

## 1

### Diagnosis of IgA nephropathy (IgAN)

IgAN can only be diagnosed with a kidney biopsy. There are no validated diagnostic serum/urine biomarkers. The differential diagnosis of IgA dominant glomerulonephritis includes primary IgAN, IgA vasculitis, cirrhosis, inflammatory bowel disease and infection-related GN. (Figure 1)

## 2

### Prognosis

The International IgAN Prediction Tool helps determine the risk of a 50% decline in eGFR or progression to kidney failure up to 6.7 years from the time of kidney biopsy to inform shared decision-making with patients (available at QxMD<sup>1</sup>). (Figure 2) There are no validated prognostic serum/urine biomarkers other than eGFR and proteinuria.

## 3

### Treatment for all patients with primary IgAN

Initial management is supportive care including lifestyle modification (smoking cessation, weight control, regular exercise and dietary sodium restriction), blood pressure control and maximum tolerated RAS blockade.

## 4

### Identification of patients at high risk of progression

Assess risk of progression regularly in all patients with IgAN. High risk is currently defined as persistent proteinuria >1 g/d despite 3 months of stable, optimized supportive care. While proteinuria reduction to under 1 g/d is a surrogate marker of improved kidney outcome in IgAN, persistent proteinuria 0.5–1 g/d is also likely to increase the risk of progression. (Figure 3)

## 5

### Management of high-risk patients on optimized supportive care

Neither the International IgAN Prediction Tool nor the Oxford Classification MEST-C score alone can be used to determine the likely impact of any particular treatment regimen. Unless inclusion in a clinical trial is possible, glucocorticoid therapy may reduce the risk of kidney failure in IgAN. If glucocorticoids are considered, the risk of treatment-emergent toxicity, in particular serious infectious complications, must be discussed with the patient, particularly those with an eGFR <50ml/min/1.73 m<sup>2</sup>. (Figure 3)

## 6

### Ethnicity-specific alternative treatment options

There are data to support the use of mycophenolate mofetil (MMF) in Chinese patients as a glucocorticoid-sparing agent. Tonsillectomy is widely performed in Japan with supportive data for Japanese patients. (Figure 3)

## 7

### IgAN variants – rapidly progressive glomerulonephritis

Rarely patients with IgAN can develop a rapidly progressive glomerulonephritis (RPGN) associated with extensive crescent formation. These cases should be treated with cyclophosphamide and glucocorticoids in the same way as ANCA-associated vasculitis. The presence of crescents in a kidney biopsy without a concomitant change in serum creatinine does not constitute a RPGN.

## 8

### IgAN variants – AKI and nephrotic syndrome

Acute kidney injury can complicate episodes of severe visible hematuria. Treatment is supportive. IgAN rarely presents with nephrotic syndrome. In such cases, electron microscopy may be otherwise consistent with minimal change disease (MCD) and patients should be treated in accordance with the guidelines for MCD.

## 9

### IgAN in children

The International IgAN Prediction Tool has been validated for use in children. Management of children with IgAN is similar to adults except for a lower threshold for the early use of glucocorticoids. Long term follow-up is essential, even after complete remission, as children can relapse after many years of stable disease.

## 10

### IgA vasculitis

Diagnosis is often made on clinical criteria alone but a kidney biopsy should always be considered in patients with an RPGN, proteinuria >1 g/d, and/or impaired kidney function. Glucocorticoids should not be used to prevent nephritis in patients with isolated extrarenal IgAV. Management for adults and children should follow the guidance for IgAN.

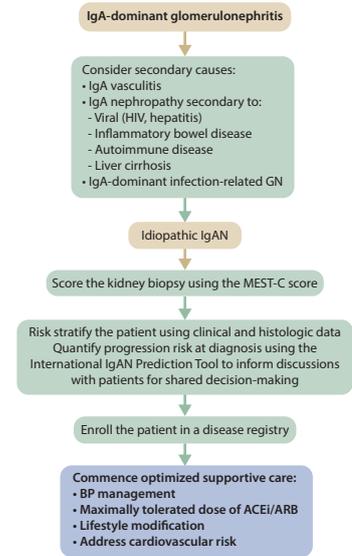


Figure 1

Estimated GFR at biopsy.....ml/min/1.73 m <sup>2</sup>
Systolic blood pressure at biopsy.....mmHg
Diastolic blood pressure at biopsy.....mmHg
Proteinuria at biopsy.....g/day
Age at biopsy.....years
Race Caucasian Chinese Japanese Other
Use of ACE inhibitor or ARB at the time of biopsy No Yes
MEST M-score 0 1
MEST E-score 0 1
MEST S-score 0 1
MEST T-score 0 1 2
Immunosuppression use at or prior to biopsy No Yes

Figure 2

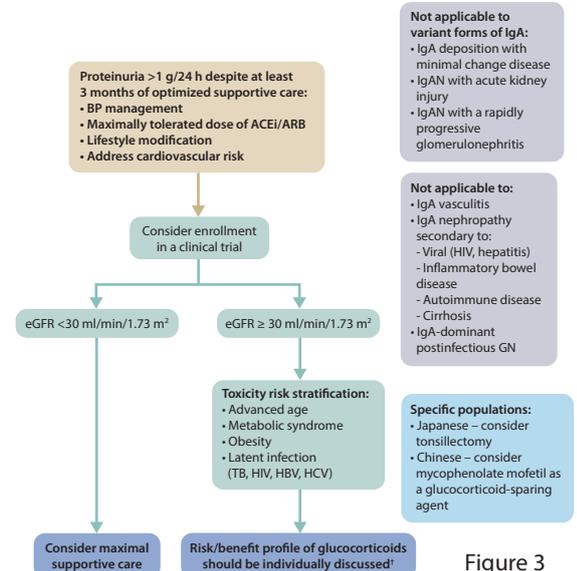


Figure 3

<sup>1</sup> [https://qxmd.com/calculate/calculator\\_499/international-igan-prediction-tool-at-biopsy-adults?branch\\_match\\_id=656546875419766679](https://qxmd.com/calculate/calculator_499/international-igan-prediction-tool-at-biopsy-adults?branch_match_id=656546875419766679)