Response to: Timely and individualized use of immunosuppression is associated with favourable outcomes in paediatric IgA Vasculitis Nephritis

Eda Didem Kurt-Şükür¹ · Kjell Tullus²

Received: 15 December 2021 / Accepted: 16 December 2021 / Published online: 19 January 2022
© Crown 2022

Dear Editor,

We read with great interest the letter from Dr Chan and colleagues [1] on our paper on IgA vasculitis with nephritis [2]. They do bring up some important points.

The knowledge on when and how to treat IgA vasculitis (IgAV) is still very scarce and controversial. The very recent guidance from Kidney Disease Improving Global Outcomes (KDIGO) do still give very little space to IgAV and mainly refer to data from adult studies in IgA nephropathy (IgAN) [3]. They recommend that oral prednisolone/prednisone or pulsed intravenous methylprednisolone should be used in children with mild or moderate IgAV nephritis. Children with IgAVN with nephrotic syndrome and/or rapidly deteriorating kidney function are treated in the same way as those with rapidly progressive IgAN, i.e. with cyclophosphamide. Interestingly, none of these statements is backed up with any reference to the literature [3].

In their letter, Dr Chan et al. present much better long-term outcome compared to our study. No child was left with impaired kidney function and only three children out of 21 had ongoing low-grade proteinuria. In contrast, 59% of the children in our study had CKD at last follow-up. The reason for this difference is interesting to discuss.

In Hong Kong, all children were treated with prednisolone, while 76% of our children had that treatment. In our children, the first-line immunosuppressive (IS) treatment was very variable, with mycophenolate mofetil (MMF) being given to 40%, cyclophosphamide 12% with a total of 71% having had any immunosuppressive agent. The Chinese

children did in a majority get a CNI (67%) while the worst cases (33%) were treated with cyclophosphamide pulses.

There was a significant difference in severity of symptoms at onset between the two groups. Among the Chinese children, 19% showed nephrotic syndrome at onset while this was present in 55% of the British cohort.

As to be expected from a London cohort, there was variable ethnicity with about half of the children having European decent. The children from Hong Kong were all Chinese. The aforementioned KDIGO guidance interestingly gives separate advice to Chinese patients with IgAN and suggests the use of MMF while no IS is recommended for any of other ethnic groups if they do not experience a rapidly progressive disease.

In summary, we would suggest that the main reason for the better outcome in the Hong Kong children was a milder disease at onset while we cannot rule out that a more active treatment and the choice of a CNI instead of MMF (in contradiction to KDIGO guidance) also played a role. We hope that a cohort of 1175 children with IgAVN that we recently have collected will shed some further light on this.

References

1. Lee MH, Chan EY, Ma AL (2021) Timely and individualized use of immunosuppression is associated with favourable outcomes in paediatric IgA Vasculitis Nephritis. *Pediatr Nephrol*. <https://doi.org/10.1007/s00467-021-05405-0>
2. Kurt-Şükür ED, Sekar T, Tullus K (2021) Biopsy-proven Henoch-Schönlein purpura nephritis: a single center experience. *Pediatr Nephrol* 36:1207–1215. <https://doi.org/10.1007/s00467-020-04809-8>
3. KDIGO (2021) KDIGO 2021 clinical practice guideline for the management of glomerular diseases. *Kidney Int* 100:S125–S127

✉ Kjell Tullus
Kjell.Tullus@gosh.nhs.uk

¹ Department of Pediatrics, Division of Nephrology, Hacettepe University School of Medicine, Ankara, Turkey

² Nephrology, GOSH, Great Ormond Street Hospital, London, UK