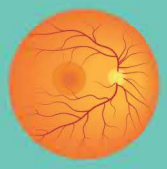
C3 Glomerulopathy

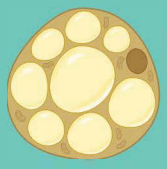
ISN INTERNATIONAL SOCIETY OF NEPHROLOGY

Presentation

Proteinuria/ Microscopic hematuria



Retinal drusen



Lipodystrophy

Histology

Mesangial proliferative, membrano-proliferative, endocapillary proliferative pattern with or without crescents

Dominant C3 deposition ≥2 orders of magnitude over other immune reactants

C3GN: Amorphous mesangial ± subendothelial and subepithelial deposits

DDD: Dense osmiophilic mesangial and intramembranous deposits

Treatment

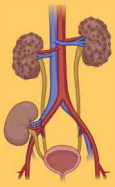
Optimal blood pressure control with ACE inhibitors/ARBs

Corticosteroids and mycophenolate mofetil in moderate to severe disease

Eculizumab - doubtful role

Natural Course

30-50% of adults reach kidney failure within 10 years of diagnosis



Allograft failure within 10 years in 50% of recurrences

Histological recurrence as high as 90%

ACEi: Angiotensin Converting Enzyme Inhibitor; **ARB**: Angiotensin Receptor Blocker; **C3GN**: C3 Glomerulonephritis; **DDD**: Dense Deposit Disease

•Goodship, Timothy H J et al. "Atypical hemolytic uremic syndrome and C3 glomerulopathy: conclusions from a "KDIGO Controversies Conference." *Kidney intl* vol. 91,3 (2017): 539-551. doi:10.1016/j.kint.2016.10.005
•KDIGO Glomerular Diseases Work Group. "KDIGO 2021 Clinical Practice Guideline for the Management of Glomerular Diseases." *Kidney intl* vol. 100,4S (2021): S1-S276. doi:10.1016/j.kint.2021.05.021

For funding and support information, see: https://www.theisn.org/initiatives/toolkits/complement-mediated-kidney-disease-toolkit/#Support