Summary of September 2010 ICD-9-CM Coordination and Maintenance Committee Meeting

The ICD-9-CM Coordination and Maintenance (C&M) Committee, cosponsored by the Centers for Disease Control and Prevention (CDC) and the Centers for Medicare and Medicaid Services (CMS), met on September 15-16, 2010 in Baltimore, MD. Donna Pickett, RHIA, from CDC, and Patricia Brooks, RHIA, from CMS, cochaired the meeting.

Several ICD-10 topics were discussed, included a code set freeze, modifications to the General Equivalence Mappings (GEMs), and ICD-10-based MS-DRGs. Proposed modifications to ICD-9-CM and ICD-10-CM/PCS were also presented and are summarized below.

This summary does not include all of the details of the code proposals or all of the recommendations made at the meeting. For complete details, review the minutes and code proposals posted on the CMS and NCHS websites. Diagnosis code proposals and the minutes from the diagnosis portion of the meeting are posted on the CDC website and can be accessed at the following link: http://www.cdc.gov/nchs/icd/icd9cm_maintenance.htm. Procedure code proposals and the minutes from the procedure portion of the meeting can be found at the CMS website and can be accessed at the following link: http://www.cms.hhs.gov/ICD9ProviderDiagnosticCodes/03_meetings.asp.

The proposed code modifications, if approved by CMS and CDC, would go into effect with discharges on or after October 1, 2011. None of the code proposals presented at this meeting is being considered for implementation on April 1, 2011.

Suggestions for procedure code proposals to be considered at a future Coordination and Maintenance Committee, as well as comments on procedure proposals presented at the September meeting, may be emailed to Pat Brooks at Patricia.brooks2@cms.hhs.gov or mailed to: Centers for Medicare & Medicaid Services, CMM, HAPG, Division of Acute Care, Mail Stop C4-08-06, 7500 Security Boulevard, Baltimore, Maryland 21244-1850.

Suggestions for diagnosis code proposals for consideration at a future Coordination and Maintenance Committee, as well as comments on diagnosis proposals presented at the September meeting, may be emailed to Donna Pickett at dfp4@cdc.gov or mailed to: Donna Pickett, National Center for Health Statistics, 3311 Toledo Road, room 2402, Hyattsville, Maryland 20782.
The deadline for receipt of public comments on revisions to the GEMs (for the 2011 version of the GEMs) is November 12, 2010.

The deadline for receipt of public comments on the code proposals presented at the September meeting is November 19, 2010.

The next meeting of the ICD-9-CM Coordination and Maintenance Committee is scheduled for March 9-10, 2011 and will be held at the CMS building in Baltimore, MD. New code proposals for inclusion on this agenda must be received by January 7, 2011.

ICD-10-CM/PCS Topics

Final Decision on Partial Code Freeze for ICD-9-CM and ICD-10-CM/PCS

The ICD-9-CM Coordination and Maintenance Committee will implement a partial freeze of the ICD-9-CM and ICD-10 (ICD-10-CM and ICD-10-PCS) codes prior to the implementation of ICD-10 on October 1, 2013. The partial freeze will be implemented as follows:

- The last regular, annual updates to both ICD-9-CM and ICD-10 code sets will be made on October 1, 2011.
- On October 1, 2012, there will be only limited code updates to both the ICD-9-CM and ICD-10 code sets to capture new technologies and diseases as required by section 503(a) of Pub. L. 108-173.
- On October 1, 2013, there will be only limited code updates to ICD-10 code sets to capture new technologies and diagnoses as required by section 503(a) of Pub. L. 108-173. There will be no updates to ICD-9-CM, as it will no longer be used for reporting.
- On October 1, 2014, regular updates to ICD-10 will begin.

The ICD-9-CM Coordination and Maintenance Committee will continue to meet twice a year during the partial freeze. At these meetings, the public will be asked to comment on whether or not requests for new diagnosis or procedure codes should be created based on the criteria of the need to capture a new technology or disease. Any code requests that do not meet the criteria will be evaluated for implementation within ICD-10 on and after October 1, 2014 once the partial freeze has ended.

The GEMs are not part of the code freeze; they will continue to be updated annually.

ICD-10-CM/PCS GEMs 2010 Version Update

Section 10109(c) of the Patient Protection and Affordable Care Act of 2010 (ACA) requires the Secretary of Health and Human Services to task the C&M Committee to convene a meeting before January 1, 2011, to receive stakeholder input regarding the crosswalk between ICD-9 and ICD-10 for the purpose of making appropriate revisions to the crosswalk. Section 10109(c) further requires that any revised crosswalk be treated as a code set for which a standard has been adopted by the Secretary and that revisions to this crosswalk be posted to the CMS website. The morning of the first day of the September C&M Committee meeting was used to fulfill the ACA
requirements regarding public input on crosswalk revisions. No other meeting will be convened by the C&M Committee for this purpose.

CMS clarified that the GEMs (but not the Reimbursement Mappings) are the “crosswalk” referenced in the ACA legislation.

Additional documentation for using the GEMs, called “GEMs documentation for technical users,” was developed for clinical analysts and information technology users and has been posted on the CMS website. This document specifies GEMs entry inclusion criteria and provides examples; discusses GEMs flags in detail and provides examples; answers other frequently asked technical questions; and discusses translation rules for obstetrics and angioplasty.

The updated 2010 GEMs were posted on the CMS website for public comment. The updated files contain all changes to date in response to public comment and internal review. Also, a comprehensive review of the ICD-9-CM to ICD-10-CM and ICD-10-PCS GEM files was conducted, and entries were streamlined as needed to conform to inclusion criteria as defined in the GEMs documentation for technical users.

The public was encouraged to provide comments on the GEMs, both at the C&M Committee meeting as well as in writing following the meeting. Commenters expressed their appreciation for the GEMs as a valuable transition tool. Other commenters provided specific suggestions for modifications of the GEMs.

The 2011 update of the diagnosis and procedure GEMs, as well as updated Reimbursement Mappings, are expected to be posted in January 2011.

Interested parties and other stakeholders should submit their written comments on the GEMs by November 12, 2010 to Pat Brooks (CMS) and Donna Pickett (CDC).

**MS-DRG Impact Analysis**

A report on the impact of the transition to ICD-10 on Medicare inpatient hospital payments was developed by 3M Health Information Systems and presented at the C&M Committee meeting. The results of this impact analysis showed that the conversion to a native ICD-10 version of MS-DRGs (which is how CMS converted the MS-DRGs) will have a minimal impact on aggregate payments to hospitals and the distribution of payments across hospitals. Mapping ICD-10 data back to ICD-9-CM and using the ICD-9-CM version of MS-DRGs would produce a greater impact on aggregate payments to hospitals and the distribution of payments across hospitals. Any mapping will inherently produce less consistent results because a single choice between mapping alternatives is uniformly applied across all DRGs, whereas in a native conversion context, specific judgment can be used to independently assign translation alternatives on a DRG by DRG basis.

If payers do not convert their core payment and claims adjudication systems to native ICD-10 versions and instead use an ICD-10 to ICD-9-CM mapping in order to continue to use their existing ICD-9-CM based systems, there are potential biases and unintended results of such an approach. This is especially true if a payer attempts to use a single uniform mapping across all
systems. Fortunately, as evidenced by the development of the native ICD-10 version of the MS-DRGs, CMS appears to be moving toward creating native ICD-10 versions of its systems instead of mapping ICD-10 data to ICD-9-CM and continuing to use ICD-9-CM based systems.

A complete report of the payment impact analysis can be found with the materials for the C&M Committee posted on the CMS website.

**Version 28.0 ICD-10 MS-DRGs**

CMS expects to post version 28.0 ICD-10 MS-DRGs definitions manual in February 2011 and to release the version 28.0 ICD-10 grouper in March 2011.

**ICD-10-PCS Update**

The 2011 ICD-10-PCS update contains 72,131 codes. Examples of code revisions made in response to public input are the addition of a new qualifier, Humeral Surface, to the shoulder joint body part to specify partial shoulder replacement and the streamlining of device values for interbody spinal fusion devices.

The updated 2011 ICD-10-PCS files will be posted in October 2010 (except for the GEMs and Reimbursement Mappings, which will be posted in January 2011).

The ICD-10-PCS guidelines were reviewed and revised by the Cooperating Parties (American Health Information Hospital Association, American Hospital Association, CMS, and CDC) and the 2011 version was posted on the CMS website in June 2010. The guidelines are also included in the ICD-10-PCS Reference Manual, Appendix B.

**ICD-10-CM Update**

The 2011 ICD-10-CM update (including the coding guidelines) will be posted in January 2011. This update will include a machine-readable ICD-10-CM file in XML format.

**Diagnoses**

**ICD-9-CM Proposals**

**Infection Following Transfusion**

The Food and Drug Administration (FDA) Center for Biologics Evaluation and Research requested a new code for infection following transfusion, infusion, or injection of blood and blood products. An instructional note under this code would indicate that human immunodeficiency virus (HIV) disease (042) should be coded first. Attendees expressed concern about the length of time the proposed code would continue to be used, since the transfusion-associated infection may be present for many years (as in the case of HIV).
Anaphylactic Reaction and Other Serum Reaction

The FDA Center for Biologics Evaluation and Research requested unique codes for anaphylactic reactions due to administration of blood and blood products and for anaphylactic reactions due to vaccination. They also requested new codes for other serum reactions due to administration of blood and blood products and for other serum reactions due to vaccination. The titles of existing codes would also be modified to reflect “anaphylactic reaction” rather than “anaphylactic shock” to clarify that all anaphylactic reactions are included in these codes, even when there is no shock.

Anaphylactic reactions are a type I hypersensitivity reaction, involving IgE antibodies, and occurring immediately with exposure. Other serum reactions can also occur, including type II and type III hypersensitivity reactions, which both involve IgG antibodies. There are certain types of transfusion reactions and drug reactions that exemplify type II hypersensitivity reactions. Examples of type III hypersensitivity reactions include serum sickness and Arthus reaction. Serum sickness is a hypersensitivity reaction to a protein in serum, generally occurring one to three weeks after exposure. It may involve fever, itching, and rash, potentially with other manifestations, some of which include joint pain and swollen lymph nodes.

Mesh Erosion/Mesh Exposure

New codes for mesh erosion and mesh exposure were proposed. Two options were offered. The first option would create two codes for mesh erosion and mesh exposure in category 998, Other complications of procedures, NEC, which could be used by any surgical specialty. The second option would create a new subcategory for mechanical complication of implanted vaginal mesh in category 629, Other disorders of female genital organs, with two codes for erosion of implanted vaginal mesh to surrounding organ or tissue and exposure of implanted vaginal mesh into vagina. Mesh erosion and exposure can both be present at the same time.

In an abdominal sacral colpopexy, a graft is used to suspend the upper vagina to the anterior longitudinal ligament of the sacrum. Synthetic graft material used to suspend the apex of the vagina to the anterior longitudinal ligament of the sacrum has been associated with mesh erosion and subsequent pelvic infection (due to the erosion into surrounding organs or tissue). Treatment for the erosion usually requires surgical removal of the mesh. Exposure of the mesh, into the vagina, can also occur. Though this is a less severe condition which can be treated nonsurgically sometimes, it does have potential for infection to develop. Mesh and mesh patches are also used to repair ventral (incisional) hernias caused by thinning or stretching of scar tissue that forms after surgery. Mesh erosion is a known complication that results in bowel perforations and/or chronic intestinal fistulae.

Attendees expressed concern that the terminology may be problematic because the terms may have different meanings in other clinical circumstances or when used by other surgical specialties. For example, this type of erosion isn’t the same as a pacemaker erosion through the skin. It was suggested that inclusion terms and index entries may need to be added to ensure proper code usage.

Malnutrition

The American Dietetic Association and the American Society for Parenteral and Enteral Nutrition have requested several new codes and instructional notes to update the classification of malnutrition. The existing ICD-9-CM codes for malnutrition are outdated and do not reflect the
current standard of care or understanding of malnutrition-disease interaction. Under this proposal, new codes would be created for:

- Severe malnutrition in acute injury
- Severe malnutrition in acute illness
- Severe malnutrition in chronic illness
- Severe malnutrition in environmental and social circumstances
- Severe malnutrition related to other disorders

Instructional notes would indicate that the underlying condition should be sequenced first.

Attendees suggesting deleting “frailty” from the “code first” notes under the proposed new codes. It was also suggested that the proposed code titled “severe malnutrition related to other disorders” be changed to “severe malnutrition in other disorders.” Objections were raised to the proposed indexing of both severe malnutrition not due to a specified underlying condition and malnutrition NOS to code 263.9, Unspecified protein-calorie malnutrition.

Lymphangioleiomyomatosis

A unique code for lymphangioleiomyomatosis has been proposed in category 516, Other alveolar and parietoalveolar pneumonopathy. Lymphangioleiomyomatosis, also known as lymphangiomyomatosis or LAM, is a rare, frequently fatal lung disease that affects women almost exclusively. It may occur sporadically (not inherited), and may also occur in patients with tuberous sclerosis complex (inherited). LAM is characterized by the infiltration of the lung with neoplastic smooth muscle cells of unknown origin and cystic destruction of lung tissue.

Although ICD-9-CM has an index entry for lymphangiomyomatosis with an instructional note to “see Neoplasm, connective tissue, uncertain behavior,” the clinical behavior of this condition is more consistent with an interstitial lung disease rather than a neoplasm.

Dementia Unspecified With and Without Behavioral Disturbance

It has been requested that fifth digits be added to code 294.8, Other persistent mental disorder due to conditions classified elsewhere, to identify the presence or absence of behavioral disturbance when the etiology of the dementia is not known.

Elective C-Sections Prior to 39 Weeks

The American College of Obstetricians and Gynecologists has requested a new code in category 644, Early or threatened labor, for late preterm onset of labor, with delivery. An inclusion term would be added indicating that onset (spontaneous) of labor after 37 weeks of gestation but before 39 weeks gestation, with delivery, is included in this code.

A quality measure involves looking at elective deliveries done prior to 39 weeks gestation. Planned repeat cesarean section deliveries performed prior to 39 weeks fall into this category. However, often these deliveries occur earlier than planned because the patient presents at 37-38 weeks gestation in labor, and the physician determines that it is best to go ahead and deliver at that time rather than to try to take measures to wait until the 39th week. Therefore, a unique code was proposed to identify the onset of labor at 37-38 weeks gestation with delivery.
Attendees noted that the semantics and placement of the code are problematic, since seemingly conflicting terms of “late” and “preterm” are used in the proposed code title, and the proposed placement of this code is in a category for early or threatened labor. Suggested alternatives to the code proposal included creating codes indicating the number of weeks and with or without active labor, or creating V codes indicating whether the patient was in labor or not.

**Personal History of Gestational Diabetes**

A new code for personal history of gestational diabetes has been requested.

**Encounter for Fetal Viability Ultrasound/Personal History of Ectopic Pregnancy**

The American College of Obstetricians and Gynecologists has requested a new code for pregnancy with inconclusive fetal viability in subcategory V23.4, Pregnancy with other poor obstetric history. “Encounter to determine fetal viability of pregnancy” would be an inclusion term under this code. Patients previously confirmed as pregnant in very early weeks may return within a few weeks, and if the fetal heartbeat cannot be heard, an ultrasound may be necessary to confirm that the pregnancy is viable. The proposed code would provide a means to accurately describe these encounters. A C&M meeting attendee questioned the appropriateness of the proposed placement of the code because the subcategory describes pregnancy with poor obstetric history, and the inconclusive fetal viability described in the proposed code refers to the current pregnancy.

A new code for pregnancy with history of ectopic pregnancy has also been requested.

**Adult Pulmonary Langerhans Cell Histiocytosis (PLCH)**

A unique code for Adult Pulmonary Langerhans Cell Histiocytosis (PLCH) has been requested. This condition is a rare interstitial lung disorder of unknown etiology that occurs almost exclusively in smokers. In adults, pulmonary involvement with Langerhans’ cell histiocytosis usually occurs as a single-system disease and is characterized by focal Langerhans’ cell granulomas infiltrating and destroying distal bronchioles. Since the clinical behavior of adult PLCH is consistent with an interstitial lung disease, rather than a neoplasm or a metabolic disorder, the new code has been proposed in category 516, Other alveolar and parietoalveolar pneumonopathy.

**Acquired Absence of Joint**

A new subcategory for acquired absence of joint has been proposed. Codes for acquired absence of hip joint, knee joint, and other joint would be created in this subcategory. Inclusion terms would indicate that these codes include acquired absence of joint following explantation of joint prosthesis, with or without presence of antibiotic-impregnated cement spacer.

**Glaucoma Severity Staging**

A new subcategory to identify the glaucoma stage has been proposed. Treatment at early stages of glaucoma typically results in better outcomes and uses fewer resources than patients who present with more severe stages.
New codes for open angle glaucoma with borderline findings, high risk, primary angle closure glaucoma without glaucoma damage, and family history of glaucoma have also been proposed. As part of this proposal, the title of code 365.01 would be revised to state “open angle with borderline findings, low risk.”

Meeting attendees suggested that separate codes should be created for indeterminate and unspecified glaucoma stage.

**Corticobasal Degeneration**

A unique code for corticobasal degeneration has been proposed. Corticobasal degeneration is a neurodegenerative disease that is manifested by both a movement disorder and cognitive impairment. The cognitive symptoms resemble those of the frontotemporal dementias, especially loss of executive function, visuospatial and number processing, and language impairment. The movement disorder most often presents asymmetrically and may include akinetic-rigid syndrome, myoclonus, or dystonia. Patients may also have alien limb syndrome, apraxia, and cortical sensory loss.

**Pulmonary Arteriovenous Malformation and Pulmonary Atresia**

New codes have been requested for pulmonary artery coarctation and atresia and pulmonary arteriovenous malformation. A pulmonary arteriovenous malformation is an abnormal communication between pulmonary arteries and pulmonary veins. While they are most often congenital, they can also be acquired. They may also be called pulmonary arteriovenous aneurysms or pulmonary arteriovenous fistulae.

Narrowing of the pulmonary artery is called coarctation or stenosis. Complete failure of the pulmonary valve to form, with the origin of the pulmonary artery not connecting to the heart, may be called pulmonary artery atresia or agenesis. This is generally seen only along with a patent ductus arteriosus.

**Complications of Stem Cell Transplant**

There is no unique code for complications of stem cell transplants in ICD-9-CM. Two options for handling these complications were presented at the C&M meeting. The first option would involve creation of a unique code for complications of stem cell transplants. In the second option, complications of stem cell transplants would be indexed to existing code 996.85, Complications of transplanted organ, Bone marrow.

The greatest concentration of blood stem cells is in the bone marrow. However, it is possible to move blood stem cells out of the bone marrow into the bloodstream or “peripheral blood” where they can be collected and used instead of bone marrow for the transplant. Umbilical cord blood also contains blood stem cells that can be used for transplant. With the availability of stem cell growth factors, most hematopoietic stem cell transplantation procedures are now performed using stem cells collected from the peripheral blood, rather than from the bone marrow. Collecting peripheral blood stem cells provides a bigger graft, does not require that the donor be subjected to general anesthesia to collect the graft, results in a shorter time to engraftment, and may provide for a lower long-term relapse rate.
**Pseudobulbar Affect**

A unique code for pseudobulbar affect has been proposed. A “code first” note would indicate that the code for the underlying cause should be sequenced first. It was suggested at the C&M meeting that “if applicable” should be added to the “code first” note to clarify that the underlying cause should be sequenced first only if it has been identified.

Pseudobulbar affect is a neurologic condition caused by underlying structural damage in the brain which triggers involuntary, frequent, and disruptive outbursts of crying or laughing that are out of proportion or incongruent to the patient’s underlying emotional state. The pathophysiology of pseudobulbar affect is widely believed to involve injury to the neurologic pathways that regulate affect.

**Reportable Malignant Skin Cancers**

The New York State Cancer Registry has requested that ICD-9-CM be expanded to allow for the classification of reportable skin cancers that are currently included under category 173, Other malignant neoplasm of skin. The vast majority of skin cancers are either basal or squamous cell, neither of which are reportable conditions to central cancer registries. It has been proposed that codes under category 173 be expanded to the 5th digit level to allow for the differentiation of non-reportable (basal and squamous cell) and reportable skin cancers. Under this proposal, every code in category 173 would be split into two separate codes – basal cell/squamous cell carcinoma and other malignant neoplasm.

**Disorders Due to Intrinsic Circulating Anticoagulants, Antibodies, or Inhibitors**

An expansion of code 286.5, Hemorrhagic disorder due to intrinsic circulating anticoagulants, has been requested to specifically capture disorders due to iatrogenic anticoagulants, acquired hemophilia, and antiphospholipid antibody with hemorrhagic disorder. Hyperheparinemia, which is an inclusion term under the proposed code for disorders due to iatrogenic anticoagulants, is an iatrogenic disorder related to elevated heparin levels from being given too much heparin. Acquired hemophilia, or secondary hemophilia, is a disorder in which antibodies to a coagulation factor develop, usually coagulation factor VIII. This is also called autoimmune hemophilia. Systemic lupus erythematosus (SLE) inhibitor or lupus anticoagulant is an antibody directed against protein phospholipid complexes. There are certain other antiphospholipid antibodies that when present are a risk factor for thromboembolic disease, but patients may also be asymptomatic. While these antibodies do not typically cause hemorrhagic disease, there are some reported cases where such antibodies appeared to be related to bleeding. When a hypercoagulable state is present, code 289.81, Primary hypercoagulable state, should be used instead of the proposed new code. Presence of antibody without any diagnosis should be assigned code 795.79, Other and unspecified nonspecific immunological findings.

**Interstitial Lung Diseases of Childhood**

The American Thoracic Society and the American College of Chest Physicians have requested new codes for interstitial lung diseases of childhood. Both options would involve creation of a new subcategory for interstitial lung diseases of childhood in category 516, Other alveolar and parietoalveolar pneumonopathy. In the first option, new codes would be created for: neuroendocrine cell hyperplasia of infancy; pulmonary interstitial glycogenosis; surfactant
mutations of the lung; and alveolar capillary dysplasia with vein misalignment. The second option would involve specification of each surfactant mutation of the lung, with specific codes for: neuroendocrine cell hyperplasia of infancy; pulmonary interstitial glycogenosis; surfactant protein B mutation of the lung; surfactant protein C mutation of the lung; surfactant associated ATP binding cassette A3 mutation of the lung; surfactant associated thyroid transcription factor 1 mutations of the lung; and alveolar capillary dysplasia with vein misalignment.

Neuroendocrine cell hyperplasia of infancy is an interstitial lung disease associated with prolonged oxygen use for years in children, starting in the first year of life and persistsing with mild symptoms into adolescence. Pulmonary interstitial glycogenosis results from the proliferation of a poorly defined clear cell population that contains glycogen in the alveolar interstitium, resulting in significant thickening of this space and marked diffusion abnormalities for oxygen. This disorder can occur as the primary finding or in association with premature lung disease or congenital heart disease. The surfactant mutations of the lung are a group of disorders that are a leading indication for pediatric lung transplantation. Children with these disorders most commonly present in the newborn period, but may present in later childhood with unknown chronic lung disease. Each surfactant mutation has a characteristic clinical presentation, course and prognosis. Alveolar capillary dysplasia with vein misalignment is a developmental lung disorder involving rapidly progressive respiratory failure and severe pulmonary hypertension that progresses to death in the first 2 months of life despite therapeutic interventions for pulmonary hypertension, advanced ventilation strategies, and extracorporeal membrane oxygenation.

Meeting attendees raised the question as to whether the proposed codes for these conditions should be more appropriately located in chapter 15. However, apparently some of these diseases don’t develop until after the perinatal period. Another commenter noted that coders may look for medical record documentation specifying that the condition is “of childhood” in order to assign one of the proposed codes.

**Idiopathic Pulmonary Fibrosis**

The American Thoracic Society and the American College of Chest Physicians have requested unique codes for idiopathic pulmonary fibrosis and idiopathic interstitial pneumonia, not otherwise specified. Idiopathic pulmonary fibrosis is a distinctive type of chronic fibrosing interstitial pneumonia of unknown cause. Onset of symptoms is usually gradual, with dyspnea the most prominent and disabling symptom. A nonproductive cough is usually present and may be paroxysmal. The patient’s age at onset is usually greater than 50 years.

**Nonspecific Interstitial Pneumonitis**

The American Thoracic Society and the American College of Chest Physicians have requested a unique code for idiopathic non-specific interstitial pneumonitis. Nonspecific interstitial pneumonitis is a type of idiopathic interstitial pneumonia. Onset of this disease is often between ages 40-50 years. Breathlessness, cough, and fatigue are common symptoms, and many patients experience weight loss. The physician presenting this topic at the C&M meeting clarified that “nonspecific” in the proposed code title is a pathologic description, not a general descriptive phrase, and so it would be documented in the medical record.
Acute Interstitial Pneumonia

The American Thoracic Society and the American College of Chest Physicians have requested a specific code for acute interstitial pneumonia. Acute interstitial pneumonia is a rapidly progressive and histologically distinct form of interstitial pneumonia. It occurs over a wide age range, with a mean age of approximately 50 years old. There is no effective treatment and mortality rates are high. Most deaths occur between one and two months of the onset of illness.

A C&M meeting attendee commented that radiologists may read an x-ray as indicating an interstitial process, and some physicians may then describe the patient’s condition as an acute interstitial pneumonia rather than an acute atypical pneumonia. Therefore, pneumonia that is not actually an acute interstitial pneumonia may end up classified to the proposed new code. The presenter noted that a chest x-ray would not provide sufficient information to establish a diagnosis of true acute interstitial pneumonia and that a lung biopsy would be needed.

Respiratory Bronchiolitis-associated Interstitial Lung Disease

The American Thoracic Society and the American College of Chest Physicians have requested a unique code for respiratory bronchiolitis-associated interstitial lung disease. This condition is the clinical manifestation of interstitial lung disease associated with the pathologic lesion of respiratory bronchiolitis. Respiratory bronchiolitis is a histopathologic lesion found in cigarette smokers and is characterized by the presence of pigmented intraluminal macrophages within respiratory bronchioles. It is rarely symptomatic and is usually associated with no more than minor small airway dysfunction. However, in rare cases the condition presents as a form of interstitial lung disease with significant pulmonary symptoms, abnormal pulmonary function, and imaging abnormalities. It is then described as respiratory bronchiolitis-associated interstitial lung disease.

Lymphocytic Interstitial Pneumonia

The American Thoracic Society and the American College of Chest Physicians have requested a specific code for idiopathic lymphoid interstitial pneumonia. Lymphocytic interstitial pneumonia, or lymphoid interstitial pneumonia, is characterized by the infiltration of the pulmonary interstitium with lymphocytes and plasma cells. Onset of this condition is often slow with gradually increasing cough and breathlessness over three or more years. Fever, weight loss, chest pain, and arthralgia are occasionally found. It is more common in women. And although lymphocytic interstitial pneumonia may present at any age, it is most typically diagnosed in the fifth decade.

The addition of instructional notes under 516.8, Other specified alveolar and parietoalveolar pneumonopathies, indicating that the underlying cause of pneumonopathy should be coded first, if applicable, and that an additional E code for drug-induced or toxic pneumonopathy should be used, if applicable, has also been proposed.

Cryptogenic Organizing Pneumonia

The American Thoracic Society and the American College of Chest Physicians have requested a new code for cryptogenic organizing pneumonia. The term “cryptogenic organizing pneumonia” is preferred over previous names, such as bronchiolitis obliterans with organizing pneumonia.
(BOOP). In cryptogenic organizing pneumonia, patients typically present with an illness of relatively short duration (usually less than three months) with variable degrees of cough and dyspnea. The majority of patients recover completely on administration of oral corticosteroids, but a significant number relapse within one to three months when the corticosteroids are reduced or stopped. This condition develops twice as frequently in smokers as non-smokers, and the mean age of onset is 55 years old.

**Desquamative Interstitial Pneumonia**

The American Thoracic Society and the American College of Chest Physicians have requested a specific code for desquamative interstitial pneumonia. This condition involves accumulation of macrophages in alveoli. It is more common in men than in women. Insidious onset of dyspnea and dry cough over weeks or months is typical, and patients may progress to respiratory failure. The main feature that distinguishes desquamative interstitial pneumonia from respiratory bronchiolitis-associated interstitial lung disease is that it affects the lung in a uniform diffuse manner and lacks the bronchiolocentric distribution seen in respiratory bronchiolitis-associated interstitial lung disease. The prognosis for desquamative interstitial pneumonia is generally good, as most patients improve with smoking cessation and corticosteroids.

**ICD-9-CM Diagnosis Addenda**

Proposed ICD-9-CM diagnosis addenda changes were reviewed. Highlights of the proposed revisions include (these are only proposed at this time – they have not been finalized):

- Revision of title of chapter 5 to state “Mental and Behavioral Disorders;”
- Addition of Excludes note for “berry aneurysm, nonruptured (437.3)” under code 430, Subarachnoid hemorrhage, nonruptured;
- Revision of “code first” note under code 536.3, Gastroparesis, to indicate that the underlying disease should be coded first if applicable;
- Addition of Index entries for:
  - Disease, Fournier’s disease, female (616.89);
  - Disease, microvillus atrophy (751.5);
  - Encephalopathy, metabolic, drug induced (349.82);
  - Leukoencephalopathy, arteriosclerotic (437.0);
  - Lipodermatosclerosis (729.39);
  - Osteomalacia, oncogenic (275.8);
  - Pancytopenia, with myelodysplastic syndrome (see Syndrome, myelodysplastic);
  - Syndrome, superior, semi-circular canal dehiscence (386.8).
- Revision of Index entries for:
  - Disease, iron metabolism (275.09);
  - Disease, Sweeley-Klionsky (272.7);
  - Encephalopathy, due to drugs (349.82);
  - Hemochromatosis, diabetic (275.03);
  - Infiltrate, lung, and Infiltrate, pulmonary (793.1).
ICD-10-CM Proposals

Opioids Expansion
An extensive expansion of the poisoning, adverse effect, and underdosing codes has been proposed in order to specifically identify various opioid molecules. The creation of a 7th character for category F11, Opioid related disorders, to identify the type of opioid has also been proposed. Meeting attendees expressed concern about the level of detail in the proposal, including whether this level of detail is appropriate for a classification system and whether medical record documentation would support identification of specific opioid molecules.

Weeks of Gestation of Pregnancy
A new category, Z35, has been proposed to capture the exact number of weeks of gestation of pregnancy. While the trimester identified in the obstetric codes is still important, the specific gestational age is now considered a more precise indicator of risk. A commenter noted that there is a conflict between the definition of trimesters in chapter 15 and the placement of the code for week 28 in the proposal (i.e., week 28 is part of the 3rd trimester in chapter 15 and part of the 2nd trimester in the proposal). Also, a suggestion was made to provide beginning and ending date limits for each code to make it abundantly which gestational ages fall within each code. Another individual suggested placing all of the proposed codes into a single subcategory, instead of spreading them across a category, to conserve space for future coding needs.

Weeks of Gestation for Newborn
An expansion of the codes for prematurity of newborn has been requested in order to capture the specific week of gestational age. As was also suggested for the weeks of gestation of pregnancy proposal, a suggestion was made to provide beginning and ending date limits for each code.

Benign Neoplasm of Genitourinary Organs
The American Urological Association has requested the creation of unique codes for benign lipomatous neoplasm of kidney and other genitourinary organ. Angiomyolipomas are a common occurrence in male patients in their 50s. While benign, they can cause symptoms, including spontaneous bleeding.

Urethral False Passage
A unique code for urethral false passage has been proposed. A false urethral passage is a generally traumatic occurrence caused by instrumentation or catheterization and causes a bypass of the normal urethra. The intervention is involves either catheter drainage or allowing the false passage to heal on its own.

Nodular Prostate
New codes for nodular prostate with and without lower urinary tract symptoms have been proposed.


Inflammatory Disease of Prostate

Deletion of codes for acute and chronic prostatitis with and without hematuria has been proposed. Urologists do not look for hematuria with prostatitis.

Deletion of codes is allowed prior to, but not after, ICD-10 implementation.

Cyst of the Prostate

A unique code for cyst of prostate has been requested. Cysts of prostate are generally asymptomatic, but may lead to perineal discomfort.

Acquired and Congenital Torsion of Penis

Unique codes for acquired and congenital torsion of penis have been requested. Congenital torsion of the penis is a common occurrence in pediatric patients and occurs in the first trimester in utero. An acquired torsion of penis may occur as a result of the repair of a hypospadias condition.

Cyst of the Epididymis

A unique code for cyst of epididymis has been requested. This condition is a fluid-filled growth on the epididymis that may remain asymptomatic or may cause perineal pain.

Hidden Penis

A new code for hidden penis has been requested in chapter 17.

Personal History of Malignant Neoplasm of Ureter

Creation of a code for personal history of malignant neoplasm of ureter has been proposed. A patient with a personal history of ureteral cancer needs to be monitored closely, as there can be a recurrence on the contralateral ureter or a secondary cancer in the bladder may develop.

Visual Agnosia and Related Conditions

A unique code for visual agnosia has been proposed. Inclusion terms for prosopagnosia and simultanagnosia (asimultagnosia) would appear under the new code. These conditions are symbolic dysfunctions. It was suggested that an instructional note be added to indicate that any known underlying condition should be coded first.

Displacement/Dislocation of Internal Hip Prosthesis

The American Academy of Orthopedic Surgeons has requested that the titles of codes T84.022, Dislocation of internal right knee prosthesis, and T84.023, Dislocation of internal left knee prosthesis, be revised to state “Instability” instead of “Dislocation.” Complete dislocation of prosthetic knees is extremely rare, whereas instability of prosthetic knees is a common indication for revision surgery.
Gastroparesis
A unique code for gastroparesis has been requested. An instructional note under this code would indicate that the underlying disease should be coded first if known. Gastroparesis is a chronic disorder of the stomach characterized by abnormal motility and delayed gastric emptying. Symptoms of gastroparesis include early satiety, bloating, epigastric and upper abdominal pain, chronic nausea, and frequent vomiting.

The most common cause of gastroparesis is diabetes. Other causes include infections, endocrine disorders, connective tissue disorders such as scleroderma, autoimmune conditions, neuromuscular diseases, cancer, some forms of chemotherapy and radiation therapy, and surgery of the upper intestinal tract. Although gastroparesis is frequently due to diabetes or another known underlying condition, in about one-third to one-half of cases, no underlying diagnosis is identified.

Meeting attendees commented that the proposed Excludes2 note for diabetic gastroparesis is confusing, particularly in conjunction with the “code first” note under the proposed new code.

ICD-10-CM Diagnosis Addenda
Proposed ICD-10-CM diagnosis addenda changes were reviewed. These proposed changes include:

- Deletion of inclusion term for “circumscribed brain atrophy” under code G31.01, Pick’s disease (it was suggested that revision of the inclusion term to state “frontotemporal circumscribed brain atrophy” be considered as an alternative).

Procedures (ICD-9-CM)
Implantable Hemodynamic Monitoring System
New implantable hemodynamic monitoring systems have been developed that involve the implantation of a sensor and wireless communication with the sensor via an external monitor. A new code for insertion of implantable wireless pressure sensor for intracardiac or great vessel hemodynamic monitoring has been proposed.

Commenters noted that if a new code is created, the proposed code title and instructional notes need to more clearly distinguish it from the existing codes for implantation of an implantable hemodynamic monitoring device (codes 00.56-00.57).

Endovascular Embolization with Head or Neck Vessel Reconstruction
A new code has been proposed for endovascular embolization with head or neck vessel vascular remodeling support. Terms that might be seen in the medical record to describe these devices include Pipeline device, PED (Pipeline embolization device), PVR (parent vessel reconstruction) device, and embolization stent (even though the device is not technically a stent).

Endovascular embolization with coils and adjunctive use of stents has been an effective technique for occluding certain types of aneurysms. However, results over time have indicated that the occlusion achieved is often incomplete and may lack durability, with the possibility of
some degree of recanalization taking place. This puts patients at higher risk for continued disease progression and possible aneurysm rupture, and necessitates long-term periodic follow-up.

To address durability, the focus of treatment has begun to shift from basic occlusion of the aneurysm to techniques that actually restructure the diseased vessel. This can be done without the use of coils. A cylindrical mesh stent-like device is implanted within the artery across the aneurysm neck. The degree of metal surface area over the aneurysm neck directs the flow of blood away from the aneurysm. This reduces the stress on the aneurysm wall. The metal surface also serves as scaffolding, triggering growth of endothelial cells across the neck of the aneurysm. A thick membrane forms, completely remodeling and covering the aneurysm neck. This newly constructed vessel wall seals the aneurysm and excludes it from circulation. Depending on the length of the vessel, multiple devices may be deployed in an overlapping technique, one within the other. In time, the devices become incorporated into the vessel wall and are covered by an endothelial lining.

These aneurysm devices are sometimes referred to as stents or stent-like devices. However, they are technically different from conventional stents. Vessel reconstruction devices such as the Pipeline™ Embolization Device are stand-alone devices that treat the aneurysm by rebuilding the vessel from within. The devices are catheter deployed and angioplasty is not involved in the procedure.

Until a new code is created, code 39.72, Endovascular embolization or occlusion of head and neck vessels, should be used for procedures involving the Pipeline™ device.

**Fenestrated Endograft Repair of Abdominal Aortic Aneurysm**

Creation of a unique code for endovascular implantation of fenestrated graft(s) in abdominal aorta has been proposed. The title of existing code 39.71, Endovascular implantation of graft in abdominal aorta, would be revised to limit its use to non-fenestrated grafts.

Fenestrated endograft repair is a new technology for treating abdominal aortic aneurysms in a subset of patients who are not candidates for using standard endovascular devices. Fenestrated endografts can seal above the renal arteries while maintaining access and uninterrupted blood flow to branch vessels of the aorta. The fenestrated endograft is a modular or component system. The main body of the graft consists of two parts: the proximal tubular graft, with precisely located holes (fenestrations) and/or cutouts from the proximal margin (scallops) of the graft material along with a bare proximal stent with barbs to provide fixation, and a distal bifurcated graft body. An iliac leg component, which couples with the main bifurcated body, completes the basic fenestrated endograft.

In a standard endovascular repair of an abdominal aortic aneurysm, it is rarely necessary to stent a branch vessel. In a fenestrated endovascular procedure, most often both renal arteries require stenting and sometimes mesenteric vessels as well. The fenestrated portion of the endovascular graft is first partially deployed in the aorta, with the physician making sure to align the fenestrations of the stent graft body with the origins of the arterial ostia. Guidewires are then passed through the fenestrations in the stent graft body into the relevant arteries. This allows for
catheterization of the arteries and subsequent stenting to assure the stent graft fenestrations remain aligned with the visceral arteries over time.

Meeting attendees noted that a default needs to be designated for those instances when the graft isn’t specified as fenestrated or non-fenestrated.

**Removal of Contrast Dye**

A unique code for catheter-based removal of contrast media has been requested. CMS recommends not creating a new code and that no code should be assigned for the contrast material.

In patients with pre-existing moderate to severe chronic kidney disease, exposure to contrast media can result in a further reduction in kidney function, or contrast-induced nephropathy. Many patients at the highest risk for contrast-induced nephropathy may forego coronary diagnostic procedures because the risk of further kidney injury may outweigh the treatment benefits of a potential interventional procedure. The CINCOR™ Contrast Removal System is a new technology to assist the physician in the removal of contrast dye during a coronary angiogram or percutaneous intervention procedure. This system utilizes a catheter in the venous drainage area of the heart (coronary sinus) to remove contrast media before it is exposed to the kidneys.

Since the clinical trial for this technology is not expected to be completed until mid-2012, C&M meeting attendees commented that it may be too early to consider creating a new code.

**ICD-9-CM Procedure Addenda**

Proposed procedure addenda changes were reviewed. Highlights of the proposed revisions include (these are only proposed at this time – they have not been finalized):

- Addition of inclusion term for “mini-bronchoalveolar lavage [mini-BAL]” under code 33.24, Closed [bronchoscopic] biopsy of bronchus (and deletion of the Excludes note under code 33.24);
- Deletion of Excludes note for “bronchoalveolar lavage [BAL] (33.24)” under code 33.29, Other diagnostic procedures on lung and bronchus;
- Movement of Excludes note for “laparoscopic total abdominal hysterectomy (68.41)” from subcategory 68.4, Total abdominal hysterectomy, to code 68.49, Other and unspecified total abdominal hysterectomy;
- Revision and addition of Index entries for hysterectomy to clarify that:
  - Laparoscopic total abdominal hysterectomy is classified to code 68.41,
  - Laparoscopic radical abdominal hysterectomy is classified to code 68.61,
  - Laparoscopic supracervical hysterectomy is classified to code 68.31,
  - Laparoscopic vaginal assisted hysterectomy is classified to code 68.51,
  - Laparoscopic radical vaginal assisted hysterectomy is classified to code 68.71,
  - Hysterectomy, not otherwise specified, is classified to code 68.9.