textbf{at least one of the following}:
      1. Recurrent images, thoughts, or dreams
      2. Flashbacks
      3. Experience of reliving the event
      4. Distress on exposure to reminders of the event
   \textit{and}

2. The disturbance lasts at least two days and a maximum of 4 weeks and occurs within 4 weeks of event (Post-Traumatic Stress Disorder (PTSD) is considered otherwise.)
**280158 OBSESSIVE COMPULSIVE DISORDER**
Characterized by recurrent obsessions (repetitive thoughts) or compulsions (repetitive behaviors) that cause marked distress and consume more than an hour a day. The person (but not necessarily children) is aware the thoughts and behaviors are excessive.

1. Obsessions are defined by **all of the following**:
   a. Recurrent, persistent thoughts perceived as intrusive and cause anxiety
   b. The thoughts are not simply excessive worry about real problems
   c. The person attempts to suppress the thoughts or counteract them with other thoughts or actions
   d. The person realizes that the thoughts are products of his own mind and are not imposed from outside (psychotic thoughts, thought insertion)

2. Compulsions are defined by **both of the following**:
   a. Repetitive behaviors (hand washing, ordering, checking) or mental acts (counting, repeating words silently) are done in response to an obsession or done according to rigid rules
   b. The behaviors or mental acts are aimed at preventing distress

**280168 ANXIETY DISORDER DUE TO GENERAL MEDICAL CONDITION**
Characterized by anxiety that is judged to be due to physiologic effects of a medical condition

**ATTENTION DEFICIT AND DISRUPTIVE DISORDERS (USUALLY BEGINNING IN CHILDHOOD)**

**280178 ATTENTION DEFICIT HYPERACTIVITY DISORDER**
Characterized by early onset of problems of attention, impulse control, or hyperactivity.

1. **Six or more of the following** symptoms of attention:
   a. Often fails to give close attention or makes careless mistakes
   or
   b. Often has difficulty sustaining attention to tasks or play activities
   or
   c. Often seems to not listen when spoken to directly
   or
   d. Often does not follow through on directions
   or
   e. Often has difficulty organizing tasks or activities
   or
   f. Avoids, dislikes, or refuses to engage in activities that require sustained focus
   or
   g. Often loses things
   or

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h. Is often distracted by external stimuli

or

i. Is often forgetful

or

2. Six or more of the following symptoms of hyperactivity/impulsivity:
   a. Often fidgets or squirms in seat
   or
   b. Often leaves seat in classroom
   or
   c. Runs or climbs excessively in inappropriate situations
   or
   d. Often has difficulty playing quietly
   or
   e. Is often “on the go” as if “driven by a motor”
   or
   f. Often talks excessively
   or
   g. Often blurts out answers
   or
   h. Has difficulty waiting turns
   or
   i. Often interrupts or intrudes on others

and

3. Symptoms must be at level to cause impairment in more than one setting and some aspect of symptoms were present prior to age seven years.

280188  CONDUCT DISORDER

1. Characterized by a persistent and repetitive pattern of behavior in which the basic rights of others or major age-appropriate rules/norms are violated as evidenced by three or more of the following symptoms:
   a. Often bullies, threatens or intimidates others
   or
   b. Initiates fights
   or
   c. Has used a weapon to cause serious physical harm
   or
   d. Has been physically cruel to people
   or
   e. Has been physically cruel to animals
   or
   f. Has stolen while confronting a victim
   or
   g. Has forced someone into sexual activity
   or
h. Deliberate fire setting to cause damage
   or
i. Deliberate destruction of property
   or
j. Has broken into someone’s house or car
   or
k. Lies to obtains goods or favors
   or
l. Has stolen goods (more than trivial value) without confronting victim
   or
m. Curfew violations prior to age 13
   or
n. Running away from home overnight at least twice
   or
o. Often truant beginning before age 13

and

2. The behavior disturbance causes major difficulty in social, academic, or occupational settings.

280198  OPPOSITIONAL DEFIANT DISORDER
Characterized by a pattern of negative, hostile or defiant behavior lasting at least 6 months with four or more of the following symptoms:
1. Often loses temper
   or
2. Often argues with adults
   or
3. Often actively defies or refuses to comply with adult requests or rules
   or
4. Often deliberately annoys others
   or
5. Often blames others for mistakes
   or
6. Is often touchy or easily annoyed by others
   or
7. Is often angry or resentful
   or
8. Is often spiteful or vindictive.
**COGNITIVE DISORDERS**

**280208 DEMENTIA**
Characterized by the development of multiple cognitive deficits that are due to direct physiologic effects of a general medical condition, persisting effects of substance abuse or to multiple etiologies.
The multiple cognitive deficits are manifest by **both of the following:**
1. Memory impairment including inability to learn new information or to recall previously learned information

   and

2. **At least one of the following** cognitive problems:
   a. Aphasia (language disturbance)
   b. Apraxia (impaired ability to carry out motor functions despite intact motor functioning)
   c. Agnosia (failure to recognize or identify objects despite intact sensory function)
   d. Disturbance in executive functioning (planning, organizing, sequencing, or abstracting).

**280218 DELIRIUM**
Characterized by an acute, fluctuating disturbance of consciousness and change in cognition due to the direct effects of a general medical condition, effects of a substance (intoxication or withdrawal), exposure to a toxin or a combination of these as manifest by **all of the following:**
1. Disturbance of consciousness (reduced awareness of environment) with reduced ability to focus attention

   and

2. A change in cognition (memory deficits, disorientation, language disturbance) or perceptual disturbance that is not better accounted for by a pre-existing dementia

   and

3. The disturbance develops over a short period of time (hours to days) and fluctuates during the course of the day.

   and

4. There is evidence the disturbance is caused by direct physiologic effects of medical condition, substance or other physiologic impairment.

**EATING DISORDERS**

**280228 ANOREXIA NERVOSA**
Characterized by **all of the following:**
1. Refusal to maintain body weight above minimal norms for age and height or failure to gain expected weight during growth period

   and

2. Intense fear of gaining weight even though underweight
3. Disturbed body image, undue influence of weight or shape on self-evaluation or denial of seriousness of current low body weight and
4. Amenorrhea in post-menarchal females

280238 BULIMIA NERVOSA
Characterized by all of the following:
1. Recurrent episodes of binge eating (large amounts of food eaten in relatively short time frame and a sense of lack of control over eating during the episode and
2. Recurrent inappropriate compensatory behavior to prevent weight gain (self-induced vomiting, misuse of laxatives, fasting, excessive exercise) and
3. Binge eating and compensatory behaviors occur at least twice a week for three months

MOOD DISORDERS

280248 MAJOR DEPRESSIVE DISORDER
1. Characterized by five or more of the following symptoms present during a 14-day period with at least one symptoms being depressed/irritable mood or loss of interest in pleasurable activities:
   a. Depressed mood for most of the day; for children and adolescents can be persistently irritable mood
   b. Diminished interest or pleasure in usual activities
   c. Marked decrease or increase in appetite
   d. Daily sleep problems (increased, decreased, or lack of restful sleep)
   e. Observable restlessness or slowed behavior
   f. Fatigue, loss of energy
   g. Feelings of worthlessness, excessive guilt
   h. Decreased concentration
   i. Recurrent thoughts of death, including suicidal thoughts and
2. The symptoms must cause distress or impairment and are not the direct effect of medication or physiologic illness.

280258 DYSTHYMIC DISORDER
A chronic form of depressive disorder with similar, although usually milder symptoms of depression that have been present for at least a year (two years in adults).
280268  **BIPOLAR DISORDER**

1. Characterized by cycles of disturbed mood that include episodes of depressive episodes (see symptoms above) and manic episodes. Manic episodes include abnormally and persistently elevated, expansive, or irritable mood lasting at least seven days with **three or more of the following** symptoms to a significant degree:
   a. Inflated self esteem or grandiosity
   b. Decreased need for sleep (not inability to get to sleep)
   c. More talkative than usual, pressured need to keep talking
   d. Flight of ideas or racing thoughts
   e. Distractibility
   f. Increased activity levels
   g. Excessive involvement in pleasurable activities that have high potential for painful consequences (sexual indiscretions, foolish business investments, unrestrained spending sprees)

2. The symptoms are severe enough to cause marked impairment in functioning or require hospitalization to prevent harm to self or others.

3. The symptoms are not due to direct effects of medication or drugs nor a part of a medical illness.

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280278  **PAIN DISORDER**

Characterized by **all of the following**:

1. Pain in one or more sites as the predominant focus of clinical presentation and is of sufficient severity to warrant clinical attention

2. Pain causes significant distress or impairment in social, occupational, or other important areas

3. Psychological factors are judged to have an important role in the onset, severity, exacerbation, or maintenance of the pain

4. The pain is not intentionally produced or feigned (as in malingering)

5. The pain is not better accounted for as a part of a mood, anxiety, or psychotic disorder.
PERVASIVE DEVELOPMENTAL DISORDERS

280288 AUTISTIC DISORDER

Characterized by severe and pervasive impairment in development of social interactions, communication skills, and other behaviors as beginning before age three years and manifested by six or more of the following (with at least two symptoms of (1) impaired social interactions and (2) at least one communication problem and (3) stereotyped behavior problem:

1. Qualitative impairment in social interaction
   At least two of the following:
   a. Marked impairment in use of non-verbal interactions such as eye contact, facial expressions, or body postures
   or
   b. Failure to develop developmentally appropriate peer relationships
   or
   c. Lack of social or emotional reciprocity

2. Qualitative impairments in communication
   At least one of the following:
   a. Delay or lack of spoken language without attempt to compensate through gesture
   or
   b. If speech exists there is marked impairment in conversation
   or
   c. Stereotyped and repetitive use of language in idiosyncratic ways
   or
   d. Lack of spontaneous make-believe or imitative play

3. Restrictive repetitive and stereotyped behaviors, interests, or activities
   At least one of the following:
   a. Encompassing preoccupation with one or more patterns of interest that is abnormal in intensity or focus
   or
   b. Inflexible adherence to nonfunctional routines or rituals
   or
   c. Stereotyped movements (e.g., handwringing, spinning, or hand flapping)
   or
   d. Persistent preoccupation with parts of objects
280298  **ASPERGER’S DISORDER**

Characterized by qualitative abnormalities in development of social interactions, communication and stereotyped interests but speech is preserved (as opposed to Autistic Disorder). The use of language may be idiosyncratic with odd use of language. Asperger’s Disorder is sometimes referred to as “high functioning autism.”

1. Symptoms of qualitatively impaired social interactions
   
   **At least two of the following:**
   
   a. Marked impairment in use of non-verbal behaviors
   
   *or*
   
   b. Failure to develop developmentally appropriately peer relationships
   
   *or*
   
   c. Lack of spontaneous sharing of enjoyment, interests, or achievements with other people
   
   *or*
   
   d. Lack of social or emotional reciprocity

   **and**

2. Symptom of restrictive repetitive or stereotyped behavior or interests

   **At least one of the following:**

   a. Encompassing preoccupation with one or more restricted behaviors or interests
   
   b. Inflexible adherence to specific nonfunctional routines or rituals
   
   c. Stereotyped and repetitive movements (e.g., handwringing, spinning, or hand flapping)
   
   d. Persistent preoccupation with parts of objects

---

**PSYCHOTIC DISORDERS**

280308  **SCHIZOPHRENIA**

1. Characterized the presence of **two or more of the following** symptoms of psychosis for a significant portion of a one-month period:

   a. Delusions
   
   *or*
   
   b. Hallucinations
   
   *or*
   
   c. Disorganized speech (derailment or incoherence)
   
   *or*
   
   d. Grossly disorganized or catatonic behavior
   
   *or*
   
   e. Flattened affect, social isolation, marked inactivity

   **and**

2. The symptoms are severe enough to cause major disturbance of functioning in one or more major areas of life (personal, school, work, etc). Usually there is a gradual several month period of deterioration prior to development of full psychotic symptoms.
PSYCHOSIS (PSYCHOTIC SYMPTOMS)
May be present in other mental illness including depressive disorders, manic episodes, and substance induced disorders, among others.

SUBSTANCE-RELATED DISORDERS
Substance-related disorders are categorized by the specific substance(s), possible direct physiologic effects of the substance (intoxication, psychosis, withdrawal, etc.) and by either abuse or dependence.

Commonly abused substances include:
- Alcohol
- Amphetamine
- Cannabis
- Cocaine
- Hallucinogens
- Inhalants
- Nicotine
- Opioids
- Phencyclidine
- Sedative/Hypnotic/Anxiolytics

SUBSTANCE ABUSE
Characterized by a maladaptive pattern of substance use leading to clinically significant impairment or distress manifested by one or more of the following in a 12-month period:
1. Recurrent substance use resulting in failure to fulfill major role obligations at work, school, or home
or
2. Recurrent substance use in situations in which it is physically hazardous (e.g., driving)
or
3. Recurrent substance-related legal charges
or
4. Continued substance use despite having experienced negative or recurrent social or interpersonal consequences of substance use.

SUBSTANCE DEPENDENCE
Characterized by a maladaptive pattern of substance use leading to impairment manifested by three or more of the following in the same 12-month period:
1. Tolerance for the substance, both of the following:
   a. A need for increasing amounts to achieve intoxication or desired effect
      and
   b. Diminished effect with continued use of the same amount of substance
or
2. Withdrawal symptoms, both of the following:
   a. The characteristic withdrawal syndrome for the substance
      and
   b. The same or closely related substance is taken to avoid withdrawal symptoms
or
3. Substance is taken in larger amounts or over longer period than was intended
   or
4. Persistent desire or unsuccessful efforts to cut down or control substance use
   or
5. Great deal of time is devoted to obtain substances or recover from their effects
   or
6. Important social occupational or recreational activities are given up or reduced because of
   substance use
   or
7. The substance use is continued despite knowledge of continuing negative effects of the
   substance.
**APPENDIX 100 - Diagnoses Appendix**

**XIX. RENAL DISORDERS**

**GLOMERULONEPHRITIS, IMMUNE MEDIATED**, e.g., IgA Nephritis, Lupus-like Nephritis

290011 CONFIRMED
Demonstrated by kidney biopsy

**HEMOLYTIC UREMIC SYNDROME**

290021 CONFIRMED
Acute renal failure associated with thrombocytopenia and microangiopathic hemolytic anemia, normal coagulation tests and the absence of neurologic disease, with characteristic histopathologic features by biopsy.

**INTERSTITIAL NEPHRITIS**

290031 CONFIRMED
Decreasing renal function associated with inflammation of the tubules and the spaces between the tubules and the glomeruli, demonstrated by kidney biopsy

290032 PROBABLE
Decreasing renal function with eosinophils in urine by Wright’s stain or Hansel’s stain, and possibly with peripheral eosinophilia. Typically associated with exposure to a drug such as: penicillins and cephalosporins, sulfonamides and sulfonamide-containing diuretics, NSAIDs, rifampin, phenytoin, and allopurinol and proton pump inhibitors.

**NEPHROCALCINOSIS**

290041 CONFIRMED
Deposition of calcium and oxalate or phosphate in the renal tubules and interstitium as demonstrated by radiographic studies

**NEPHROLITHIASIS**

290051 CONFIRMED
One of the following:
1. Presence of calculi in the kidney as demonstrated radiographic studies
   *or*
2. By passing a stone in the urine

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290052  PROBABLE
Both of the following:
1. Clinical syndrome only with typical intermittent flank pain, hematuria, but no stone recovered
2. Negative radiographic studies or radiographic studies not done

**NEPHROPATHY, DRUG-INDUCED**, specify drug
For tenofovir or adefovir– see PROXIMAL RENAL TUBULAR ACIDOSIS DYSFUNCTION (PRTD), in association with ART (Fanconi-Like Syndrome)

290061  CONFIRMED
In association with Indinavir or atazanavir or other suspected agents: renal insufficiency in association with characteristic crystalluria demonstrated by polarizing filters, in the absence of other causes of crystalluria

**NEPHROPATHY, HIV-ASSOCIATED**

290071  CONFIRMED
Collapsing glomerulosclerosis demonstrated by kidney biopsy

290072  PROBABLE
Both of the following:
1. Renal insufficiency in HIV-infected individuals in association with proteinuria (defined as protein excretion greater than (>) 1.0 gm (gram) per 24-hours or a Urine Protein to Urine Creatinine ratio greater than (>) 1.0)
2. No other likely causes of kidney disease including chronic hepatitis (B or C), diabetes, obstructive kidney disease, or connective tissue diseases

**NEPHROTIC SYNDROME**

290081  CONFIRMED
All of the following:
1. Urine protein excretion greater than (>) 3.0 grams per 24-hours (or Urine Protein to Urine Creatinine ratio greater than (>) 3.0)
2. Hypoalbuminemia (albumin less than (<) three g/dL (grams/deciliter))
3. Peripheral edema
PROXIMAL RENAL TUBULAR ACIDOSIS DYSFUNCTION (PRTD), in association with tenofovir and adefovir or other suspected agents (Fanconi-Like Syndrome)

290091 CONFIRMED
Both of the following:
1. A reduction in creatinine clearance (CrCl) by at least 25% (percent) from the CrCl at the start of the current ART regimen which includes a suspect drug such as tenofovir or adefovir

and
2. Evidence of proximal tubular injury defined as fulfilling at least two of the following conditions within 14 days of the finding of reduced CrCl:
   a. New onset or worsening of pre-existing proteinuria (greater than or equal to (\(\geq\)) 1+) on urinary dipstick

   or
   b. New onset or worsening of pre-existing glycosuria greater than or equal to (\(\geq\)) 1+ on urinary dipstick with corresponding serum glucose of less than (<) 200 mg/dL (milligrams/deciliter)

   or
   c. Low serum potassium, less than (<) 3.0 mEq/L (milliequivalents/liter)

   or
   d. Low serum bicarbonate, less than (<) 19 mEq/L in subjects with CrCL greater than or equal (\(\geq\)) to 25 mL/min (milliliters/minute)

   or
   e. Low serum phosphorus less than (<) 2.0 mg/dL (milligrams/deciliter)

NOTE: CrCl should be estimated using the Cockcroft-Gault equations as: Cockcroft-Gault: CrCl (ml/min) = ((140 – Age) x (Weight in kg) x (0.85 if female))/(72 x (Serum creatinine in mg/dL))

RENAL FAILURE, ACUTE

290101 CONFIRMED
Sudden inability of the kidney to appropriately regulate fluid and electrolyte homeostasis, resulting in an elevation of nitrogen waste products and serum creatinine

RENAL FAILURE, CHRONIC, specify with dialysis or without dialysis

290111 CONFIRMED
Progressive irreversible loss of renal function

For renal function, the glomerular filtration rate (GFR) can be mildly decreased (less than (<) 60ml/min/1.73m² (milliliters/minute/cubic meter)) to severely decreased (less than (<) 10ml/min/1.73m²).
## RENAL INSUFFICIENCY, ACUTE

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>290121</td>
<td>CONFIRMED Increases in serum creatinine to values greater than (&gt;\ ) 1.5 mg/dl (milligrams/deciliter) (or greater than (&gt;\ ) 1.0 - 1.3 times the upper limit of normal (ULN)) that return to normal values within three months</td>
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## RENAL INSUFFICIENCY, CHRONIC

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<th>Code</th>
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<tbody>
<tr>
<td>290131</td>
<td>CONFIRMED Increases in serum creatinine to values greater than (&gt;\ ) 1.5 milligrams per deciliter (mg/dl) (or greater than (&gt;\ ) 1.0 - 1.3 times the upper limit of normal (ULN) that persist for greater than (&gt;\ ) three months.</td>
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## RENAL TUBULAR ACIDOSIS

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<tbody>
<tr>
<td>290141</td>
<td>CONFIRMED Syndrome in which renal tubular defect leads to an inability to maintain normal plasma bicarbonate manifested by metabolic acidosis, specify cause</td>
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</tbody>
</table>

**NOTE:** See also PROXIMAL RENAL TUBULAR ACIDOSIS DYSFUNCTION in this section

## RHABDOMYOLYSIS

<table>
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<th>Description</th>
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<tbody>
<tr>
<td>290151</td>
<td>CONFIRMED Disorder involving injury to the kidney caused by toxic effects of the contents of injured muscle cells. Kidney function decreased and muscle enzymes increased (CK, aldolase).</td>
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## PERSISTENT ALBUMINURIA

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<th>Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>290161</td>
<td>CONFIRMED Defined as the documentation of at least two sequentially abnormal urine albumin measurements demonstrating microalbuminuria or as defined by an albumin/creatinine ratio (ACR) greater than or equal (≥\ ) 30 - 299 mg/g (milligrams/gram), 3.4 to 34 mg/mmol (milligrams/millimoles) for microalbuminuria, or a urinary albumin concentration greater than or equal to (≥\ ) 10 mg/L (milligrams/liter).</td>
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## PERSISTENT PROTEINURIA

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<th>Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>290171</td>
<td>CONFIRMED Defined as the documentation of at least two sequentially abnormal urine protein measurements, where the urine protein/creatinine ratio (nucleic acid amplification test) is greater than (&gt;\ ) 0.2.</td>
</tr>
</tbody>
</table>
XX. OTHER DISORDERS

300012 ARTHRITIS, JUVENILE IDIOPATHIC, formerly referred to Juvenile Rheumatoid Arthritis
Including but not limited to Still’s Disease, pauciarticular JIA, or systemic onset JIA.

Clinical diagnosis made by an experienced practitioner.

ARTHRITIS, ADULT NONINFECTIOUS

300022 CRYSTAL-INDUCED ARTHRITIS
300032 GOUTY ARTHRITIS
300042 OSTEOARTHRITIS
300052 REACTIVE ARTHRITIS
300062 RHEUMATOID ARTHRITIS
300072 ARTHRITIS, ADULT NONINFECTIOUS NOT LISTED ABOVE, EXCLUDES septic arthritis and HIV-associated arthritis

For each: Clinical diagnosis made by experienced practitioner

ASTHMA, ACUTE

300081 CONFIRMED
Both of the following:
1. Sudden onset of wheezing and cough; may or may not be associated with a concurrent infection, drug exposure, or exposure to another antigen
   and
2. Episode is self-limited; may respond to treatment

ASTHMA, CHRONIC

300091 CONFIRMED
Both of the following:
1. Two or more recurring episodes of sudden onset of wheezing and cough; may or may not be associated with a concurrent infection, drug exposure or exposure to another antigen
   and
2. Episode is self-limited; may respond to treatment
BONE FRACTURES

NOTE: All bone fractures are considered CONFIRMED.

300101 TRAUMATIC BONE FRACTURE AS THE RESULT OF A FALL FROM STANDING HEIGHT OR LESS, specify body site or specific bone (e.g., left arm, left leg, right femur, left clavicle, etc.)

300111 TRAUMATIC BONE FRACTURE FROM A HARD FALL, specify body site or specific bone (e.g., left arm, left leg, right femur, left clavicle, etc.)

300121 TRAUMATIC BONE FRACTURE FROM A CAR ACCIDENT OR OTHER SEVERE TRAUMA, specify body site or specific bone (e.g., left arm, left leg, right femur, left clavicle, etc.)

300131 NON-TRAUMATIC BONE FRACTURE, specify body site or specific bone (e.g., left arm, left leg, right femur, left clavicle, etc.)

300141 BONE FRACTURE OF UNKNOWN MECHANISM, specify body site or specific bone (e.g., left arm, left leg, right femur, left clavicle, etc.)

300151 NON-HEALING BONE FRACTURE, specify body site or specific bone (e.g., left arm, left leg, right femur, left clavicle, etc.)

BONE NECROSIS, AVASCULAR NECROSIS, specify site (e.g., hip, shoulder, knee) and either bone necrosis or avascular necrosis

300161 CONFIRMED
Both of the following:
1. Radiographic diagnosis, ideally confirmed by MRI or CT
2. No active infectious or other cause of bone abnormality

300162 PROBABLE
Radiographic diagnosis only

300178 CATARACT
CHRONIC OBSTRUCTIVE PULMONARY DISEASE (COPD)

300181 CONFIRMED
Prolonged or persistent respiratory dysfunction resulting in oxygenation or CO2 elimination at an inappropriate rate

300198 CONJUNCTIVITIS, ALLERGIC

FEVER OF UNKNOWN ORIGIN

300201 CONFIRMED
One of the following:
1. For adults (age greater than or equal to $\geq$ 15 years):
   Both of the following:
   a. A temperature higher than ($>$) 99.5°F (37.5°C) that lasts for more than one month
   and
   b. No obvious source despite appropriate investigation.
   or
2. For children (age less than ($<$) 15 years):
   Both of the following:
   a. Fever greater than ($>$) 99.5°F (37.5°C) for longer than one month
   and
   b. No diagnosis is apparent after initial outpatient or hospital evaluation that includes a careful history and physical examination and initial laboratory assessment

FIBROSIS, PULMONARY

300211 CONFIRMED
Both of the following:
1. Restrictive pulmonary physiology on PFT (Pulmonary Function Test) with CT imaging showing honeycomb changes along with interstitial thickening
   and
2. Lung biopsy showing extracellular collagen deposition consistent with pulmonary fibrosis

300212 PROBABLE
Both of the following:
1. Restrictive pulmonary physiology on PFT (Pulmonary Function Test)
   and
2. CT imaging showing honeycomb changes along with interstitial thickening
300408 INJECTION SITE REACTION

Any localized or systemic signs/symptoms that occur following an injection:
  • Localized signs/symptoms are signs/symptoms that occur at or near the injection site, including any of the following:
    o Ecchymosis
    o Edema,
    o Erythema,
    o Hematoma
    o Induration,
    o Nodule
    o Pain,
    o Swelling,
    o Tenderness
    o or any other localized sign/symptom not listed here that is thought to be related to an injection reaction (e.g., itching, warmth at or around site, hypersensitivity reaction, blistering).

  • Systemic signs/symptoms including any of the following:
    o Arthralgia
    o Chills
    o Fatigue
    o Fever greater than or equal to grade 1
    o Headache
    o Malaise
    o Myalgia
    o Nausea
    o Vomiting
    o Regional/localized lymphadenopathy
    o or any other systemic sign/symptom not listed here that is thought to be related to an injection reaction (e.g., dizziness).

MYOSITIS, specify type (pathogen, drug or other related) and location.

300221 CONFIRMED
Both of the following:
1. Inflammation of muscle tissue associated with signs and symptoms such as muscle pain or tenderness.

and
2. At least one of the following:
   a. Muscle enzymes levels are elevated (e.g., aldolase or CPK)
   or
   b. Characteristic findings are seen on biopsy, if performed

300222 PROBABLE
Inflammation of muscle tissue associated with signs and symptoms such as muscle pain or tenderness
<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Code</th>
<th>Description</th>
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</thead>
<tbody>
<tr>
<td><strong>NYSTAGMUS</strong></td>
<td>300231</td>
<td>Involuntary beating movement of the eyes</td>
</tr>
<tr>
<td><strong>OSTEONECROSIS, specify site</strong></td>
<td>300241</td>
<td>Necrosis of bone documented radiographically or with other bone testing</td>
</tr>
<tr>
<td><strong>OSTEOPENIA/OSTEOPOROSIS - MEN GREATER THAN OR EQUAL TO (≥) 50 YEARS OF AGE AND POSTMENOPAUSAL WOMEN</strong></td>
<td>300251</td>
<td>Bone mineral density (BMD) or bone mineral content (BMC) between one and 2.5 standard deviations (SD) below the young adult (gender-matched) reference population mean (T-scores between -1 and -2.5)</td>
</tr>
<tr>
<td></td>
<td>300261</td>
<td>Bone mineral density (BMD) or bone mineral content (BMC) 2.5 standard deviations (SD) or more below the young adult (gender-matched) reference population mean (T-scores less than or equal to (≤) -2.5)</td>
</tr>
<tr>
<td></td>
<td>300271</td>
<td>Bone mineral density (BMD) or bone mineral content (BMC) 2.5 standard deviations (SD) or more below the young adult (gender-matched) reference population mean (T-scores less than or equal to (≤) -2.5), in the presence of one or more fragility fractures.</td>
</tr>
<tr>
<td><strong>OSTEOPENIA/OSTEOPOROSIS-MEN LESS THAN (&lt;) 50 YEARS OF AGE, PREMENOPAUSAL WOMEN, ADOLESCENTS, AND CHILDREN</strong></td>
<td>300281</td>
<td>Bone mineral density (BMD) or bone mineral content (BMC) less than or equal to (≤) 2.0 standard deviations (SD) below the age-, gender-, and body size-matched reference population mean (Z-scores less than or equal to (≤) -2.0)</td>
</tr>
</tbody>
</table>
300291 OSTEOPOROSIS, severe
-Men less than (<) 50 years of age, premenopausal women, 
adolescents and children
Bone mineral density (BMD) or bone mineral content (BMC) less than or equal to (≤) 2.0 
standard deviations (SD) below the age-, gender-, and body size-matched reference population 
mean (Z-scores less than or equal to (≤) -2.0), and a history of clinically significant fractures 
(either one long-bone fracture of the lower extremity, or vertebral compression fracture, or two 
or more long-bone fractures of the upper extremities)

OPTIC NERVE ATROPHY (ONA)

300301 CONFIRMED
Both of the following:
1. A permanent visual impairment caused by damage to the optic nerve 
   and
2. Proven on fundoscopic exam

OPTIC NERVE HYPOPLASIA

300311 CONFIRMED
Both of the following:
1. A reduction in the number of optic nerve fibers within the optic nerve 
   and
2. Proven on fundoscopic exam

PIGMENTARY RETINOPATHY (RETINITIS PIGMENTOSA), specify either pigmentary 
retinopathy or retinitis pigmentosa

300321 CONFIRMED
Characteristic findings seen on fundoscopic exam by an experienced clinician

PNEUMONITIS, INTERSTITIAL

300331 CONFIRMED
Both of the following:
1. Symptoms of dyspnea with classic CT findings of interstitial thickening, with or without 
ground glass opacities, and with restrictive physiology on Pulmonary Function Test 
   and
2. Lung biopsy evidence of usual interstitial pneumonia (UIP), desquamative interstitial 
pneumonia (DIP), respiratory bronchiolitis associated interstitial lung disease (RB-ILD), 
Acute interstitial pneumonia (AIP, A.K.A. Hamman Rich), nonspecific interstitial 
pneumonia (NSIP), and cryptogenic organizing pneumonia (COP; formerly idiopathic 
BOOP)

300332 PROBABLE
Both of the following:
1. Symptoms of dyspnea with classic CT findings of interstitial thickening, with or without ground glass opacities

   and

2. Restrictive physiology on Pulmonary Function Test

**PNEUMOTHORAX**, specify which lung.

<table>
<thead>
<tr>
<th>Code</th>
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<tbody>
<tr>
<td>300341</td>
<td>CONFIRMED Collapse of the lung</td>
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**300358 RHINITIS, ALLERGIC**

**RETINITIS**, specify type (See also CMV RETINITIS in the Pathogen Diagnoses Section)

<table>
<thead>
<tr>
<th>Code</th>
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<tbody>
<tr>
<td>300368</td>
<td>PATHOGEN RELATED RETINITIS, specify pathogen</td>
</tr>
<tr>
<td>300378</td>
<td>DRUG RELATED RETINITIS, specify drug</td>
</tr>
<tr>
<td>309008</td>
<td>OTHER RETINITIS, specify</td>
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</table>

**WASTING SYNDROME**

**ADULTS (AGE GREATER THAN OR EQUAL TO (≥) 15 YEARS)**

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<thead>
<tr>
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<td>300381</td>
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<tr>
<td></td>
<td>Both of the following:</td>
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<tr>
<td></td>
<td>1. Documented weight loss greater than (&gt; 10% (percent) of body weight</td>
</tr>
<tr>
<td></td>
<td>and</td>
</tr>
<tr>
<td></td>
<td>2. At least one of the following:</td>
</tr>
<tr>
<td></td>
<td>a. Two or more unformed stools negative for pathogens</td>
</tr>
<tr>
<td></td>
<td>or</td>
</tr>
<tr>
<td></td>
<td>b. Documented temperature of greater than (&gt; 99.5°F (37.5°C) with no other cause of disease, negative blood culture, negative malaria slide, and normal or unchanged chest X-ray</td>
</tr>
</tbody>
</table>

<table>
<thead>
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<td>300382</td>
<td>PROBABLE</td>
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<tr>
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<td>Both of the following:</td>
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<tr>
<td></td>
<td>1. Unexplained involuntary weight loss (greater than (&gt; 10% (percent) baseline body weight) with obvious wasting or body mass index less than (&lt;) 18.5 kg/m² (kilograms/meter squared)</td>
</tr>
<tr>
<td></td>
<td>and</td>
</tr>
<tr>
<td></td>
<td>2. At least one of the following:</td>
</tr>
<tr>
<td></td>
<td>a. Unexplained chronic diarrhea (loose or watery stools three or more times daily reported for longer than one month)</td>
</tr>
<tr>
<td></td>
<td>b. Reports of fever or night sweats for more than one month without other cause and lack of response to antibiotics or antimalarial agents; malaria must be excluded in malarious areas</td>
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</tbody>
</table>
CHILDREN (AGE LESS THAN (<) 15 YEARS)

300391 CONFIRMED
Documented weight for height or weight for age of more than negative three (-3) standard deviations (SD) from the mean with or without edema

300392 PROBABLE (WHO-4)
Both of the following:
1. Persistent weight loss, stunting wasting or malnutrition not explained by poor or inadequate feeding, other infections and not adequately responding in two weeks to standard therapy
   and
2. At least one of the following:
   a. Visible severe wasting of muscles, with or without edema of both feet
   or
   b. Weight-for-height of negative three (-3) standard deviations (SD) from the mean, as defined by WHO Integrated Management of Childhood Illness guidelines.

OTHER DIAGNOSIS, any other diagnosis that is not listed in the appendix; specify

309011 Confirmed diagnosis criteria may include:
1. A compatible clinical syndrome plus any diagnostic evidence that supports the diagnosis
   or
2. There may be the diagnostic test confirmation of the diagnosis without a clinical syndrome. Examples of diagnostic tests are: Microbiologic; cytologic, or histologic evidence that supports the diagnosis or any currently-approved diagnostic test.

309012 Probable diagnosis criteria may include:
1. A compatible clinical syndrome
   or
2. One or more test results from procedures generally accepted as supportive of the diagnosis in standard medical practice
   or
3. Initiation or recommendation of specific therapy when appropriate
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<td>THROMBOCYTOPENIC PURPURA, IMMUNE</td>
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<td>THROMBOPHILEBITIS, OVARIAN VEIN</td>
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<tr>
<td>THROMBOPHILEBITIS, SEPTIC PELVIC</td>
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<tr>
<td>THROMBOSIS, DEEP VEIN, ASYMTOMATIC</td>
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## APPENDIX 100 - Diagnoses Appendix

<table>
<thead>
<tr>
<th>Code</th>
<th>Diagnosis</th>
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<tbody>
<tr>
<td>11</td>
<td>THROMBOSIS, DEEP VEIN, SYMPTOMATIC</td>
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<td>12</td>
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<tr>
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<td>TOXOPLASmosis, CONGENITAL</td>
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<td>THYROID CANCER</td>
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<td>TRACHEA CANCER</td>
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<td>URINARY TRACT INFECTION</td>
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