Why children with IgA vasculitis nephritis still lack targeted therapies

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Immunoglobulin A vasculitis (IgAV), formerly known as Henoch-Schönlein purpura, is the most common form of vasculitis in childhood, often leading to renal involvement known as IgAV nephritis. Despite its prevalence and potential for long-term kidney damage, current clinical guidance offers little in the way of specific, evidence-based treatments for this vulnerable population.

The recently updated Kidney Disease: Improving Global Outcomes (KDIGO) 2025 Clinical Practice Guideline for the Management of Immunoglobulin A Nephropathy (IgAN) and IgA Vasculitis (IgAV) underscores this critical unmet need, noting a distinct lack of major clinical trials in children with IgAV.1

KEY TAKEAWAYS

- The Pivot: The KDIGO 2025 guideline update for IgAN and IgAV explicitly states a lack of major clinical trials in children with IgAV, leading to no new specific treatment recommendations for this group.
- The Data: The guideline's executive summary highlights that "little has changed for special situations of IgA-dominant immune complex glomerular diseases such as... children with IgAN or IgAV, given the lack of major clinical trials in these patient populations."1
- The Action: Clinicians managing children with IgAV nephritis must continue to rely on extrapolated data and general principles, as no new targeted therapies or management strategies have emerged from dedicated pediatric trials.

IgA vasculitis nephritis, a serious complication of IgAV, can lead to chronic kidney disease and end-stage renal failure in children. The disease is characterised by IgA deposition in the mesangium of the glomeruli, triggering an inflammatory response. While the systemic manifestations of IgAV, such as rash, arthralgia, and abdominal pain, are often self-limiting, renal involvement dictates the long-term prognosis. Identifying and managing IgAV nephritis early is crucial to preserving kidney function, but the optimal therapeutic approach for children remains elusive.1

The KDIGO 2025 guideline, summarised by Floege, Barratt, and Cook in Kidney International, represents a significant update for IgAN management, but it explicitly acknowledges the persistent void in pediatric IgAV.1 The guideline's primary focus shifted to more liberal kidney biopsy policies and stricter proteinuria control for IgAN, aiming for less than 0.5 g/d, ideally less than 0.3 g/d, alongside a stable estimated glomerular filtration rate. These recommendations, however, largely apply to adult IgAN and do not translate directly to pediatric IgAV nephritis due to distinct disease characteristics and progression patterns in children.1

The Adult IgAN Blueprint, Not Pediatric IgAV

For adult IgAN, the KDIGO 2025 guideline introduces a major new concept: initiating treatment with therapies that both prevent or reduce pathogenic IgA production and manage the consequences of existing nephron loss.1 Approaches to the first aim include targeted-release budesonide (Nefecon) or reduced-dose systemic corticosteroid therapy. In Chinese patients, mycophenolate mofetil is also suggested. These therapies directly target the underlying immune dysregulation thought to drive IgAN progression.1

The second aim, managing nephron loss, involves more generic strategies applicable to various forms of chronic kidney disease. These include healthy lifestyle education, renin-angiotensin system blockers, sodium-glucose cotransporter-2 inhibitors, and/or dual endothelin angiotensin receptor blockers. These interventions primarily focus on mitigating proteinuria, controlling blood pressure, and slowing the decline in kidney function, regardless of the primary etiology.1

But the guideline authors, including Jürgen Floege from RWTH Aachen University and Jonathan Barratt from the University of Leicester, are clear: "Little has changed for special situations of IgA-dominant immune complex glomerular diseases such as... children with IgAN or IgAV, given the lack of major clinical trials in these patient populations."1

This statement highlights a critical gap in evidence. While adult IgAN has seen new therapeutic developments, pediatric IgAV nephritis has not benefited from dedicated, large-scale clinical trials that could inform specific treatment recommendations.

Why the Pediatric Data Gap Persists

The absence of major clinical trials in children with IgAV is a complex issue. Pediatric populations are inherently challenging for clinical research due to ethical considerations, smaller patient numbers, and the need for age-appropriate formulations and endpoints. IgAV nephritis, while common among childhood vasculitides, is still a relatively rare disease compared to adult conditions, making large-scale, adequately powered trials difficult to recruit. This often leads to reliance on observational data or extrapolation from adult studies, which may not accurately reflect disease biology or treatment response in children.

The current standard of care for severe IgAV nephritis in children often involves corticosteroids, sometimes combined with immunosuppressants like cyclophosphamide or azathioprine, based largely on expert opinion and small, uncontrolled studies. These treatments carry significant side effects, and their long-term efficacy and safety in children are not fully established through rigorous clinical trials. The lack of specific guidance means clinicians must weigh the potential benefits against the risks of broad immunosuppression without the backing of robust pediatric data. For a comprehensive overview of general pediatric conditions, the Oxford Handbook of Paediatrics remains a valuable resource.

The Unanswered Questions

The KDIGO 2025 guideline's executive summary provides a quick reference for the most important changes in IgAN and IgAV management.1 However, for pediatric IgAV, the summary essentially confirms a status quo of uncertainty. The guideline encourages a more liberal kidney biopsy policy for IgAN, which could potentially be beneficial in children to better characterise the extent of renal involvement and guide therapy. But without specific pediatric trial data on how different biopsy findings correlate with treatment response in children, this remains an area requiring further investigation.

The lack of progress in pediatric IgAV nephritis trials means that the field continues to operate without a clear

understanding of optimal proteinuria targets, the role of renin-angiotensin system blockers in younger patients, or the potential utility of newer IgAN-specific therapies like targeted-release budesonide in children. Whether these adult-focused interventions could prevent or reduce pathogenic IgA production in children with IgAV nephritis, or if their safety profiles are acceptable, remains entirely unknown. The next generation of trials must address these fundamental questions to finally provide evidence-based care for children with this potentially devastating kidney disease.

CLINICAL IMPLICATIONS

The KDIGO 2025 guideline's explicit acknowledgment of the lack of major clinical trials in pediatric IgAV nephritis is not merely an observation; it is a stark indictment of the research landscape. Clinicians managing these children are left to navigate complex decisions without the robust evidence base that now exists for adult IgAN. This means continued reliance on broad-spectrum immunosuppression, often with significant side effects, simply because no targeted alternatives have been adequately tested in this population.

The industry's focus on adult indications, while understandable from a market perspective, leaves a critical gap for pediatric rare diseases. Developing therapies for children with IgAV nephritis requires dedicated investment in smaller, ethically complex trials. Without this, the 'new concepts' in IgAN management, such as therapies preventing pathogenic IgA production, will remain out of reach for children, perpetuating a two-tiered system of care.

For general practitioners and specialists alike, the message is clear: the management of pediatric IgAV nephritis remains an area of significant unmet need. Until specific trials address this population, treatment decisions will continue to be based on extrapolated data and clinical experience rather than definitive evidence. This situation demands a concerted effort from researchers, pharmaceutical companies, and funding bodies to prioritise pediatric studies.

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REFERENCES

1. 1. Floege J, Barratt J, Cook HT. Executive summary of the KDIGO 2025 Clinical Practice Guideline for the Management of Immunoglobulin A Nephropathy (IgAN) and Immunoglobulin A Vasculitis (IgAV). Kidney Int. 2025;107(1):1-14.

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