Welcome and Announcements
  Donna Pickett, MPH, RHIA
  Co-Chair, ICD-9-CM Coordination and Maintenance Committee

ICD-9-CM Timeline

Chronic total occlusion of coronary artery
  Kirk Garratt, M.D.

Non-Hodgkin’s Lymphomas
  Luis Fayad, M.D.
  M.D. Anderson Cancer Center

Normal pressure hydrocephalus (NPH)
  Michael Williams, M.D.

Counseling for natural family planning
  Joseph B. Stanford, M.D., MSPH
  Leslie Chorun, M.D.
  American Academy of Fertility Care Professionals

Endosseous dental implant failure

Hypoxia of newborn, Hypoxic ischemic encephalopathy [HIE] and related newborn issues

Family history of sudden cardiac death

Human Herpesvirus Infections, including Human Herpesvirus 6 (HHV-6)
  Encephalitis

Corticoadrenal Insufficiency Including Hypoaldosteronism

Bandemia

Stevens-Johnson syndrome

Long term use of other drugs

Restless legs syndrome

Secondary diabetes mellitus

Botulism not associated with food poisoning

Vulvar intraepithelial neoplasia I and II [VIN I and II]

Multiple endocrine neoplasia [MEN type I, type IIA, type IIB]

Anal sphincter tear

Addenda
ICD-9-CM Coordination and Maintenance Committee Meeting
March 23-24, 2006

ICD-9-CM TIMELINE

A timeline of important dates in the ICD-9-CM process is described below:

January 3, 2006  On-line registration opens for the March 23 – 24, 2006 ICD-9-CM Coordination and Maintenance Committee meeting at:
http://www.cms.hhs.gov/events/

January 23, 2006  Deadline for requestors: Those members of the public requesting that topics be discussed at the March 23 –March 24, 2006 ICD-9-CM Coordination and Maintenance Committee meeting must have their requests to CMS for procedures and NCHS for diagnoses by this date.

February, 2006  Tentative agenda for the Procedure part of the March 23, 2006 ICD-9-CM Coordination and Maintenance Committee meeting posted on CMS homepage as follows:
http://www.cms.hhs.gov/paymentsystems/icd9

Tentative agenda for the Diagnosis part of the March 24, 2006 ICD-9-CM Coordination and Maintenance Committee meeting posted on NCHS homepage as follows:

Federal Register notice announcing March 23 – March 24, 2006 ICD-9-CM Coordination and Maintenance Committee Meeting will be published. This will include the tentative agenda.

March 17, 2006  Because of increased security requirements, those wishing to attend the March 23 – March 24, 2006 ICD-9-CM Coordination and Maintenance Committee meeting must register for the meeting online at: http://www.cms.hhs.gov/events. Attendees must register online by March 17, 2006; failure to do so may result in lack of access to the meeting.

March 23-24, 2006  ICD-9-CM Coordination and Maintenance Committee Meeting. Those who wish to attend the ICD-9-CM Coordination and Maintenance Committee meeting must have registered for the meeting online by March 17, 2006. You must bring an official form of picture identification (such as a driver’s license) in order to be admitted to the building.

April 1, 2006  There will not be any new ICD-9-CM codes implemented on April 1, 2006 to capture new technology.
April 2006  Notice of Proposed Rulemaking to be published in the Federal Register as mandated by Public Law 99-509. This notice will include the final ICD-9-CM diagnosis and procedure codes for the upcoming fiscal year. It will also include proposed revisions to the DRG system on which the public may comment. The proposed rule can be accessed at: http://www.cms.hhs.gov/providers/hipps/frnotices.asp

April 2006  Summary report of the Procedure part of the March 23, 2006 ICD-9-CM Coordination and Maintenance Committee meeting will be posted on CMS homepage as follows: http://www.cms.hhs.gov/paymentsystems/icd9

Summary report of the Diagnosis part of the March 24, 2006 ICD-9-CM Coordination and Maintenance Committee meeting report will be posted on NCHS homepage as follows: http://www.cdc.gov/nchs/icd9.htm

April 14, 2006  Deadline for receipt of public comments on proposed code revisions discussed at the March 23-24, 2006 ICD-9-CM Coordination and Maintenance Committee meeting for implementation on October 1, 2006 to capture new technology.

June 2006  Final addendum posted on web pages as follows: Diagnosis addendum at - http://www.cdc.gov/nchs/icd9.htm


July 28, 2006  Deadline for requestors: Those members of the public requesting that topics be discussed at the September 28-29, 2006 ICD-9-CM Coordination and Maintenance Committee meeting must have their requests to CMS for procedures and NCHS for diagnoses by this date.

August, 2006  Hospital Inpatient Prospective Payment System final rule to be published in the Federal Register as mandated by Public Law 99-509. This rule will also include all the final codes to be implemented on October 1, 2006. This rule can be accessed at: http://www.cms.hhs.gov/providers/hipps/frnotices.asp
August 2006  Tentative agenda for the Procedure part of the September 28 – 29, 2006 ICD-9-CM Coordination and Maintenance Committee meeting will be posted on CMS homepage at - http://www.cms.hhs.gov/paymentsystems/icd9

Tentative agenda for the Diagnosis part of the September 28 – 29, 2006 ICD-9-CM Coordination and Maintenance Committee meeting will be posted on NCHS homepage at - http://www.cdc.gov/nchs/icd9.htm

Federal Register notice for the September 28 – 29, 2006 ICD-9-CM Coordination and Maintenance Committee Meeting will be published. This will include the tentative agenda.

September 24, 2006  Because of increased security requirements, those wishing to attend the September 28-29, 2006 ICD-9-CM Coordination and Maintenance Committee meeting must register for the meeting online at: http://www.cms.hhs.gov/events. Attendees must register online by September 24, 2006; failure to do so may result in lack of access to the meeting.

Sept. 28-29, 2006  ICD-9-CM Coordination and Maintenance Committee Meeting. Those who wish to attend the ICD-9-CM Coordination and Maintenance Committee meeting must have registered for the meeting online by September 24, 2006. You must bring an official form of picture identification (such as a driver’s license) in order to be admitted to the building.


October, 2006  Summary report of the Procedure part of the September 28-29, 2006 ICD-9-CM Coordination and Maintenance Committee meeting posted on CMS homepage at - http://www.cms.hhs.gov/paymentsystems/icd9

October 7, 2006  Deadline for receipt of public comments on proposed code revisions discussed at the September 29 – 30, 2006 ICD-9-CM Coordination and Maintenance Committee meetings for implementation on April 1, 2007 to capture new technology.

October 2006  Summary report of the Procedure part of the September 29 – 30, 2006 ICD-9-CM Coordination and Maintenance Committee meeting will be posted on CMS homepage as follows: 
http://www.cms.hhs.gov/paymentsystems/icd9

Summary report of the Diagnosis part of the September 29 – 30, 2006 ICD-9-CM Coordination and Maintenance Committee meeting report will be posted on NCHS homepage as follows: 

October 13, 2006  Deadline for receipt of public comments on proposed code revisions discussed at the September 28-29, 2006 ICD-9-CM Coordination and Maintenance Committee meeting for implementation on October 1, 2007 to capture new technology.

Early Nov., 2006  Any new ICD-9-CM codes required to capture new technology that will be implemented on April 1, 2007 will be announced. Information on any new codes to be implemented on April 1, 2007 will be posted on the following websites:
Procedure at http://www.cms.hhs.gov/paymentsystems/icd9
Diagnosis addendum at http://www.cdc.gov/nchs/icd9.htm

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NCHS Classifications of Diseases web page:

Please consult this web page for updated information.
**Topic: Chronic total occlusion of coronary artery**

A complete blockage of a coronary artery which has been present for an extended duration is known as a chronic total occlusion of the coronary artery. Collateral flow may avoid myocardial infarction, despite the chronic total occlusion of the coronary artery. However, this flow could not likely increase much during exercise, so would be likely to greatly limit activity.

There is increased risk of myocardial infarction or death, for individuals with chronic total occlusion of a coronary artery. Correcting this is beneficial. However, passing a guide wire through a chronic total coronary occlusion is more difficult than for other coronary stenosis. Chronic total occlusion of a coronary artery may be treated with angioplasty or stent placement, usually with a drug eluting stent. Advances in treatment have been made in recent years, with methods developed specifically for handling chronic total coronary occlusions.

It would be beneficial to be able to specifically track the diagnosis of chronic total coronary artery occlusion. Two options for coding are presented below. While the second option shows excludes notes between proposed new codes and existing ones, it would be possible to have both of these at the same time, in different coronary arteries. For that matter, it would be possible to have more than one of the current codes at 414.0 together. However, these situations might lead to confusion.

These changes were requested by Abbott.

**TABULAR MODIFICATION**

**Option 1.** Create a new code for chronic total occlusion to be used in addition to the current coronary atherosclerosis code (414.00-414.07).

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>414</td>
<td>Other forms of chronic ischemic heart disease</td>
</tr>
<tr>
<td>414.0</td>
<td>Coronary atherosclerosis</td>
</tr>
</tbody>
</table>

Add Use additional code to identify chronic total occlusion of coronary artery (414.2)
New Code 414.2 Chronic total occlusion of coronary artery

Code first coronary atherosclerosis (414.00-414.07)

Excludes: acute coronary occlusion with myocardial infarction (410.00 – 410.92)
acute coronary occlusion without myocardial infarction (411.81)

**Option 2.** Restructure the atherosclerosis codes to provide for a subcategory for atherosclerosis without chronic total occlusion and a subcategory for that with chronic total occlusion.

414 Other forms of chronic ischemic heart disease

Revise title 414.0 Coronary atherosclerosis without chronic total occlusion

Add Excludes: coronary atherosclerosis with chronic total occlusion (414.20 – 414.27)

New sub-category 414.2 Coronary atherosclerosis with chronic total occlusion

Excludes: coronary atherosclerosis without chronic total occlusion (414.00-414.07)

New code 414.20 Of unspecified type of vessel, native or graft

New code 414.21 Of native coronary artery

New code 414.22 Of autologous biological bypass graft

New code 414.23 Of nonautologous biological bypass graft

New code 414.24 Of artery bypass graft
Internal mammary artery

New code 414.25 Of unspecified type of bypass graft
Bypass graft NOS

New code 414.26 Of native coronary artery of transplanted heart

New code 414.27 Of bypass graft (artery) (vein) of transplanted heart
Topic: Non-Hodgkin’s Lymphomas

Non-Hodgkin’s lymphomas are a heterogeneous group of malignant lymphomas, the only common feature being the absence of the giant Reed-Sternberg cells characteristic of Hodgkin’s disease. They arise from the lymphoid components of the immune system, and present a clinical picture broadly similar to that of Hodgkin’s disease, except that the disease is initially more widespread, with the most common manifestation being painless enlargement of one or more peripheral lymph nodes. The main cell found in lymphoid tissue is the lymphocyte, of which there are two main types, B lymphocytes (B cells), and T lymphocytes (T cells). B cell lymphomas are much more common, accounting for 85% of cases on non-Hodgkin’s lymphoma in the United States.

The non-Hodgkin’s lymphoma disease process is complex. There are over 30 subtypes of non-Hodgkin’s lymphoma, including Mantle cell, mucosa associated lymphoid tissue [MALT], and primary central nervous system lymphoma. Though more specific designations of the behavior have been defined with new morphology terms and synonyms, the ICD-9-CM has not been updated to accommodate these changes. The MD Anderson Cancer Center has requested that the non-Hodgkin’s lymphoma codes in the ICD-9-CM be updated to allow for more current classification of these malignancies. Though it is not allowable to delete or modify existing code titles, it is being proposed that certain updates to the non-Hodgkin’s lymphoma codes be made based on the proposal submitted by MD Anderson Cancer Center.

TABULAR MODIFICATIONS

<table>
<thead>
<tr>
<th>Revise</th>
<th>200</th>
<th>Lymphosarcoma and reticulosarcoma and other specified malignant tumors of lymphatic tissue</th>
</tr>
</thead>
</table>
| New code | 200.3 | Marginal zone lymphoma  
Extranodal marginal zone B cell lymphoma  
Mucosa associated lymphoid tissue [MALT]  
Nodal marginal zone B cell lymphoma  
Splenic marginal zone B cell lymphoma |
| New code | 200.4 | Mantle cell lymphoma |
| New code | 200.5 | Primary central nervous system lymphoma |
| New code | 200.6 | Anaplastic large cell lymphoma |
| New code | 200.7 | Large cell lymphoma |
| 202 | Other malignant neoplasms of lymphoid and histiocytic tissue |
| New code | 202.7 | Peripheral T cell lymphoma |
**Topic: Normal pressure hydrocephalus (NPH)**

Normal pressure hydrocephalus (NPH) is a treatable disorder of gait impairment, subcortical dementia and urinary urgency and incontinence associated with impaired cerebrospinal fluid (CSF) circulation and ventriculomegaly. NPH results from a disruption in the CSF circulation leading to gradual enlargement of the ventricles and emergence of symptoms. This syndrome, when secondary to disease processes including subarachnoid hemorrhage, traumatic brain injury, cerebral infarction, and meningitis, is referred to as secondary NPH, appropriately coded as communicating hydrocephalus, 331.3. In patients without known etiologies (2/3 of all cases), it is called idiopathic NPH (INPH), also coded as 331.3.

Many common disorders of aging cause the individual components of the INPH triad of cognitive, gait, and urinary problems. Because these symptoms are ubiquitous in the elderly, evaluation of suspected NPH requires consideration of the differential diagnosis of all three symptoms simultaneously. It is common for patients with NPH to have multifactorial causes of dementia, gait impairment, or incontinence, such as vascular or degenerative dementia, Parkinsonism, cervical or lumbar stenosis, peripheral neuropathy, arthritis, bladder instability, or prostate enlargement. Careful screening for these conditions is important because shunt surgery will only improve symptoms related to NPH. Public awareness of INPH has increased in part due to a television and internet media campaign, and efforts of the Hydrocephalus Association, a patient advocacy group.

While there are no findings on CT or MR imaging studies of the brain that are sufficient alone to diagnose INPH, ventricular enlargement is necessary to establish the diagnosis of INPH for patients with appropriate symptoms. Consensus guidelines to help physicians diagnose this condition were developed and published in Neurosurgery in 2005. Highly recommended are tests of the patient’s response to short-term removal of CSF either by lumbar puncture, or several days CSF drainage via temporary spinal catheter.

The treatment for INPH is surgical diversion of CSF. This is accomplished by implanting a shunt to drain CSF either from the intracranial ventricular system or the lumbar subarachnoid space to a distal site, such as the peritoneal or pleural cavity or the venous system, where the CSF can be reabsorbed. A 2001 meta-analysis found 59% improved immediately after shunting (range 24-100%), while two major studies in 2005 found that 75-90% of patients selected on the basis of response to controlled CSF drainage improved after shunt surgery. Early diagnosis and treatment improves this chance of recovery. If the condition is not treated the symptoms will worsen.

Currently this condition is not specifically indexed in ICD-9-CM, although upon review of the tabular it would likely be assigned to existing code 331.3, Communicating hydrocephalus. The impact of treating NPH is difficult to evaluate if there is no unique diagnosis code for the condition. The American Academy of Neurology has requested that a unique code be created for this condition using the following tabular modification.
<table>
<thead>
<tr>
<th>New code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>331.5</td>
<td>Normal pressure hydrocephalus</td>
</tr>
<tr>
<td>331.5</td>
<td>Excludes: congenital hydrocephalus (741.0, 742.3)</td>
</tr>
<tr>
<td>331.5</td>
<td>secondary normal pressure hydrocephalus (331.3)</td>
</tr>
<tr>
<td>331.5</td>
<td>spina bifida with hydrocephalus (741.0)</td>
</tr>
</tbody>
</table>

331.3 Communicating hydrocephalus
- Add Secondary normal pressure hydrocephalus

331.4 Obstructive hydrocephalus
- Revise Excludes: congenital hydrocephalus (741.0, 742.3)
- Add idiopathic normal pressure hydrocephalus (331.5)
- Add normal pressure hydrocephalus (331.5)
- Add spina bifida with hydrocephalus (741.0)
**Topic: Counseling for natural family planning**

Natural methods of birth regulation are being provided both on a national and international level in hospitals and other ambulatory care settings. In the U.S., major methods in use include the Billings Ovulation Method, Creighton Model Fertility Care System, Standard Days Method, Two-Day Method and the Sympto-thermal Method.

Currently the tabular ICD-9-CM does not include natural methods of birth regulation as a specific form of management either for birth control or procreative management. Existing codes V25.09 and V26.4 have been used as the closest available codes.

A request was received from the American Academy of Fertility Care Professionals to create new codes for encounters related to natural methods of birth regulation including counseling and instruction, follow up surveillance and procreative management.

The following tabular modifications are proposed:

**Option 1:**

<table>
<thead>
<tr>
<th>TABULAR MODIFICATION</th>
</tr>
</thead>
<tbody>
<tr>
<td>V25</td>
</tr>
<tr>
<td>Encounter for contraceptive management</td>
</tr>
<tr>
<td>V25.0</td>
</tr>
<tr>
<td>General counseling and advice</td>
</tr>
</tbody>
</table>

New code  
V25.04    Counseling and instruction in natural method birth control

| V26                  |
| Procreative management |
| V26.4                |
| General counseling and advice |

New code  
V26.40    Procreative counseling and advice, unspecified

New code  
V26.41    Procreative counseling and advice using natural method birth regulation

New code  
V26.49    Other procreative management counseling and advice
**Option 2:**

**TABULAR MODIFICATION**

<table>
<thead>
<tr>
<th>Revise</th>
<th>V25</th>
<th>Encounter for contraceptive family planning management</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>V25.0</td>
<td>General counseling and advice</td>
</tr>
<tr>
<td>New code</td>
<td>V25.04</td>
<td>Counseling and instruction in natural method of family planning</td>
</tr>
<tr>
<td></td>
<td>V25.4</td>
<td>Surveillance of previously prescribed family planning methods</td>
</tr>
<tr>
<td>New code</td>
<td>V25.44</td>
<td>Natural method of family planning</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>V26</th>
<th>Procreative management</th>
</tr>
</thead>
<tbody>
<tr>
<td>V26.4</td>
<td>General counseling and advice</td>
</tr>
<tr>
<td>New code</td>
<td>V26.40</td>
</tr>
<tr>
<td>New code</td>
<td>V26.41</td>
</tr>
<tr>
<td>New code</td>
<td>V26.49</td>
</tr>
</tbody>
</table>
Topic: Endosseous dental implant failure

A dental implant is an artificial tooth root that holds a replacement tooth or bridge. There are two types of implants currently in use, endosteal and subperiosteal. Endosteal implants, the more common type, are implanted in the jaw bone. Each implant holds one or more prosthetic teeth. Subperiosteal implants are placed on top of the jaw with the metal framework’s post protruding through the gum to hold the prosthesis. Subperiosteal implants are used for patients who are unable to wear conventional dentures and who have minimal bone height.

Dental implants are a good option to replace a lost tooth or teeth. Adequate bone in the jaw is needed to support the implant. It is also important to establish an adequate level of oral health for placement and maintenance of dental implants. As with natural teeth, implants require conscientious at home oral care and regular dental visits. Under proper conditions, and with diligent patient maintenance, implants can last a lifetime. Long-term studies continue to show improving success rates for implants.

However, there are a small number of implants that do fail. There are two types of failures, pre-osseointegration and post-osseointegration. Pre-osseointegration failure occurs when the implant fails to achieve integration with the surrounding bone and soft tissue. These failures to osseointegrate are most commonly related to placement of the implant into bone of poor quality (including previously irradiated bone), hemorrhagic complications, and iatrogenic causes.

Post-osseointegration failures are either biological or mechanical. Biological failure includes periodontal infection (peri-implantitis), caused by poor oral hygiene, lack of attached gingiva, or occlusal trauma caused by not enough support for the forces that the implants were subjected to, i.e., weak bone, too few implants, poor prosthetic design, and parafunctional habits, to name a few. Mechanical failure is due to fracture or the implant body itself and any failures of the prosthesis that cause the loss of the implant.

New ICD-9-CM codes have recently been implemented for complications and failures of dental restorations and endodontic treatment. It is now being proposed that similar codes be created for failed dental implants.
ICD-9-CM Coordination and Maintenance Committee Meeting
March 23-24, 2006

TABULAR MODIFICATIONS

525 Other diseases and conditions of the teeth and supporting structures

New subcategory 525.7 Endosseous dental implant failure

New code 525.71 Osseointegration failure of dental implant
Failure of dental implant due to poor bone quality
Hemorrhagic complications of dental implant placement
Iatrogenic failure of dental implant
Pre-integration failure of dental implant NOS
Pre-osseointegration failure of dental implant

New code 525.72 Post-osseointegration biological failure of dental implant
Failure of dental implant due to lack of attached gingiva
Failure of dental implant due to occlusal trauma caused by poor prosthetic design
Failure of dental implant due to parafunctional habits
Failure of dental implant due to periodontal infection (peri-implantitis)
Failure of dental implant due to poor oral hygiene

New code 525.73 Post-osseointegration mechanical failure of dental implant
Failure of dental prosthesis causing loss of dental implant
Fracture of dental implant

Excludes: cracked tooth (521.81)
fractured dental restorative material with loss of material (525.64)
fractured dental restorative material without loss of material (525.63)
fractured tooth (873.63, 873.73)
<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>525.79</td>
<td>Other endosseous dental implant failure</td>
</tr>
<tr>
<td></td>
<td>Dental implant failure NOS</td>
</tr>
<tr>
<td>996</td>
<td>Complications peculiar to certain specified procedures</td>
</tr>
<tr>
<td>Add</td>
<td>Excludes: endosseous dental implant failures (525.71-525.79)</td>
</tr>
</tbody>
</table>
Topic: Hypoxia of newborn, Hypoxic ischemic encephalopathy [HIE] and related newborn issues

This topic was presented in September 2005. The current proposal is based on the initial proposal from the American Academy of Pediatrics, with subsequent input from the American College of Obstetricians and Gynecologists.

Traditional theories of the etiology of neonatal neurologic injury have focused on the hospital portion of the labor and delivery because it is available for careful and minute by minute observation. This represents a limited portion of the complete gestation and has inappropriately led to a series of conclusions on the etiology of brain injury in the newborn and young child that focused almost strictly on the intrapartum period.

The nomenclature associated with these “diagnoses” has also been problematic, with traditional terminology applied that are technically incorrect descriptors of the fetal/neonatal condition and establishing an accepted “etiology” of the later injury that assumed a cause and effect relationship.

For example the terminology currently used to describe fetal encephalopathic injury and death is antiquated and imprecise. The term “hypoxia” actually refers to a deficiency of oxygen reaching the tissues of the body, while “hypoxemia” means deficient oxygenation of the blood. Asphyxia, from the Greek, actually means stopping of the pulse but has come to be associated with hypoxia and hypercapnia.

As our understanding of perinatal cerebral injury has become clearer, it is obvious that the older terminology can no longer apply. The actual cause of the morbidity and mortality in this condition is due to ischemic injury from hypoxemia, hypercapnia and acidosis. While it is normal for these conditions to occur during the normal birth process, when it leads to brain damage the result is hypoxic-ischemic encephalopathy (HIE).

HIE also has well defined clinical definitions (mild, moderate, and severe) based on clinical presentation and imaging findings.

Since some of these conditions can exist during the perinatal period but are unrelated to the birthing process, additional changes were recommended to accommodate these conditions, unrelated to the birth process.

Because of the need to correctly identify these potentially devastating conditions accurately, the following changes to ICD-9-CM have been recommended.

Note: it has been requested that these changes be effective October 1, 2006. Thus, comments are needed by April 14, 2006.
TABULAR MODIFICATIONS

768  Intrauterine hypoxia and birth asphyxia

Add  Excludes:  acidemia NOS of newborn (775.81)
      acidosis NOS of newborn (775.81)
      cerebral ischemia NOS (779.2)
      hypoxia NOS of newborn (770.88)
      mixed metabolic and respiratory acidosis of newborn (775.81)
      respiratory arrest of newborn (770.87)

Revise  768.3 Fetal distress first noted during labor and delivery, in liveborn infant
Revise  Fetal metabolic acidemia first noted during labor and delivery, in liveborn infant

768.5 Severe birth asphyxia

Add  Excludes:  hypoxic-ischemic encephalopathy (HIE) (768.7)

768.6 Mild or moderate birth asphyxia

Add  Excludes:  hypoxic-ischemic encephalopathy (HIE) (768.7)

New code  768.7 Hypoxic-ischemic encephalopathy (HIE)

Delete  Hypoxia NOS, in liveborn infant

768.9 Unspecified birth asphyxia in liveborn infant

770 Other respiratory conditions of fetus and newborn

770.8 Other respiratory problems after birth

Add  Excludes:  mixed metabolic and respiratory acidosis of newborn (775.81)

New code  770.87 Respiratory arrest of newborn
New code  770.88 Hypoxemia of newborn
             Hypoxia NOS of newborn
<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>775</td>
<td>Endocrine and metabolic disturbances specific to the fetus and newborn</td>
</tr>
<tr>
<td>Revise</td>
<td>775.8 Other transitory neonatal endocrine and metabolic disturbances</td>
</tr>
<tr>
<td>Delete</td>
<td>Amino-acid metabolic disorders described as transitory</td>
</tr>
<tr>
<td>New code</td>
<td>775.81 Other acidosis of newborn</td>
</tr>
<tr>
<td></td>
<td>Acidemia NOS of newborn</td>
</tr>
<tr>
<td></td>
<td>Acidosis of newborn NOS</td>
</tr>
<tr>
<td></td>
<td>Mixed metabolic and respiratory acidosis of newborn</td>
</tr>
<tr>
<td>New code</td>
<td>775.89 Other neonatal endocrine and metabolic disturbances</td>
</tr>
<tr>
<td></td>
<td>Amino-acid metabolic disorders described as transitory</td>
</tr>
<tr>
<td>779</td>
<td>Other and ill-defined conditions originating in the perinatal period</td>
</tr>
<tr>
<td>779.2</td>
<td>Cerebral depression, coma, and other abnormal cerebral signs</td>
</tr>
<tr>
<td>Add</td>
<td>Cerebral ischemia NOS of newborn</td>
</tr>
<tr>
<td>779.8</td>
<td>Other specified conditions originating in the perinatal period</td>
</tr>
<tr>
<td>New code</td>
<td>779.85 Cardiac arrest of newborn</td>
</tr>
</tbody>
</table>
Topic: Family history of sudden cardiac death

Heart disease causes more deaths in the U.S. than any other disease, close to 700,000 deaths annually. While ischemic heart disease is the most common cause of death related to heart disease, other causes include heart failure, hypertensive heart disease, conduction disorders or arrhythmias, cardiomyopathy, and valvular heart disease.

Tracking family history of deaths due to heart disease can be helpful in assessing risk of development of a similar heart problem. This can be of use in a number of types of heart disease, including heart failure, hypertensive heart disease, certain conduction disorders or arrhythmias, certain cardiomyopathies, and certain types of valvular heart disease. A family history of death due to ischemic heart disease should be coded to V17.3, Family history of ischemic heart disease.

A specific code was requested for family history of sudden cardiac death, by a private cardiology practice.

TABULAR MODIFICATION

V17 Family history of certain chronic disabling diseases

V17.4 Other cardiovascular diseases

New code V17.40 Family history of cardiovascular diseases, unspecified

New code V17.41 Family history of sudden cardiac death

Excludes: Family history of sudden cardiac death due to ischemic heart disease (V17.3)

New code V17.49 Family history of other cardiovascular diseases
Topic: Human Herpesvirus Infections, including Human Herpesvirus 6 (HHV-6) Encephalitis

Human herpesvirus 6 (HHV-6) is a beta herpesvirus with two recognized variants, A and B. It was initially called human B-lymphotropic virus. Primary infection with HHV-6B causes roseola infantum or exanthem subitum, a common childhood exanthema. HHV-6 may reactivate and cause problems in the immune suppressed, those with AIDS or transplant recipients.

HHV-6 is extremely neurotropic, and neuroinvasion is documented even in primary infection in infants. It may cause encephalitis, and other neurological disorders. There may be a connection of HHV-6 with pediatric febrile seizures, in some cases. There have been postulated involvement of HHV-6 in multiple sclerosis and chronic fatigue syndrome.

Human herpesvirus 7 (HHV-7) is another beta herpesvirus, which also causes roseola infantum in infants, and can reactivate and cause disease in those who are immunosuppressed. The other human beta herpesvirus is cytomegalovirus.

Roseola infantum, or exanthema subitum, is coded to 057.8, Other specified viral exanthemata. However, reoccurrence of HHV-6 or HHV-7 would usually not involve the viral exanthema, and would not be appropriately coded here. HHV-6 encephalitis would now be coded to 049.8, Other specified non-arthropod-borne viral diseases of central nervous system.

Other human herpesviruses are classified into the alpha human herpesviruses, and the gamma human herpesviruses. The alpha human herpesviruses include herpes simplex virus type 1 and type 2, and varicella-zoster virus. The gamma human herpesviruses include Epstein-Barr virus and human herpesvirus 8 (HHV-8). The gamma human herpesviruses are frequently latent in lymphatic cells. HHV-8 is also known as Kaposi’s sarcoma-associated herpesvirus, and it is associated with development of Kaposi’s sarcoma in immune suppressed individuals. There is evidence that it was transmitted with HIV in a concurrent epidemic in the 1980s. In addition to the association with Kaposi’s sarcoma, HHV-8 is linked to certain lymphomas, and other neoplastic disease.

There is a need to be able to specifically identify infections with human herpesvirus 6, particularly with encephalitis, and also human herpesvirus 7, and to classify human herpesvirus 8. The HHV-6 Foundation requested consideration of human herpesvirus 6 and human herpesvirus 7, and also of a specific code for human herpesvirus 6 encephalitis.
ICD-9-CM Coordination and Maintenance Committee Meeting
March 23-24, 2006

TABULAR MODIFICATION

Option 1

049 Other non-arthropod-borne viral diseases of central nervous system

049.8 Other specified non-arthropod-borne viral diseases of central nervous system

Add

Excludes: human herpesvirus 6 encephalitis (058.12)

054 Herpes simplex

054.3 Herpetic meningoencephalitis

Add

Excludes: human herpesvirus 6 encephalitis (058.12)

057 Other viral exanthemata

057.8 Other specified viral exanthemata

Delete

Dukes (-Filatow) disease
Exanthema subitum [sixth disease]
Fourth disease
Parascalatica
Pseudoscalarata
Roseola-infantum

Delete

Add

Excludes: Exanthema subitum [sixth disease] (058.0)
Roseola-infantum (058.0)
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New section

OTHER HUMAN HERPESVIRUSES (058)

New category

058  Other human herpesvirus

Excludes:  cytomegalovirus (078.5)
             Epstein-Barr virus (075)
             herpes NOS (054.0-054.9)
             herpes simplex (054.0-054.9)
             herpes zoster (053.0-053.9)
             human herpesvirus NOS (054.0-054.9)
             human herpesvirus 1 (054.0-054.9)
             human herpesvirus 2 (054.0-054.9)
             human herpesvirus 3 (052.0-053.9)
             human herpesvirus 4 (075)
             human herpesvirus 5 (078.5)
             varicella (052.0-052.9)
             varicella-zoster virus (052.0-053.9)

New code 058.0  Roseola infantum, unspecified
             Exanthema subitum [sixth disease], unspecified

Add  Excludes:  Roseola infantum due to human herpesvirus 6 (058.11)
             Roseola infantum due to human herpesvirus 7 (058.21)

New subcategory

058.1  Human herpesvirus 6

New code 058.10  Human herpesvirus 6, unspecified

New code 058.11  Roseola infantum due to human herpesvirus 6
             Exanthema subitum [sixth disease] due to human herpesvirus 6

Add  Excludes:  Roseola infantum, unspecified (058.0)

New code 058.12  Human herpesvirus 6 encephalitis

New code 058.19  Other human herpesvirus 6 infection
<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>058.2</td>
<td>Human herpesvirus 7</td>
</tr>
<tr>
<td>058.20</td>
<td>Human herpesvirus 7, unspecified</td>
</tr>
<tr>
<td>058.21</td>
<td>Roseola infantum due to human herpesvirus 7</td>
</tr>
<tr>
<td>058.29</td>
<td>Other human herpesvirus 7</td>
</tr>
<tr>
<td>058.3</td>
<td>Human herpesvirus 8</td>
</tr>
<tr>
<td>058.4</td>
<td>Kaposi’s sarcoma-associated herpesvirus</td>
</tr>
</tbody>
</table>

**Option 2**

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>049</td>
<td>Other non-arthropod-borne viral diseases of central nervous system</td>
</tr>
<tr>
<td>049.8</td>
<td>Other specified non-arthropod-borne viral diseases of central nervous system</td>
</tr>
<tr>
<td>054</td>
<td>Herpes simplex</td>
</tr>
<tr>
<td>054.3</td>
<td>Herpetic meningoencephalitis</td>
</tr>
<tr>
<td>057</td>
<td>Other viral exanthemata</td>
</tr>
<tr>
<td>057.8</td>
<td>Other specified viral exanthemata</td>
</tr>
<tr>
<td>Delete</td>
<td>Dukes (-Filatow) disease</td>
</tr>
<tr>
<td>Delete</td>
<td>Exanthema subitum [sixth disease]</td>
</tr>
<tr>
<td>Delete</td>
<td>Fourth disease</td>
</tr>
<tr>
<td>Delete</td>
<td>Parascarlatina</td>
</tr>
<tr>
<td>Delete</td>
<td>Pseudoscarlatina</td>
</tr>
<tr>
<td>Add</td>
<td>Roseola infantum</td>
</tr>
<tr>
<td>Add</td>
<td>Excludes: Exanthema subitum [sixth disease] (058.10-058.12) Roseola infantum (058.10-058.12)</td>
</tr>
</tbody>
</table>
New section

OTHER HUMAN HERPESVIRUSES (058)

New category

058 Other human herpesvirus

Excludes: cytomegalovirus (078.5)
Epstein-Barr virus (075)
herpes NOS (054.0-054.9)
herpes simplex (054.0-054.9)
herpes zoster (053.0-053.9)
human herpesvirus NOS (054.0-054.9)
human herpesvirus 1 (054.0-054.9)
human herpesvirus 2 (054.0-054.9)
human herpesvirus 3 (052.0-053.9)
human herpesvirus 4 (075)
human herpesvirus 5 (078.5)
varicella (052.0-052.9)
varicella-zoster virus (052.0-053.9)

New subcategory 058.1 Roseola infantum

New code

058.10 Roseola infantum, unspecified
Exanthema subitum [sixth disease], unspecified

New code

058.11 Roseola infantum due to human herpesvirus 6
Exanthema subitum [sixth disease] due to human herpesvirus 6

New code

058.12 Roseola infantum due to human herpesvirus 7
Exanthema subitum [sixth disease] due to human herpesvirus 7

New subcategory 058.2 Other human herpesvirus encephalitis

Excludes: herpes encephalitis NOS (054.3)
herpes simplex encephalitis (054.3)
human herpesvirus encephalitis NOS (054.3)
simian B herpes virus encephalitis (054.3)

New code

058.21 Human herpesvirus 6 encephalitis

New code

058.29 Other human herpesvirus encephalitis
Human herpesvirus 7 encephalitis
ICD-9-CM Coordination and Maintenance Committee Meeting  
March 23-24, 2006

<table>
<thead>
<tr>
<th>New subcategory</th>
<th>Code</th>
<th>Description</th>
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</thead>
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<tr>
<td>058.8</td>
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<td>Other human herpesvirus infections</td>
</tr>
<tr>
<td>New code</td>
<td>058.81</td>
<td>Human herpesvirus 6 infection</td>
</tr>
<tr>
<td>New code</td>
<td>058.82</td>
<td>Human herpesvirus 7 infection</td>
</tr>
<tr>
<td>New code</td>
<td>058.89</td>
<td>Other human herpesvirus infection</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Human herpesvirus 8 infection</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Kaposis’s sarcoma-associated herpesvirus infection</td>
</tr>
</tbody>
</table>
Topic: Corticoadrenal Insufficiency Including Hypoaldosteronism

Corticoadrenal insufficiency is related to decreased function of the adrenal cortex, which produces cortisol and aldosterone. Decreased production of cortisol results in a glucocorticoid deficiency. This can cause a range of signs and symptoms, including malaise, loss of appetite, orthostatic hypotension, weight loss, anemia, pre-renal azotemia, hyperpigmentation, and hyponatremia. If aldosterone is also affected, hyperkalemia may also occur. In more acute cases, agitation, confusion, fever, and abdominal pain may be found, and if untreated, it may progress to coma and death. Diagnosis is confirmed by challenge with adrenocorticotropic hormone (ACTH), and testing for a lack of response in plasma cortisol level.

Levels of ACTH are also tested, to assess for ACTH dependence. ACTH-dependent glucocorticoid deficiency is caused by dysfunction of the hypothalamus or pituitary gland, or it can result from adrenal suppression from taking glucocorticoids. ACTH-independent glucocorticoid deficiency is caused by disordered adrenal function (primary adrenal insufficiency). Primary adrenal insufficiency may be caused by destruction of the adrenal cortex, due to tuberculosis or autoimmune disease, referred to as Addison’s disease. There are other genetic and metabolic disorders which may cause primary adrenal insufficiency, including amyloidosis, congenital adrenal hypoplasia, and familial glucocorticoid insufficiency.

Mineralocorticoid deficiency results in hyponatremia, hyperkalemia, and mild metabolic acidosis. These can lead to profound weakness and cardiac arrhythmias. This may be caused by combined deficiency of cortisol and aldosterone, so testing will usually first exclude this with ACTH challenge test. Next, testing will check for aldosterone level, and if this is low, isolated hypoaldosteronism is diagnosed.

It would be beneficial to have distinct codes for glucocorticoid deficiency and mineralocorticoid deficiency. This proposal came from NCHS staff.
TABULAR MODIFICATION

255 Disorders of adrenal glands

255.4 Corticoadrenal insufficiency

Delete
- Addisonian crisis
- Addison’s disease NOS
- Adrenal atrophy (autoimmune)
- Adrenal calcification
- Adrenal crisis
- Adrenal hemorrhage
- Adrenal infarction
- Adrenal insufficiency NOS

New code 255.41 Glucocorticoid deficiency
- Addisonian crisis
- Addison’s disease NOS
- Adrenal atrophy (autoimmune)
- Adrenal calcification
- Adrenal crisis
- Adrenal hemorrhage
- Adrenal infarction
- Adrenal insufficiency NOS
- Combined glucocorticoid and mineralocorticoid deficiency
- Corticoadrenal insufficiency NOS

New code 255.42 Mineralocorticoid deficiency
- Hypoaldosteronism
Topic: Bandemia

White blood cell counts may be elevated for a number of reasons, and in particular, neutrophil counts are often considered. However, in some cases the white blood cell count may be normal, but there are an excess of immature white blood cells, or band cells. This is referred to as bandemia. It is frequently present in cases of bacterial infection.

In cases where a diagnosis of infection has not been established, but a bandemia is present, it would be useful to have an ICD-9-CM code to specifically identify the bandemia. This request is from the American Academy of Pediatrics.

TABULAR MODIFICATION

288 Diseases of white blood cells

288.6 Elevated white blood cell count

New code 288.66 Bandemia
Stevens-Johnson syndrome is a form of erythema multiforme that affects the mucous membranes of the mouth and eyes. It has systemic effects, and can involve the nose and anus, as well as the rest of the gastrointestinal system, and also the heart, lungs, kidneys, and genitals. Hemorrhagic crusts may be noted on the lips. It is also called erythema multiforme major.

Erythema multiforme involves concentric target or bull’s eye lesions, called erythema iris or herpes iris. Erythema multiforme may also be due to reaction to a drug, such as penicillin, or to an infection, such as recurrent herpes simplex.

The severity of Stevens-Johnson syndrome is much worse than erythema multiforme without mucous membrane involvement, and it can cause death. It would be useful to have a specific code for Stevens-Johnson syndrome. This proposal is from NCHS staff.

**TABULAR MODIFICATION**

695  Erythematous conditions

695.1  Erythema multiforme

Delete

- Erythema iris
- Herpes iris
- Lyell's syndrome
- Scalded skin syndrome
- Stevens-Johnson syndrome
- Toxic epidermal necrolysis

New code  695.10  Erythema multiforme, unspecified

- Erythema iris
- Herpes iris

New code  695.11  Stevens-Johnson syndrome

New code  695.19  Other erythema multiforme

- Lyell's syndrome
- Scalded skin syndrome
- Toxic epidermal necrolysis
**Topic: Long term use of other drugs**

Tamoxifen (also known as Nolvadex®) is a drug in the family of antiestrogens. It is used to treat breast cancer. In addition it is used to prevent breast cancer in women who are at a high risk of developing it. It works by blocking the effects of the hormone estrogen in the breast. It has been used for about 20 years as an adjuvant or additional therapy following primary treatment of early stage breast cancer. It has been shown to reduce the chance of developing a recurrence of breast cancer.

If a patient is still undergoing treatment for breast cancer the code for the neoplasm of the breast would be assigned and sequenced before this new code. If the patient is taking the drug as a long term prophylactic, to prevent recurrence, then the appropriate history of cancer code would be assigned followed by this new code.

To be able to identify patients who are taking this drug it has been suggested to create a new code using the following modifications to the tabular.

**Tabular Modification**

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>V07</td>
<td>Need for isolation and other prophylactic measures</td>
</tr>
<tr>
<td>V07.3</td>
<td>Other prophylactic chemotherapy</td>
</tr>
</tbody>
</table>

New code V07.32 Prophylactic administration of antiestrogen agents

Raloxifene (also known as Evista®) is a drug in the class of drugs known as selective estrogen receptor modulators (SERMs). It is used in the prevention of osteoporosis in postmenopausal women but is also used as a cancer prevention drug. Long term use of this class of drugs will be indexed to code V07.39, Other prophylactic chemotherapy.
**Topic: Restless legs syndrome**

Restless legs syndrome (RLS) is a sensory-motor disorder characterized by unpleasant sensations in the legs and an uncontrollable urge to move, when at rest, in an effort to relieve these feelings. RLS sensations are often described by people as burning, creeping, tugging, pain associated with the desire to move the legs. It does affect the ability to sleep as it occurs most often at night. Currently no etiology has been found to cause RLS though a number of medical conditions have been associated with it including: neuropathies, radiculopathies, end-stage renal disease, Parkinson’s disease, rheumatoid arthritis and diabetes. There have been recent findings showing a relationship between anemia, low serum ferritin levels and RLS, however, this correlation is still being studied.

Treatment options range from non-pharmacological (hot baths, muscle stretching, massage, moderate exercise) to pharmacologic folate, vitamin C, and B12. These treatments may improve some results but are not consistent. Dopamine agonist therapy as well as Levo-dopa are used first line for the primary form of RLS.

Currently this condition is indexed to and listed as an inclusion term in ICD-9-CM code 333.99, Other extrapyramidal diseases and abnormal movement disorders. To be able to better distinguish these patients, from those with other conditions indexed to code 333.99, the Centers for Medicare and Medicaid Services (CMS) has requested that a new code be created for restless leg syndrome using the following tabular modifications:

<table>
<thead>
<tr>
<th>TABULAR MODIFICATION</th>
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</thead>
<tbody>
<tr>
<td>333</td>
</tr>
<tr>
<td>333.9</td>
</tr>
<tr>
<td>New code</td>
</tr>
<tr>
<td>333.99</td>
</tr>
<tr>
<td>Delete</td>
</tr>
</tbody>
</table>
**Topic: Secondary diabetes mellitus**

The American Association of Pediatrics (AAP) had requested a code to identify secondary diabetes mellitus specifically for cystic fibrosis (CF) patients who develop diabetes mellitus as a result of the CF. Diabetes mellitus can also result from other specific disease processes, such as Cushing’s syndrome, pancreatitis, malignant neoplasm, and certain genetic disorders. According to the “Report of the Expert Committee on the Diagnosis and Classification of Diabetes Mellitus, 1997”, secondary diabetes is considered neither type I or type II diabetes mellitus and are grouped as “other specific types”.

Currently, the diabetes mellitus codes in category 250 provide fifth-digits for type I and type II diabetes, but there is no code or fifth-digit to indicate diabetes secondary to another condition. Previous advice given in AHA’s Coding Clinic has been to code the underlying condition followed by 251.8, Other specified disorders of pancreatic internal secretion. Additionally, the advice stated that codes from category 250 are not to be used for secondary diabetes mellitus.

In ICD-10-CM secondary diabetes mellitus is classified to category E08, Diabetes mellitus due to underlying condition. This category is included in the range of categories for diabetes mellitus.

It was proposed at a previous C&M meeting to create two new fifth-digits at category 250, Diabetes mellitus, for secondary diabetes. This proposal was extremely unpopular with both attendees at the C&M meeting, and within CDC, so the proposal was not approved for implementation. However, the AAP, as well as others, would still like secondary diabetes mellitus to be included in the classification.

At this time a new proposal is being presented for a new category for secondary diabetes that parallels category 250. All of the manifestation codes that apply to category 250 would also apply to the new category for secondary diabetes mellitus. The distinction between category 250 and the new category would be that the new category would be coded secondary to the underlying condition that is responsible for the secondary diabetes. This sequencing rule would comply with the etiology/manifestation convention of the classification. A note would instruct coders to sequence the underlying condition before the secondary diabetes codes. Additionally, code V58.67, Long term current use of insulin, would be assigned for those patients requiring insulin.

This proposal does not include fifth-digits for the new codes, nor does it include the concept of controlled or uncontrolled. All corresponding index entries, such as the entry for steroid induced diabetes, would also be modified. Should this proposal be approved, the official coding guidelines would be updated to provide instruction on the coding of secondary diabetes mellitus.
The proposal as presented here is an abbreviated version. The full proposal will be available with this topic packet as it is posted on the NCHS website.

**TABULAR MODIFICATIONS**

157  Malignant neoplasm of pancreas

Add  Use additional code to identify associated secondary diabetes mellitus, if applicable (249.0-249.9)

New  249  Diabetes mellitus due to underlying condition

Category  Secondary diabetes mellitus

Code first underlying condition, such as:
- Cushing’s syndrome (255.0)
- Cystic fibrosis (277.00-277.09)
- Malignant neoplasm of pancreas (157.0-157.9)
- Pancreatitis (577.0, 577.1)

Use additional code to identify any associated insulin use (V58.67)

New code  249.0  Diabetes mellitus due to underlying condition without mention of complication
  Diabetes (mellitus) due to underlying condition without mention of complication or manifestation classifiable to 249.1-249.9
  Diabetes (mellitus) due to underlying condition NOS

New code  249.1  Diabetes mellitus due to underlying condition with ketoacidosis
  Diabetes mellitus due to underlying condition with diabetic acidosis without mention of coma
  Diabetes mellitus due to underlying condition with diabetic ketosis without mention of coma

New code  249.2  Diabetes mellitus due to underlying condition with hyperosmolarity
  Diabetes mellitus due to underlying condition with hyperosmolar (nonketotic) coma
New code 249.3  Diabetes mellitus due to underlying condition with other coma
  Diabetes mellitus due to underlying condition with diabetic coma (with ketoacidosis)
  Diabetes mellitus due to underlying condition with diabetic hypoglycemic coma
  Diabetes mellitus due to underlying condition with insulin coma NOS

Excludes: diabetes mellitus due to underlying condition with hyperosmolar coma (249.2)

New code 249.4  Diabetes mellitus due to underlying condition with renal manifestations

Use additional code to identify manifestation, as:
  chronic kidney disease (585.1-585.9)
  diabetic nephropathy NOS (583.81)
  diabetic nephrosis (581.81)
  intercapillary glomerulosclerosis (581.81)
  Kimmelstiel-Wilson syndrome (581.81)

New code 249.5  Diabetes mellitus due to underlying condition with ophthalmic manifestations

Use additional code to identify manifestation, as:
  diabetic blindness (369.00-369.9)
  diabetic cataract (366.41)
  diabetic glaucoma (365.44)
  diabetic macular edema (362.07)
  diabetic retinal edema (362.07)
  diabetic retinopathy (362.01-362.07)

New code 249.6  Diabetes mellitus due to underlying condition with neurological manifestations

Use additional code to identify manifestation, as:
  diabetic amyotrophy (358.1)
  diabetic gastroparalysis (536.3)
  diabetic gastroparesis (536.3)
  diabetic mononeuropathy (354.0-355.9)
  diabetic neurogenic arthropathy (713.5)
  diabetic peripheral autonomic neuropathy (337.1)
  diabetic polyneuropathy (357.2)
New code  249.7  Diabetes mellitus due to underlying condition with peripheral circulatory disorders

Use additional code to identify manifestation, as:
  diabetic gangrene (785.4)
  diabetic peripheral angiopathy (443.81)

New code  249.8  Diabetes mellitus due to underlying condition with other specified manifestations
  Diabetic hypoglycemia
  Hypoglycemic shock

Use additional code to identify manifestation, as:
  any associated ulceration (707.10-707.9)
  diabetic bone changes (731.8)

Use additional E code to identify drug, if due to therapeutic drug use (sequencing issue with this situation)

New code  249.9  Diabetes mellitus due to underlying condition with unspecified complication

250  Diabetes mellitus

Add  Excludes:  diabetes mellitus due to underlying condition (249.0-249.9)
  secondary diabetes mellitus (249.0-249.9)

250.8  Diabetes with other specified manifestations

Delete  Use additional E code to identify cause, if drug-induced

251  Other disorders of pancreatic internal secretion

251.0  Hypoglycemic coma

Revise  Excludes:  hypoglycemic coma in diabetes mellitus (249.3, 250.3)

251.1  Other specified hypoglycemia

Excludes:  hypoglycemia:

Revise  in diabetes mellitus (249.8, 250.8)
Topic: Botulism not associated with food poisoning

Botulism, neuromuscular poisoning from Clostridium botulinum toxin, occurs in three forms, food borne, wound, and infant botulism. C. botulinum is an anaerobic, gram-positive bacillus with seven types of distinct neurotoxins, four of which affect humans.

In food borne botulism, toxin produced in contaminated food is eaten. Type A and B toxins are highly poisonous proteins resistant to digestion by GI enzymes. Approximately 50% of food borne outbreaks in the U.S. are caused by type A toxin, followed by types B and E. Type A toxin occurs predominantly west of the Mississippi river, type B in the eastern states, and type E in Alaska and the Great Lakes area.

C. botulinum spores are highly heat-resistant and may survive boiling for several hours. Toxins are readily destroyed by heat and cooking at or above 176 degrees F for 30 minutes. Home canned foods are the most common source, but commercially prepared foods account for 10% of outbreaks.

Onset of food borne botulism is within 18 to 36 hours after ingestion. Nausea, vomiting, abdominal cramps, and diarrhea frequently precede neurologic symptoms. Neurologic symptoms are characteristically bilateral and symmetric, beginning with the cranial nerves and followed by descending weakness and paralysis.

Wound botulism results from traumatic injury or a deep puncture wound. It is often caused by abscesses due to self injection of illegal drugs. It is manifested by neurologic symptoms, but without GI symptoms. Classically, symptoms begin within 2 weeks of the initial trauma or wound, but onset is much less predictable in injection drug use.

Infant botulism occurs most often in infants <6 months old. It results from the ingestion of C. botulinum spores that colonize in the large intestine with toxin production in vivo. Constipation is present initially in 90% of patients prior to the neuromuscular paralysis. Severity ranges from mild lethargy and slowed feeding to severe hypotonia and respiratory insufficiency. Most cases are idiopathic, though C. botulinum spores are common in the environment. Parents are advised not to feed honey to a child which may contain spores.

After absorption the toxins interfere with release of acetylcholine at peripheral nerve endings. The greatest threat to life from botulism is respiratory impairment and its complications. Patients should be hospitalized and closely monitored. Improvements in intensive care medicine have reduced the mortality rate to <10%.

Currently the only ICD-9-CM code for botulism is 005.1, Botulism food poisoning. It is being proposed that a new code 040.83, Clostridium botulinum, be created to be used for wound botulism and infant botulism.
ICD-9-CM Coordination and Maintenance Committee Meeting
March 23-24, 2006

TABULAR MODIFICATIONS

005  Other food poisoning (bacterial)

    Revise          005.1  Botulism food poisoning
                      Botulism NOS

    Add  Excludes: infant botulism (040.83)
                      wound botulism (040.83)

040  Other bacterial diseases

040.8  Other specified bacterial diseases

    New code  040.83  Other specified botulism
                      Infant botulism
                      Non-food borne intoxication due to toxins of
                      Clostridium botulinum [C. botulinum]
                      Wound botulism

                      Excludes: food poisoning due to toxins of Clostridium
                      botulinum (005.1)

771  Infections specific to the perinatal period

    Add  Excludes: infant botulism (040.83)
**Topic: Vulvar intraepithelial neoplasia I and II [VIN I and II]**

Unique codes for vulvar intraepithelial neoplasia I and II [VIN I] and [VIN II] have been requested from the American College of Obstetricians and Gynecologists (ACOG) in keeping with the unique code that exist for cervical intraepithelial neoplasia I and II [CIN I] and [CIN II]. VIN I and VIN II are currently indexed to code 624.8, Other specified noninflammatory disorders of vulva and perineum. ACOG has requested that the new codes be created under subcategory 624.0, Dystrophy of vulva, as dystrophy and dysplasia are approximately synonymous.

**TABULAR MODIFICATIONS**

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<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
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<tbody>
<tr>
<td>624</td>
<td>Noninflammatory disorders of vulva and perineum</td>
</tr>
<tr>
<td>624.0</td>
<td>Dystrophy of vulva</td>
</tr>
<tr>
<td>Delete</td>
<td>Kraurosis of vulva</td>
</tr>
<tr>
<td></td>
<td>Leukoplakia of vulva</td>
</tr>
<tr>
<td>Excludes:</td>
<td>carcinoma in situ of vulva (233.3)</td>
</tr>
<tr>
<td>Add</td>
<td>severe dysplasia of vulva (233.3)</td>
</tr>
<tr>
<td>Add</td>
<td>vulvar intraepithelial neoplasia III [VIN III] (233.3)</td>
</tr>
<tr>
<td>New code</td>
<td>624.01</td>
</tr>
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<td>Mild dysplasia of vulva</td>
</tr>
<tr>
<td>New code</td>
<td>624.02</td>
</tr>
<tr>
<td></td>
<td>Moderate dysplasia of vulva</td>
</tr>
<tr>
<td>New code</td>
<td>624.09</td>
</tr>
<tr>
<td></td>
<td>Kraurosis of vulva</td>
</tr>
<tr>
<td></td>
<td>Leukoplakia of vulva</td>
</tr>
</tbody>
</table>
Multiple endocrine neoplasia [MEN type I, type IIA, type IIB]

Multiple endocrine neoplasia [MEN] syndromes are a group of genetically distinct familial diseases involving adenomatous hyperplasia and malignant tumor formation in several endocrine glands. MEN is also referred to as multiple endocrine adenomatosis, and familial endocrine adenomatosis. Three distinct syndromes, MEN I, MEN IIA, and MEN IIB, have been identified, though there is some overlap between them. Conditions associated with MEN syndromes can appear in infants, or in patients as old as 70. Because these syndromes are almost always inherited, any person with a family member who has MEN needs to be tested as well for both the genetic defect and any of the possible conditions associated with the syndrome.

Multiple endocrine neoplasia, type I [MEN I], also referred to as Wermer’s syndrome, is characterized by tumors of the parathyroid glands, pancreatic islet cells, and pituitary gland. MEN I patients also commonly have kidney stones and peptic ulcer disease. Multiple endocrine neoplasia, type IIA [MEN IIA], also referred to as Sipple’s syndrome, is characterized by medullary carcinoma of the thyroid, pheochromocytomas, which usually raises blood pressure, sometimes to severe levels, and hyperparathyroidism. Almost all patients with MEN type IIA have medullary thyroid cancer. MEN type IIB has similar features to type IIA, but with the additional distinct feature of mucosal neuromas. The medullary thyroid cancers associated with type IIB tend to develop at an early age, they have been found in infants as young as three months, and tends to grow faster and spread more rapidly than in type IIA disease. Type IIB disease has been found in patients with no known family history of MEN.

Currently, Wermer’s syndrome [MEN type I] is indexed in ICD-9-CM to code 258.0, Polyglandular activity in multiple endocrine adenomatosis. Sipple’s syndrome [MEN type IIA] is indexed to code 193, Malignant neoplasm of thyroid gland. Neither of these codes adequately classifies these complex syndromes. It is being proposed that unique codes be created for the three types of MEN, as well as codes for genetic susceptibility to MEN syndromes, and a family history of MEN syndromes.
TABULAR MODIFICATIONS

193  Malignant neoplasm of thyroid gland
     Sipple’s syndrome

Delete

258  Polyglandular dysfunction and related disorders

258.0  Polyglandular activity in multiple endocrine adenomatosis
        Wermer’s syndrome

Delete

Add  Multiple endocrine neoplasia [MEN] syndromes

Add  Use additional codes to identify all malignancies and other
     conditions associated with the syndromes

New code  258.01  Multiple endocrine neoplasia [MEN] type I
        Wermer’s syndrome

New code  258.02  Multiple endocrine neoplasia [MEN] type IIA
        Sipple’s syndrome

New code  258.03  Multiple endocrine neoplasia [MEN] type IIB

V18  Family history of certain other specific conditions

V18.1  Other endocrine and metabolic conditions

New code  V18.11  Family history of multiple endocrine neoplasia
                [MEN] syndrome

New code  V18.19  Other endocrine and metabolic conditions

V84  Genetic susceptibility to disease

V84.0  Genetic susceptibility to malignant neoplasm

New code  V84.05  Genetic susceptibility to malignant neoplasms
                of endocrine glands
              Genetic susceptibility to multiple endocrine neoplasia [MEN]
Topic: Anal sphincter tear

Currently, the only code for anal sphincter tear associated with delivery is that included with a third degree perineal laceration. However, anal sphincter tears can occur during delivery independent of third degree lacerations, and such tears may not be identified until they complicate a subsequent delivery. In addition to being a complicating factor in a delivery, anal sphincter tears are responsible for fecal incontinence. Fecal incontinence may be the first symptom that leads to a diagnosis of an old, nonhealed anal sphincter tear in non-gravid patients. The American Academy of Obstetricians and Gynecologists (ACOG) has requested a series of codes, and other modifications, for the various anal sphincter tears in gravid and nongravid patients.

Included in the proposal are code 624.41, Anal sphincter tear (healed) (old), for non-gravid patients being seen for the complications of an old tear, and code 664.6 Anal sphincter tear complicating delivery, not associated with third-degree perineal laceration. It is also being proposed that the inclusion term anal sphincter tear (healed) (old) be added under code 654.8, Congenital or acquired abnormalities of vulva, for patients with known tears that are complicating pregnancy. Each of these codes would be excluded from each other.

TABULAR MODIFICATIONS

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>624</td>
<td>Noninflammatory disorders of vulva and perineum</td>
</tr>
<tr>
<td>624.4</td>
<td>Old laceration or scarring of vulva</td>
</tr>
<tr>
<td>New code</td>
<td>624.41</td>
</tr>
<tr>
<td></td>
<td>Use additional code for any associated fecal incontinence (787.6)</td>
</tr>
<tr>
<td></td>
<td>Excludes: anal sphincter tear (healed) (old) complicating pregnancy, childbirth, and the puerperium (654.8)</td>
</tr>
<tr>
<td>New code</td>
<td>624.49</td>
</tr>
<tr>
<td>654</td>
<td>Abnormality of organs and soft tissues of pelvis</td>
</tr>
<tr>
<td>Add</td>
<td>Excludes: trauma to perineum and vulva complicating current delivery (664.0-664.9)</td>
</tr>
<tr>
<td></td>
<td>654.8</td>
</tr>
<tr>
<td>Add</td>
<td>Anal sphincter tear (healed) (old)</td>
</tr>
</tbody>
</table>
664 Trauma to perineum and vulva during delivery

664.2 Third-degree perineal laceration

Add Excludes: anal sphincter tear during delivery not associated with third-degree perineal laceration (664.6)

New code 664.6 Anal sphincter tear complicating delivery, not associated [0,1,4] with third-degree perineal laceration

Excludes: third-degree perineal laceration (664.2)
ADDENDA

TABULAR

233 Carcinoma in situ of breast and genitourinary system

233.1 Cervix uteri
Add Adenocarcinoma in situ of cervix

250 Diabetes mellitus

250.6 Diabetes with neurological manifestations

Use additional code to identify manifestation, as:
diabetic amyotrophy (358.1 353.1)

353 Nerve root and plexus disorders

353.1 Lumbosacral plexus lesions
Add Code first any associated underlying disease, such as:
diabetes mellitus (250.6)

358 Myoneural disorders

358.1 Myasthenic syndromes in diseases classified elsewhere
Delete Amyotrophy from stated cause classified elsewhere

Delete Code first underlying disease, as:
diabetes mellitus (250.60)

438 Late effects of cerebrovascular disease

438.8 Other late effects of cerebrovascular disease
Revise 438.89 Other late effects of cerebrovascular disease
528 Diseases of the oral soft tissues, excluding lesions specific for gingiva and tongue

528.7 Other disturbances of oral epithelium, including tongue

Revise Exclude: leukokeratosis NOS (702.8)

784 Symptoms involving head and neck

784.9 Other symptoms involving head and neck

Add Feeling of foreign body in throat

V58 Encounter for other and unspecified procedures and aftercare

V58.6 Long-term (current) drug use

Revise Long-term (current) use of other medications

Add Other high-risk medications

V58.7 Aftercare following surgery to specified body systems, not elsewhere classified

V58.78 Aftercare following surgery of the musculoskeletal system NEC

Add Excludes: orthopedic aftercare (V54.01-V54.9)

V64 Persons encountering health services for specific procedures, not carried out

V64.0 Vaccination not carried out

Add Vaccination not carried out because of caregiver refusal

Add Guardian refusal

Add Parent refusal

V74 Special screening examinations for bacterial and spirochetal diseases

Add Sexually transmitted diseases
ADDENDA

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with inflammation  -  -  459.33
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Add Q wave 410.9 – see also Infarct, myocardium, by site

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Add malignant (M9080/3) – see Neoplasm, by site, malignant