IMMUNOSUPPRESSIVE THERAPIES IN CHILDREN WITH
BIOPSY-PROVEN IGA VASCULITIS NEPHRITIS: A
TERTIARY CENTRE EXPERIENCE

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Background IgA Vasculitis Nephritis (IgAVN) can lead to severe presentation including nephrotic syndrome. Data pertaining to the treatment outcomes of IgAVN with persistent moderate or nephrotic range proteinuria in children are, however, limited.

Objectives The aim of this study is to determine the response to immunosuppressive therapies in this patient population.

Methods We conducted a retrospective review on all children presenting with IgAV before 18 years between January 2009 and December 2019 in the Paediatric Nephrology Centre in Hong Kong. Patients with biopsy-proven IgAVN developing persistent moderate or severe nephrotic-range proteinuria despite ACE-inhibitor (ACEi), and followed for 24 months or more were included. Patient demographics, clinical and laboratory data, therapies received, and treatment outcomes were evaluated.

Results Of the 177 patients with IgAV, 42 children developed proteinuria. 21 Chinese patients (76% boy) had persistent proteinuria despite ACEi and kidney biopsy confirmed IgAVN at a median age of 8.5 years (IQR 5.8–11.2). At baseline, 3 (14%), 14 (66%), 3 (14%) and 1 (4%) patients had moderate proteinuria, nephrotic-range proteinuria, nephrotic syndrome and nephritic-nephrotic syndrome, respectively. All patients had normal kidney function, except one child with an estimated GFR of 31 ml/min/1.73 m². Median urine protein to urine creatinine ratio (UPCR) was 4.4 mg/mg (IQR 2.4–9.0) and serum albumin was 32 g/L (IQR 28–33.5). Histological findings were classified according to International Study of Kidney Diseases in Children (ISKDC): Class II (n=5, 24%), Class IIIa (n=9, 42%), Class IIIb (n=6, 29%), Class IV (n=1, 5%).

All patients received corticosteroid at a median time of 33 days (IQR 12–52) since kidney involvement. Whereas 7 children (33%) with severe disease received monthly intravenous cyclophosphamide as induction therapy, 12 patients (57%) and 2 patients (10%) received calcineurin inhibitors and azathioprine, respectively. The maintenance therapy consisted of corticosteroid and one additional immunosuppression, including calcineurin inhibitors (n=16, 76%), azathioprine (n=4, 19%) and mycophenolate mofetil (n=1, 5%).

Over a median follow-up period of 3.6 years (IQR 2.8–5.6), 18 patients (86%) attained complete remission at a median of 139.5 days (IQR 102–225) since immunosuppressants initiation. The other 3 patients achieved partial remission. Three patients (14%) relapsed in 7.5 months (IQR 1.2–16.2) following complete remission but resolved promptly with treatments. At last follow-up, all patients had normal kidney function and the median UPCR was 0.11 mg/mg (IQR 0.10–0.16).

Conclusions Immunosuppressive therapies were associated with favourable renal outcomes in children with biopsy-proven IgAVN presented with persistent moderate or nephrotic range proteinuria despite ACEi. Further studies are required to determine the optimal treatments in this patient population.

HOW EFFECTIVE ARE WE COMMUNICATING WITH OUR PAEDIATRIC ONCOLOGY PATIENTS?

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Background Given the sensitive and intricate nature of oncological cases, especially in paediatric age group, it is important to have specific and strategic ways of discussing and communicating diagnosis and treatment plan. Especially being aware that to achieve the trust and faith of our patients and relatives in paediatric oncology, effective communication is key.

Thus, emphasis of both verbal and non-verbal communication with oncology patient cannot be over emphasised.

Objectives Our survey highlighted an assessment of how our communication of diagnosis and care has been with emphasis on striving for improvement. Examples of communications asked about includes, how well the diagnosis was discussed the first time, and how much of the information given was fully understood in the easiest possible manner.

Also, emphasis was placed on regular care given with each admission and how well our team communicated our line of management provided at each step of the way.

There were other areas covered in the questionnaire including support from community nurses, tertiary centres, play specialists and medications given.