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Diagnosis of IgA vasculitis-associated nephritis (IgAVN)

IgAVN can only be diagnosed with a kidney biopsy, which should be performed in adults with suspected IgAV if there are signs of significant end-organ tissue damage, proteinuria ≥ 0.5 g/d persistent for >4 weeks, kidney function impairment, or

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Prognosis

Limited registry data have identified uncontrolled hypertension and the amount of proteinuria at presentation, and hypertension and mean proteinuria during follow-up as predictors of a poor kidney outcome in adults with IgAV. The Oxford Classification MEST-C score E1 lesions were strongly associated with initial improvement followed by progressive decline in kidney function in patients treated with immunosuppression. The International IgAN Prediction Tools are not designed to determine the prognosis of IgAVN.

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Prevention of IgAVN

Systemic glucocorticoids should not be used to prevent nephritis in patients with isolated extrarenal IgAV.

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Focus of the management of patients with IgAN who are at risk of progressive loss of kidney function

Proteinuria ≥ 0.5 g/d (while on or off treatment) identifies a patient with IgAVN at increased risk of progressive loss of kidney function. The focus of the management in most patients should be to *simultaneously*: i) prevent or reduce IgA-IC formation and IgA-IC-mediated glomerular injury (whether this requires lifelong or intermittent therapy is currently unknown) and ii) manage the consequences of existing IgAN-induced nephron loss (likely lifelong).

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Prevention of IgA-IC mediated injury

Prevention of IgA-IC-mediated injury should incorporate treatments with proven anti-inflammatory effects and ideally should be used in combination with treatments that prevent or reduce IgA-IC formation. In patients who wish to try systemic glucocorticoids after a discussion of the risks and benefits of each drug, a reduced-dose regimen as described for IgAN should be employed with antimicrobial prophylaxis.

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Managing the consequences of IgAVN-induced nephron loss

All patients should receive extensive lifestyle advice on smoking and vaping cessation, weight control, dietary sodium restriction (<2 g/d), and regular exercise. Blood pressure should be controlled to a target of $\leq 120/70$ mm Hg using a RASi as the first-choice drug intervention. Measures to reduce glomerular hyperfiltration and the impact of proteinuria on the tubulointerstitium should include use of RASi and SGLT2i, singly or in combination.

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Other considerations in managing IgAVN

All patients should be offered participation in a clinical trial if one is available. There is insufficient evidence to support the use of the Oxford Classification MEST-C score in determining which drug should be commenced in these patients and there is insufficient evidence to base treatment decisions on the presence and number of crescents in the kidney biopsy. The International IgAN Prediction Tools cannot be used to determine the likely impact of any particular treatment regimen. Dynamic assessment of patient risk over time should be performed, as decisions regarding immunosuppression may change.

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Special situations: IgAV with RPGN

Patients with rapidly progressive IgAV should be offered treatment in accordance with the KDIGO 2024 Guideline for ANCA-Associated Vasculitis. There are insufficient data to determine the efficacy of plasma exchange in IgAVN with RPGN.

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Special situations: IgAV in children

The KDIGO 2025 Guideline for the Management of IgAV has harmonized its guidance with that of the 2025 IPNA recommendations available at <https://link.springer.com/article/10.1007/s00467-024-06502-6>

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Research recommendations

Unlike IgAN, there are currently few clinical trials of novel therapies in IgAVN. It is recommended that those agents currently being evaluated in patients with IgAN should also be tested for safety and efficacy in IgAVN in adults and children. In addition, a better understanding of the natural history and pathogenesis of IgAVN is warranted.