Clinicopathologic Changes in Children with Immunoglobulin A Nephritis and Henoch–Schnlein Purpura Nephritis after Cyclosporine A and Angiotensin–converting Enzyme Inhibitor Treatment

Purpose: To investigate the clinicopathologic effects of cyclosporine A (CsA) in children with diseases characterized by mesangial immunoglobulin A deposits such as immunoglobulin A nephropathy (IgAN) and Henoch–Schnlein purpura nephritis (HSPN).

Methods: We retrospectively reviewed the clinicopathologic outcomes of 54 children (IgAN, 36; HSPN, 18) treated with CsA. The starting dose of CsA was 5 mg/kg per day, and it was administered in 2 divided doses. The degree of proteinuria and pathologic changes in renal biopsies were evaluated before and after CsA treatment.

Results: The mean protein to creatinine ratio decreased from 3.7±1.5 to 0.6±0.4 (P<0.001), and 32 (59.2%) children achieved complete remission of proteinuria after 1-year CsA treatment. Among the 54 children, 24 maintained normal renal function and 25 exhibited microscopic hematuria or proteinuria at the end of CsA treatment. In the HSPN group, 3 children whose initial biopsies indicated class IIIb disease showed class II disease on follow-up, and the follow-up biopsies of 2 children who had class II disease indicated the same class II disease. In the IgAN group, cortical tubular atrophy occurred in 1 child, and no child with IgAN had cortical interstitial fibrosis or tubular atrophy after 1-year CsA treatment. No significant complications were found in the children treated with CsA.

Conclusion: Our findings indicate that CsA treatment is effective and beneficial...
in reducing massive proteinuria and preventing progression to end-stage renal failure in children with glomerular diseases characterized by IgA deposits, such as IgAN and HSPN, within 1 year of treatment.

**Key words:** IgA deposit, IgA nephropathy, Henoch-Schönlein purpura nephritis, cyclosporine A

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**Introduction**

Both immunoglobulin A nephropathy (IgAN) and Henoch-Schönlein purpura nephritis (HSPN), affecting glomerular function and structures, are pathologically characterized by mesangial deposits of IgA. Among several immunoglobulins, IgA is unique for its capability to form multimers, and circulating IgA-containing immune complexes have been demonstrated in both illnesses related to the deposits of IgA and IgA-containing immune complexes in the glomerular mesangium [1].

Characterized by prominent diffuse mesangial IgA deposits on immunofluorescence microscopy, primary IgA nephropathy was initially thought to be a rare but benign cause of recurrent hematuria [2]. However, the clinical course of IgAN is variable, with some patients having stable renal function over decades and others developing hypertension, nephrotic syndrome, and chronic renal failure [2]. A number of clinical and histological prognostic markers, including impairment of renal function, severe proteinuria, and high blood pressure, predict a poor outcome [3].

HSPN, a leucocytoclastic vasculitis, is caused by mainly IgA1-mediated inflammation of small vessels [4-7]. Although isolated microscopic hematuria is the more frequent clinical presentation of HSPN, nephritic syndrome and acute nephritic syndrome, often with hypertension, are observed in about half of the cases [4-7]. Renal involvement occurs in 30–90% of HSP patients [8]. The severity of renal symptoms at onset is known to be the most prognostic factor for HSP in children; patients presenting with nephrotic-range proteinuria or severe nephritis have the highest risk of unfavorable prognosis [9].

Patients with IgAN or HSPN should be treated properly at an early stage to prevent rapid progress to renal insufficiency. Although the management of the two diseases characterized by IgA deposits still remains elusive, recent studies have suggested that cyclosporin A (CsA) might have a beneficial effect in children with severe IgA deposits [10-15]. However, the number of patients included in those studies was limited, and the glomerular changes in IgAN were categorized according to the Haas classification [13]. Therefore, we aimed to investigate clinicopathologic effects of CsA on children with glomerular IgA deposits by histologically classifying HSPN according to the International Study of Kidney Disease in Childhood and IgAN according to the Oxford classification instead of the Haas classification.

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**Materials and Methods**

1. **Patients**

We retrospectively reviewed a total of 54 patients (IgAN:HSPN=36:18; M:F=32:22) treated with CsA from 2005 to 2012 in our hospital. Of 42 HSP patients that we reviewed, 25 were younger than 10 years of age, and 23 HSP patients were boys. Among the 42 HSP patients, 18 children developed HSPN and were treated with CsA due to nephrotic-range proteinuria. Nephrotic-range proteinuria was defined as IgAN or HSPN patients had proteinuria greater than 40 mg/m²/hr. Six IgAN and six HSPN patients underwent repeated renal biopsies before and after CsA treatment, respectively. Both of the patients with IgAN and HSPN showed microscopic hematuria at diagnosis. Thirty children experienced nephrotic syndrome after diagnosis of IgAN and HSPN, and the remaining patients showed nephrotic-range proteinuria except two IgAN patients.

2. **Treatment protocol**

CsA was administered to the patients with diseases
characterized with IgA deposits such as IgAN or HSPN when the patients showed nephrotic syndrome or nephrotic-range proteinuria. Angiotensin-converting enzyme inhibitor (ACEi) was concurrently used in all patients during the period of treatment with CsA, where as corticosteroids were not used for the treatment of the patients. The starting dose of CsA was 5 mg/kg/day in two divided doses, and the desired drug level was maintained between 100 and 200 ng/mL. CsA levels were measured every 2 weeks at the beginning of treatment and every two months during the maintenance period.

3. Renal biopsy

Renal specimens of HSP patients were graded by the histological classification of ISKDC: grade I, minor glomerular abnormalities; grade II, pure mesangial proliferation (a: focal; b: diffuse); grade III, minor glomerular abnormalities or mesangial proliferation with crescents/segmental lesion in 50% crescents (a: focal; b: diffuse mesangial proliferation); grade IV, mesangial proliferation with crescents/segmental lesions in 50-75% glomeruli (a: focal; b: diffuse mesangial proliferation); grade V, mesangial proliferation with crescents/segmental lesions in >75% glomeruli (a: focal; b: diffuse mesangial proliferation); grade VI, membranoproliferative-like lesion (10). For an active lesion index, the scoring was assessed as follows: 1) mesangial proliferation: normal (0), slight (1), moderate (2), and severe (3); 2) necrosis or cellular crescents: 0% (0), 1-20% (1), 20-50% (2), >50% of glomeruli. The chronicity index was evaluated as follows: 1) fibrous crescents or global sclerosis: 0% (0), 1-20% (1), 20-50% (2), >50% of glomeruli. The chronicity index was evaluated as follows: 1) fibrous crescents or global sclerosis: 0% (0), 1-20% (1), 20-50% (2), >50% of glomeruli (3); 2) tubular atrophy associated with interstitial fibrosis: 0% (0), 1-20% (1), 20-50% (2), >50% of cortical area (3).

Clinicopathologic classification with pathologic features predictive of disease progression was graded using the Oxford Classification system (2009) of IgAN. The key pathologic features to be reported on renal biopsy were scored as follows: 1) mesangial hypercellularity: mean ≤4 mesangial cells/mesangial area (M0) and mean 4 or more mesangial cells/mesangial area (M1); 2) segmental glomerulosclerosis or adhesions present (S1) or absent (S0); 3) endocapillary proliferation present (E1) or absent (E0); 4) cortical interstitial fibrosis and tubular atrophy 0-25% (T0), 26-50% (T1), and >50% (T2); 5) total number of glomeruli with changes: endocapillary hypercellularity, extracapillary proliferation, global glomerulosclerosis, and segmental glomerulosclerosis (11, 12). Each pathologist from two different universities completed a scoring sheet for every biopsy and these data were used to obtain a consensus for each parameter.

4. Clinical status evaluation

The clinical status of each patient at diagnosis and at the most recent observation was classified as follows: 1) state A, normal physical examination, urine, and renal function; 2) state B1, microscopic hematuria without proteinuria; 3) state B2, proteinuria <40 mg/m²/hr with or without hematuria; 4) state C, active renal disease characterized by hypertension or proteinuria of 40 mg/m²/hr or greater, with glomerular filtration rate (GFR) of 60 mL/min/1.73m² or greater.

5. Statistical analysis

Statistical analyses were performed with SPSS for Windows version 18.0 (SPSS, Chicago, Illinois, USA). Descriptive statistics were expressed as mean±standard deviation or as a median with ranges. Differences between parameters before and after CsA treatment were compared with the Wilcoxon signed rank test and non-parametric statistical methods. All differences were considered statistically significant at P<0.05.

Results

The mean age of the patients was 10±4.2 year (range 2.0-16) at the time of HSPN or IgAN diagnosis (Table 1). No patient showed renal failure, hypertension, or acute nephritic features at onset. Among 54 patients with microscopic hematuria, 36 (66.7%) had gross hematuria at the time of the initial examination (Table 1). Mean duration of CsA treatment was 11.7±5.6 months (range
Angiotensin-converting enzyme inhibitor was maintained for the duration of CsA treatment (mean 14.5 ±24.2 months). Mean period of 3.0±2.6 months (range 1.3–12.6) was needed to normalize massive proteinuria after treatment of CsA. The median duration of follow-up was 3.7±3.1 year (range 1.7–15.2) from the onset of disease.

The mean serum levels of total protein and albumin increased from 6.5±0.8 g/dL to 7.1±0.4 g/dL (P<0.001) and from 2.8±0.6 g/dL to 4.3±0.2 g/dL (both P<0.001), respectively, after CsA treatment (Table 2). Mean cholesterol level decreased significantly from 205.9±77.9 mg/dL to 166.0±44.0 mg/dL (P<0.001), whereas serum creatinine level increased from 0.6±0.2 mg/dL to 0.7±0.2 mg/dL (P=0.005). Of note, the uric acid level significantly increased from 4.8±1.3 to 5.3±1.6 (P=0.004) (Table 2).

The pretreatment and follow-up renal biopsy results for 6 IgAN and 6 HSPN children are shown in Table 3. After 1-year treatment of CsA, one child with HSPN had ISKDC IIIb at the second biopsy from initial IIIa. Three children whose initial biopsies were class IIIb had class II disease on follow-up, and follow-up biopsies of two children with class II had the same class II in HSPN (Table 3). According to Oxford classification of IgAN, one children had M1S1, one had M1E1, two had M1, and two remained within normal range from the initial to the follow-up biopsies. Progressed cortical tubular atrophy occurred in only one child, and no children with IgAN progressed to cortical interstitial fibrosis or tubular atrophy due to CsA treatment (Table 3). As shown in Table 3, one year of oral CsA treatment was not associated with a marked increase in mean activity index without an increase in chronicity. None of the renal biopsy classifications worsened in either HSPN or in IgAN.

At the latest follow-up after the end of CsA treatment,

**Table 1. Clinical Characteristics of Children with Immunoglobulin A Nephritis and Henoch-Schönlein Purpura Nephritis**

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Total patients</th>
<th>N=54 (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>IgAN:HSPN</td>
<td>36:18</td>
<td>59.3:33.3</td>
</tr>
<tr>
<td>Sex</td>
<td>32:22</td>
<td>59.3:40.7</td>
</tr>
<tr>
<td>Age (years)</td>
<td>10±4.2</td>
<td></td>
</tr>
<tr>
<td>Microscopic hematuria</td>
<td>54 (100)</td>
<td></td>
</tr>
<tr>
<td>Gross hematuria</td>
<td>36 (66.7)</td>
<td></td>
</tr>
<tr>
<td>Duration of CsA (months)</td>
<td>11.7±5.6</td>
<td></td>
</tr>
<tr>
<td>Duration of ACEI (months)</td>
<td>14.5±24.2</td>
<td></td>
</tr>
<tr>
<td>Follow-up duration (years)</td>
<td>3.7±3.1</td>
<td></td>
</tr>
</tbody>
</table>

**Table 2. Comparison of Laboratory Data before and after the Treatment of Cyclosporin A**

<table>
<thead>
<tr>
<th>Variables</th>
<th>Pre-treatment</th>
<th>Post-treatment</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total protein (g/dL)</td>
<td>5.5±0.8</td>
<td>7.1±0.4</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Albumin (g/dL)</td>
<td>2.8±0.6</td>
<td>4.3±0.2</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Na (mMol/L)</td>
<td>139.5±2.7</td>
<td>140.2±1.7</td>
<td>0.040</td>
</tr>
<tr>
<td>K (mMol/L)</td>
<td>4.2±0.4</td>
<td>4.4±0.4</td>
<td>0.111</td>
</tr>
<tr>
<td>Cl (mMol/L)</td>
<td>104.4±3.3</td>
<td>104.8±2.0</td>
<td>0.691</td>
</tr>
<tr>
<td>BUN (mg/dL)</td>
<td>14.0±4.2</td>
<td>14.4±4.8</td>
<td>0.851</td>
</tr>
<tr>
<td>Creatinine (mg/dL)</td>
<td>0.6±0.2</td>
<td>0.7±0.2</td>
<td>0.005</td>
</tr>
<tr>
<td>Cholesterol (mg/dL)</td>
<td>205.9±77.9</td>
<td>166.0±44.0</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Uric acid (mg/dL)</td>
<td>4.8±1.3</td>
<td>5.3±1.6</td>
<td>0.004</td>
</tr>
</tbody>
</table>

**Fig. 1.** Elementary renal lesions observed in patients with IgA nephropathy are shown representatively according to the Oxford classification: a) segmental sclerosis (S1); b) mesangial hypercellularity (M1); and c) tubular atrophy/interstitial fibrosis >50% (T2).
24 patients returned to normal urinalysis and maintained normal renal function (state A), 8 patients had microscopic hematuria without proteinuria (state B1), and 17 patients had proteinuria of $>40 \text{ mg/m}^2/\text{hr}$ (state B2). The proportion of children with active renal disease with proteinuria of $>40 \text{ mg/m}^2/\text{hr}$ (state C) was decreased from 30 (55.6%) to 5 (9.3%). Side effects of CsA treatment, including gingival hypertrophy and trichosis, were minimal, and no one withdrew from CsA treatment because of the absence of severe side effects or complications. The protein to creatinine ratio significantly decreased from $3.7\pm1.5$ to $0.6\pm0.4$ after CsA treatment ($p<0.001$) (Fig. 3). Fig. 4 exhibits that if the basal level of proteinuria was set to 1, a 50% reduction in proteinuria was achieved 2.6 months after treatment with CsA. CsA treatment was well tolerated in all patients, and no patients required drug discontinuation or had any serious adverse effects that warranted hospitalization.

**Fig. 2.** Elementary renal lesions observed in patients with HSP nephritis are shown representatively according to the ISKDC classification: a) minimal glomerular abnormality; b) pure mesangial proliferation; and c) mesangial proliferation with crescent formation (arrow).

**Table 3.** Follow-up Histological Data in 6 HSPN and 6 IgA Patients Treated with Cyclosporin A

<table>
<thead>
<tr>
<th>HSPN case</th>
<th>1st Biopsy</th>
<th>2nd Biopsy</th>
<th>HSPN case</th>
<th>1st Biopsy</th>
<th>2nd Biopsy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Glomeruli (n)</td>
<td>ISKDC</td>
<td>AI</td>
<td>CI</td>
<td>TI</td>
<td>Glomeruli (n)</td>
</tr>
<tr>
<td>1</td>
<td>32</td>
<td>II</td>
<td>2/0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>2</td>
<td>25</td>
<td>IIb</td>
<td>3/0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>3</td>
<td>64</td>
<td>IIa</td>
<td>1/0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>4</td>
<td>43</td>
<td>IIb</td>
<td>2/1</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>5</td>
<td>29</td>
<td>IIb</td>
<td>2/1</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>IgAN case</th>
<th>1st Biopsy</th>
<th>2nd Biopsy</th>
<th>IgAN case</th>
<th>1st Biopsy</th>
<th>2nd Biopsy</th>
</tr>
</thead>
<tbody>
<tr>
<td>CIF</td>
<td>CTA</td>
<td>Oxford CS</td>
<td>CIF</td>
<td>CTA</td>
<td>Oxford CS</td>
</tr>
<tr>
<td>1</td>
<td>0</td>
<td>0</td>
<td>M1E0S1T0</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>2</td>
<td>0</td>
<td>0</td>
<td>M1E1S0T0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>3</td>
<td>0</td>
<td>0</td>
<td>M0E0S0T0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>4</td>
<td>0</td>
<td>0</td>
<td>M0E0S0T0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>5</td>
<td>0</td>
<td>0</td>
<td>M1E0S0T0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>6</td>
<td>0</td>
<td>0</td>
<td>M0E0S0T0</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

Abbreviations: Bx, biopsy; ISKDC, International Study of Kidney Disease in Childhood; AI, activity index; CI, chronicity index; TI, tubulointerstitial change; CIF, cortical interstitial fibrosis; CTA, cortical tubular atrophy; CS, classification system.
Discussion

As previously suggested [16, 17], IgAN and HSPN represent a spectrum of clinical and pathologic presentations of the same or a similar disorder, probably sharing a common pathogenesis. The features supporting this hypothesis include reports on the occurrence of these two diseases in the same family [18, 19], patients presenting with the combination of symptoms of HSP and IgAN [20, 21], and the indistinguishable nature of the renal histopathologic lesion [22]. However, according to our pathologic experiences, the two diseases are distinguishable not only in clinical presentations but also in pathology. Particularly, electron microscopy reveals that much larger one or two electron dense deposits are located in paramesangium and near the capillary wall in IgAN, whereas characteristically small and scattered electron dense deposits are often within and throughout the mesangial regions in HSPN [23]. This pathognomonic feature gives us a clue to pathologically differentiate and diagnose the two different diseases of IgA deposits.

Although the majority of patients with IgAN have good prognosis, 20–30% or sometimes up to 40% of patients will progress to ESRD over the course of 10–20 years [1, 2, 24]. These patients who are at high risk of progressive renal disease need to take a more aggressive therapeutic approach. Both clinical and histologic features at first biopsy should thoroughly be evaluated to determine whether the patients with IgAN are at high risk of progressive renal disease. While HSP is often a benign and a self-limited condition, approximately 30–90% of pediatric patients develop nephritis within 4 to 6 weeks of the initial presentation [8]. HSP is the most common cause of crescentic glomerulonephritis in children, which often manifests hematuria and proteinuria of variable intensity [25].

The treatment of severe IgAN and HSPN is still controversial, and recommendations are based on small, often uncontrolled series. Some studies have reported favorable results of treatments including intravenous administration of methylprednisolone followed by oral treatment with corticosteroids, corticosteroids in combination with azathioprine, cyclophosphamide with/without anticoagulants, plasmapheresis, and mycophenolate mofetil [26–28]. Nevertheless, there have been only a few studies of CsA treatment for IgAN or HSPN, respectively. Thus, the main goal of our study was to demonstrate the clinical efficacy of CsA in glomerular diseases characterized by IgA deposits and particularly to determine whether there were any severe glomerular histologic changes after CsA use according to the ISKDC and the Oxford classification.

Our study showed that CsA treatment has antiproteinuric effect in patients with glomerular injury by IgA deposits. In our study, there was a patient (HSPN, patient 3) who showed histological changes from ISKDC IIIa to IIIb, although he achieved clinical remission in nephritic-range proteinuria. In the remaining patients, 3 patients

![Fig. 3. Protein to creatinine ratio before and after cyclosporine treatment. Protein to creatinine ratio decreased from 3.7±1.5 to 0.6±0.4 (P<0.001).](image-url)

![Fig. 4. Proteinuria was reduced to the half of its pretreatment level after 2.6 months of CsA treatment (P<0.001).](image-url)
(HSPN, patients 2, 4, and 5) showed improvements in histological grades (ISKDC IIIb to II in 3 patients) and 2 patients (HSPN, patients 1 and 6) showed the same histological grades (ISKDC II to II in 2 patients) with clinical improvements. In IgAN cases, all patients showed same histological grades with clinical improvements. These results are in accordance with those of the previous studies. According to a study by Shin et al. [13], nine patients achieved stable remission after treatment with CsA during a mean follow-up of 4.6 years in 14 pediatric IgAN patients with nephrotic-range proteinuria. In addition, the laboratory findings exhibited a decline in the rate of 24-hr urinary protein excretion and a significant increase in serum albumin after CsA treatment. Furthermore, immunofluorescent analysis at the first and second biopsies showed a significant reduction of mesangial IgA deposits in seven (50%) of the 14 patients. A controlled study with a small number of patients by Lai et al. [29], also demonstrated a >50% reduction in proteinuria in 83% of 12 cyclosporine-treated IgAN patients and decreased IgA levels in 80% as well as increased serum albumin. Ronkainen et al. also reported that in seven pediatric HSPN patients with nephrotic-range proteinuria, four patients achieved stable remission after treatment with CsA during a mean follow-up of 6.0 years [9]. Consistent with these previous studies, the protein to creatinine ratio and urinary protein excretion in 24-h collection significantly decreased after CsA treatment in our study. However, our participants were different from those of previous studies in that our patients, who did not respond to prednisolone, did not receive any other immunosuppressive agents such as cyclophosphamide or azathioprine before CsA treatment.

The mechanism of CsA immunosuppressive action in glomerular diseases is still elusive. One possible explanation for the immunological basis of CsA efficacy is related to its ability to inhibit the secretion of cytokines by infiltrating T cells and macrophages [30]. In addition, CsA also stabilizes the podocyte actin cytoskeleton, resulting in reducing massive proteinuria [31]. Although CsA is a very effective immunosuppressive agent for steroid-dependent and steroid-resistant nephritic syndrome [32], nephrotoxicity is one adverse effect that limits its long-term use. Significantly increased creatinine level might result from CsA treatment in our study [33]. Notably, CsA-induced toxicity is related to the duration of treatment (>24 or 36 months) and dose of CsA [34, 35]. All of our patients were carefully observed and monitored during the treatment period, and the trough level of CsA was maintained between 100–200 ng/mL during the entire period. Due to relatively low CsA trough levels, there were no significant side effects that warranted hospitalization or discontinuation of treatment.

The ideal duration of CsA treatment for patients with renal disease characterized by IgA deposits is not known. Our results indicated that treatment duration of 2.6 months was necessary to achieve a 50% reduction of proteinuria from levels at the initial diagnosis. Thus, based on the result, we suggest that CsA should be treated for at least three months or more to achieve the goal of reducing massive proteinuria in patients with glomerular injury by IgA deposits.

The present study has some limitations: 1) this was a retrospective study, and the sample size was small; 2) the study had relatively short duration of CsA treatment and follow-up period; and 3) selection bias may have existed in that only mild patients were recruited at renal biopsy. 4) we were not able to analyze the date of control group receiving ACEi alone due to unavailability of date. All our patients received an ACEi, which is known to provide antiproteinuria and renoprotective effect against deterioration in renal function in various renal diseases [36]. Despite these limitations, the current data clearly show that patients with nephrotic-range proteinuria can effectively and safely be treated with CsA for less than one year. Also, the current pathologic results reveal that no patients treated with CsA for less than one year progress to renal deterioration and aggravation in the glomerular diseases. In other words, CsA is not inferior to other immunosuppressive agents in the treatment of patient with IgA deposits without causing renal cytotoxicity in one-year usage.

In conclusion, our study indicates that CsA can be used to reduce massive proteinuria without any pathologic deterioration within one year. Additionally, our findings suggest that CsA treatment should be maintained for
at least three months or more to obtain a satisfactory
effect on decreasing massive proteinuria in glomerular
injury associated with IgA deposits, such as IgAN and
HSPN.

Conflict of interest

The authors have no financial conflicts of interest.

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