



Diagnosis	Cohort	Inclusion Criteria	Exclusion Criteria	Date of Diagnosis
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





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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 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





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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
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





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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 <b>not</b> required.	None stated	Date that PCR first equalled or exceeded 10 log 4
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





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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
Cytomegalovirus (CMV) Post Transplant	CMV Post Transplant	Kidney transplant recipients from 2016 onwards. Patients whose transplants have failed can be included.  and  CMV infection in first year after kidney transplantation	Multi-organ transplant recipients	The date the first clinical diagnosis of CMV infection in the first year after kidney transplantation was made.
Dent Disease	Tubulopathy	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





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Drug induced hypomagnesemia	Tubulopathy	Drug induced hypomagnesemia	None stated	Date that clinical diagnosis was first made
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





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Familial renal glucosuria SLC5A2	Tubulopathy	Familial renal glucosuria  Genetically confirmed homozygous pathogenic variant in SLC5A2	None stated	Date that clinical diagnosis was first made
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	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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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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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
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





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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 Type 1  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
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





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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	lgA 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





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Inherited Renal Cancer Syndrome	Renal Cancer Inherited	<ol> <li>A molecular or clinical diagnosis according to standard criteria of any of the following conditions:</li> <li>Von Hippel Lindau disease (VHL) OMIM 193300</li> <li>PTEN hamartoma tumour syndrome (Cowden syndrome) OMIM 158350</li> <li>Birt Hogg Dube syndrome (BHD) OMIM 135150</li> <li>Hereditary leiomyomatosis and renal cell cancer syndrome(HLRCC) OMIM 150800</li> <li>Succinate dehydrogenase-related tumour predisposition syndrome</li> <li>BAP1-related tumour predisposition syndrome OMIM 614327</li> <li>Hereditary Type 1 papillary renal cell carcinoma syndrome (MET oncogene) OMIM 605074</li> <li>Two or more cases in first degree relatives of any type of renal cancer without an established molecular or clinical diagnosis</li> <li>Bilateral, multiple primary renal cancers of any histopathological type with or with a family history</li> </ol>	None stated	Date of molecular or clinical diagnosis according to standard criteria
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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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	Tubulopathy	Lowe Syndrome	None Stated	Date that the clinical label of Lowe Syndrome first applied
Membranoproliferative glomerulonephritis  Mesangiocapillary glomerulonephritis  Dense Deposit Disease  C3 Glomerulonephritis  C3 Glomerulopathy	MPGN	<ul> <li>Child or adult with histological finding of:</li> <li>MPGN Type I</li> <li>Dense Deposit Disease (morphological pattern may or may not be MPGN)</li> <li>Other pattern of MPGN</li> <li>C3 Glomerulonephritis (Characterised by C3 deposits in the absence of immunoglobulin with electron dense deposits (morphological pattern may or may not be MPGN)</li> <li>Unclassified GN with capillary wall immune deposits</li> </ul>	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 <b>or</b> Mitochondrial Cytopathy	None Stated	Date that clinical diagnosis was first made





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Monoclonal Gammopathy of Renal Significance	MGRS	Renal biopsy proven confirmation of:  AH 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 Inght chain proximal tubulopathy, non crystalline Monoclonal Immunoglobulin Deposition Disease (MIDD; includes Light Chain Deposition Disease - LCDD; Heavy Chair Deposition Disease - HCDD) 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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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 & Ciliopathy	Histological or radiological features of Nephronophthisis  Genetic diagnosis of Nephronophthisis or Nephronophthisis-related ciliopathy  Conditions include (but are not limited to): Meckel–Gruber syndrome (MKS) Senior–Løken syndrome (SLS) Alström syndrome Orofaciodigital syndrome (OFD) Ellis–van Creveld syndrome (EVC) Short-rib thoracic dysplasias (SRPS) (including Jeune asphyxiating thoracic dystrophy) Mainzer–Saldino syndrome Sensenbrenner syndrome (cranioectodermal dysplasia)	None stated	Date that histological /radiological or genetic diagnosis was first made





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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
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





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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.
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





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Pseudohypoaldosteronism type 2D	Tubulopathy	Pseudohypoaldosteronism type 2D  Genetically confirmed homozygous pathogenic variant in KLHL3	None stated	Date that clinical diagnosis was first made
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 ciliopathies other than ADPKD, ARPKD or Nepronophthisis	ARPKD, NPHP & Ciliopathy	Histological or radiological features of renal ciliopathy  Genetic diagnosis of renal ciliopathy.  Conditions include (but are not limited to): Bardet Biedl Syndrome (BBS) Joubert Syndrome (JS)	Please recruit patients with ADPKD to the ADPKD cohort	Date that histological / radiological or genetic diagnosis was first made





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Renal pseudohypoaldosteronism type	Tubulopathy	Renal pseudohypoaldosteronism type 1  Genetically confirmed homozygous pathogenic variant in NR3C2	None stated	Date that clinical diagnosis was first made
Retroperitoneal Fibrosis	Retroperitone al 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





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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





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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  Variable vessel Vasculitis  • Behçet's disease  • Cogan's syndrome  Single organ Vasculitis  • Isolated aortitis  • 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