

# C3G IS AN ULTRA-RARE KIDNEY DISEASE THAT CAN RESULT IN TRANSPLANT OR DIALYSIS<sup>1-5</sup>

## COMPLEMENT-MEDIATED DISEASE



Dysregulation of the **alternative complement pathway** is the **primary driver of C3G**<sup>3,6</sup>

C3 deposits in the glomerular mesangium and along capillary walls can cause inflammation and injury<sup>7,8</sup>

## MAY PROGRESS RAPIDLY



Patients can be diagnosed with 1 of 2 subtypes of C3G—DDD or C3GN<sup>9</sup>

✓ DDD is usually associated with pediatric cases and may progress more quickly than C3GN

Regardless of subtype, ~50% of patients progress to kidney failure within 10 years<sup>4</sup>

## HETEROGENOUS IN CLINICAL PRESENTATION AND PROGNOSIS<sup>1,2,5,9</sup>



Patients may present with low to heavy levels of proteinuria and hematuria

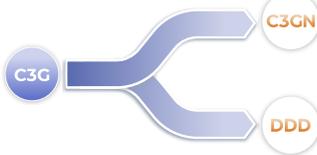
C3G can be acute, recurrent, or rapidly progressive



C3, complement 3; C3G, complement 3 glomerulopathy; C3GN, complement 3 glomerulonephritis; DDD, dense deposit disease.

Patient portrayal.

# ACCURATE CLASSIFICATION AND CODING OF C3G MAY BE IMPORTANT IN THE MANAGEMENT OF YOUR PATIENTS<sup>10</sup>



**C3G** and its subtypes were previously classified as MPGN until being reclassified as C3GN and DDD in the latest ICD-10-CM codes.

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### DDD and recently added C3GN ICD-10-CM codes

Code	Description
N00.A	Acute nephritic syndrome with C3GN
N00.6	Acute nephritic syndrome with DDD
N01.A	Rapidly progressive nephritic syndrome with C3GN
N01.6	Rapidly progressive nephritic syndrome with DDD
N02.A	Recurrent and persistent hematuria with C3GN
N02.6	Recurrent and persistent hematuria with DDD
N03.A	Chronic nephritic syndrome with C3GN
N03.6	Chronic nephritic syndrome with DDD
N04.A	Nephrotic syndrome with C3GN
N04.6	Nephrotic syndrome with DDD
N05.A	Unspecified nephritic syndrome with C3GN
N05.6	Unspecified nephritic syndrome with DDD
N06.A	Isolated proteinuria with C3GN
N06.6	Isolated proteinuria with DDD
N07.A	Hereditary nephropathy, not elsewhere classified with C3GN
N07.6	Hereditary nephropathy, not elsewhere classified with DDD

C3G

Previously classified as MPGN, **C3G** was recently reclassified as its own disease state to better acknowledge the causative factors of the disease.<sup>8</sup>

C3G, complement 3 glomerulopathy; C3GN, complement 3 glomerulonephritis; DDD, dense deposit disease; GN, glomerulonephritis; ICD-10-CM, International Classification of Diseases, Tenth Revision, Clinical Modification; MPGN, membranoproliferative glomerulonephritis.

**References:** 1. Schena FP, Esposito P, Rossini M. A narrative review on C3 glomerulopathy: a rare renal disease. *Int J Mol Sci.* 2020;21(2):525. doi:10.3390/ijms21020525 2. Smith RJH, Alexander J, Barlow PN, et al. New approaches to the treatment of dense deposit disease. *J Am Soc Nephrol.* 2007;18:2447-2456. doi:10.1681/ASN.2007030356 3. C3 glomerulopathy: dense deposit disease and C3 glomerulonephritis. National Organization for Rare Disorders (NORD). Accessed September 24, 2022. <https://rarediseases.org/rare-diseases/c3-glomerulopathy-dense-deposit-disease-and-c3-glomerulonephritis/> 4. Martin B, Smith RJH. C3 glomerulopathy. In: Adam MP, Mirzaa GM, Pagon RA, et al, eds. *GeneReviews*<sup>®</sup> [Internet]. University of Washington; 2007. Updated April 5, 2018. Accessed July 28, 2022. <https://www.ncbi.nlm.nih.gov/books/NBK1425/> 5. Medjeral-Thomas NR, O'Shaughnessy MM, O'Regan JA, et al. C3 glomerulopathy: clinicopathologic features and predictors of outcome. *Clin J Am Soc Nephrol.* 2014;9(1):46-53. doi:10.2215/CJN.04700513 6. Caravaca-Fontán F, Lucientes L, Caverio T, Praga M. Update on C3 glomerulopathy: a complement-mediated disease. *Nephron.* 2020;144(6):272-280. doi:10.1159/000507254 7. Willows JW, Brown M, Sheerin NS. The role of complement in kidney disease. *Clin Med (London).* 2020;20(2):156-160. doi:10.7861/clinmed.2019-0452 8. Sethi S, Vriese ASD, Fervenza FC. Acute glomerulonephritis. *Lancet.* 2022;399:1646-1663. doi:10.1016/S0140-6736(22)00461-5 9. Smith RJH, Appel GB, Blom AM, et al. C3 glomerulopathy - understanding a rare complement-driven renal disease. *Nat Rev Nephrol.* 2019;15(3):129-143. doi:10.1038/s41581-018-0107-2 10. Centers for Disease Control and Prevention. ICD10 Coordination and Maintenance Committee Meeting Diagnosis Agenda. March 5-6, 2019, Part 2. Accessed October 12, 2023. [https://www.cdc.gov/nchs/icd/icd10cm\\_maintenance.htm](https://www.cdc.gov/nchs/icd/icd10cm_maintenance.htm)