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Challenge Your Thinking

Epidemiology of C3G and primary IC-MPGN: A systematic literature review and meta-analysis

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CONCLUSIONS

- This systematic literature review and meta-analysis indicates C3G and primary IC-MPGN have similar epidemiological profiles
- Based on data from Europe, Asia and South America, pooled estimated **prevalence** was **17.6–17.8 per million population (pmp)** and **incidence** was **0.4–0.5 cases per 10^6 person-years (PY)**
- Correcting for disease-specific mortality, which addresses the limitations of previously calculated estimates, reduced prevalence of both conditions to ~12.5 pmp
- These data support that C3G and primary IC-MPGN are **ultra-rare (or ultra-orphan) diseases**, and emphasise the unmet need for **disease-modifying targeted therapies** that can effectively prevent renal damage and reduce the risk of kidney failure in patients with C3G or primary IC-MPGN

INTRODUCTION

- C3G and primary IC-MPGN are heterogeneous, rare, complement-mediated kidney diseases[1,2]
- Up to 50% of individuals with C3G or primary IC-MPGN progress to irreversible kidney failure within a decade of diagnosis, requiring dialysis or transplantation.[1,3] Most people (~89%) experience disease recurrence after transplantation, with allograft loss occurring frequently[2,4]
- The poor long-term outcomes and disease-related complications of C3G and primary IC-MPGN create substantial clinical, economic and healthcare burdens[5]
- Robust epidemiological data are essential for informing healthcare planning, resource allocation and public health policy making[6]

AIM

- To conduct an SLR on the epidemiology of C3G and primary IC-MPGN and synthesise the data using a frequentist meta-analytic approach

METHODS

Systematic literature review

- PubMed, Embase and Cochrane databases were searched to identify real-world studies reporting prevalent or incident cases of C3G and/or primary IC-MPGN between 01 January 2010 and 30 June 2025. This analysis reflects an updated search conducted after the ERA 2026 abstract submission
- Studies were excluded if they described secondary IC-MPGN, case reports, or case series without a definable denominator
- Data extracted from eligible studies included number of cases, referral population size, study duration and reported study-level incidence/prevalence
- Quality assessment was performed using STROBE criteria[7]

Meta-analysis

- All prevalence and incidence estimates were harmonised to common units of pmp and cases per 10^6 PY, respectively
- A frequentist meta-analysis was conducted using a restricted maximum likelihood (REML)-weighted random-effects model; pooled estimates were reported with 95% CIs
- Mortality-adjusted prevalence was calculated by applying a pooled estimated correction factor to the unadjusted prevalence estimates
- Heterogeneity was assessed using the I^2 statistic, Cochran's Q and the between-study variance τ^2; publication bias was evaluated using Egger's regression test

Sensitivity analyses

- To determine the influence of: (1) excluding individual studies; and (2) excluding either adult or paediatric age cohort studies

RESULTS

Study characteristics

- The searches identified 7727 records; after removing duplicates, 5615 records underwent title/abstract screening and 155 records assessed by full-text review. Overall, 11 studies reporting data for 32–620 patients from Europe, Asia and South America were included in the meta-analysis; 8/11 studies met ≥80% of STROBE checklist items

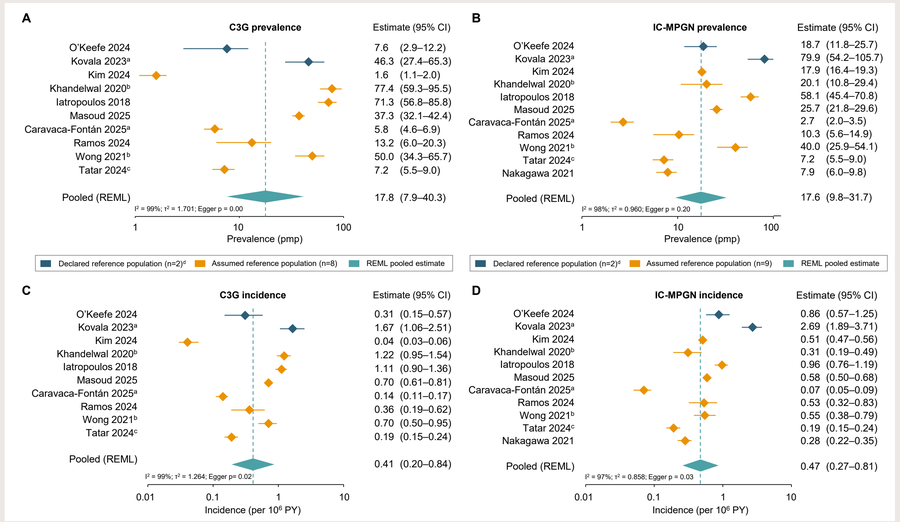
Meta-analysis

- Pooled estimated prevalence rates (95% CI) for C3G and primary IC-MPGN were 17.8 (7.9–40.3) and 17.6 (9.8–31.7) pmp, respectively (**Figure 1 A & B**). Incidence rates (95% CI) were 0.41 (0.20–0.84) and 0.47 (0.27–0.81) per 10^6 PY, respectively (**Figure 1 C & D**)
- Substantial heterogeneity in prevalence and incidence rates was observed between studies (I^2, 97–99%; Cochran's Q, 399–956 [p<0.001 for all]; τ^2, 0.86–1.70)
- Disease-specific mortality, derived from five studies, estimated a pooled correction factor of 0.71 (95% CI 0.55–0.88) with no heterogeneity. The mortality-adjusted prevalence rates (95% CI) for C3G and primary IC-MPGN were 12.7 (5.4–29.7) and 12.6 (6.7–23.7) pmp, respectively, corresponding to ~30% lower than the unadjusted rates

Sensitivity analyses

- Individual studies generally had minimal influence on pooled estimates. The change in prevalence and incidence by excluding individual studies ranged from −2.7 to 5.7 pmp and −0.06 to 0.12 per 10^6 PY, respectively
- Excluding the two paediatric studies reduced pooled estimates of C3G prevalence (Δ −4.7 pmp) and incidence (Δ −0.07 per 10^6 PY). Excluding the two adult studies minimally decreased C3G prevalence (Δ −0.40 pmp) and incidence (Δ −0.01 per 10^6 PY). Excluding studies based on age subgroups had a relatively similar impact on primary IC-MPGN prevalence and incidence

Figure 1. Forest plots of prevalence (A & B) and incidence (C & D) for C3G and primary IC-MPGN



Data reported on log scale with 95% CI error bars. Dashed line represents pooled mean estimates.
[a]Adult population. [b]Paediatric population. [c]Tatar 2024 reported 66 patients with primary MPGN as a combined cohort without subtype split; the meta-analysis applied the same case count to both C3G and primary IC-MPGN. [d]Only 2/11 studies (O'Keefe and Kovala) explicitly declared their reference populations; the remaining 9 required assumptions about catchment coverage over study periods of up to 32 years.
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DISCLOSURES

FC-F: reports consultancy and/or honoraria from Alexion, Apellis, AstraZeneca, Bayer, Novartis, Otsuka, STADA and Sobi. **CR**, **VC** and **JN**: employees of Sobi and may hold stock or stock options. **CDP**: is employed by Digital Health Labs, which received funding to conduct analyses for this study. **DPG**: is supported by the St Peter's Trust for Kidney, Bladder and Prostate research and has consulted or presented for Alexion, Apellis, Bayer, Boehringer Ingelheim, CSL Vifor, GSK, Kyowa Kirin, Novartis, Otsuka, Sanofi and Sobi.

ABBREVIATIONS

C3G, C3 glomerulopathy, CI, confidence interval; ERA, European Renal Association; IC-MPGN, immune-complex membranoproliferative glomerulonephritis; pmp, per million population; PY, person-years; REML, restricted maximum likelihood; SLR, systematic literature review; STROBE, Strengthening the Reporting of Observational Studies in Epidemiology.

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