- **Glomerulopathies (93548.0)**
  - Nephrotic syndrome without extrarenal manifestations (567564.0)
    - Nephrotic syndrome, idiopathic (357502.0)
      - Idiopathic steroid-sensitive nephrotic syndrome (69061.0)
      - Idiopathic steroid-sensitive nephrotic syndrome with secondary steroid resistance (567546.0)
      - Idiopathic steroid-resistant nephrotic syndrome (567548.0)
        - Idiopathic multidrug-resistant nephrotic syndrome (567550.0)
        - Idiopathic steroid-resistant nephrotic syndrome with sensitivity to second-line immunouppressive therapy (567552.0)
    - Nephrotic syndrome, genetic (564127.0)
      - Genetic steroid-resistant nephrotic syndrome (SRNS) (656.0)
      - Congenital nephrotic syndrome, Finnish type (839.0)
      - Congenital nephrotic syndrome, no genetic cause specified (97556.0)
  - Glomerulopathy as part of a syndromic disorder (567562.0)
    - Denys-Drash syndrome (220.0)
    - Frasier syndrome (347.0)
    - Galloway-Mowat syndrome (2065.0)
    - Schimke immuno-osseous dysplasia (1830.0)
    - Pierson syndrome (2670.0)
    - Nail-patella syndrome (2614.0)
    - Leigh syndrome with nephrotic syndrome (255249.0)
    - Nephrotic syndrome, familial, steroid-resistant (SRNS), with sensorineural deafness (280406.0)
    - MYH9-related disease (182050.0)
    - Familial steroid-resistant nephrotic syndrome with adrenal insufficiency (506334.0)
    - Nephrotic syndrome-deafness-pretibial epidermolysis bullosa syndrome (300333.0)
    - Congenital nephrotic syndrome-interstitial lung disease-epidermolysis bullosa syndrome (306504.0)
    - Hypotrichosis-lymphedema-telangiectasia-renal defect syndrome (69735.0)
    - Autosomal dominant intermediate Charcot-Marie-Tooth disease type E (93114.0)
    - Severe oculo-renal-cerebellar syndrome (2715.0)
    - Action myoclonus-renal failure syndrome (163696.0)
    - Fibronectin glomerulopathy (84090.0)
    - Lipoprotein glomerulopathy (329481.0)
    - Collagen type III glomerulopathy (84087.0)
    - Congenital membranous nephropathy due to maternal anti-neutral endopeptidase alloimmunization (69063.0)
    - Nephrotic syndrome, syndromic, not otherwise specified (9999972.0)
  - IgA nephropathy (34145.0)
  - Membranous glomerulonephritis (97560.0)
  - Collagenopathies (inc. Alport syndrome) (544590.0)
    - Alport syndrome (63.0)
      - Alport syndrome, X-linked (88917.0)
      - Alport syndrome, X-linked with diffuse leiomyomatosis (1018.0)
      - Alport syndrome, autosomal dominant (88918.0)
      - Alport syndrome, autosomal recessive (88919.0)
      - Alport syndrome, digenic (9999965.0)
      - Microscopic (including familial) hematuria (9999976.0)
      - Collagenopathy, not further specified (9999977.0)
    - HANAC syndrome (73229.0)
  - Primary membranoproliferative glomerulonephritis (MPGN) (54370.0)
    - Immunoglobulin-mediated membranoproliferative glomerulonephritis (MPGN) (329903.0)
    - C3 glomerulopathy (329918.0)
      - Dense deposit disease (93571.0)
      - C3 glomerulonephritis (329931.0)
  - Glomerulopathy as part of a systemic disorder (567554.0)
    - Glomerulopathy as part of a genetic systemic disease (567556.0)

- Familial Mediterranean fever (342.0)
- Muckle-Wells syndrome (575.0)
- Hypocomplementemic urticarial vasculitis (36412.0)
- Hereditary amyloidosis with primary renal involement (85450.0)
  - AApoAI amyloidosis (93560.0)
  - ALys amyloidosis (93561.0)
  - AFib amyloidosis (93562.0)
  - AApoAII amyloidosis (238269.0)
- Autoimmune interstitial lung disease-arthritis syndrome (444092.0)

Glomerulopathy as part of a non-genetic systemic disease (567558.0)

- Systemic vasculitis associated with glomerulopathy (567560.0)
  - Systemic Lupus Erythematosus (SLE) (536.0)
  - Childhood-onset systemic lupus erythematosus (SLE) (93552.0)
  - Granulomatosis with polyangiitis (Wegener) (900.0)
  - IgA vasculitis (Henoch Schonlein nephritis, PSH) (761.0)
  - Pauci-immune glomerulonephritis (93126.0)
    - Pauci-immune glomerulonephritis with ANCA (97563.0)
    - Pauci-immune glomerulonephritis without ANCA (97564.0)
  - Microscopic polyangiitis (727.0)
  - Eosinophilic granulomatosis with polyangiitis (183.0)
  - Polyarteritis nodosa (767.0)
  - Takayasu arteritis (3287.0)
  - Giant cell arteritis (397.0)
  - Relapsing polychondritis (728.0)
  - Buerger disease (36258.0)
  - Cryoglobulinemic vasculitis (91138.0)
  - Anti-glomerular basement membrane disease (375.0)
- Mixed connective tissue disease (809.0)
- Monoclonal gammopathy of renal significance (MGRS) (9999974.0)
  - C3 glomerulopathy secondary to MGRS (9999970.0)
  - Proliferative glomerulonephritis with monoclonal immune deposits (PGNMID) (9999971.0)
- AA amyloidosis (85445.0)
- AL amyloidosis (85443.0)
  - Primary systemic amyloidosis (314701.0)
  - Primary localized amyloidosis (314709.0)
- AH amyloidosis (442582.0)
- AApoAIV amyloidosis (439232.0)
- Non-amyloid monoclonal immunoglobulin deposition disease (86861.0)
  - Heavy chain deposition disease (93556.0)
  - Light and heavy chain deposition disease (93557.0)
  - Light chain deposition disease (93558.0)
- IgG4-related kidney disease (449395.0)
- Polymyositis (732.0)
- Juvenile polymyositis (93568.0)
- Dermatomyositis (221.0)
- Juvenile dermatomyositis (93672.0)
- Beh (117.0)
- Sarcoidosis (797.0)
- Adult-onset Still disease (829.0)
- Reynolds syndrome (779.0)
- Systemic sclerosis (90291.0)
  - CREST syndrome (90290.0)
  - Diffuse cutaneous systemic sclerosis (220393.0)
  - Limited cutaneous systemic sclerosis (220402.0)
  - Limited systemic sclerosis (220407.0)
- Immunotactoid or fibrillary glomerulopathy (91137.0)
  - Non-amyloid fibrillary glomerulopathy (97566.0)
  - Immunotactoid glomerulopathy (97567.0)

Idiopathic non-lupus full-house nephropathy (567544.0)

- **Tubulopathies (93603.0)**
  - Primary renal tubular acidosis (314822.0)
    - Proximal renal tubular acidosis (47159.0)
      - Autosomal dominant proximal renal tubular acidosis (314889.0)
      - Autosomal recessive proximal renal tubular acidosis (93607.0)
    - Osteopetrosis with renal tubular acidosis (2785.0)
    - Distal renal tubular acidosis (18.0)
      - Autosomal dominant distal renal tubular acidosis (dRTA) (93608.0)
      - Autosomal recessive distal renal tubular acidosis (dRTA) (402041.0)
      - Distal renal tubular acidosis with hemolytic anemia (93610.0)
      - Auto-immune distal renal tubular acidosis (9999994.0)
        - Sjogren Syndrome (289390.0)
        - Primary biliary cholangitis (186.0)
        - Autoimmune hepatitis (2137.0)
        - Other autoimmune dRTA (9999966.0)
  - Cystinuria (214.0)
  - Hypotonia-cystinuria syndrome (163690.0)
  - Bartter syndrome (112.0)
  - Hypokalemic alkalosis, CLDN10 associated (528105.0)
  - Gitelman syndrome (358.0)
  - Gitelman-like syndrome (9999964.0)
  - Genetic primary hypomagnesemia (34526.0)
    - Familial primary hypomagnesemia with hypercalciuria and nephrocalcinosis (306516.0)
      - Familial primary hypomagnesemia with hypercalciuria and nephrocalcinosis with severe ocular involvement (2196.0)
      - Familial primary hypomagnesemia with hypercalciuria and nephrocalcinosis without severe ocular involvement (31043.0)
    - Genetic primary hypomagnesemia with normocalcuria (306522.0)
      - Familial primary hypomagnesemia with normocalciuria and normocalcemia (34527.0)
      - Isolated autosomal dominant hypomagnesemia, Glaudemans type (199326.0)
      - Primary hypomagnesemia with secondary hypocalcemia (30924.0)
    - Familial primary hypomagnesemia with hypocalcuria (306519.0)
      - Autosomal dominant primary hypomagnesemia with hypocalciuria (34528.0)
    - Primary hypomagnesemia with refractory seizures and intellectual disability (564178.0)
  - Hypomagnesaemia, drug-induced (9999992.0)
  - Fanconi syndrome, primary (3337.0)
  - Fanconi syndrome, drug induced (9999996.0)
    - Fanconi syndrome, Cisplatin induced (240863.0)
    - Fanconi syndrome, induced by other drug (9999967.0)
  - Fanconi syndrome, heavy metal induced (9999998.0)
    - Fanconi syndrome, lead induced (330015.0)
    - Fanconi syndrome, mercury induced (330021.0)
    - Faconi syndrome, induced by other heavy metal (9999997.0)
  - Fanconi syndrome, monoclonal Ig light chain-associated (91136.0)
  - Hypophosphatemic rickets (437.0)
    - X-linked hypophosphatemic rickets (XLHR) (89936.0)
    - Autosomal recessive hypophosphatemic rickets (289176.0)
    - Autosomal dominant hypophosphatemic rickets (89937.0)
    - Hereditary hypophosphatemic rickets with hypercalciuria (157215.0)
  - Hypophosphatemia, dominant, with nephrolithiasis or osteoporosis (244305.0)
  - Hypercalciuria, idiopathic (2197.0)
  - Oncogenic osteomalacia (352540.0)

Nephrogenic diabetes insipidus (223.0)
Nephrogenic syndrome of inappropriate antidiuresis (93606.0)
Tubulointerstitial nephritis and uveitis syndrome (91500.0)
Familial renal glucosuria (69076.0)
Pseudohypoaldosteronism type 1 (756.0)
- Generalized pseudohypoaldosteronism type 1 (171876.0)
- Renal pseudohypoaldosteronism type 1 (171871.0)

EAST syndrome (199343.0)
Arthrogryposis-renal dysfunction-cholestasis (ARC) syndrome (2697.0)
Autosomal dominant hypocalcemia (428.0)
Hereditary renal hypouricemia (94088.0)

- **Metabolic nephropathies (93593.0)**
  - Dent disease (1652.0)
    - Dent disease type 1 (CLCN5-related) (93622.0)
    - Dent disease type 2 (OCRL-related) (93623.0)
  - Lowe syndrome (534.0)
  - Nephropathic cystinosis (213.0)
    - Infantile nephropathic cystinosis (411629.0)
    - Juvenile nephropathic cystinosis (411634.0)
  - Primary hyperoxaluria (416.0)
    - Primary hyperoxaluria type 1 (PH1) (93598.0)
    - Primary hyperoxaluria type 2 (PH2) (93599.0)
    - Primary hyperoxaluria type 3 (PH3) (93600.0)
  - Autosomal recessive infantile hypercalcemia (300547.0)
  - Amelogenesis imperfecta-nephrocalcinosis syndrome (1031.0)
  - Fabry disease (324.0)
  - Tubulopathy due to mitochondrial oxidative phosphorylation disorder (223713.0)
  - Methylmalonic acidemia (without homocystinuria) (293355.0)
    - Methylmalonic acidemia, Vitamin B12-responsive (28.0)
    - Methylmalonic acidemia, Vitamin B12-unresponsive (27.0)
  - Glycogen storage disease due to glucose-6-phosphatase deficiency (364.0)
  - Glycogen storage disease due to GLUT2 deficiency (2088.0)
  - Lysinuric protein intolerance (470.0)
  - Hereditary xanthinuria (3467.0)
    - Xanthinuria type I (93601.0)
    - Xanthinuria type II (93602.0)
  - Hypoxanthine-guanine phosphoribosyltransferase deficiency (206428.0)
  - Phosphoribosylpyrophosphate synthetase superactivity (3222.0)
  - LCAT deficiency (650.0)
  - Adenine phosphoribosyltransferase deficiency (976.0)
  - Galactosemia (352.0)
  - Imerslund-Gräsbeck syndrome (35858.0)
  - Donnai-Barrow syndrome (2143.0)

- **Thrombotic microangiopathies (93573.0)**
  - Hemolytic uremic syndrome (HUS) (544458.0)
    - Infection-related hemolytic uremic syndrome (HUS) (544482.0)
      - Shiga toxin-associated hemolytic uremic syndrome (STEC-HUS) (90038.0)
      - Streptoccous pneumoniae-associated hemolytic uremic syndrome (HUS) (544493.0)
    - Atypical hemolytic uremic syndrome (aHUS) (2134.0)
      - Atypical hemolytic uremic syndrome (aHUS) with anti-factor H antibodies (93581.0)
      - Atypical hemolytic uremic syndrome (aHUS) with complement gene abnormality (544472.0)
      - Atypical hemolytic uremic syndrome (aHUS), not further specified (9999987.0)
    - Atypical hemolytic uremic syndrome (aHUS) due to Methylcobalamin deficiency, type CblC (79282.0)
    - Atypical hemolytic uremic syndrome (aHUS) with DGKE deficiency (357008.0)
  - Thrombotic thrombocytopenic purpura (TTP) (54057.0)
    - Congenital thrombotic thrombocytopenic purpura (TTP) (93583.0)
    - Acquired thrombotic thrombocytopenic purpura (TTP) (93585.0)
  - Systemic Lupus Erythematosus associated TMA (SLE) (536.1)
  - Childhood-onset systemic lupus erythematosus associated TMA (SLE) (93552.1)
  - De novo thrombotic microangiopathy after kidney transplantation (244275.0)

- **Renal or urinary tract malformations (93545.0)**
  - Non-syndromic renal or urinary tract malformation (93546.0)
    - Renal agenesis (411709.0)
      - Renal agenesis, unilateral (93100.0)
      - Renal agenesis, bilateral (1848.0)
    - Renal hypoplasia (93101.0)
      - Renal hypoplasia, unilateral (97361.0)
      - Renal hypoplasia, bilateral (97362.0)
    - Oligomeganephronia (2260.0)
    - Renal dysplasia (93108.0)
      - Renal dysplasia, unilateral (93172.0)
      - Renal dysplasia, bilateral (93173.0)
    - Multicystic dysplastic kidney (1851.0)
      - Unilateral multicystic dysplastic kidney (97363.0)
      - Bilateral multicystic dysplastic kidney (97364.0)
    - Medullary sponge kidney (1309.0)
    - Renal tubular dysgenesis (3033.0)
      - Renal tubular dysgenesis of genetic origin (97369.0)
      - Drug-related renal tubular dysgenesis (97368.0)
      - Renal tubular dysgenesis due to twin-twin transfusion (97367.0)
    - Vesicoureteric reflux (VUR), high-grade (9999969.0)
    - Vesicoureteric reflux (VUR), familial (289365.0)
    - Fetal lower urinary tract obstruction (LUTO) (435365.0)
      - Posterior urethral valve (PUV) (93110.0)
      - Atresia of urethra (105.0)
      - Prune belly syndrome (2970.0)
      - Anterior urethral valve (435372.0)
    - Congenital hydronephrosis (2190.0)
    - Congenital primary megaureter (617.0)
      - Ureteropelvic junction (UPJ) obstruction (bilateral or in solitary kidney) (9999985.0)
      - Congenital primary megaureter, obstructed form (238646.0)
      - Congenital primary megaureter, refluxing form (238650.0)
      - Congenital primary megaureter, refluxing and obstructed (9999986.0)
      - Congenital primary megaureter, nonrefluxing and unobstructed form (238654.0)
      - Primary megaureter, adult-onset form (238642.0)
    - Neurogenic bladder, congenital or acquired (9999968.0)
    - Megacystis-megaureter syndrome (238637.0)
    - Exstrophy-epispadias complex (322.0)
      - Bladder exstrophy (93930.0)
      - Cloacal exstrophy (93929.0)
      - Epispadias (93928.0)
  - Syndromic renal or urinary tract malformation (93547.0)
    - 17q12 microdeletion syndrome (261265.0)
    - Acro-pectoro-renal dysplasia (956.0)
    - Acroosteolysis dominant type (955.0)
    - Acrorenal syndrome (971.0)
    - Alagille syndrome (52.0)
    - Aniridia-renal agenesis-psychomotor retardation syndrome (1064.0)
    - AREDYLD (acral-renal-ectodermal-dysplasia-lipoatrophic-diabetes) syndrome (1133.0)
    - Bardet-Biedl syndrome (renal / urinary tract malformation) (110.1)
    - Beckwith-Wiedemann syndrome (116.0)
    - BOR (branchio-oto-renal) syndrome (107.0)
    - Cat-eye syndrome (195.0)
    - Caudal regression sequence (3027.0)
    - CHARGE syndrome (138.0)
    - Cornelia de Lange syndrome (96095.0)

Di George syndrome (22q11.2 deletion) (567.0)
EEC (Ectrodactyly-ectodermal dysplasia-cleft lip/palate) syndrome (1896.0)
Fraser syndrome (2052.0)
HDR (Hypoparathyroidism-deafness-renal disease) syndrome (2237.0)
Holoprosencephaly-radial heart renal anomalies syndrome (3186.0)
Infundibulopelvic stenosis-multicystic kidney syndrome (1849.0)
Jeune syndrome (renal / urinary tract malformation) (474.1)
Kallmann syndrome (478.0)
Mayer-Rokitansky-K (3109.0)
Meckel syndrome (renal / urinary tract malformation) (564.1)
Megacystis-microcolon-intestinal hypoperistalsis syndrome (2241.0)
Multicentric carpo-tarsal osteolysis with or without nephropathy (2774.0)
Noonan syndrome (648.0)
Ochoa syndrome (2704.0)
Pallister-Hall syndrome (672.0)
RCAD (Renal cysts and diabetes) syndrome (93111.0)
Renal coloboma syndrome (1475.0)
Renal nutcracker syndrome (71273.0)
Rubinstein-Taybi syndrome (783.0)
Schinzel-Giedion syndrome (798.0)
SERKAL syndrome (139466.0)
Simpson-Golabi-Behmel syndrome (373.0)
Smith-Lemli-Opitz syndrome (818.0)
Structural heart defects-renal anomalies (SHDRA) syndrome (689822.0)
Thyrocerebrorenal syndrome (3327.0)
Townes-Brocks syndrome (857.0)
Trisomy 13 (3378.0)
Trisomy 18 (3380.0)
Turner syndrome (881.0)
VACTERL/VATER association (887.0)
WAGR syndrome (893.0)
BNAR (Bifid nose, anorectal and renal anomalies) syndrome (217266.0)
Kabuki Syndrome (2322.0)

Hereditary cystic renal diseases (ciliopathies) (93587.0)

- Autosomal dominant tubulointerstitial kidney disease (ADTKD) (34149.0)
  - UMOD-related autosomal dominant tubulointerstitial kidney disease (ADTKD) (88950.0)
  - MUC1-related autosomal dominant tubulointerstitial kidney disease (ADTKD) (88949.0)
  - REN-related autosomal dominant tubulointerstitial kidney disease (ADTKD) (217330.0)
  - HNF1B-related autosomal dominant tubulointerstitial kidney disease (ADTKD) (93111.2)
  - Autosomal dominant tubulointerstitial kidney disease (ADTKD) NOS - Not further specified (9999999.0)
- Autosomal dominant polycystic kidney disease (ADPKD) (730.0)
- Autosomal dominant polycystic kidney disease type 1 with tuberous sclerosis (ADPKD-TSC) (88924.0)
- Tuberous sclerosis complex (TSC) (805.0)
- Von Hippel-Lindau disease (vHL) (892.0)
- Autosomal recessive polycystic kidney disease (ARPKD) (731.0)
- Nephronophthisis (655.0)
  - Infantile nephronophthisis (93591.0)
  - Juvenile nephronophthisis (93592.0)
  - Late-onset nephronophthisis (93589.0)
- Bardet-Biedl syndrome (110.0)
- Jeune syndrome (ciliopathies) (474.0)
- Joubert syndrome with oculorenal defect (2318.0)
- Joubert syndrome with renal defect (220497.0)
- OFD (Orofaciodigital) syndrome type 1 (2750.0)
- Senior-Loken syndrome (3156.0)
- Senior-Boichis syndrome (84081.0)
- Saldino-Mainzer (conorenal) syndrome (140969.0)
- Meckel syndrome (ciliopathies) (564.0)
- Cerebrorenodigital (Meckel-like) syndrome (1396.0)
- Cranioectodermal dysplasia (Sensenbrenner syndrome) (1515.0)
- Ellis van Creveld syndrome (289.0)
- Alström Syndrome (64.0)
- Renal-hepatic-pancreatic dysplasia (294415.0)
- RHYNS (retinitis pigmentosa, hypopituitarism, nephronophthisis, skeletal dysaplasia) syndrome (140976.0)
- Neonatal diabetes-congenital hypothyroidism-congenital glaucoma-hepatic fibrosis-polycystic kidneys syndrome (79118.0)
- Karyomegalic interstitial nephritis (401996.0)
- Lethal fetal brain malformation-duodenal atresia-bilateral renal hypoplasia syndrome (444069.0)
- Lethal fetal cerebrorenogenitourinary agenesis/hypoplasia syndrome (439897.0)

- **Rare causes of hypertension (93618.0)**
  - Renal artery stenosis, congenital (97598.0)
  - Genetic hypertension involving tubular dysfunction (156629.0)
    - Apparent mineralocorticoid excess (320.0)
    - Familial hyperaldosteronism (235936.0)
      - Familial hyperaldosteronism type I (glucocorticoid-sensitive hypertension) (403.0)
      - Familial hyperaldosteronism type II (404.0)
      - Familial hyperaldosteronism type III (251274.0)
    - Williams syndrome (904.0)
    - Neurofibromatosis type 1 (636.0)
    - Pseudohypoaldosteronism type 2 (757.0)
    - Liddle Syndrome (526.0)
  - Brachydactyly-arterial hypertension syndrome (1276.0)

Paediatric Chronic Kidney Disease (10000007.0)
Paediatric Dialysis (10000008.0)
Paediatric Transplantation (10000009.0)