IgA Nephropathy (IgAN): "Hot Topics" in Disease Management

Toolkit for Health Care Professionals

This toolkit was developed by Skymedcare in collaboration with Dana Rizk, MD (University of Alabama at Birmingham, Birmingham, AL), Vladimir Tesar, MD (Charles University, Prague, Czech Republic), and Jonathan Barratt, MD (University of Leicester, Leicester, UK). This activity was supported by an independent medical grant from Calliditas Therapeutics. You can access this activity's two patient vignettes and accompanying roundtable discussion at www.my-ime.com.

What is IgAN?[1–3]

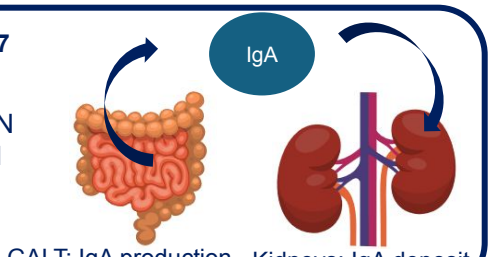
- Most common type of primary glomerulonephritis worldwide
- Up to 75% of patients progress to kidney failure within 20 years
- More prevalent in males than in females
- Incidence peaks between 2nd and 3rd life decade

How does IgAN present?[3,4]

- Highly diverse course
- Ranges from asymptomatic urinary abnormalities to rapidly progressive glomerulonephritis with kidney failure
- Common clinical features: episodes of visible hematuria (often associated with upper respiratory tract infection); proteinuria; hypertension
- Variable progression after diagnosis

Primary IgAN: causes and pathophysiology[3,5–7]

- Genetic and environmental factors influence risk of developing IgAN
- Production of pathogenic IgA forms by the gut-associated lymphoid tissue (GALT) → formation of IgA immune complexes → glomerular IgA accumulation → activation of pro-inflammatory and profibrotic pathways in the kidneys



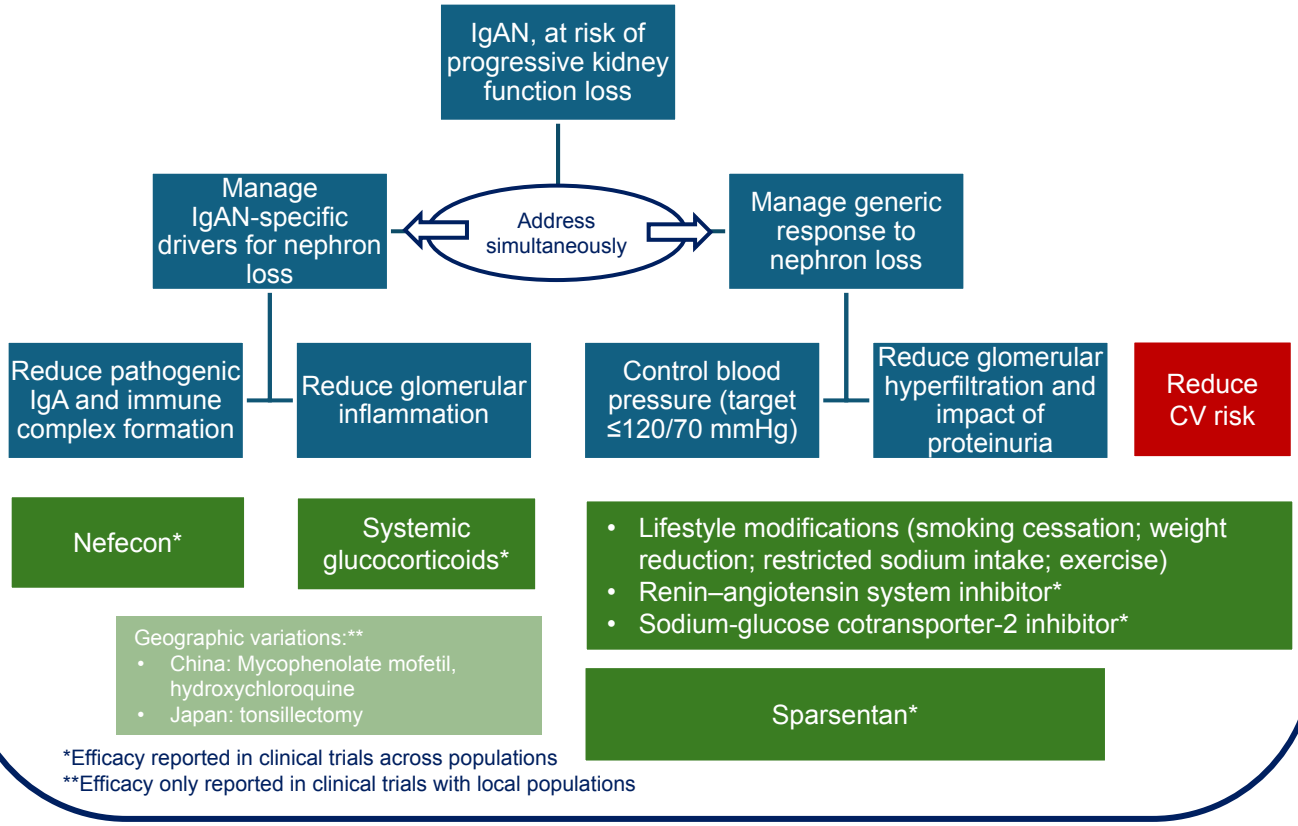
How is IgAN diagnosed?[6,8,9]

- Kidney biopsy is the gold standard for the diagnostic evaluation of glomerular diseases
- A diagnosis of IgAN can only be made by kidney biopsy
- No validated diagnostic serum or urine biomarkers are available
- Early diagnosis and treatment are crucial to prevent progressive nephron loss

Diagnostic process:

1. Perform kidney biopsy in all adults with proteinuria ≥0.5 g/day (or equivalent) who have a suspicion of IgAN and no contraindication for kidney biopsy
2. IgAN is diagnosed: Assess for possible secondary causes (e.g., liver cirrhosis, inflammatory bowel disease)
3. Primary IgAN: Determine the Oxford MEST-C score (mesangial [M] and endocapillary [E] hypercellularity, segmental sclerosis [S], interstitial fibrosis/tubular atrophy [T], crescents [C])
4. Quantify progression risk at diagnosis using the International IgAN Prediction Tool: **https://www.mdcalc.com/calc/10533/international-iga-nephropathy-prediction-tool**

IgAN treatment[6]



*Efficacy reported in clinical trials across populations
**Efficacy only reported in clinical trials with local populations

Rare presentations of IgAN

Nephrotic syndrome:[6,8]
- Edema; hypoalbuminemia; nephrotic-range proteinuria >3.5 g/day
- Treatment follows recommendations for minimal change disease or mesangioproliferative glomerulonephritis

Acute kidney injury (AKI):[6]
- Severe visible hematuria; commonly associated with upper respiratory tract infection
- Consider repeat kidney biopsy in patients without improvement in kidney function within 2 weeks following cessation of hematuria
- Immediate management in case of visible hematuria: supportive care for AKI

Rapidly progressive IgAN:[6,10]
- ≥50% decline in estimated glomerular filtration rate in ≤3 months
- Offer treatment with cyclophosphamide and systemic glucocorticoids

CAVE

When treating women of childbearing age, take contraindications regarding pregnancy and breastfeeding of various drug classes into account![6]

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