IgA Nephropathy & IgA Vasculitis

Diagnosis, risk stratification, IgAN-specific drug therapy, special situations, IgA vasculitis nephritis, and pediatric management.

2025 KDIGO Clinical Practice Guideline
Management of IgA Nephropathy (IgAN) and IgA Vasculitis (IgAV)
KDIGO IgAN/IgAV Work Group · Kidney Int 2025;108(4S):S1–S71

FORMAT *Condensed from recommendations + practice points across Chapters 1 (IgAN) and 1.7–1.10 (IgAV).*

1 Diagnosis & Initial Assessment

–	GUIDANCE
Biopsy	Required — no validated serum/urine biomarker for diagnosis. Consider in adults with proteinuria ≥0.5 g/d (or equivalent) where IgAN is possible and biopsy not contraindicated
Secondary causes	Rule out before labeling idiopathic: IgA vasculitis, HIV/hepatitis, IBD, autoimmune disease, liver cirrhosis, infection-related GN
MEST-C score	Score every primary IgAN biopsy per revised Oxford Classification (M, E, S, T, C)
Registry & support	Enroll in a disease registry if available; provide patient advocacy/education resources

2 Prognosis & Risk Stratification

–	GUIDANCE
Prediction tool	International IgAN Prediction Tool (at-biopsy or 1–2y post-biopsy; adult and pediatric versions) quantifies 7-y risk of 50% eGFR decline/kidney failure — use to inform shared decision-making
Biomarkers	No validated prognostic biomarker beyond eGFR and proteinuria
Tool limits	Not validated to select a specific treatment regimen or to determine prognosis in IgAV

3 At-Risk Definition & Treatment Goals

–	GUIDANCE
At risk	Proteinuria ≥0.5 g/d (or equivalent) while on or off treatment → treatment or additional treatment should be considered
Target	Reduce loss of kidney function toward physiologic rate (<1 mL/min/yr); keep proteinuria <0.5 g/d, ideally <0.3 g/d
Lifestyle	Sodium <2 g/d; smoking/vaping cessation; weight control; endurance exercise
BP target	≤120/70 mm Hg
Clinical trial	Always consider enrollment given expanding treatment options

4 IgAN-Specific Drug Therapy

–	GUIDANCE
RASi	Optimized maximally tolerated ACEi or ARB in all patients with IgAN (1B)
Nefecon	9-month course suggested for at-risk patients (2B) — targeted-release budesonide; response may not be sustained; approval/availability varies globally
Glucocorticoids	If Nefecon unavailable: reduced-dose regimen + antimicrobial prophylaxis (2B) — methylprednisolone 0.4 mg/kg/d (max 32 mg/d) ×2mo, taper 4 mg/mo, total 6–9mo. Higher toxicity risk: eGFR <30, diabetes, obesity, latent infection, active PUD, uncontrolled psychiatric illness, osteoporosis, cataracts
Sparsentan	Suggested for at-risk patients (2B) — DEARA; do not combine with a RASi (already combines both mechanisms)
SGLT2i	Suggested for at-risk patients (2B) — uncertain value in younger patients with eGFR >60

5 Other Agents — Not Recommended

–	GUIDANCE
No evidence	Antiplatelets, anticoagulants, azathioprine, calcineurin inhibitors, rituximab — not recommended. Cyclophosphamide not recommended unless rapidly progressive IgAN
MMF	Chinese patients: may use as glucocorticoid-sparing agent — 3 RCTs show benefit. Non-Chinese: insufficient evidence for monotherapy
Hydroxychloroquine	Chinese patients: consider if high risk despite optimized RASi. Not evaluated in non-Chinese patients
Tonsillectomy	Recommended in Japanese guidelines; should not be performed for IgAN in non-Japanese patients

6 Special Situations in IgAN

–	GUIDANCE
Nephrotic syndrome	Rare — if biopsy shows MCD-like features, treat per KDIGO 2021 Glomerular Diseases MCD chapter; if mesangioproliferative features coexist, manage as at-risk IgAN
AKI	With visible hematuria (often post-URI): supportive care; rebiopsy if no improvement 2wk after hematuria resolves
RPGN	≥50% eGFR decline in ≤3mo after excluding AAV/anti-GBM/reversible causes → biopsy → treat with cyclophosphamide + glucocorticoids per KDIGO 2024 ANCA guideline. Insufficient evidence for rituximab
Pregnancy planning	Preconception counseling for all women of childbearing potential — stop RASi/SGLT2i/sparsentan/Nefecon/glucocorticoids before conception; enalapril if RASi needed while breastfeeding

7 IgA Vasculitis (IgAV) — Nephritis

–	GUIDANCE
Diagnosis	No agreed adult diagnostic criteria; kidney biopsy required for IgAVN. Biopsy if proteinuria ≥0.5 g/d >4wk, kidney impairment, or RPGN. Screen for secondary causes/malignancy
Prognosis	Uncontrolled HTN and proteinuria (presentation and follow-up) predict poor outcome; E1 lesion predicts initial improvement then decline
Isolated extrarenal	Do NOT use glucocorticoids to prevent nephritis (1B)
At-risk IgAVN	No proven anti-IgA-IC drug (unlike IgAN); BP ≤120/70 with RASi first-line ± SGLT2i; glucocorticoids only after shared decision-making, reduced-dose + antimicrobial prophylaxis if used
RPGN	Treat per KDIGO 2024 ANCA guideline; consider plasma exchange for life/organ-threatening extrarenal (pulmonary, GI, skin) involvement

8 Pediatric IgAN & IgAV (<18y)

–	GUIDANCE
IgAN biopsy	Perform if persistent/recurrent hematuria + PCR >500 mg/g (×2, 1–2wk apart) or nephrotic-range proteinuria/reduced eGFR; consider if PCR 200–500 mg/g (×3)
IgAN treatment	RAS blockade + sodium <3–5 g/d + BP <90th percentile if proteinuria >200 mg/d. Steroids if PCR 500–1000 despite 3–6mo RASi, PCR >1000 despite 4wk RASi, or active MEST-C; IV methylprednisolone pulses for higher-risk/crescentic (C2) disease
IgAN follow-up	Target proteinuria ≤200 mg/d, BP <90th percentile; lifelong yearly monitoring — relapse possible after years
IgAV/IgAVN	Urinary monitoring monthly ≥6mo from presentation; biopsy if PCR 1000–2000 mg/g (2–4wk) or 200–500 mg/g (>4wk); ACEi/ARB if proteinuria >3mo; steroids if nephrotic-range or RPGN with ISKDC ≥II; monitor ≥5y

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