~~N01 Rapidly progressive nephritic syndrome [See before N00 for subdivisions ]~~

~~***Includes:*** rapidly progressive:~~

~~· glomerular disease~~

~~· glomerulonephritis~~

~~· nephritis~~

~~***Excludes:*** nephritic syndrome NOS ( N05.- )~~

~~*Definition: worsening renal function in the setting of acute nephritic syndrome, often with macroscopic haematuria.*~~

N021 Nephrotic syndrome [See before N00 for subdivisions ]

*Includes:* congenital nephrotic syndrome

lipoid nephrosis

*Definition: albuminuria or proteinuria within nephrotic range, often with gross edema, hypoalbuminemia. If duration less than 90 days, then code acute kidney disease, if duration is greater than 90 days, then concurrently code CKD.*

~~N03 Unspecified nephritic syndrome [See before N00 for subdivisions ]~~

~~***Includes:*** glomerular disease~~
~~glomerulonephritis~~ ~~NOS~~
~~nephritis~~
~~nephropathy NOS and renal disease NOS with morphological lesion specified in .0-.8 before N00.-~~
~~***Excludes:*** nephropathy NOS with no stated morphological lesion ( N28.9 )~~
~~renal disease NOS with no stated morphological lesion ( N28.9 )~~
~~tubulo-interstitial nephritis NOS ( N12 )~~

N02 Recurrent and persistent haematuria [See before N00 for subdivisions ]



***Includes:*** haematuria:
· benign (familial)(of childhood)
· with morphological lesion specified in .0-.8 before N00.-

***Excludes:*** haematuria NOS ( R31 )

N063 Isolated proteinuria with specified morphological lesion [See before N00 for subdivisions ]

***Includes:*** proteinuria (isolated)(orthostatic)(persistent) with morphological lesion specified in .0-.8 before N00.-
***Excludes:*** proteinuria:
· NOS ( R80 )
· Bence Jones ( R80 )
· gestational ( O12.1 )
· isolated NOS ( R80 )
· orthostatic NOS ( N39.2 )
· persistent NOS ( N39.1 )

New code N05 Congenital or infantile nephrotic syndrome

New code N05.1 Congenital Nephrotic Syndrome

*proteinuria present before 3 months of age*

New code N05.2 Infantile Nephrotic Syndrome

*proteinuria develops between 3-12 months of age*

Revised N07 Hereditary nephropathy, not elsewhere classified

~~*Excludes:* Alport's syndrome ( Q87.8 )~~
~~hereditary amyloid nephropathy ( E85.0 )~~

~~nail patella syndrome ( Q87.2 )~~
~~non-neuropathic heredofamilial amyloidosis ( E85.0 )~~

• Alport syndrome (Q87.8)

• Fabry (-Anderson) disease (E75.2†)

• Thin basement membrane disease ()

• Hereditary amyloidosis E85.0

• Nail patella syndrome (Q87.2)

• Hereditary FSGS ()

Revised N08* Glomerular disorders in diseases classified elsewhere

*Includes:* nephropathy in diseases classified elsewhere
***Excludes:*** renal tubulo-interstitial disorders in diseases classified elsewhere ( N16.-* )

Revised N08.0* Glomerular disorders in infectious and parasitic diseases classified elsewhere

Glomerular disorders in:
· Plasmodium malariae malaria ( B52.0+ )
· mumps ( B26.8+ )
· schistosomiasis [bilharziasis] ( B65.-+ )
· septicaemia ( A40-A41+ )
· strongyloidiasis ( B78.-+ )
· syphilis ( A52.7+ )

• Post-infectious glomerulonephritis ()

• Hepatitis B virus ()

• Hepatitis C virus () ( cryoglobulinaemia ( D89.1+ ))

• Shunt nephropathy ()



• Hemorrhagic fever with renal syndrome (epidemic hemorrhagic fever, EHF)

• Human immunodeficiency virus ()

• Leptospirosis ()

• Leishimaniasis

• Trichinosis

• Endocarditis/SBE

• Parvovirus

• EBV

Revised N08.1* Glomerular disorders in neoplastic diseases:

• multiple myeloma /plasma cell neoplasia ( C90.0+ )
• lymphoplasmacytic lymphoma (Waldenström's macroglobulinaemia )( C88.0+ )

• cryoglobinemia related to lymphona

Revised N08.2* Glomerular disorders in blood diseases and disorders involving the blood system Glomerular disorders in:

· disseminated intravascular coagulation ( D65+ )
· haemolytic-uraemic syndrome ( D59.3+ )
· sickle-cell disorders ( D57.-+ )

· thrombotic thrombocytopenic purpura ( M31.1+· cryoglobulinaemia ( D89.1+ )

Revised N08.3* Glomerular disorders in diabetes mellitus ( E10-E14+ with common fourth character .2)

Revised N08.4* Glomerular disorders in other endocrine, nutritional and metabolic diseases

Glomerular disorders in:
· amyloidosis ( E85.-+ ) (other than related to plasma cell dyscrasias)
· Fabry(-Anderson) disease ( E75.2+ )
· lecithin cholesterol acyltransferase deficiency ( E78.6+ )

Revised N08.5* Glomerular disorders in systemic or auto-immune disorders

Glomerular disorders in:
• Goodpasture's disease ( M31.0+ ) (note to classifiers : Note M31.0 is not correct , which is hypersensitivy angitis, which is not goodpasture's disease)
• microscopic polyangiitis ( M31.7+ )
• Wegener's granulomatosis ( M31.3+ )

• Churg-Strauss syndrome ()

• systemic lupus erythematosus ( M32.1+ )
• Immunotactoid nephropathy ()

• Dense deposit disease

Revised N08.8* Glomerular disorders secondary to chemicals or drugs

includes drugs such as gold, penicillamine



**ICD-11 Acute Kidney Diseases and Disorders (N17)**

**Version date:** **October 25, 2010**

**Contact person:** **Lesley Stevens MD MS**

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The current ICD-9-CM codes for acute renal failure require a specific diagnosis to be made in order to code. This is contrast to the new Kidney Disease International Global Outcomes (KDIGO) Evidence Based Guidelines on acute kidney injury (AKI), as well as current clinical practice. Indeed, there has been much vocal concern by clinicians and the coding community about the confusing state of the codes given that clinicians first recognize an acute impairment in kidney function and then proceed to assign a specific diagnosis. Here, we propose revisions to the ICD-9-CM classification system that reflect the current understanding and definitions of acute kidney disease and AKI. In this document we first briefly describe the concepts of acute vs chronic kidney disease, the new KDIGO definitions and staging system for AKI and then propose modification to the current codes which we expect will substantially decrease the current confusion.

**Overview of Acute and Chronic Kidney Diseases and disorders**

Acute kidney diseases and disorders (AKD) is a global problem. It is common and occurs in the community, in the hospital where it is common on medical, surgical, pediatric, and oncology wards, and in ICUs. It imposes a heavy burden of illness, it is a predictor of immediate and long-term adverse outcomes, and has an associated high cost with its requirements for intensive evaluation and management. Individuals with chronic kidney disease (CKD) are especially susceptible to AKD which, in turn, may act as a promoter of progression of the underlying CKD. AKD is amenable to early detection and potential prevention, and therefore important to be recognized by clinicians and health care systems.

**Definition of AKI, AKD and CKD and Staging of AKI**

The definition of AKD has not been well standardized until recently. As shown in figure 1, AKI is a subset of AKD, and AKI and AKD can occur in patients with CKD.

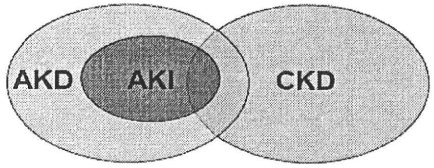


**Figure 1. Conceptual model for integration of AKI, CKD, and AKD.** Overlapping ovals show the relationships among AKI, AKD, and CKD. AKI is a subset of AKD. Both AKI and AKD without AKI can be superimposed upon CKD. Individuals without AKI, AKD, or CKD have no known kidney disease (NKD), not shown here. AKD, acute kidney diseases and disorders; AKI, acute kidney injury; CKD, chronic kidney disease.

AKI is defined as:

- Increase in SCr by >0.3 mg/dl within 48 hours; or
- Increase in SCr by >1.5-fold above baseline, which is known or presumed to have occurred within 7 days; or
- Urine volume <0.5 ml/kg/h for 6 hours or more.

AKD is defined as

- Reduction in kidney function for **less than 90 days** as evident by:
  - AKI; or
  - Reduction in GFR < 60 ml/min per 1.73 $m^2$; or
  - Decrease in GFR by ≥35%; or
  - Increase in SCr by >50%
- Kidney damage **for less than 90 days**

CKD is defined as

- Reduction in GFR < 60 that is present for **greater than** 90 days; or
- Rvidence of kidney damage that is present for **greater than** 90 days

Importantly, these definitions for AKI, AKD and CKD can be made irrespective of etiology. Similar to CKD, AKI and AKD encompasses various etiologies, including



acute tubular necrosis, acute interstitial nephritis, acute glomerular and vasculitic renal diseases, prerenal azotemia, and acute postrenal obstructive nephropathy. It is recognized that more than one of these conditions may coexist in the same patient; the manifestations and clinical consequences of AKI can be quite similar regardless of etiology; and that even mild, reversible, AKI has important clinical consequences, including increased risk of death [1,2].

AKI is classified based on both rate of change of kidney function (indicated by change in GFR or serum creatinine or by magnitude of urine output). Table 1 shows the stages of AKI. Each stage has different clinical action plan associated with it

**Table 1: Staging system for AKI**

| Stage | Scr | Urine output |
|---|---|---|
| 1 | ≥1.5-1.9 times baseline<br>OR<br>0.3 mg/dl increase | <0.5 ml/kg/h for 6-12 hours |
| 2 | ≥2.0-2.9 times baseline | <0.5 ml/kg/h for ≥12 hours |
| 3 | ≥3.0 times baseline<br>OR<br>increase in SCr to ≥4.0 mg/dl<br>OR<br>RRT<br>In patients <18 years, decrease in eGFR to <35 ml/min per 1.73 $m_2$ | <0.3 ml/kg/h for ≥24 hours<br>OR<br>Anuria for ≥12 hours |

**Proposal for modification of the Acute Renal Failure Codes (N10) AKI codes**

The current codes for acute renal failure (N17) are primarily based on an etiology and pathology. However, as described above, that is in contrast to clinical practice and recent Clinical Practice Guidelines, and is a source of confusion. We therefore

propose removal of specific etiologies and pathologies from N17 and revise them so that they reflect the definition and stages of AKI and AKD. Instructions will be added to this code to also code for etiology of the acute kidney disease. Since all specific etiologies and pathologies listed in N17 are of the tubular-interstitial component of the kidney, they wll be removed from N17 and specified with a third digit in N10 (acute tubulo-interstitual diseases). We therefore also propose modification to N10 to add this etiology.

Revise N17 Acute ~~renal failure~~ kidney disease and disorders

*Excludes:*

*following labor and delivery (669.3)*

*posttraumatic (958.5)*

*that complicating:*

*abortion (634-638 with .3, 639.3)*

*ectopic or molar pregnancy (639.3)*

Comment: Use additional code to identify: clinical diagnosis, dialysis status (V45.1), kidney transplant status (V42), hypertension (XXX), and diabetes status (XXX)

New code N17.1 Acute kidney injury, Stage 1 (mild)

New code N17.2 Acute kidney injury, Stage 2 (moderate)

New code N17.3 Acute kidney injury, Stage 3 (severe)

New code N17.4 Acute kidney disease, requiring dialysis

New code N17.5 Other acute kidney disease *Excludes AKI.* Use additional codes to specify causes of kidney disease (581, 582, 583)

584.51 Decreased GFR <90 days



584.52 Markers of kidney damage <90 days

Remove ~~N17.0 With lesion of tubular necrosis~~

~~Lower nephron nephrosis~~

~~Renal failure with (acute) tubular necrosis~~

~~Tubular necrosis:~~

~~NOS~~

~~acute~~

Remove ~~N17.1 With lesion of renal cortical necrosis~~

Remove ~~N17.2 With lesion of renal medullary [papillary] necrosis~~

~~Necrotizing renal papillitis~~

Revise N17.8 Other acute ~~renal failure~~ kidney injury

Revise N17.9 Acute ~~renal failure~~ kidney injury, unspecified

Revise N10 Acute tubulo-interstitial nephritis

Use additional code (B95-B97), if desired, to identify infectious agent.

New code N10.1 Acute tubular necrosis

New code N10.2 Acute intersitial nephritis

New code N10.3 Acute cortical necrosis

New code N10.4 Acute medullary necrosis

New code N10.5 Acute pyelonephritis

**ICD-11 – Revisions to Chronic Kidney Disease (N18)**

**Version date:** **October 25, 2010**



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**Rationale**

In some countries, 'Code 585 (ICD-9) or N18 (ICD-10), Chronic renal failure, has been re-titled and expanded to reflect the new clinical practice standard for treating what is now referred to as chronic kidney disease (CKD). We propose that these same changes be included in ICD-11.

The nonspecific term 'chronic renal failure' has been used to mean any degree of kidney failure including end stage renal disease (ESRD). It has also been used interchangeably with the nonspecific term 'chronic renal insufficiency'. Similar non-specific terms are used in other languages. Based on the clinical practice guidelines developed by the National Kidney Foundation and supported by other international organizations, including Kidney Disease International Global Outcomes (KDIGO), the non-specific terms have been replaced with the more specific 'Chronic kidney disease', with stages of GFR used to classify the different stages of CKD. This widely accepted staging system was incorporated into these revisions and is proposed to be incorporated into ICD-11.

Stage 2, code N18.2, equates to mild CKD, stage 3, code N18.3, equates to moderate CKD, and stage 4, code N18.4, equates to severe CKD. End stage renal disease (ESRD), code N118.6, is defined as CKD stage 5 with dialysis status. It is important to distinguish between the terms "End Stage Renal Disease" (ESRD) and CKD Stage 5 (GFR < 15). ESRD refers to patients treated with dialysis or transplantation. Patients who are on chronic dialysis therapy should be coded as N18.6. Patients who have GFR estimated at < 15 but are not being treated with chronic dialysis therapy, should be coded N18.5. The terms chronic renal failure, chronic uremia, and chronic renal insufficiency are indexed to code N18.9, Chronic kidney disease, unspecified. Providers should be educated in the use and value of these new codes to greatly improve patient data. They should be encouraged to document the stage of CKD instead of using nonspecific terms.

A kidney transplant does not fully restore kidney function, therefore, patients who have undergone a kidney transplant are defined as having CKD but this does not necessarily indicate failure or rejection. Code Z94.0, Kidney replaced by transplant, may be assigned with the appropriate CKD code, based on the patient's post-transplant stage. The use additional code note under category N18 provides this instruction.

Patients with CKD following a transplant should not be assumed to have transplant failure or rejection unless it is documented by the provider. If documentation supports the presence of failure or rejection, then it is appropriate to assign code T86.1X, complication of kidney transplant followed by the appropriate CKD code. The renal work group is suggesting revisions to the sections on complications of kidney transplant. For each of those complications, we would expect that the appropriate CKD code be assigned.

Patients with CKD may also suffer from other serious conditions, most commonly diabetes mellitus and hypertension. The sequencing of the CKD code in relationship to codes for other contributing conditions is based on the conventions in the tabular list.

**Revisions**

**N18 Chronic ~~renal failure~~ Kidney Disease**

~~Includes: chronic uraemia~~

~~- diffuse sclerosing glomerulonephritis~~

~~Excludes: chronic renal failure with hypertension ( I12.0 )~~

Comment: Use additional code to identify: clinical diagnosis, dialysis status (XX), kidney transplant status (XXX), hypertension (XXX), and diabetes status (XXX), as well as complications of CKD (eg anemia)



~~**N18.0 End-stage renal disease**~~

Revise N18.1 Chronic kidney disease, Stage 1 - Kidney damage with normal or increased GFR (90 ml/min per 1.73 $m^2$)

Revise N18.2 Chronic kidney disease, Stage 2 - Kidney damage with mild decreased GFR (60-89 ml/min per 1.73 $m^2$)

Revise N18.3 Chronic kidney disease, Stage 3 -Moderate: decreased GFR (30-59 ml/min per 1.73 $m^2$)

Revise N18.4 Chronic kidney disease, Stage 4 - Severe, decreased GFR (15-29 ml/min per 1.73 $m^2$)

Revise N18.5 Chronic kidney disease, Stage 5 – Kidney failure, GFR < 15 ml/min per 1.73 $m^2$, not on dialysis

Revise N18.6 ESRD

Synonyms: Chronic uraemia

Revise N18.7 Chronic kidney disease, not specified

Revise N18.8 Other chronic ~~**renal failure**~~ kidney disease

~~Uraemic:~~

~~- neuropathy+ ( G63.8* )~~
~~- pericarditis+ ( I32.0* )~~

Revise N18.9 Chronic ~~**renal failure**~~ unspecified

**ICD-11: Renal tubulo-interstitial diseases (N10-N16)**

**Version date:** **October 25, 2010**

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**Rationale**

The codes for chronic interstitial nephritis are comprehensive and consistent with current clinical practice. We are not suggesting any changes to them. We are suggesting changes to the acute tubulo-interstitial codes are to make them consistent with revisions to acute renal failure (N17).

**Revisions**

Revise N10 Acute tubulo-interstitial nephritis

Use additional code (B95-B97), if desired, to identify infectious agent.

New code N10.1 Acute Tubular Necrosis

New code N10.2 Acute Interstitial Nephritis

New code N10.3 Acute cortical necrosis

New code N10.4 Acute medullary necrosis

New code N10.5 Acute pyelonephritis

N11 Chronic tubulo-interstitial nephritis

*Includes:* chronic:
· infectious interstitial nephritis
· pyelitis
· pyelonephritis

Use additional code (B95-B97), if desired, to identify infectious agent.

N11.0 Nonobstructive reflux-associated chronic pyelonephritis

Pyelonephritis (chronic) associated with (vesicoureteral) reflux

*Excludes:* vesicoureteral reflux NOS ( N13.7 )



N11.1 Chronic obstructive pyelonephritis

Pyelonephritis (chronic) associated with:

· anomaly
· kinking
· obstruction of
· stricture

pelviureteric junction
pyeloureteric junction
ureter

*Excludes:* calculous pyelonephritis ( N20.9 )
obstructive uropathy ( N13.- )

N11.8 Other chronic tubulo-interstitial nephritis

Nonobstructive chronic pyelonephritis NOS

N11.9 Chronic tubulo-interstitial nephritis, unspecified

Chronic:
· interstitial nephritis NOS
· pyelitis NOS
· pyelonephritis NOS

N12 Tubulo-interstitial nephritis, not specified as acute or chronic

Interstitial nephritis NOS
Pyelitis NOS
Pyelonephritis NOS

*Excludes:* calculous pyelonephritis ( N20.9 )

N13 Obstructive and reflux uropathy

*Excludes:* calculus of kidney and ureter without hydronephrosis ( N20.- )
congenital obstructive defects of renal pelvis and ureter ( Q62.0-Q62.3 )
obstructive pyelonephritis ( N11.1 )

N13.0 Hydronephrosis with ureteropelvic junction obstruction

*Excludes:* with infection ( N13.6 )

N13.1 Hydronephrosis with ureteral stricture, not elsewhere classified

*Excludes:* with infection ( N13.6 )

N13.2 Hydronephrosis with renal and ureteral calculous obstruction

*Excludes:* with infection ( N13.6 )

N13.3 Other and unspecified hydronephrosis

*Excludes:* with infection ( N13.6 )

N13.4 Hydroureter

*Excludes:* with infection ( N13.6 )

N13.5 Kinking and stricture of ureter without hydronephrosis

*Excludes:* with infection ( N13.6 )

N13.6 Pyonephrosis

Conditions in N13.0-N13.5 with infection
Obstructive uropathy with infection

Use additional code (B95-B97), if desired, to identify infectious agent.

N13.7 Vesicoureteral-reflux-associated uropathy

Vesicoureteral reflux:
· NOS
· with scarring

*Excludes:* reflux-associated pyelonephritis ( N11.0 )

N13.8 Other obstructive and reflux uropathy



N13.9 Obstructive and reflux uropathy, unspecified

Urinary tract obstruction NOS

N14 Drug- and heavy-metal-induced tubulo-interstitial and tubular conditions

*Use additional external cause code (Chapter XX), if desired, to identify toxic agent.*

N14.0 Analgesic nephropathy

N14.1 Nephropathy induced by other drugs, medicaments and biological substances

N14.2 Nephropathy induced by unspecified drug, medicament or biological substance

N14.3 Nephropathy induced by heavy metals

N14.4 Toxic nephropathy, not elsewhere classified

N15 Other renal tubulo-interstitial diseases

N15.0 Balkan nephropathy

Balkan endemic nephropathy

N15.1 Renal and perinephric abscess

N15.8 Other specified renal tubulo-interstitial diseases

N15.9 Renal tubulo-interstitial disease, unspecified

Infection of kidney NOS

*Excludes:* urinary tract infection NOS ( N39.0 )

N16* Renal tubulo-interstitial disorders in diseases classified elsewhere

N16.0* Renal tubulo-interstitial disorders in infectious and parasitic diseases classified elsewhere

Renal tubulo-interstitial disorders (due to)(in):
· brucellosis ( A23.-+ )
· diphtheria ( A36.8+ )
· salmonella infection ( A02.2+ )
· septicaemia ( A40-A41+ )
· toxoplasmosis ( B58.8+ )

N16.1* Renal tubulo-interstitial disorders in neoplastic diseases

Renal tubulo-interstitial disorders in:
· leukaemia ( C91-C95+ )
· lymphoma ( C81-C85+ , C96.-+ )
· multiple myeloma ( C90.0+ )

N16.2* Renal tubulo-interstitial disorders in blood diseases and disorders involving the immune mechanism

Renal tubulo-interstitial disorders in:
· mixed cryoglobulinaemia ( D89.1+ )
· sarcoidosis ( D86.-+ )

N16.3* Renal tubulo-interstitial disorders in metabolic diseases

Renal tubulo-interstitial disorders in:
· cystinosis ( E72.0+ )
· glycogen storage disease ( E74.0+ )
· Wilson's disease ( E83.0+ )

N16.4* Renal tubulo-interstitial disorders in systemic connective tissue disorders

Renal tubulo-interstitial disorders in:
· sicca syndrome [Sjögren] ( M35.0+ )
· systemic lupus erythematosus ( M32.1+ )

N16.5* Renal tubulo-interstitial disorders in transplant rejection ( T86.-+ )

N16.8* Renal tubulo-interstitial disorders in other diseases classified elsewhere

**ICD-11 Revisions to Renal Osteodystrophy**



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**Rationale**

Patients with Chronic kidney disease have several complications of decreased kidney function. One of them is mineral and bone disease (CKD-MBD), which was previously referred to as renal osteodystrophy. In the past, the focus of diagnosis and management of this syndrome was only on bone. With increased understanding of the pathophysiology, it is now clear that this is a systemic disorder, with consequences outside of bone. Thus, the term renal osteodystrophy (which means bone abnormalities of renal disease) is inadequate to define the full spectrum of abnormalities that occur in CKD-MBD. Below is a brief description of the definition of CKD-MBD, followed by suggested revisions to the classification system

As kidney function declines, there is a progressive deterioration in mineral homeostasis, leading to abnormalities in several hormones and minerals, including parathyroid hormone, vitamin D, fibroblast growth factor 23 (FGF-23), phosphate, serum calcium, and others. These hormones and minerals are vital to the regulation of both bone formation during growth as well as bone structure and function during adulthood. As a result, bone abnormalities are found in virtually all patients with kidney failure and in majority of patients with earlier stages of kidney disease. There is also extra-skeletal calcification that occurs and which leads to other morbidity and mortality of patients with CKD. Kidney Disease International Global Outcomes (KDIGO), an international organization, formulated a Work Group to develop a uniform definition, diagnostic criteria for definition, evaluation and management plans (Kidney International 2009. Vol 76 Suppl 113). The sum of disorders related to these impairments are referred CKD-MBD. The definition is listed in Table 1.

Table 1 | KDIGO classification of CKD-MBD and renal osteodystrophy

Definition of CKD-MBD

A systemic disorder of mineral and bone metabolism due to CKD manifested by either one or a combination of the following:

- Abnormalities of calcium, phosphorus, PTH, or vitamin D metabolism.
- Abnormalities in bone turnover, mineralization, volume, linear growth, or strength.
- Vascular or other soft-tissue calcification.

Definition of renal osteodystrophy

- Renal osteodystrophy is an alteration of bone morphology in patients with CKD.
- It is one measure of the skeletal component of the systemic disorder of CKD-MBD that is quantifiable by histomorphometry of bone biopsy.

CKD, chronic kidney disease; CKD-MBD, chronic kidney disease-mineral and bone disorder; KDIGO, Kidney Disease: Improving Global Outcomes; PTH, parathyroid hormone. Adapted with permission from Moe et al.[2]

To account for the complete definition and pathology of CKD-MBD, we have expanded the codes under renal osteodystrophy. In ICD-10, this is listed as part of disorders of impaired renal tubular function. We suggest separating CKD-MBD from disorders of impaired tubular function. See suggestions below. Note that the biochemical parameters only include those measured in clinical practice right now, but we expect that other markers (eg FGF-23) may be available in the future, and these should be incorporated into this classification system.

**Revisions**

| | | |
|---|---|---|
| Revise | N25 | ~~Disorders resulting from impaired renal tubular function~~ Chronic kidney disease- mineral and bone disease |
| Revise | N25.0 | Abnormal bone (synonyms Renal osteodystrophy~~)~~ |
| New code | | N25.01 Abnormal mineralization |
| New code | | N25.02 Abnormal turnover |
| New code | | N25.03 Abnormal volume |
| New code | | N25.04 Suspected renal osteodystrophy |
| | | *Biopsy not performed* |
| New code | | N25.08 Renal osteodystrophy Other |
| New code | | N25.09 Renal osteodystrophy unknown |



| | | |
|---|---|---|
| Revise | N25.1 | Biochemical abnormalities ~~Nephrogenic diabetes insipidus~~ |
| New code | | N25.11 Hypercalcemia |
| New code | | N25.12 Hypocalcemia |
| New code | | N25.13 Hyperphosphatemia |
| New code | | N25.14 Hypophosphatemia |
| New code | | N25.15 High iPTH |
| New code | | N25.16 Low iPTH |
| New code | | N25.17 Low vitamin D |
| New code | | N25.18 High alkaline phosphatase |
| New code | | N25.19 Low alkaline phosphatase |
| New code | N25.3 | Calcification abnormalities |
| New code | | N25.31 Calcification of the vasculature |
| | | N25.32 Calcification of the valves |
| New code | | N25.33 Calcifications of the soft tissue |
| New code | | N25.24 Calciphylaxis |
| New code | | N25.28 Other calcification |
| New code | | N25.29 Calcification, unknown |

| | |
|---|---|
| N26 | Phosphate-losing tubular disorders |
| | Renal: |
| | • rickets |
| | • short stature |
| N27 | Nephrogenic diabetes inspidus |

| | | |
|---|---|---|
| | N28 | Renal tubular acidosis |
| New code | | N27.1 proximal |
| New code | | N27.2 distal type I |
| New code | | N27.3 distal type IV |
| New code | | N27.8 Other |
| New code | | N27.9 Unknown |



**ICD-11: Cystic Disease (Q61)**

**Version date:** **October 25, 2010**

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**Rationale**

The ICD-10 code for Cystic Kidney Disease (Q61) is limited for three reasons. First, it does not describe the breadth and understanding of current genetics as pertains to cystic kidney disease. Second, there is no organization of the cystatic conditions. Third, it excludes specific conditions

In the revised codes, we link genetics to the underlying conditions where the genetics are known. The codes are categorized according to structural components of the kidney – namely glomerular, tubular or non-tubular structure.

We have included cysts related to cancer here . We recognize that this may be primarily placed within the oncology section, with linkage to the Renal section.

**Revisions**

Q61 Cystic kidney disease

~~*Excludes:* acquired cyst of kidney ( N28.1 )~~
~~Potter's syndrome ( Q60.6 )~~

Remove ~~Q61.0 Congenital single renal cyst~~

~~Cyst of kidney (congenital)(single)~~

Remove ~~Q61.1 Polycystic kidney, autosomal recessive~~

~~Polycystic kidney, infantile type~~

Remove ~~Q61.2 Polycystic kidney, autosomal dominant~~

~~Polycystic kidney, adult type~~

Remove ~~Q61.3 Polycystic kidney, unspecified~~

Remove ~~Q61.4 Renal dysplasia~~

Remove ~~Q61.5 Medullary cystic kidney~~

~~Sponge kidney NOS~~

Remove ~~Q61.8 Other cystic kidney diseases~~

~~Fibrocystic:~~
~~- kidney~~
~~- renal degeneration or disease~~

Remove ~~Q61.9 Cystic kidney disease, unspecified Meckel-Gruber syndrome~~

New code Q61.0 Cystic Kidney Disease, NOS

New code Q61.1 Hereditary Cystic Kidney Disease, Dominant Inheritance, NOS

New code Q61.11 Autosomal Dominant Polycystic Kidney Disease

New code Q61.12 Orofacial Digit Syndrome Type 1

New code Q61.13 Tuberous Sclerosis Complex

New code Q61.14 Von Hippel-Lindau

New code Q61.15 Glomerulocystic Disease



New code Q61.16 Familial Renal Hamartoma associated with Hyperparathyroidism – Jaw Tumor Syndrome

New code Q61.17 Cystic Kidney Disease, Dominant Inheritance associated with a syndrome

*Including: Alagille*

*Beckwith-Wiedemann*

*Brancio-oto-renal*

*Noonan*

*Pallister-Hall*

*Townes-Brock*

New code Q61.2 Hereditary Cystic Kidney Disease, Recessive inheritance, NOS

New code Q61.20 Autosomal Recessive Polycystic Kidney Disease

New code Q61.21 Hereditary Cystic Kidney Disease, Recessive Inheritance associated with a syndrome

*Including: Meckel-Gruber*

*Zellweger*

*Ellis van Crevald*

*Ivemark*

*Smith-Lemli-Optiz*

*Pierson*

*Elejalde*

New code Q61.3 Hereditary Cystic Kidney Disease with Interstitial Nephritis,

Dominant Inheritance, NOS

New code Q61.30 Medullary Cystic Kidney Disease Type 1

New code Q61.31 Medullary Cystic Kidney Disease Type 2

*Include;' Uromodulin ( Familial Hyperuricemic Nephropathy)*

New code Q61.4 Hereditary Cystic Kidney Disease with Interstitial Nephritis,

Recessive Inheritance, NOS

New code Q61.40 Nephronophthisis

New code Q61.41 Joubert Syndrome

New code Q61.42 Bardet-Biedl Syndrome

New code Q61.43 Jeune's Syndrome

New code Q61.44 Alstrom Syndrome

New code Q61.5 Hereditary Cystic Kidney Disease, Chromosomal Abnormalities, NOS

New code Q61.50 Hereditary Cystic Kidney Disease, Known Chromosomal Abnormality

*Include*

*Trisomy 9, 13 [Pateu],*

*Trisomy 18 [Edwards]*

*Trisomy 21 [Down]*

*22q11 deletion [Velocardiofacial]*

*Trisomy 45 X0 [Turner*



*Trisomy Triplodies*

New code Q61.6 Non-Hereditary Cystic Kidney Disease, NOS

New code Q61.60 Simple Cyst

New code Q61.61 Medullary Sponge Kidney

New code Q61.62 Cysts associated with chronic kidney disease

New code Q61.63 Hypokalemia

New code Q61.64 Trauma

New code Q61.65 Localized or Unilateral Cystic Disease

*Comment: Hard to be certain re making a diagnosis of this; but this coding would preclude a known Ash-Upmark or Unilateral AD or ARPKD obviously*

Revise Q61.7 Renal Cystic Dysplasia *(RDP comment: Of tubular origin? always known)*

New code Q61.70 Multicystic Dysplastic Kidney

New code Q61.71 Hepatocyte Nuclear Factor 1-Beta Mutation

New code Q61.72 Renal Cystic Dysplasia due to renal tract obstruction

*Code for obstructive uropathy*

New code Q61.73 Renal Cystic Dysplasia due to vesicoureteral reflux

*Code for vescucouretal reflux*

New code Q61.8 Renal Cystic Disease, Non-Tubular Origin, NOS

New code Q61.80 Cystic Disease of Renal Sinus

New code Q61.81 Perirenal Lymphangioma

New code Q61.82 Subscapular and Perirenal Urinoma

New code Q61.83 Pyelocalyceal Cyst

New code Q61.9 Renal Cystic Neoplasms, NOS

New code Q61.90 Cystic Renal Cell Carcinoma

New code Q61.91 Multilocular Cystic Nephroma

New code Q61.92 Cystic Partially Differentiated Nephroblastoma

New code Q61.93 Mixed Epithelial and Stromal Tumor



**ICD-11: Revisions to Urine Laboratory Findings**

**Version date:** **October 25, 2010**

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**Rationale**

We suggest that the classification be revised such that urine findings be classified together. Previously the classification included a category called isolated proteinuria. These were not consistent with practice and disease. For example, it is not possible to have Benes Jones proteinuria that is isolated. However, we do feel that it is important to be code for the presence of Benes Jones proteinuria. We therefore suggest revising the codes that such that the findings in the urine can be documented separately from the etiology. Changes to hematuria have been made so that they are comprehensive and can describe all observed forms of hematuria, along with its origin, timing and miscropic nature.

**Title proteinuria (INCLUDES ALBUMINURIA). (incorporates the original N06, B39.1, N39.2 R80)**

Revise X01~~R80 Isolated~~ proteinuria

~~Albuminuria NOS~~

~~Bence Jones proteinuria~~

~~Proteinuria NOS~~

~~Excludes:~~

- ~~gestational~~
- ~~isolated, with specified morphological lesion (Q12.1)~~
- ~~orthostatic (N06.)~~
- ~~persistent (N39.1)~~

1 Glomerular origin (albuminuuria if pathology known, the code along with glomerular diseases in N00)

2 Benes Jones protein
3 Gestational
4 Orthostatic
5 Isolated NOS

INSTRUCTIONS *assign*

*2cd decimal 1=intermittent;*

*2=persistent*

*3=unlknown;*

Revise

~~N02 Recurrent and persistent~~ haematuria ~~(See before N00 for subdivisions)~~ (Note incorporates original R31 and N02)

~~Includes: haematuria~~

- ~~benign (familial) (of childhood)~~
- ~~with morphological lesion specified in .0-.8 before N00.-~~

~~Excludes: haematuria NOS (R31)~~

~~R31 Unspecified haematuria~~

~~Excludes: recurrent or persistent haematuria (N02.-)~~

INSTRUCTIONS *assign*

1st decimal place: Known or presumed origin of the hematuria

1 Glomerular in origin (if pathology known, then code along with glomerular diseases in N00)
2 Tubular in origin
3 Urinary tract in origin
4 Urinary tract infection
5 Not specified

2cd decimal : Timing and persistence



1=intermittent

2=persistent

3=unlknown

3RD decimal: Microscopy

1=microscopic

2=macroscopic

No change

X03 ~~R81~~ Glycosuria

Excludes: renal glycosuria (E74.8)

Remove

~~R82 Other abnormal findings in urine~~

~~Excludes: haematuria (R31)~~

~~0 Chyluria~~

- ~~Excludes: filarial chyluria (B74.1)~~

~~1 Myoglobunuria~~

~~2 Biliuria~~

~~3 Haemoglobinuria~~

~~Excludes: haemoglobinuria~~

- ~~due to haemolysis from external causes NEC (D59.6)~~
- ~~paroxysmal nocturnal [Machiafava-Micheli] (D59.5)~~

New code

X04 Chyluria

Excludes:

- filarial chyluria (original B74.-)

New code

X05 Myoglobinuria

New code

X06 Biliuria

New code X07 Haemoglobinuria

No change X08 ~~R82.4~~ Acetonuria

- Ketonuria

Revise X09 ~~R82.5~~ Elevated urine levels of drugs, medicaments and biological substances

Elevated urine levels of:

- catecholamines
- ~~indoleacetic acid~~
- ~~17-ketosteroids~~
- ~~steroids~~

No change X10 ~~R82.6~~ Abnormal urine levels of substances chiefly nonmedical as to source

- Abnormal urine level of heavy metals

Remove ~~R82.7 Abnormal findings on microbiological examination of urine~~

- ~~Positive culture findings~~

Comment: to be removed and added to infection

No change X11 ~~R82.8~~ Abnormal findings on cytological and histological examination of urine

No change X12 ~~R82.9~~ Other and unspecified abnormal findings in urine

- ~~Cells and casts in urine~~
- ~~Crystalluria~~
- ~~Melanuria~~



## *T86.1 Kidney transplant failure and rejection*

***Kidney transplant revisions***

**T86 Failure and rejection of transplanted organs and tissues**

~~**T86.1 Kidney transplant failure and rejection**~~

**T86.1 Complications of kidney transplant**

T86.11 Primary graft nonfunction

T86.12 Graft rejection

*use additional code to code acute or chronic kidney disease*

T86.121 Acute cellular

T86.122 Acute antibody mediated

T86.123 Chronic (transplant glomerulopathy)

T86.13 Non rejection causes of graft dysfunction

*use additional code to code acute or chronic kidney disease*

T86.131 BK nephropathy

T86.132 Interstitial fibrosis and tubular atrophy

T86.133 Recurrent disease in allograft

T86.134 De novo disease in allograft

T86.138 Other specified abnormality of graft function

T86.139 Abnormality of graft function, unspecified.

T86.14 Post transplant infections

*Use additional code (B95–B97), to identify infectious agent.*

Opportunistic infection

T86.15 Other specified complications

T86.150 Obstruction

T86.151 Post transplant renal artery stenosis

T86.152 New onset diabetes after transplantation

T86.153 Post transplant malignancy

*Use additional code (C00-C97), to identify type of neoplasm*

T86.154 Post transplant lymphoproliferative disorder

Polyclonal hyperplasia

Monoclonal proliferation (lymphoma)

## Z52 Donors of organs and tissues

***Excludes:*** examination of potential donor (Z00.5)

Z52.4 Kidney donor

## Z94 Transplanted organ and tissue status

***Includes:*** organ or tissue replaced by heterogenous or homogenous transplant

***Excludes:*** complications of transplanted organ or tissue - see Alphabetical Index

presence of:

- vascular graft (Z95.-)
- xenogenic heart valve (Z95.3)

Z94.0 Kidney transplant status

Z94.00 Kidney transplant status, graft from living donor

Z94.01 Kidney transplant status, graft from deceased donor

Z94.02 Kidney transplant recipient with a failed graft (back on dialysis)

V94.09 Kidney transplant status, graft status unspecified

## Y83 Surgical operation and other surgical procedures as the cause of abnormal reaction of the patient, or of later complication, without mention of misadventure at the time of the procedure



Y83.0 Surgical operation with transplant of whole organ

Y83.00 Kidney

Y83.01 Pancreas

Y83.1 Surgical operation with implant of artificial internal device

Y83.2 Surgical operation with anastomosis, bypass or graft

Y83.3 Surgical operation with formation of external stoma

Y83.4 Other reconstructive surgery

Y83.5 Amputation of limb(s)

Y83.6 Removal of other organ (partial) (total)

Y83.8 Other surgical procedures

Y83.9 Surgical procedure, unspecified

**Z99.2 Renal dialysis modality (status)**

**Rationale**

Although changes to Chapter XXI, *Factors influencing health status and contact with health services* (Z00–Z99), are not considered a priority for the alpha draft of ICD-11, the Nephrology working group strongly recommend the following changes to the renal dialysis status codes for ICD-11.

There is a large and growing population of dialysis dependent patients worldwide, with 6-8% annual cumulative growth rates in developed countries and much higher growth rates in the developing world with increasing access to renal dialysis therapies and the global increase in chronic diseases such as diabetes and hypertension. These patients have a very high morbidity, resulting in many heath care encounters, and a high mortality both of which are, to varying degrees, influenced by the modality (type) of dialysis and the access used for dialysis. A separate code for these patients, allowing sub-coding for modality and access, could be used for comparisons of morbidity, mortality, cost analysis, and health care planning. As such dialysis status is an important denominator, which could be used in determining complication rates for the various modalities, especially in the morbidity, mortality, primary care and patient safety use cases for ICD-11. For other use cases, where less granularity is required, or where insufficient documentation is provided, the proposed codes are able to be aggregated to a higher level with less detail.

**Revisions**

Add The following sixth-character subdivisions are for use with categories Z99.2., to identify the access device

.1 arteriovenous fistula

.2 arteriovenous graft synthetic

.3 ateriovenous graft biologic (porcine, cadaveric)

.4 cuffed hemodialysis catheter (tunnelled)



.5 uncuffed hemodialysis catheter (untunnelled)

.6 peritoneal dialysis catheter

.7 other specified access device

.8 unspecified access device

***Note:*** If chronic haemodialysis or peritoneal dialysis, assign a code based on patient's overall status. That is, if generally nocturnal, use Z95.01, even if the patient is having an intermittent session in centre on a particular day. For acute, status can vary with each treatment

New code **Z95.0 Chronic haemodialysis**

New code Z95.00 Standard chronic haemodialysis

New code Z95.01 Frequent chronic haemodialysis (includes daily and nocturnal)

New code Z95.08 Other chronic haemodialysis

New code Z95.09 Unspecified chronic haemodialysis

New code **Z95.1 Acute haemodialysis**

New code Z95.10 Acute intermittent haemodialysis

New code Z95.11 Continuous Haemofiltration

New code Z95.12 Continuous Haemodiafiltration

New code Z95.13 Slow long efficiency dialysis (SLED)

New code Z95.18 Other acute haemodialysis

New code Z95.90 Unspecified acute haemodialysis

New code **Z95.2 Peritoneal dialysis**

New code Z95.20 Continuous ambulatory peritoneal dialysis

| | |
|---|---|
| New code | Z95.21 Intermittent peritoneal dialysis |
| New code | Z95.22 Automated peritoneal dialysis |
| New code | Z95.28 Other peritoneal dialysis |
| New code | Z95.29 Unspecified peritoneal dialysis |

## B Complications of dialysis - Injury and External Cause TAG

**Rationale**

A sub-group of the Nephrology working group of the Internal Medicine TAG has considered changes for the area of dialysis (status codes) and complications of dialysis.

The latter overlaps with the responsibility of the Injury and External causes TAG, therefore we wish to outline the revisions requested by this group.

### Diagnosis codes

The main types of complications arising from dialysis and related procedures, such as thrombosis, haemorrhage, infection, embolism etc already exist in ICD-10, therefore no major changes are requested. From a classification perspective, we are interested to know if the diagnosis complication codes are still going to be split, ie end of chapter codes and injury chapter codes, or if this structure will change for ICD-11.

***Comment from expert reviewer: Should include dialysis specific access complications, and specify for eg inflow, outflow central stenosis***

### External cause codes

The concepts that are important to capture for external causes in relation to dialysis are listed below. We have not attempted to add these concepts to the current structure of ICD-10 as track changes, due to the current overlap of diagnosis/external cause concepts currently in this area, which require a major overhaul.



**Revisions**

- insertion/presence/removal of vascular access device, with the following specificity:

  Untunnelled uncuffed internal jugular vein hemodialysis catheter

  Untunnelled uncuffed femoral vein hemodialysis catheter

  Tunnelled cuffed hemodialysis catheter

  Arteriovenous fistula

  Arteriovenous conduit/graft

  Other hemodialysis vascular access device

  Unspecified hemodialysis vascular access device

- insertion/presence/removal of peritoneal dialysis catheter

  Cuffed flexible catheter for peritoneal dialysis

  Uncuffed rigid catheter for peritoneal dialysis ***Comment from expert reviewer: is this used anymore? Should it be deleted?***

- the use of dialysate (substance/drug related) – this may need to be covered by the Table of drugs and chemicals, and split on whether on correct or incorrect usage.
  **I am not sure what this means**

  the overall procedure of dialysis, ie an event occurring during or after dialysis and regarded as due to dialysis, with the following specificity in relation to the type of dialysis: **I am not sure what this will be used**

  Haemodialysis

  Haemofiltration

  Haemodiafiltration

  Slow long efficiency dialysis (SLED)