| **Diagnosis** | **Cohort** | **Inclusion Criteria** | **Exclusion Criteria** | **Date of Diagnosis** |
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| **Congenital anomalies of the Kidneys and Urinary Tract** | CAKUT | Paediatric and adult patients with structural malformations of the kidneys and lower urinary tract. Eligible patients will have radiologically confirmed urinary tract malformations and fulfil at least one of the following criteria:  1. Congenital kidney malformations (including congenital renal tumours)  2. Congenital ureter and bladder abnormalities (e.g., Prune Belly syndrome, Ureterocele, Bladder exstrophy)  3. Congenital bladder outlet obstruction (e.g., Posterior Urethral Valves)  4. Cloacal malformations  This may include the following SNOMED-CT terms:  Bladder exstrophy (61758007); Cloacal Exstrophy (20815007); Congenital Anomaly Of The Kidney (44513007); Congenital Anomaly Of The Urinary Tract Proper (118642009); Congenital Hydronephrosis (16297002); Congenital Megaureter (718485003); Congenital Posterior Urethral Valves (253900005); Congenital vesico-ureteric junction obstruction (1155732005); Cystic Renal Dysplasia (16507009); Duplex Kidney (44796002); Ectopic Kidney (1230270001); Ectopic Ureter (95575002); Exstrophy Epispadias Complex (5187006); Pelviureteric Junction Obstruction (204942005); Prune Belly Syndrome (204949001); Renal Agenesis (32659003); Renal Dysplasia (12818004); Renal Hypoplasia (197811007); Ureterocele ; Vesicoureteric Reflux | None | Date of radiologically confirmed urinary tract malformation |
| **Adenine Phosphoribosyltransferase Deficiency (APRT-D)** | APRT Deficiency | APRT Deficiency confirmed  Abolished APRT enzyme activity or confirmed disease-causing mutation | None, if APRT Deficiency not confirmed | Date that clinical diagnosis was first made |
| **Lupus Nephritis** | Lupus Nephritis | Diagnosis of Lupus Nephritis within the last five years confirmed by biopsy and to provide classification and overlapping classification where applicable | None | Date that clinical diagnosis was first made |
| **Alport Syndrome and Type IV collagenopathies** | Alport | Alport Syndrome definite or probable  Alport carrier definite or probable  Female heterozygote for X-linked Alport Syndrome (COL4A5)  Heterozygote for autosomal Alport Syndrome (COL4A3, COL4A4)  Thin basement membrane nephropathy | None stated | Date that clinical diagnosis was first made |
| **APOL1 disease, suspected or confirmed** | CKD-Africa Genes | People of African or Afro-Caribbean ancestry with CKD (KDIGO definition), >18 years  including: Focal segmental glomerulosclerosis (primary or secondary) on renal biopsy; Non-diabetic and non-immunological kidney disease with no other confirmed cause | None stated | Date that clinical diagnosis was first made |
| **Autoimmune distal renal tubular acidosis** | Tubulopathy | Autoimmune distal renal tubular acidosis | None stated | Date that clinical diagnosis was first made |
| **Autosomal dominant distal renal tubular acidosis** | Tubulopathy | Autosomal dominant distal renal tubular acidosis  Genetically confirmed heterozygous pathogenic variant in SLC4A1 | None stated | Date that clinical diagnosis was first made |
| **Autosomal recessive distal renal tubular acidosis** | Tubulopathy | Autosomal recessive distal renal tubular acidosis  Genetically confirmed homozygous pathogenic variant in ATP6V0A4, ATP6V1B1 or FOXI1 | None stated | Date that clinical diagnosis was first made |
| **Autosomal recessive proximal renal tubular acidosis** | Tubulopathy | Autosomal recessive proximal renal tubular acidosis with ocular abnormalities and intellectual disability  Genetically confirmed homozygous pathogenic variant in SLC4A4 | None stated | Date that clinical diagnosis was first made |
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| **Diagnosis** | **Cohort** | **Inclusion Criteria** | **Exclusion Criteria** | **Date of Diagnosis** |
| **Bartter Syndrome types 1 and 2** | Tubulopathy | Bartter Syndrome, infantile onset  Hypokalaemic alkalosis, infantile onset without hypertension  Hypokalaemic alkalosis, infantile onset with raised renin | Acidosis  Persistent Hyperkalaemia | Date that clinical diagnosis was first made |
| **Bartter Syndrome type 3**  **Gitelman Syndrome** | Tubulopathy | Bartter Syndrome type 3  Gitelman Syndrome  Hypokalaemic alkalosis with hypomagnesaemia  Hypokalaemic alkalosis with raised renin  Hypokalaemic alkalosis without hypertension | Acidosis  Hyperkalaemia | Date that clinical diagnosis was first made |
| **Bartter Syndrome Type 4** | Tubulopathy | Bartter Syndrome, infantile onset with deafness  Hypokalaemic alkalosis, infantile onset without hypertension with deafness  Hypokalaemic alkalosis, infantile onset with raised renin, with deafness | Acidosis  Persistent Hyperkalaemia | Date that clinical diagnosis was first made |
| **BK Nephropathy** | BK Nephropathy | Significant BK viraemia, with polymerase chain reaction (PCR) greater than or equal to 10 log 4 copies per ml.  A confirmatory biopsy is **not** required. | None stated | Date that PCR first equalled or exceeded 10 log 4 |

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| **Diagnosis** | **Cohort** | **Inclusion Criteria** | **Exclusion Criteria** | **Date of Diagnosis** |
| **Calciphylaxis** | Calciphylaxis | Any patient with a diagnosis of clinical diagnosis of Calciphylaxis; tissue diagnosis not required | None stated | Date that the diagnosis was made by a nephrologist or dermatologist |
| **Cystinosis (Nephropathic Cystinosis)** | Cystinosis | Cystinosis | None stated | Date that biochemical testing first showed an elevated level of white blood cell cysteine |
| **Cystinuria** | Cystinuria | Biochemically proven cystine kidney stone  Urinary cystine level > 3X reference range of the laboratory it was taken in  Cystine crystals in the urine (biochemically proven) | Another cause of proximal tubular dysfunction accounting for the raised cystine level e.g. Fanconi's syndrome | Date that any of the inclusion criteria first occurred |
| **Dent Disease** | Dent & Lowe | Dent Disease | None stated | Date that the clinical label of Dent Disease was first applied |
| **Dominant hypophosphatemia with nephrolithiasis or osteoporosis** | Tubulopathy | Dominant hypophosphatemia with nephrolithiasis or osteoporosis  Genetically confirmed heterozygous pathogenic variant in SLC34A1, SLC9A3R1, SLC34A3 | None stated | Date that clinical diagnosis was first made |
| **Drug induced Fanconi syndrome** | Tubulopathy | Drug induced Fanconi syndrome | None stated | Date that clinical diagnosis was first made |
| **Drug induced hypomagnesemia** | Tubulopathy | Drug induced hypomagnesemia | None stated | Date that clinical diagnosis was first made |

| **Diagnosis** | **Cohort** | **Inclusion Criteria** | **Exclusion Criteria** | **Date of Diagnosis** |
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| **Drug induced Nephrogenic Diabetes Insipidus** | Tubulopathy | Drug induced Nephrogenic Diabetes Insipidus | None stated | Date that clinical diagnosis was first made |
| **EAST syndrome (Epilepsy, Ataxia, Sensorineural deafness, Tubulopathy)** | Tubulopathy | Gitelman/Bartter-type syndrome in childhood with epilepsy /ataxia | Normal CNS examination | Date that clinical diagnosis was first made |
| **End stage kidney disease of unknown cause** | CKD-Africa Genes | People of African or Afro-Caribbean ancestry with CKD (KDIGO definition), >18 years | Known cause of kidney disease identified (unless Sickle cell Nephropathy or APOL1 disease) | Date that clinical diagnosis was first made |
| **Fabry Disease** | Fabry | Confirmed diagnosis of Fabry Disease | None stated | Date that genetic diagnosis was made and/or, for males, the date that low alpha gal levels were first recorded |
| **Familial Hypomagnesaemia with hypercalciuria and nephrocalcinosis CLDN16/19** | Tubulopathy | Familial Hypomagnesaemia with Hypercalciuria and Nephrocalcinosis  Genetically confirmed homozygous pathogenic variant in CLDN 16/19 | None stated | Date that clinical diagnosis was first made |
| **Familial primary hypomagnesemia with hypocalcuria FXYD2** | Tubulopathy | Familial primary hypomagnesemia with hypocalciuria  Genetically confirmed homozygous pathogenic variant in FXYD2 | None stated | Date that clinical diagnosis was first made |
| **Familial primary hypomagnesemia with normocalcuria EGF** | Tubulopathy | Familial primary hypomagnesemia with normocalcuria  Genetically confirmed homozygous pathogenic variant in EGF | None stated | Date that clinical diagnosis was first made |
| **Familial renal glucosuria SLC5A2** | Tubulopathy | Familial renal glucosuria  Genetically confirmed homozygous pathogenic variant in SLC5A2 | None stated | Date that clinical diagnosis was first made |

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| **Diagnosis** | **Cohort** | **Inclusion Criteria** | **Exclusion Criteria** | **Date of Diagnosis** |
| **Fanconi Renotubular syndrome 1 (FRTS1)** | Tubulopathy | Fanconi Renotubular syndrome 1 | None stated | Date that clinical diagnosis was first made |
| **Fanconi Renotubular syndrome 2 (FRTS2)** | Tubulopathy | Fanconi Renotubular syndrome 2  Genetically confirmed homozygous pathogenic variant in SLC34A1 | None stated | Date that clinical diagnosis was first made |
| **Fanconi Renotubular syndrome 3 (FRTS3)** | Tubulopathy | Fanconi Renotubular syndrome 3  Genetically confirmed homozygous pathogenic variant in EHHADH | None stated | Date that clinical diagnosis was first made |
| **Fibromuscular Dysplasia** | Fibromuscular Dysplasia | Diagnosis of FMD established on radiological or histological grounds  FMD of any arterial bed | None stated | Date that FMD was diagnosed by radiological (or histological) methods |
| **Generalized pseudohypoaldosteronism type 1** | Tubulopathy | Generalized pseudohypoaldosteronism type 1  Genetically confirmed homozygous pathogenic variant in SCNN1A/ SCNN1B/SCNN1G | None stated | Date that clinical diagnosis was first made |

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| **Diagnosis** | **Cohort** | **Inclusion Criteria** | **Exclusion Criteria** | **Date of Diagnosis** |
| **Haemolytic Uraemic Syndrome - Atypical** | aHUS | Diarrhoea-negative HUS, includes congenital and familial HUS  Renal biopsy showing a TMA and/or the triad of microangiopathic haemolytic anaemia, thrombocytopenia, renal failure. | Shiga toxin associated HUS  Secondary causes:   * Drugs * Infection (HIV, pneumonia, streptococcus) * Transplantation (bone marrow, liver, lung, cardiac but not de-novo renal) * Cobalamin deficiency * SLE * APL Ab syndrome * Scleroderma * ADAMTS13 antibodies or deficiency | Date of first presentation |

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| **Diagnosis** | **Cohort** | **Inclusion Criteria** | **Exclusion Criteria** | **Date of Diagnosis** |
| **Haemolytic Uraemic Syndrome-Shiga toxin (Verocytotoxin)-associated** | STEC-HUS | Acute kidney injury (AKI) with elevated creatinine for age and/or oligoanuria (urine output <0.5ml/kg/hr over 24hr period) with either:   * Microangiopathic haemolytic anaemia (MAHA) - defined as Hgb < 10mg/dl with fragmented RBCs   or   * Thrombocytopaenia - defined as platelet count less than 130, 000 x 10 9/l   and   * Occurring with Shiga-toxin producing E Coli (STEC) infection defined as: * Positive STEC culture * Positive PCR for Stx gene directly from a faecal specimen * Positive antibodies to the lipopolysaccharide * antigen of E. coli serogroups O157, O26, O103, O111 and O145 | Septicaemia  Malignant hypertension  Primary vascular disease  Familial HUS not being part of the same | Date on which the STEC-HUS was suspected. |
| **Heavy metal induced Fanconi syndrome** | Tubulopathy | Heavy metal induced Fanconi syndrome | None stated | Date that clinical diagnosis was first made |

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| **Diagnosis** | **Cohort** | **Inclusion Criteria** | **Exclusion Criteria** | **Date of Diagnosis** |
| **Hepatocyte Nuclear Factor-1B mutation** | HNF1b | Hepatocyte nuclear factor-1B mutation  Renal cysts and diabetes (RCAD)  Inherited genetic diabetes type 2 (MODY 5). | None stated | Date of genetic diagnosis |
| **Hereditary renal hypouricemia** | Tubulopathy | Hereditary renal hypouricemia  Genetically confirmed homozygous pathogenic variant in SLC22A12, SLC2A9 | None stated | Date of genetic diagnosis |
| **Hereditary hypophosphatemic rickets with hypercalciuria** | Tubulopathy | Hereditary hypophosphatemic rickets with hypercalciuria  Genetically confirmed homozygous pathogenic variant in SLC34A3 | None stated | Date of genetic diagnosis |
| **Hyperoxaluria (Primary hyperoxaluria, Oxalosis)** | Hyperoxaluria | Primary Hyperoxaluria Type1  Primary Hyperoxaluria Type 2  Primary Hyperoxaluria Type 3  Primary Hyperoxaluria awaiting genetic confirmation (Urine oxalate excretion ≥ 0.8 mmol/1.73 m2/24 hrs)  Primary Hyperoxaluria Unclassified  Primary Hyperoxaluria Unclassified but with systemic oxalate deposition | Secondary hyperoxaluria associated with gastrointestinal disease  Renal failure without systemic oxalate deposits | Date that definitive diagnosis by genetic confirmation with gene mutation was first made.  If in doubt use the earliest date that PH was suspected or the date when treatment was first introduced |

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| **Diagnosis** | **Cohort** | **Inclusion Criteria** | **Exclusion Criteria** | **Date of Diagnosis** |
| **Hypertensive kidney disease** | **CKD-Africa Genes** | **People of African or Afro-Caribbean ancestry with CKD (KDIGO definition), >18 years** | Known cause of Kidney disease | Date that clinical diagnosis was first made |
| **Hyperuricaemic Nephropathy (Primary/Familial Hyperuricaemic nephropathy)**  **Medullary cystic kidney disease** | **ADTKD** | **Autosomal Dominant Tubulointerstitial Kidney Disease (ADTKD; previously known as FUAN)**  Familial juvenile hyperuricaemic nephropathy  Familial gouty nephropathy  Familial urate nephropathy  Familial interstitial nephropathy  Uromodulin-associated nephropathy  Medullary cystic kidney disease (type I or II) | None stated | Date that genetic confirmation was received |
| **IgA Nephropathy** | IgA Nephropathy | Biopsy proven IgA Nephropathy plus proteinuria >0.5g/ day or eGFR<60ml/min | All forms of secondary IgA nephropathy, including Henoch Schonlein purpura | Date of renal biopsy |
| **Isolated autosomal dominant hypomagnesemia, Glaudemans type** | Tubulopathy | Isolated autosomal dominant hypomagnesemia  Genetically confirmed homozygous pathogenic variant in KCNA1 | None stated | Date that clinical diagnosis was first made |

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| **Diagnosis** | **Cohort** | **Inclusion Criteria** | **Exclusion Criteria** | **Date of Diagnosis** |
| **Liddle syndrome** | Tubulopathy | Liddle syndrome  Hypertension with hypokalaemia, suppressed aldosterone  Hypertension with suppressed aldosterone  Autosomal dominant hypertension, suppressed aldosterone | Hyperaldosteronism | Date that clinical diagnosis was first made |
| **Lowe Syndrome** | Dent & Lowe | Lowe Syndrome | None Stated | Date that the clinical label of Lowe Syndrome was first applied |

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| **Diagnosis** | **Cohort** | **Inclusion Criteria** | **Exclusion Criteria** | **Date of Diagnosis** |
| **Membranoproliferative glomerulonephritis**  **Mesangiocapillary glomerulonephritis**  **Dense Deposit Disease**  **C3 Glomerulonephritis**  **C3 Glomerulopathy** | MPGN | Child or adult with histological finding of:  MPGN Type I  Dense Deposit Disease (morphological pattern may or may not be MPGN)  Other pattern of MPGN  C3 Glomerulonephritis (Characterised by C3 deposits in the absence of immunoglobulin with electron dense deposits (morphological pattern may or may not be MPGN)  Unclassified GN with capillary wall immune deposits | MPGN known to be secondary to:  Chronic bacterial infection  Hepatitis B or C infection  Malignancy  Systemic lupus erythematosus (by ACR criteria) | Date of biopsy |
| **Membranous Nephropathy** | Membranous Nephropathy | Membranous nephropathy confirmed by kidney histology | Lupus nephritis | Date of biopsy |
| **Mitochondrial Renal Disease** | Mitochondrial | Mitochondrial Disease **or** Mitochondrial Cytopathy | None Stated | Date that clinical diagnosis was first made |

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| **Diagnosis** | **Cohort** | **Inclusion Criteria** | **Exclusion Criteria** | **Date of Diagnosis** |
| **Monoclonal Gammopathy of Renal Significance** | MGRS | Renal biopsy proven confirmation of:  • AH amyloidosis\*  • AHL amyloidosis\*  • AL amyloidosis\*  • C3 glomerulonephritis with monoclonal gammopathy  • Crystalglobulinaemia  • Crystal-storing histiocytosis  • Fibrillary Glomerulonephritis  • Immunotactoid/Glomerulonephritis with Organised Microtubular Monoclonal Immunoglobulin Deposits (GOMMID)  • Intracapillary monoclonal IgM without cryoglobulin  • Intraglomerular/capillary lymphoma/leukaemia  • Light chain cast nephropathy  • Light chain proximal tubulopathy, crystalline  • Light chain proximal tubulopathy, non crystalline   * Monoclonal Immunoglobulin Deposition Disease (MIDD; includes Light Chain Deposition Disease - LCDD; Heavy Chair Deposition Disease - HCDD; and Light and Heavy Chain Deposition Disease - LHCDD) * Proliferative glomerulonephritis with monoclonal immunoglobulin deposits – PGNMID   • Thrombotic Microangiopathy with monoclonal gammopathy  • Type 1 cryoglobulinaemic Glomerulonephritis  • Unclassified MGRS  \*Patients with systemic amyloidosis may have a renal biopsy confirming AL amyloidosis or a biopsy of other tissue with confirmation of renal involvement by the UK National Amyloidosis Centre. | None Stated | Date of biopsy |

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| **Diagnosis** | **Cohort** | **Inclusion Criteria** | **Exclusion Criteria** | **Date of Diagnosis** |
| **Nephrogenic diabetes insipidus** | Tubulopathy | Nephrogenic diabetes insipidus  Genetically confirmed homozygous pathogenic variant in AVPR2, AQP2 | None stated | Date that clinical diagnosis was first made |
| **Nephrogenic syndrome of inappropriate antidiuresis** | Tubulopathy | Nephrogenic syndrome of inappropriate antidiuresis  Genetically confirmed homozygous pathogenic variant in AVPR2 | None stated | Date that clinical diagnosis was first made |
| **Nephronophthisis** | ARPKD/NPHP | Histological or radiological features of Nephronophthisis  Genetic diagnosis of Nephronophthisis or Nephronophthisis-related ciliopathy | None stated | Date that histological /radiological or genetic diagnosis was first made |

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| **Diagnosis** | **Cohort** | **Inclusion Criteria** | **Exclusion Criteria** | **Date of Diagnosis** |
| **Nephrotic Syndrome - Steroid Sensitive or Steroid Resistant**  **(Congenital nephrotic syndrome, nephrotic syndrome with focal segmental glomerulosclerosis)** | INS | Children and adults with idiopathic Nephrotic Syndrome (nephrotic range proteinuria and hypoalbuminaemia)  Congenital NS (presumed Steroid Resistance)  Childhood or adult onset with primary Steroid Resistance  Childhood or adult onset with late onset Steroid Resistance  Steroid Sensitive Nephrotic Syndrome (full or partial remission in response to steroids)  As part of a syndrome e.g. Nail Patella Syndrome and Denys-Drash Syndrome  Those with a biopsy diagnosis of FSGS or minimal change disease can be included if they fall in the above categories but biopsy is not a prerequisite for inclusion | Secondary causes of Nephrotic Syndrome   * Primary diagnosis of Glomerulonephritis (IgA Nephropathy, Membranoproliferative Glomerulonephritis, Membranous Nephropathy) * Vasculitis * Systemic Lupus Erythematosus * Diabetes * Obesity * Hypertension | Date of presentation to secondary or tertiary centre |
| **Oncogenic osteomalacia** | Tubulopathy | Oncogenic osteomalacia | None stated | Date that clinical diagnosis was first made |
| **Osteopetrosis with renal tubular acidosis** | Tubulopathy | Osteopetrosis with renal tubular acidosis  Genetically confirmed homozygous pathogenic variant in CA2 | None stated | Date that clinical diagnosis was first made |

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| **Diagnosis** | **Cohort** | **Inclusion Criteria** | **Exclusion Criteria** | **Date of Diagnosis** |
| **Polycystic Kidney Disease - Autosomal Dominant** | ADPKD | Clinical features of Autosomal Dominant Polycystic Kidney Disease meeting current image based diagnostic criteria  Clinical features compatible with ADPKD in the absence of a family history  Pathogenic or likely pathogenic PKD1 or PKD2 mutation with or without clinical features | Autosomal dominant polycystic liver disease with no evidence of renal cysts | Date that the clinical diagnosis was first made.  This may be reported by the clinician as the date of the diagnostic scan or by the patient if scans were performed at another centre |
| **Polycystic Kidney Disease - Autosomal Recessive** | ARPKD | Autosomal Recessive Polycystic Kidney Disease  Congenital Hepatic Fibrosis  Caroli Syndrome with kidney malformation or cyst | None stated | Date that clinical diagnosis was first made. |

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| **Diagnosis** | **Cohort** | **Inclusion Criteria** | **Exclusion Criteria** | **Date of Diagnosis** |
| **Pregnancy and Chronic Kidney Disease** | Pregnancy | Pregnancy in all women known to have CKD 1-5 prior to pregnancy or those with a serum creatinine >85umol/l on two occasions during pregnancy  Pregnancy in all women with renal transplants regardless of function  Pregnancy in all women with previous or current lupus nephritis regardless of function | None stated | Date of last menstrual period |
| **Primary hypomagnesemia with secondary hypocalcemia** | Tubulopathy | Primary hypomagnesemia with secondary hypocalcemia  Genetically confirmed homozygous pathogenic variant in TRPM6 | None stated | Date that clinical diagnosis was first made |
| **Pseudohypoaldosteronism type 2A** | Tubulopathy | Pseudohypoaldosteronism type 2A | None stated | Date that clinical diagnosis was first made |
| **Pseudohypoaldosteronism type 2B** | Tubulopathy | Pseudohypoaldosteronism type 2B  Genetically confirmed homozygous pathogenic variant in WNK1 | None stated | Date that clinical diagnosis was first made |
| **Pseudohypoaldosteronism type 2C** | Tubulopathy | Pseudohypoaldosteronism type 2C  Genetically confirmed homozygous pathogenic variant in WNK4 | None stated | Date that clinical diagnosis was first made |
| **Pseudohypoaldosteronism type 2D** | Tubulopathy | Pseudohypoaldosteronism type 2D  Genetically confirmed homozygous pathogenic variant in KLHL3 | None stated | Date that clinical diagnosis was first made |

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| **Diagnosis** | **Cohort** | **Inclusion Criteria** | **Exclusion Criteria** | **Date of Diagnosis** |
| **Pseudohypoaldosteronism type 2E** | Tubulopathy | Pseudohypoaldosteronism type 2E  Genetically confirmed homozygous pathogenic variant in CUL3 | None stated | Date that clinical diagnosis was first made |
| **Pure Red Cell Aplasia** | PRCA | Treatment with any injectable form of erythropoiesis stimulating agent for at least four weeks.  Haemoglobin <70 g/l without transfusion or transfusion dependence.  Normal leucocyte and platelet count  Reticulocyte count < 20.000 / mm3  Bone marrow aspirate showing well preserved myeloid and megakaryocyte development, and <5% erythroblasts.  Presence of anti-erythropoietin antibodies. | Pre-established PRCA due to myeloproliferative disorder | Date of positive antibody test |
| **Renal pseudohypoaldosteronism type 1** | Tubulopathy | Renal pseudohypoaldosteronism type 1  Genetically confirmed homozygous pathogenic variant in NR3C2 |  | Date that clinical diagnosis was first made |

| **Diagnosis** | **Cohort** | **Inclusion Criteria** | **Exclusion Criteria** | **Date of Diagnosis** |
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| **Retroperitoneal Fibrosis** | Retroperitoneal Fibrosis | Any radiologically confirmed retroperitoneal fibrosis (RPF), presumed to be 'idiopathic' or associated with primary conditions including (but not exclusively):   * Aortitis * Periaortitis * IgG4-related Vasculitis * Perivascular fibrosis * Atherosclerotic or aneurysmal disease   **Note:** There is no specific ICD code for retroperitoneal fibrosis although the diagnosis term links to two ICD codes:   * ICD10:N13.5 - Crossing vessel and stricture of ureter without hydronephrosis * ICD-9-CM 593.4 - Other ureteric obstruction | Neoplastic disease within retroperitoneal fibrosis mass defined histologically | Date of diagnostic imaging study report |
| **Sickle Cell Nephropathy** | **CKD Africa Genes** | People of African or Afro-Caribbean ancestry with CKD (KDIGO definition), >18 years  Known Sickle Cell disease with reduced kidney function, and/or blood or protein in urine with no other cause for kidney disease identified | None stated | Date that clinical diagnosis was first made |
| **Tuberous Sclerosis** | **Tuberous Sclerosis** | Clinical or molecular diagnosis of Tuberous Sclerosis Complex (TSC)  Multiple renal angiomyolipomas  Multiple renal angiomyolipomas (> 3) +/- pulmonary lymphangioleiomyomatosis (LAM) without other signs of TSC | None stated | Date that clinical diagnosis was first made |
| **Vasculitis (Primary systemic Vasculitis)** | **Vasculitis** | **Small vessel Vasculitis (ANCA associated)**  Microscopic polyangiitis (including renal limited Vasculitis)  Granulomatosis with polyangiitis (Wegener)  Eosinophilic granulomatosis with polyangiitis (Churg Strauss)  ANCA Vasculitis unclassified  **Small vessel Vasculitis (Immune complex)**  anti-GBM disease  Cryoglobulinemic Vasculitis  IgA Vasculitis (Henoch-Schönlein)  **Medium vessel Vasculitis**  Classical PAN  Kawasaki disease  **Large vessel Vasculitis**  Giant cell arteritis  Takayasu’s arteritis | None stated | Date of biopsy.  In the absence of a biopsy, the date of a positive antibody test should be used |
| **Vasculitis (Primary systemic Vasculitis)** | Vasculitis | **Variable vessel Vasculitis**  Behçet’s disease  Cogan’s syndrome  **Single organ Vasculitis**  Isolated aortitis  Primary cerebral angiitis | None stated | Date of biopsy.  In the absence of a biopsy, the date of a positive antibody test should be used |
| **Inherited Renal Cancer Syndrome** | Renal Cancer Inherited | 1. A molecular or clinical diagnosis according to standard criteria of any of the following conditions:   Von Hippel Lindau disease (VHL) OMIM 193300  PTEN hamartoma tumour syndrome (Cowden syndrome) OMIM 158350  Birt Hogg Dube syndrome (BHD) OMIM 135150  Hereditary leiomyomatosis and renal cell cancer syndrome(HLRCC) OMIM 150800  Succinate dehydrogenase-related tumour predisposition syndrome  BAP1-related tumour predisposition syndrome OMIM 614327  Hereditary Type 1 papillary renal cell carcinoma syndrome (MET oncogene) OMIM 605074   1. Two or more cases in first degree relatives of any type of renal cancer without an established  molecular or clinical diagnosis 2. Bilateral, multiple primary renal cancers of any histopathological type with or with a family history | None stated | Date of molecular or clinical diagnosis according to standard criteria |