



Diagnosis Agenda

Welcome and Announcements

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Co-Chair, ICD-9-CM Coordination and Maintenance Committee

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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:
<http://www.cdc.gov/nchs/icd9.htm>
- 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.

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- 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>
Procedure addendum at -
<http://www.cms.hhs.gov/paymentsystems/icd9>
- June 29, 2006 On-line registration opens for the September 28-29, 2006 ICD-9-CM Coordination and Maintenance Committee meeting at:
<http://www.cms.hhs.gov/events/>
- 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>

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- 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 1, 2006 New and revised ICD-9-CM codes go into effect along with DRG changes. Final addendum posted on web pages as follows:
Diagnosis addendum - <http://www.cdc.gov/nchs/icd9.htm>
Procedure addendum at - <http://www.cms.hhs.gov/paymentsystems/icd9>
- 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>
- Summary report of the Diagnosis part of the September 28-29, 2006 ICD-9-CM Coordination and Maintenance Committee meeting report posted on NCHS homepage at - <http://www.cdc.gov/nchs/icd9.htm>

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- 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:
<http://www.cdc.gov/nchs/icd9.htm>
- 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>
Code titles at <http://www.cms.hhs.gov/medlearn/icd9code.asp>
- December 4, 2006 Deadline for receipt of public comments on proposed code revisions discussed at the March 23 - 24, 2006 and September 29 - 30, 2006 ICD-9-CM Coordination and Maintenance Committee meetings for implementation on October 1, 2007.

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NCHS Classifications of Diseases web page:
<http://www.cdc.gov/nchs/icd9.htm>

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).

414 Other forms of chronic ischemic heart disease

414.0 Coronary atherosclerosis

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

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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

Revise	200	Lymphosarcoma and reticulosarcoma <u>and other specified malignant tumors of lymphatic tissue</u>
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.

TABULAR MODIFICATION

	331	Other cerebral degenerations
	331.3	Communicating hydrocephalus Secondary normal pressure hydrocephalus
Add		
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)
	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)
New code	331.5	Idiopathic normal pressure hydrocephalus Normal pressure hydrocephalus
		Excludes: congenital hydrocephalus (741.0 ,742.3)
		secondary normal pressure hydrocephalus (331.3)
		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:

TABULAR MODIFICATION

	V25	Encounter for contraceptive management
	V25.0	General counseling and advice
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

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Option 2:

TABULAR MODIFICATION

Revise	V25	Encounter for contraceptive <u>family planning</u> management
	V25.0	General counseling and advice
New code	V25.04	Counseling and instruction in natural method of family planning
	V25.4	Surveillance of previously prescribed family planning methods
New code	V25.44	Natural method of family planning
	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 with natural method of birth regulation
New code	V26.49	Other procreative management counseling and advice

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 of 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.

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)

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 <u>and delivery</u> , in liveborn infant
Revise		Fetal metabolic acidemia first noted during labor <u>and delivery</u> , 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	768.9	Unspecified birth asphyxia in liveborn infant Hypoxia NOS, 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

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	775	Endocrine and metabolic disturbances specific to the fetus and newborn
Revise	775.8	Other transitory neonatal endocrine and metabolic disturbances
Delete		Amino-acid metabolic disorders described as transitory
New code	775.81	Other acidosis of newborn Acidemia NOS of newborn Acidosis of newborn NOS Mixed metabolic and respiratory acidosis of newborn
New code	775.89	Other neonatal endocrine and metabolic disturbances Amino-acid metabolic disorders described as transitory
	779	Other and ill-defined conditions originating in the perinatal period
Add	779.2	Cerebral depression, coma, and other abnormal cerebral signs Cerebral ischemia NOS of newborn
	779.8	Other specified conditions originating in the perinatal period
New code	779.85	Cardiac arrest of newborn

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.

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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
		Parascharlatina
Delete		Pseudoscarlatina
		Roseola infantum
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

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New subcategory 058.2 Human herpesvirus 7
 New code 058.20 Human herpesvirus 7, unspecified
 New code 058.21 Roseola infantum due to human herpesvirus 7
 Exanthema subitum [sixth disease] due to human herpesvirus 7
 Add Excludes: Roseola infantum, unspecified (058.0)
 New code 058.29 Other human herpesvirus 7
 New code 058.3 Human herpesvirus 8
 Kaposi's sarcoma-associated herpesvirus

Option 2

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 (058.21)
 other human herpesvirus encephalitis (058.29)
 054 Herpes simplex
 054.3 Herpetic meningoencephalitis
 Add Excludes: human herpesvirus 6 encephalitis (058.21)
 other human herpesvirus encephalitis (058.29)
 057 Other viral exanthemata
 057.8 Other specified viral exanthemata
 Dukes (-Filatow) disease
 Delete ~~Exanthema subitum [sixth disease]~~
 Fourth disease
 Parascarlatina
 Pseudoscarlatina
 Delete ~~Roseola infantum~~
 Add Excludes: Exanthema subitum [sixth disease] (058.10-058.12)
 Roseola infantum (058.10-058.12)

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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 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

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New subcategory	058.8	Other human herpesvirus infections
New code	058.81	Human herpesvirus 6 infection
New code	058.82	Human herpesvirus 7 infection
New code	058.89	Other human herpesvirus infection Human herpesvirus 8 infection Kaposi's sarcoma-associated herpesvirus infection

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 adrenocorticotrophic 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

Topic: Stevens-Johnson syndrome

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.

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.

TABULAR MODIFICATION

V07 Need for isolation and other prophylactic measures

V07.3 Other prophylactic chemotherapy

New code V07.32 Prophylactic administration of antiestrogen agents

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:

TABULAR MODIFICATION

	333	Other extrapyramidal disease and abnormal movement disorders
	333.9	Other and unspecified extrapyramidal diseases and abnormal movement disorders
New code	333.94	Restless legs syndrome
Delete	333.99	Other Restless legs

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.

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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 Category	249	Diabetes mellitus due to underlying condition 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

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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)

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)

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- 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
- Revise Excludes: hypoglycemia:
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.

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	005	Other food poisoning (bacterial)
Revise	005.1	Botulism <u>food poisoning</u>
Add		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

	624	Noninflammatory disorders of vulva and perineum
	624.0	Dystrophy of vulva
Delete		Kraurosis of vulva
		Leukoplakia of vulva
		Excludes: carcinoma in situ of vulva (233.3)
Add		severe dysplasia of vulva (233.3)
Add		vulvar intraepithelial neoplasia III [VIN III] (233.3)
New code	624.01	Vulvar intraepithelial neoplasia I [VIN I] Mild dysplasia of vulva
New code	624.02	Vulvar intraepithelial neoplasia II [VIN II] Moderate dysplasia of vulva
New code	624.09	Other dystrophy of vulva Kraurosis of vulva Leukoplakia of vulva

Topic: 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

Delete	193	Malignant neoplasm of thyroid gland Sipple's syndrome
	258	Polyglandular dysfunction and related disorders
Delete	258.0	Polyglandular activity in multiple endocrine adenomatosis Wermer's syndrome
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

	624	Noninflammatory disorders of vulva and perineum
	624.4	Old laceration or scarring of vulva
New code	624.41	Anal sphincter tear (healed) (old)
		Use additional code for any associated fecal incontinence (787.6)
		Excludes: anal sphincter tear (healed) (old) complicating pregnancy, childbirth, and the puerperium (654.8)
New code	624.49	Other old laceration or scarring of vulva
	654	Abnormality of organs and soft tissues of pelvis
Add		Excludes: trauma to perineum and vulva complicating current delivery (664.0-664.9)
Add	654.8	Congenital or acquired abnormalities of vulva
		Anal sphincter tear (healed) (old)

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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
Revise		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
		Code first underlying disease, as: diabetes mellitus (250.60)
Delete		
	438	Late effects of cerebrovascular disease
		438.8 Other late effects of cerebrovascular disease
Revise		438.89 Other late effects of <u>cerebrovascular</u> disease

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- 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 784.99 Other symptoms involving head and neck
Feeling of foreign body in throat
- V58 Encounter for other and unspecified procedures and aftercare
- V58.6 Long-term (current) drug use
- Revise V58.69 Long-term (current) use of other medications
Other hHigh-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
- V64.05 Vaccination not carried out because of caregiver refusal
- Add Guardian refusal
- Add Parent refusal
- V74 Special screening examinations for bacterial and spirochetal diseases
- V74.5 Venereal disease
- Add Sexually transmitted diseases

ADDENDA

INDEX

Add	Abnormal, abnormality, abnormalities - see also Anomaly blood sugar 790.29
Add	Aftercare V58.9 following surgery NEC V58.49 spinal – see Aftercare, following surgery, of, specified body system
Revise	Anemia 285.9 postoperative due to (<u>acute</u>) blood loss 285.1
Add	chronic blood loss 280.0
Add	Complications chemotherapy 995.29
Add	drug NEC 995.29
Revise	Congestion, congestive- (chronic) (passive) chest <u>460</u>
Revise	lungs <u>514</u>
Add	meaning hypostatic pneumonia 514
Add	due to common cold 460
Add	nose 478.1
Add	Damage medication 995.20
Add	Deficiency, deficient short stature homeobox gene (SHOX)
Add	with
Add	dyschondrosteosis 756.59
Add	idiopathic short stature 783.43
Add	Turner syndrome 758.6
Add	Disease... liver 573.9 end stage 572.8
Add	Disorder... bleeding 286.9
Add	involuntary emotional expression (IEED) 310.8

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	Elevation			
Add	blood sugar	790.29		
Add	cholesterol	272.0		
	Fracture...			
Add	burst – see Fracture, traumatic, by site			
Add	insufficiency – see Fracture, pathologic, by site			
Revise	Gas	<u>787.3</u>		
	Gastropathy	537.9		
Add	congestive, portal	537.89		
Add	portal, hypertensive	537.89		
Add	Grief	309.0		
	History (personal) of			
Add	hysterectomy	V45.77		
Revise	Hyperactive, hyperactivity	<u>314.01</u>		
	Hypertension, hypertensive...		Malign't	Benign
	venous, chronic (asymptomatic)...		-	-
Delete	due to			Unspc
	deep vein thrombosis			459.30
	(see also Syndrome, postphlebitic)			459.10
	with			
	complication, NEC		-	-
	inflammation		-	-
	with ulcer		-	-
	ulcer		-	-
	with inflammation		-	-
Add	due to			
	deep vein thrombosis (see also			
	Syndrome, postphlebitic)		-	-
				459.10

	Infarct, infarction
	myocardium...
Add	non-Q wave 410.7
Add	Q wave 410.9 – see also Infarct, myocardium, by site
	Isoimmunization...
Add	anti-E 656.2
	Long-term (current) drug use V58.69
Add	pain killers V58.69
Add	anti-inflammatories, non-steroidal (NSAID) V58.64
Add	aspirin V58.66
Add	selective estrogen receptor modulators (SERM) V07.39
	Lymphoma...
	diffuse...
Add	large B cell 202.8
	Necrosis, necrotic
Add	colon 557.0
	Neoplasm...
Revise	N Mackenrodt's ligament ...
	Nephrosis...
Add	Finnish type (congenital) 759.89
	Pain(s)
Add	menstrual 625.3
Add	premenstrual 625.4
Revise	Paraparesis (see also <u>Paralysis</u> <u>Paraplegia</u>) 344.1
	Person (with)
Add	“worried well” V65.5
Delete	“worried well” V65.5
	Personality
	schizoid 301.20
Delete	with sexual deviation (see also <u>Deviation, sexual</u>) 302.9
Delete	antisocial 301.7
Delete	dyssoical 301.7
Add	Poison ivy, oak, sumac or other plant dermatitis 692.6
	Poisoned - see Poisoning

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Add Poisoning (acute) - see also Table of Drugs and Chemicals
water 276.6
Delete ~~Poison ivy, oak, sumac or other plant dermatitis 692.6~~

Pregnancy...
complicated (by)
Add post cesarean uterine artery clot 669.4

Prophylactic
administration of
Add drug V07.39
Add medication V07.39

Add Protection (against) (from) – see Prophylactic

Revise Regurgitation 787.03

Add Runny nose 784.99

Add Scratchy throat 784.99

Add Seizure 780.39
due to stroke 438.89

Revise Short, shortening, shortness
stature, constitutional, (hereditary) (idiopathic) 783.43

Revise Stenosis...
artery (see also Arteriosclerosis) 447.1
Add extremities 440.20

Revise Stress 308.9

Revise Swelling 782.3

Add Syndrome
hyperperfusion 997.01
Add hypothenar hammer 443.89

Add Tear...
dural 998.2

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Teratoma...

mature (M9080/0) - see Neoplasm, by site, benign

Add

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