

RaDaR Inclusion and Exclusion Criteria

Diagnosis	Cohort	Inclusion Criteria	Exclusion Criteria	Date of Diagnosis
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
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
Bartter Syndrome types 1 and 2	Hypokalaemic Alkaloses	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	Hypokalaemic Alkaloses	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

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Bartter Syndrome Type 4	Hypokalaemic Alkaloses	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
Calciophylaxis	Calciophylaxis	Any patient with a diagnosis of clinical diagnosis of Calciophylaxis; 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
EAST syndrome (Epilepsy, Ataxia, Sensorineural deafness, Tubulopathy)	Hypokalaemic Alkaloses	Gitelman/Bartter-type syndrome in childhood with epilepsy /ataxia	Normal CNS examination	Date that clinical diagnosis was first made

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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
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
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: <ul style="list-style-type: none"> • 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	<p>Acute kidney injury (AKI) with elevated creatinine for age and/or oligoanuria (urine output <0.5ml/kg/hr over 24hr period) with either:</p> <ul style="list-style-type: none"> • Microangiopathic haemolytic anaemia (MAHA) - defined as Hgb < 10mg/dl with fragmented RBCs • Thrombocytopenia - defined as platelet count less than 130, 000 x 10⁹/l <p>and/or</p> <ul style="list-style-type: none"> • 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 	<p>Septicaemia</p> <p>Malignant hypertension</p> <p>Primary vascular disease</p> <p>Familial HUS not being part of the same</p>	Date on which the STEC-HUS was suspected.
Hepatocyte Nuclear Factor-1B mutation	HNF1b	<p>Hepatocyte nuclear factor-1B mutation</p> <p>Renal cysts and diabetes (RCAD)</p> <p>Inherited genetic diabetes type 2 (MODY 5).</p>	None stated	Date of genetic diagnosis

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<p>Hyperoxaluria (Primary hyperoxaluria, Oxalosis)</p>	<p>Hyperoxaluria</p>	<p>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</p>	<p>Secondary hyperoxaluria associated with gastrointestinal disease Renal failure without systemic oxalate deposits</p>	<p>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</p>
<p>Hyperuricaemic Nephropathy (Primary/Familial Hyperuricaemic nephropathy)</p> <p>Medullary cystic kidney disease</p>	<p>ADTKD</p>	<p>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)</p>	<p>None stated</p>	<p>Date that genetic confirmation was received</p>

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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
Liddle syndrome	Hypokalaemic Alkaloses	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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<p>Membranoproliferative glomerulonephritis</p> <p>Mesangiocapillary glomerulonephritis</p> <p>Dense Deposit Disease</p> <p>C3 Glomerulonephritis</p> <p>C3 Glomerulopathy</p>	MPGN	<p>Child or adult with histological finding of:</p> <p>MPGN Type I</p> <p>Dense Deposit Disease (morphological pattern may or may not be MPGN)</p> <p>Other pattern of MPGN</p> <p>C3 Glomerulonephritis (Characterised by C3 deposits in the absence of immunoglobulin with electron dense deposits (morphological pattern may or may not be MPGN)</p> <p>Unclassified GN with capillary wall immune deposits</p>	<p>MPGN known to be secondary to:</p> <p>Chronic bacterial infection</p> <p>Hepatitis B or C infection</p> <p>Malignancy</p> <p>Systemic lupus erythematosus (by ACR criteria)</p>	Date of biopsy
Membranous Nephropathy	Membranous Nephropathy	Membranous nephropathy confirmed by kidney histology	Lupus nephritis	Date of biopsy

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Monoclonal Gammopathy of Renal Significance	MGRS	<p>Renal biopsy proven confirmation of:</p> <ul style="list-style-type: none"> • 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 <p>*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.</p>	None Stated	Date of biopsy

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Nephronophthisis	ARPKD/NPHP	<p>Histological or radiological features of Nephronophthisis</p> <p>Genetic diagnosis of Nephronophthisis or Nephronophthisis-related ciliopathy</p>	None stated	Date that histological /radiological or genetic diagnosis was first made
Nephrotic Syndrome - Steroid Sensitive or Steroid Resistant (Congenital nephrotic syndrome, nephrotic syndrome with focal segmental glomerulosclerosis)	INS	<p>Children and adults with idiopathic Nephrotic Syndrome (nephrotic range proteinuria and hypoalbuminaemia)</p> <p>Congenital NS (presumed Steroid Resistance)</p> <p>Childhood or adult onset with primary Steroid Resistance</p> <p>Childhood or adult onset with late onset Steroid Resistance</p> <p>Steroid Sensitive Nephrotic Syndrome (full or partial remission in response to steroids)</p> <p>As part of a syndrome e.g. Nail Patella Syndrome and Denys-Drash Syndrome</p> <p>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</p>	<p>Secondary causes of Nephrotic Syndrome</p> <ul style="list-style-type: none"> • 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

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<p style="text-align: center;">Polycystic Kidney Disease - Autosomal Dominant</p>	<p style="text-align: center;">ADPKD</p>	<p>Clinical features of Autosomal Dominant Polycystic Kidney Disease meeting current image based diagnostic criteria</p> <p>Clinical features compatible with ADPKD in the absence of a family history</p> <p>Pathogenic or likely pathogenic PKD1 or PKD2 mutation with or without clinical features</p>	<p>Autosomal dominant polycystic liver disease with no evidence of renal cysts</p>	<p>Date that the clinical diagnosis was first made.</p> <p>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</p>
<p style="text-align: center;">Polycystic Kidney Disease - Autosomal Recessive</p>	<p style="text-align: center;">ARPKD</p>	<p>Autosomal Recessive Polycystic Kidney Disease</p> <p>Congenital Hepatic Fibrosis</p> <p>Caroli Syndrome with kidney malformation or cyst</p>	<p>None stated</p>	<p>Date that clinical diagnosis was first made.</p>

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<p>Pregnancy and Chronic Kidney Disease</p>	<p>Pregnancy</p>	<p>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</p> <p>Pregnancy in all women with renal transplants regardless of function</p> <p>Pregnancy in all women with previous or current lupus nephritis regardless of function</p>	<p>None stated</p>	<p>Date of last menstrual period</p>
<p>Pure Red Cell Aplasia</p>	<p>PRCA</p>	<p>Treatment with any injectable form of erythropoiesis stimulating agent for at least four weeks.</p> <p>Haemoglobin <70 g/l without transfusion or transfusion dependence.</p> <p>Normal leucocyte and platelet count</p> <p>Reticulocyte count < 20.000 / mm³</p> <p>Bone marrow aspirate showing well preserved myeloid and megakaryocyte development, and <5% erythroblasts.</p> <p>Presence of anti-erythropoietin antibodies.</p>	<p>Pre-established PRCA due to myeloproliferative disorder</p>	<p>Date of positive antibody test</p>

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Retroperitoneal Fibrosis	Retroperitoneal Fibrosis	<p>Any radiologically confirmed retroperitoneal fibrosis (RPF), presumed to be 'idiopathic' or associated with primary conditions including (but not exclusively):</p> <ul style="list-style-type: none"> • Aortitis • Periaortitis • IgG4-related Vasculitis • Perivascular fibrosis • Atherosclerotic or aneurysmal disease <p>Note: There is no specific ICD code for retroperitoneal fibrosis although the diagnosis term links to two ICD codes:</p> <ul style="list-style-type: none"> • 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
Tuberous Sclerosis	Tuberous Sclerosis	<p>Clinical or molecular diagnosis of Tuberous Sclerosis Complex (TSC)</p> <p>Multiple renal angiomyolipomas</p> <p>Multiple renal angiomyolipomas (> 3) +/- pulmonary lymphangiomyomatosis (LAM) without other signs of TSC</p>	None stated	Date that clinical diagnosis was first made

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Vasculitis (Primary systemic Vasculitis)	Vasculitis	<p>Small vessel Vasculitis (ANCA associated)</p> <p>Microscopic polyangiitis (including renal limited Vasculitis)</p> <p>Granulomatosis with polyangiitis (Wegener)</p> <p>Eosinophilic granulomatosis with polyangiitis (Churg Strauss)</p> <p>ANCA Vasculitis unclassified</p> <p>Small vessel Vasculitis (Immune complex)</p> <p>anti-GBM disease</p> <p>Cryoglobulinemic Vasculitis</p> <p>IgA Vasculitis (Henoch-Schönlein)</p> <p>Medium vessel Vasculitis</p> <p>Classical PAN</p> <p>Kawasaki disease</p> <p>Large vessel Vasculitis</p> <p>Giant cell arteritis</p> <p>Takayasu's arteritis</p>	None stated	<p>Date of biopsy.</p> <p>In the absence of a biopsy, the date of a positive antibody test should be used</p>

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<p style="text-align: center;">Vasculitis (Primary systemic Vasculitis)</p>	<p style="text-align: center;">Vasculitis</p>	<p>Variable vessel Vasculitis</p> <p>Behçet's disease</p> <p>Cogan's syndrome</p> <p>Single organ Vasculitis</p> <p>Isolated aortitis</p> <p>Primary cerebral angiitis</p>	<p style="text-align: center;">None stated</p>	<p>Date of biopsy.</p> <p>In the absence of a biopsy, the date of a positive antibody test should be used</p>