

10273 Glomerulopathies

Disease / Orphacode	Autoimmune Tests
C3 glomerulonephritis 329918	<ol style="list-style-type: none"> 1) Serum C3: not tested/normal/low 2) Serum C4: not tested/normal/low/elevated 3) C3 nephritis factor: not tested/negative/positive 4) CFH autoantibodies: not tested/negative/positive
Dense deposit disease 93571	<ol style="list-style-type: none"> 1) Serum C3: not tested/normal/low 2) Serum C4: not tested/normal/low/elevated 3) C3 nephritis factor: not tested/negative/positive 4) CFH autoantibodies: not tested/negative/positive
Primary IgA nephropathy (Berger disease) 9999982	<ol style="list-style-type: none"> 1) Serum C3: not tested/normal/low 2) Serum IgA: not tested/normal/elevated
Immune complex associated membranoproliferative glomerulonephritis, with complement abnormalities 329903	<ol style="list-style-type: none"> 1) Serum C3: not tested/normal/low 2) Serum C4: not tested/normal/low/elevated 3) ANA: not tested/negative/positive 4) cANCA: not tested/negative 5) pANCA: not tested/negative 6) Cryoglobulin: not tested/negative/positive
Immune complex associated membranoproliferative glomerulonephritis, not otherwise specified 9999975	<ol style="list-style-type: none"> 1) Serum C3: not tested/normal/low 2) Serum C4: not tested/normal/low/elevated 3) ANA: not tested/negative/positive 4) cANCA: not tested/negative 5) pANCA: not tested/negative 6) Cryoglobulin: not tested/negative/positive
Membranous nephropathy 97560	<ol style="list-style-type: none"> 1) Serum C3: not tested/normal/low 2) Serum C4: not tested/normal/low/elevated 3) ANA: not tested/negative/positive 4) PLA2 antibodies: not tested/negative/positive 5) CFH autoantibodies: not tested/negative
Lupus like glomerulopathy (‘full house’ nephropathy) 567544	<ol style="list-style-type: none"> 1) Serum C3: not tested/normal/low 2) Serum C4: not tested/normal/low/elevated 3) dsDNA: not tested/negative/positive 4) ANA: not tested/negative/positive
Adult-onset Still disease 829	<ol style="list-style-type: none"> 1) ANA: not tested/negative/positive 2) RF: not tested/negative/positive
Buerger disease 36258	<ol style="list-style-type: none"> 1) ANA: not tested/negative/positive 2) Anti-beta2 Glycoprotein IgG: not tested/negative/positive 3) Anti-beta2 Glycoprotein IgM: not tested/negative/positive 4) Lupus anticoagulant: not tested/negative/positive 5) Anti-phospholipid/Anti-cardiolipin: not tested /negative/positive 6) RF: not tested /negative/positive 7) ACA: not tested /negative/positive 8) Anti-scl70: not tested/negative/positive

10273 Glomerulopathies

Dermatomyositis 221	<ol style="list-style-type: none"> 1) ANA: not tested/negative/positive 2) Anti-Mi-2: not tested/negative/positive 3) Anti-Jo-1: not tested/negative/positive
Juvenile dermatomyositis 93672	<ol style="list-style-type: none"> 1) Anti-Mi-2: not tested/negative/positive 2) Anti-Jo-1: not tested/negative/positive
Polymyositis 732	<ol style="list-style-type: none"> 1) Anti-Jo-1: not tested/negative/positive
Juvenile polymyositis 93568	<ol style="list-style-type: none"> 1) Anti-Jo-1: not tested/negative/positive
Systemic sclerosis 90291	<ol style="list-style-type: none"> 1) ANA: not tested/negative/positive 2) ACA: not tested/negative/positive 3) Anti-scl70: not tested/negative/positive 4) Anti PM-Scl: not tested/negative/positive 5) Anti-RNA polymerase III: not tested/negative/positive
Systemic Lupus Erytematosus 93552	<ol style="list-style-type: none"> 1) Serum C3: not tested/normal/low 2) Serum C4: not tested/normal/low/elevated 3) dsDNA: not tested/negative/positive 4) ANA: not tested/negative/positive 5) Anti-beta2 Glycoprotein IgG: not tested/negative/positive 6) Anti-beta2 Glycoprotein IgM: not tested/negative/positive 7) Lupus anticoagulant: not tested/negative/positive 8) Anti-Ro (SSA): not tested/negative/positive 9) Anti-RNP: not tested/negative/positive 10) Anti-phospholipid/Anti-cardiolipin: not tested/negative/positive 11) CFH autoantibodies: not tested/negative/positive
Pediatric systemic lupus erythematosus 93552	<ol style="list-style-type: none"> 1) Serum C3: not tested/normal/low 2) Serum C4: not tested/normal/low/elevated 3) dsDNA: not tested/negative/positive 4) ANA: not tested/negative/positive 5) Anti-beta2 Glycoprotein IgG: not tested/negative/positive 6) Anti-beta2 Glycoprotein IgM: not tested/negative/positive 7) Lupus anticoagulant: not tested/negative/positive 8) Anti-Ro (SSA): not tested/negative/positive 9) Anti-RNP: not tested/negative/positive 10) Anti-phospholipid/Anti-cardiolipin: not tested/negative/positive 11) CFH autoantibodies: not tested/negative/positive
Reynolds syndrome 779	<ol style="list-style-type: none"> 1) ANA: not tested/negative/positive 2) RF: not tested/negative/positive 3) ACA: not tested/negative/positive 4) Anti-Scl70: not tested/negative/positive 5) Anti-SM: not tested/negative/positive 6) Cryoglobulin: not tested/negative/positive
C3 glomerulopathy secondary to MGRS 999970	<ol style="list-style-type: none"> 1) Serum C3: not tested/normal/low

10273 Glomerulopathies

Anti-glomerular basement membrane disease 375	<ol style="list-style-type: none"> 1) cANA: not tested/negative/positive 2) pANCA: not tested/negative/positive 3) Myeloperoxidase-ANCA: not tested/negative/positive 4) Antiproteinase 3-ANCA: not tested/negative/positive 5) Anti-GBM: not tested/negative/positive
Immunoglobulin A vasculitis 761	<ol style="list-style-type: none"> 1) Serum IgA: not tested/negative/positive
Granulomatosis with polyangiitis (Wegener) 900	<ol style="list-style-type: none"> 1) cANA: not tested/negative/positive 2) pANCA: not tested/negative/positive 3) Myeloperoxidase-ANCA: not tested/negative/positive 4) Antiproteinase 3-ANCA: not tested/negative/positive 5) Anti-GBM: not tested/negative/positive
Eosinophilic granulomatosis with polyangiitis 183	<ol style="list-style-type: none"> 1) cANA: not tested/negative/positive 2) pANCA: not tested/negative/positive 3) Myeloperoxidase-ANCA: not tested/negative/positive 4) Antiproteinase 3-ANCA: not tested/negative/positive
Microscopic polyangiitis 727	<ol style="list-style-type: none"> 1) cANA: not tested/negative/positive 2) pANCA: not tested/negative/positive 3) Myeloperoxidase-ANCA: not tested/negative/positive 4) Antiproteinase 3-ANCA: not tested/negative/positive 5) Anti-GBM: not tested/negative/positive
Pauci-immune glomerulonephritis 93126	<ol style="list-style-type: none"> 1) cANA: not tested/negative/positive 2) pANCA: not tested/negative/positive 3) Myeloperoxidase-ANCA: not tested/negative/positive 4) Antiproteinase 3-ANCA: not tested/negative/positive 5) Anti-GBM: not tested/negative/positive
Polyarteritis nodosa 767	<ol style="list-style-type: none"> 1) Serum C3: not tested/normal/low 2) Serum C4: not tested/normal/low/elevated 3) ANA: not tested/negative/positive 4) Anti-beta2 Glycoprotein IgG: not tested/negative/positive 5) Anti-beta2 Glycoprotein IgM: not tested/negative/positive 6) Lupus anticoagulant: not tested/negative/positive 7) Anti-Ro (SSA): not tested/negative/positive 8) Anti-CCP: not tested/negative/positive 9) RF: Anti-Ro (SSA): not tested/negative/positive 10) pANCA: not tested/negative/positive 11) Anti-GBM not tested/negative/positive
Behcet disease 117	<ol style="list-style-type: none"> 1) Anti-beta2 Glycoprotein IgG: not tested/negative/positive 2) Anti-beta2 Glycoprotein IgM: not tested/negative/positive 3) Lupus anticoagulant: not tested/negative/positive

93609 Tubulopathies

Disease / Orphacode	Autoimmune Tests
Auto-immune distal renal tubular acidosis 93608	1) Serum C3: not tested/normal/low 2) Serum C4: not tested/normal/low/elevated 3) dsDNA: not tested/negative/positive 4) ANA: not tested/negative/positive 5) Anti-Ro (SSA): not tested/negative/positive 6) Anti-RNP: not tested/negative/positive 7) Antiphospholipid antibodies: not tested/negative/positive
Sjögren disease 289390	1) Serum C3: not tested/normal/low 2) Serum C4: not tested/normal/low/elevated 3) ANA: not tested/negative/positive 4) Anti-Ro (SSA): not tested/negative/positive 5) Anti-RNP: not tested/negative/positive 6) Antiphospholipid antibodies: not tested/negative/positive
TINU Syndrome 91500	1) ANA: not tested/negative/positive 2) RF: not tested/negative/positive 3) pANCA: ANA: not tested/negative/positive

93573 Thrombotic microangiopathy

Disease / Orphacode	Autoimmune Tests
Infection-related hemolytic uremic syndrome 54448	1) Serum C3: not tested/normal/low 2) Serum C4: not tested/normal/low/elevated 3) CFH autoantibodies: not tested/negative/positive 4) ADAMTS13 activity: not tested/10-100% 5) Shigatoxin PCR: not tested/negative/positive 6) STEC stool culture: not tested/negative/positive 7) STEC serum antibodies: not tested/negative/positive
Atypical HUS 2134	1) Serum C3: not tested/normal/low 2) Serum C4: not tested/normal/low/elevated 3) CFH autoantibodies: not tested/negative/positive 4) ADAMTS13 activity: not tested/10-100% 5) Shigatoxin PCR: not tested/negative/positive 6) STEC stool culture: not tested/negative/positive 7) STEC serum antibodies: not tested/negative/positive
Methylmalonic acidemia with homocystinuria, type cbIC 79282	1) Serum C3: not tested/normal/low 2) Serum C4: not tested/normal/low/elevated 3) CFH autoantibodies: not tested/negative/positive 4) ADAMTS13 activity: not tested/10-100% 5) Shigatoxin PCR: not tested/negative/positive 6) STEC stool culture: not tested/negative/positive 7) STEC serum antibodies: not tested/negative/positive 8) Homocysteine: not tested/negative/positive
Hemolytic uremic syndrome with DGKE deficiency 357008	1) Serum C3: not tested/normal/low 2) Serum C4: not tested/normal/low/elevated 3) CFH autoantibodies: not tested/negative/positive 4) ADAMTS13 activity: not tested/10-100% 5) Shigatoxin PCR: not tested/negative/positive 6) STEC stool culture: not tested/negative/positive 7) STEC serum antibodies: not tested/negative/positive
Thrombotic thrombocytopenic purpura 54057	1) Serum C3: not tested/normal/low 2) Serum C4: not tested/normal/low/elevated 3) CFH autoantibodies: not tested/negative/positive 4) ADAMTS13 activity: not tested/<10% 5) Shigatoxin PCR: not tested/negative/positive 6) STEC stool culture: not tested/negative/positive 7) STEC serum antibodies: not tested/negative/positive
De novo thrombotic microangiopathy after kidney transplantation 244275	1) Serum C3: not tested/normal/low 2) Serum C4: not tested/normal/low/elevated 3) CFH autoantibodies: not tested/negative/positive 4) ADAMTS13 activity: not tested/<10% 5) Shigatoxin PCR: not tested/negative/positive 6) STEC stool culture: not tested/negative/positive 7) STEC serum antibodies: not tested/negative/positive

The last update Version was performed on 16.02.2021