EMEESY
Children's Kidney Network

EMEESY annual education meeting 2026:
Glomerular disease and the Kidney Ecosystem

Friday 16 October 2026

The Barn Hotel and Spa, Marston, Nr Grantham

What do Paediatricians need to know about IgA vasculitis (HSP) nephritis in children?
The current situation

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Clinical presentation of IgA vasculitis

Clinical features

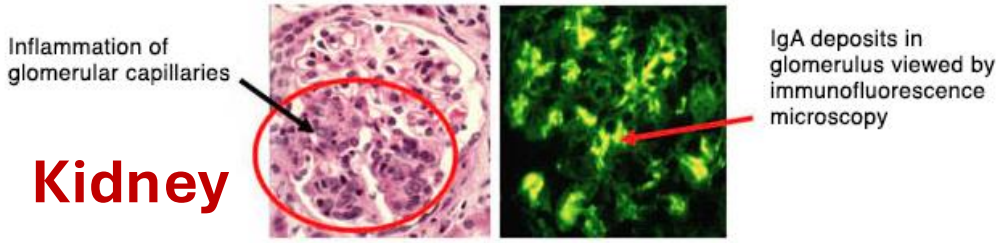




GI tract



Joints



Kidney

Age at presentation

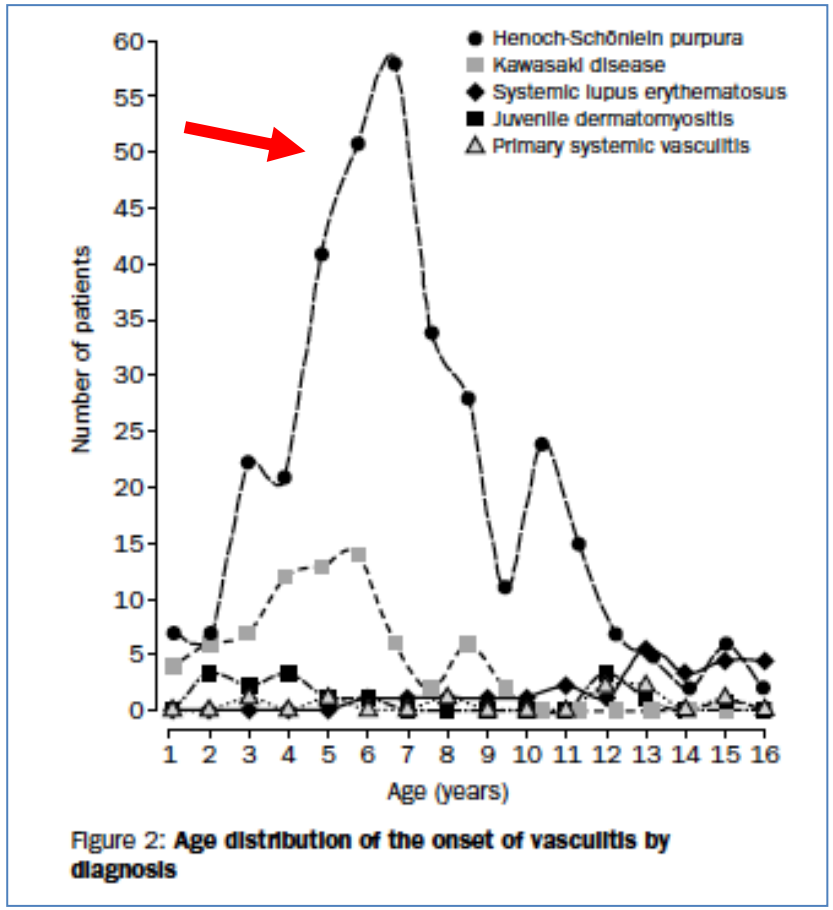


Figure 2: **Age distribution of the onset of vasculitis by diagnosis**

Classification criteria

EULAR/PRINTO/PRES criteria for Henoch–Schönlein purpura, childhood polyarteritis nodosa, childhood Wegener granulomatosis and childhood Takayasu arteritis: Ankara 2008. Part II: Final classification criteria FREE

Gardner-Medwin et al, Lancet 2002;360:1197-102

EULAR/PReS Classification of Childhood IgAV

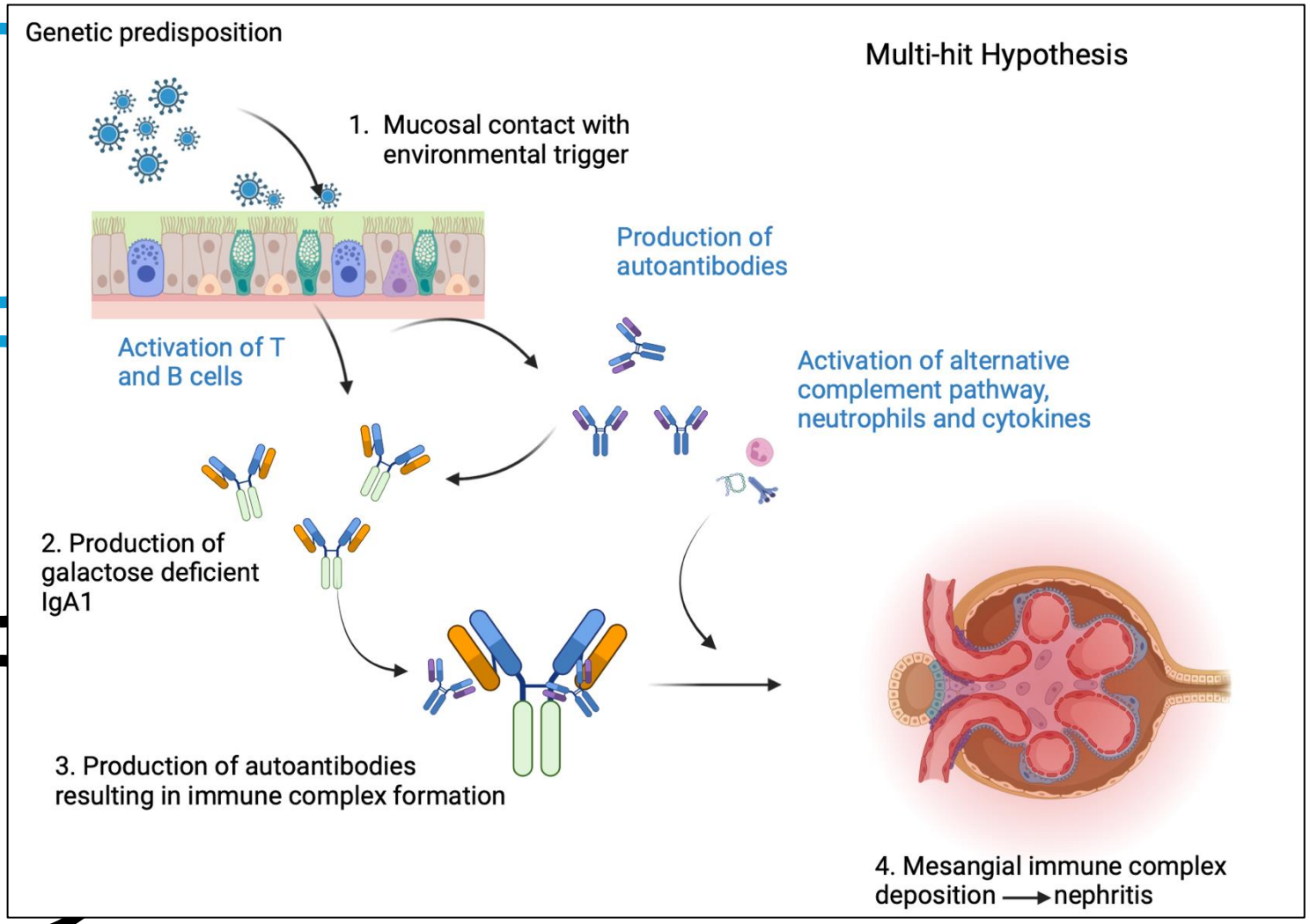
Table 1 Final EULAR/PRINTO/PRES HSP criteria (with glossary) and classification definition (sample 973)

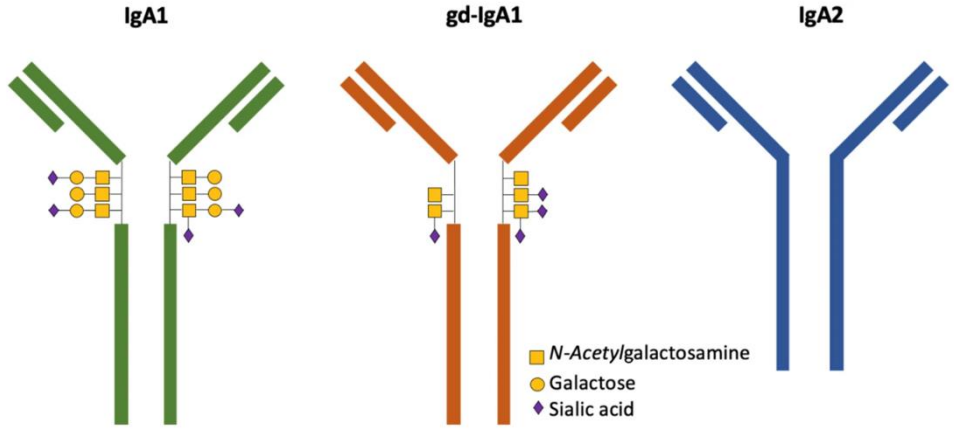
Criterion	Glossary	Sensitivity (%)	Specificity (%)	AUC (%)
Purpura (mandatory criterion)	Purpura (commonly palpable and in crops) or petechiae, with lower limb predominance, * not related to thrombocytopenia	89	86	87.5
1. Abdominal pain	Diffuse abdominal colicky pain with acute onset assessed by history and physical examination. May include intussusception and gastrointestinal bleeding	61	64	62.2
2. Histopathology	Typically leucocytoclastic vasculitis with predominant IgA deposit or proliferative glomerulonephritis with predominant IgA deposit	93	89	91.1
3. Arthritis or arthralgias	Arthritis of acute onset defined as joint swelling or joint pain with limitation on motion Arthralgia of acute onset defined as joint pain without joint swelling or limitation on motion	78	42	59.9
4. Renal involvement	Proteinuria >0.3 g/24 h or >30 mmol/mg of urine albumin/creatinine ratio on a spot morning sample Haematuria or red blood cell casts: >5 red blood cells/high power field or red blood cells casts in the urinary sediment or ≥2+ on dipstick	33	70	51.4
HSP EULAR/PRINTO/PRES Ankara 2008 classification definition: κ 0.90 (95% CI 0.84 to 0.96)	Purpura or petechiae (mandatory) with lower limb predominance* and at least one of the four following criteria: Abdominal pain Histopathology Arthritis or arthralgia Renal involvement	100	87	93.5

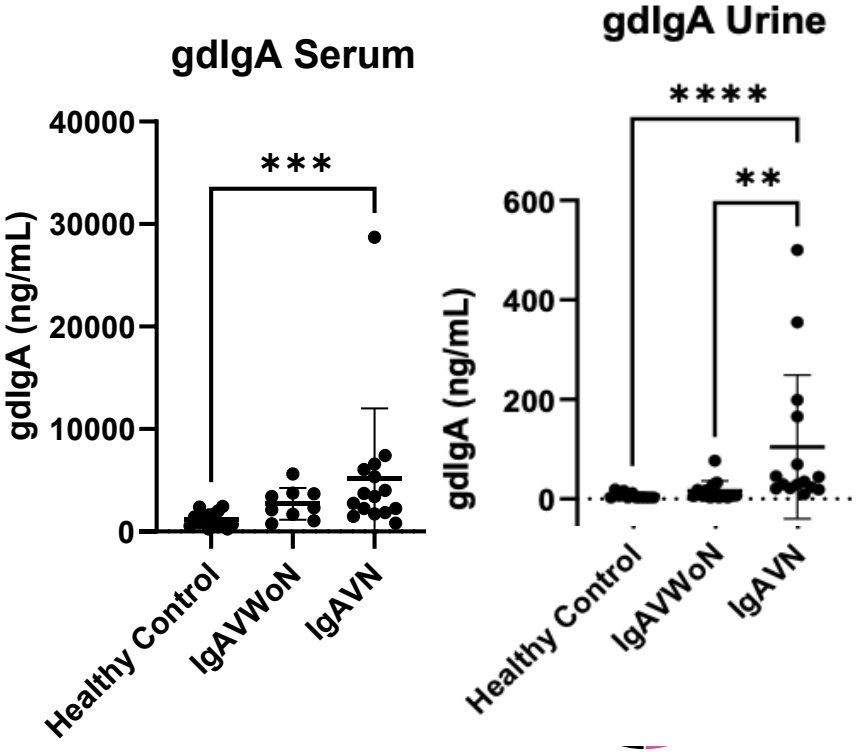
*For purpura with atypical distribution a demonstration of an IgA deposit in a biopsy is required.
AUC, area under the curve; EULAR, European League Against Rheumatism; HSP, Henoch–Schönlein purpura; PRES, Paediatric Rheumatology European Society, PRINTO, Paediatric Rheumatology International Trials Organisation.

Ozen et al, 2010

Pathophysiology



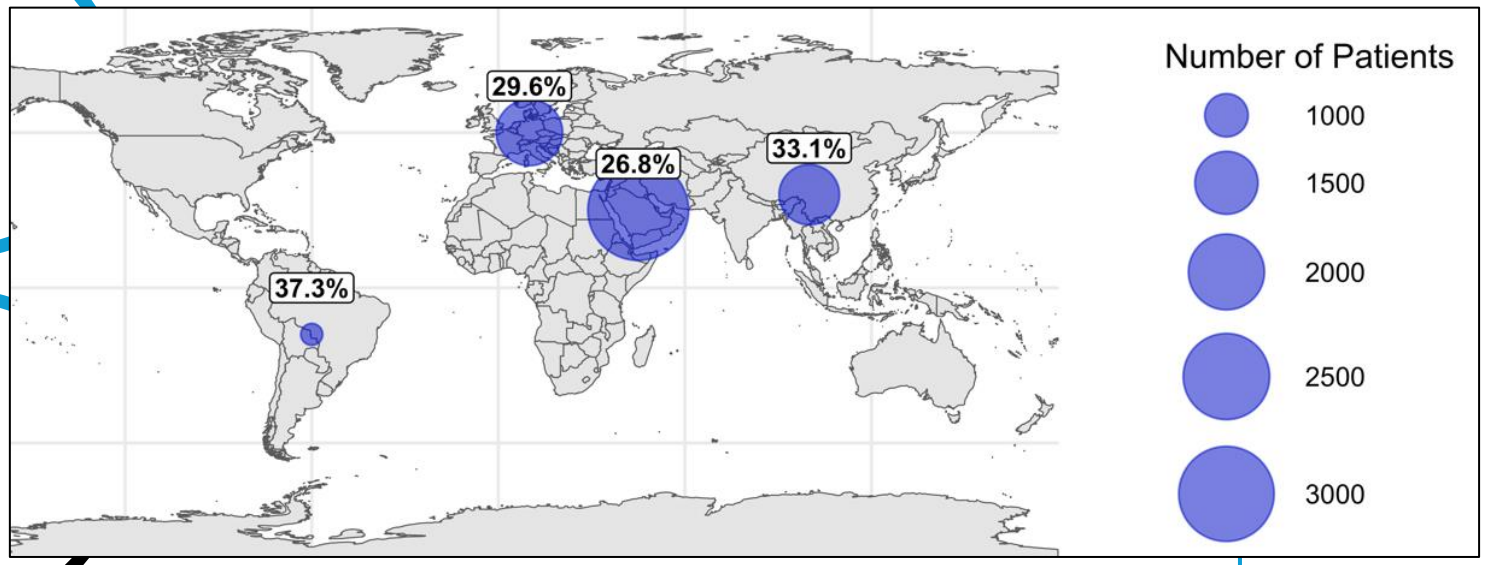




Biologically indistinguishable from IgA nephropathy using traditional measures

Suzuki et al, Kidney Int 2018; Local unpublished data.

Incidence of IgAV nephritis



Biopsy diagnosis	2021	2022	2023	2024	Average for 1 year fo 2 sites	Estimated number of cases per year
IgAVN	6	12	8	15	10.25	56.28
IgAN	6	5	6	4	5.25	28.82
lupus nephritis	2	3	7	8	5	27.45
FSGS	4	6	0	6	4	21.96
MCD	3	1	4	3	2.75	15.10
MPGN	1	5	1	4	2.75	15.10
PIGN	0	2	1	1	1	5.49
AAV	0	2	0	2	1	5.49
Membranous	0	0	2	2	1	5.49
TBMN	3	0	1	1	1.25	6.86
Alport	2	1	1	1	1.25	6.86
Mesangial hypercellularity	0	0	2	1	0.75	4.12
TMA	1	0	0	1	0.5	2.75
Vasculitic glomerulonephritis	0	0	1	0	0.25	1.37
Amyloidosis	0	0	0	1	0.25	1.37
MCD/IgAN	0	1	0	0	0.25	1.37
IgAN/PIGN	0	0	1	0	0.25	1.37
FSGS/TMA/MPGN	0	0	0	1	0.25	1.37
Total	28	38	35	51	8	43.92

Unselected cohorts of children with IgAV
(24 studies; n=7,180 children)

Overall mean incidence of nephritis = **29.9%.**

Kidney failure risk 1-2% in all patients
Rises in high risk populations

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Rare Kidney Diseases
In partnership with Kidney Research UK

Gage et al, ACR Open Rheum 2025; Ding et al, Manuscript in prep, 2026

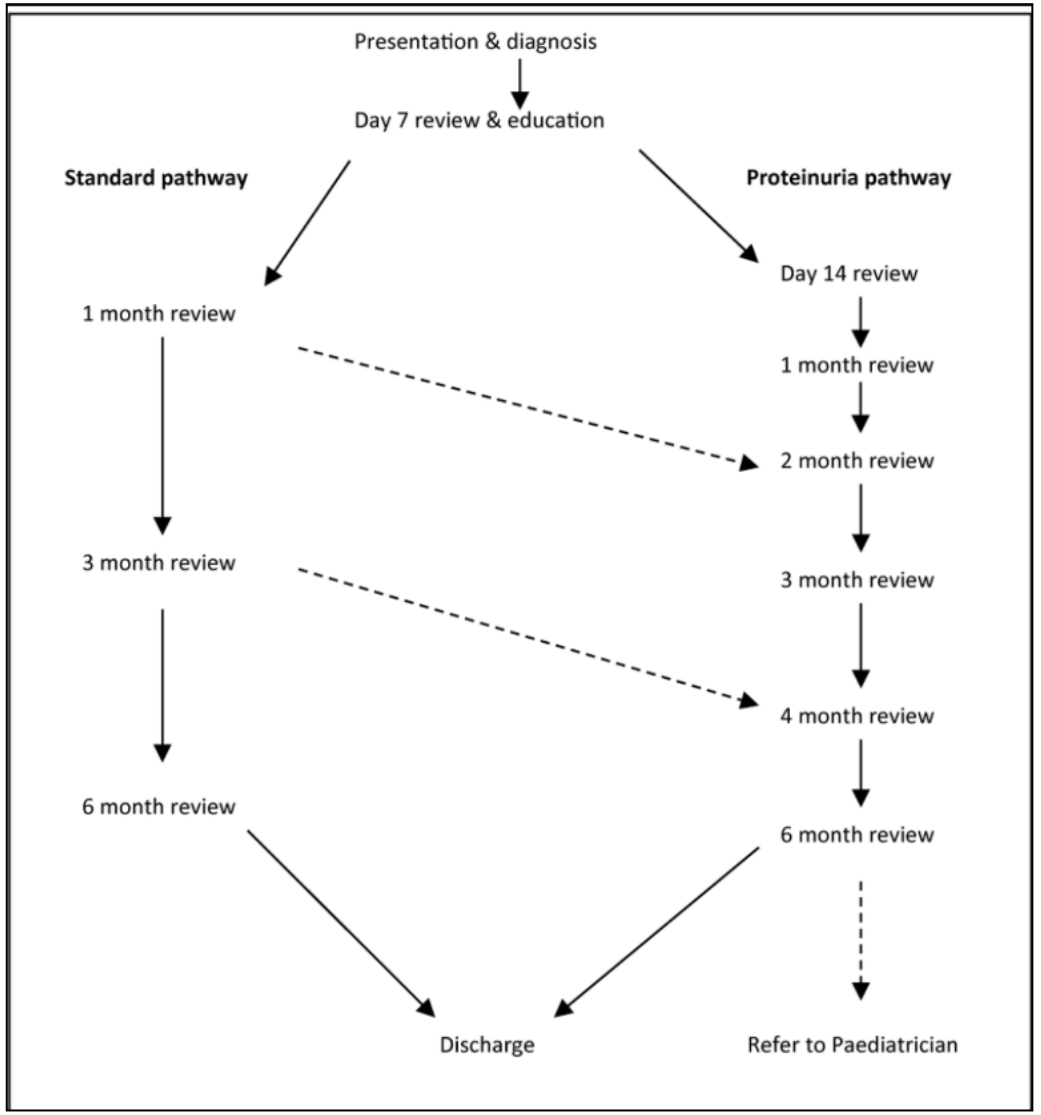
Screening for nephritis

Urine screening for all patients

Time to develop nephritis

<1 month in 68-99% and <6 months in 91-100%

Day 7 urine normal = 94-97% NPV



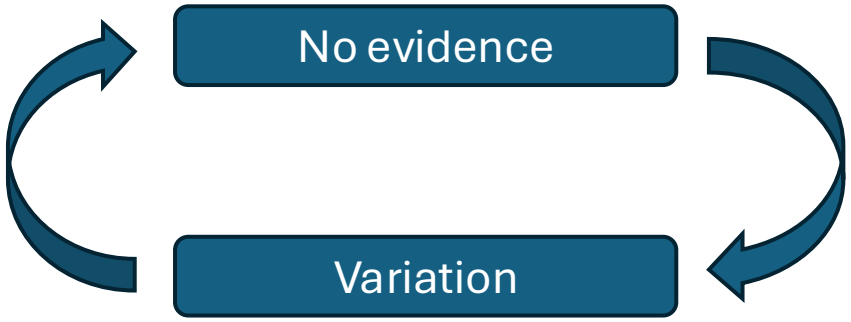
Cochrane: early steroids <u>do not prevent</u> nephritis

- 5 studies, 746 children: RR 0.74, 95% (CI 0.42 to 1.32)
- Children

Management of active nephritis by histological <u>AND</u> clinical features

Kawashima et al, Nagoya J Med Sci 2016; Watson et al, 2012

Management



UKKA
UK Kidney Association

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Home / About Us / News / IgA Vasculitis Guideline (RCPCH Endorsed)

IgA vasculitis Guideline (RCPCH endorsed)

- The initial management of IgA vasculitis (Henoch Schönlein Purpura) in children and young people
- The management of complications associated with IgA vasculitis (Henoch Schönlein Purpura) in children and young people

GUIDELINES

IPNA clinical practice recommendations for the diagnosis and management of children with IgA nephropathy and IgA vasculitis nephritis

Marina Vivarelli[1] · Susan Samuel[2] · Rosanna Coppo[3] · Jonathan Barratt[4] · Melvin Bonilla-Felix[5] · Dieter Haffner[6] · Keisha Gibson[7] · Mark Haas[8] · Maher Ahmed Abdel-Hafez[9] · Marta Adragna[10] · Paul Brogan[11] · Siah Kim[12] · Isaac Liu[13] · Zhi-Hong Liu[14] · Mukta Mantan[15] · Yuko Shima[16] · Masaki Shimuzu[17] · Qian Shen[18] · Hernan Trimarchi[19] · Deirdre Hahn[12] · Elisabeth Hodson[20] · Ken Pfister[21] · Areefa Alladin[21,22] · Olivia Boyer[23] · Koichi Nakanishi[24] on behalf of the International Pediatric Nephrology Association

European consensus-based recommendations for diagnosis and treatment of immunoglobulin A vasculitis—the SHARE initiative

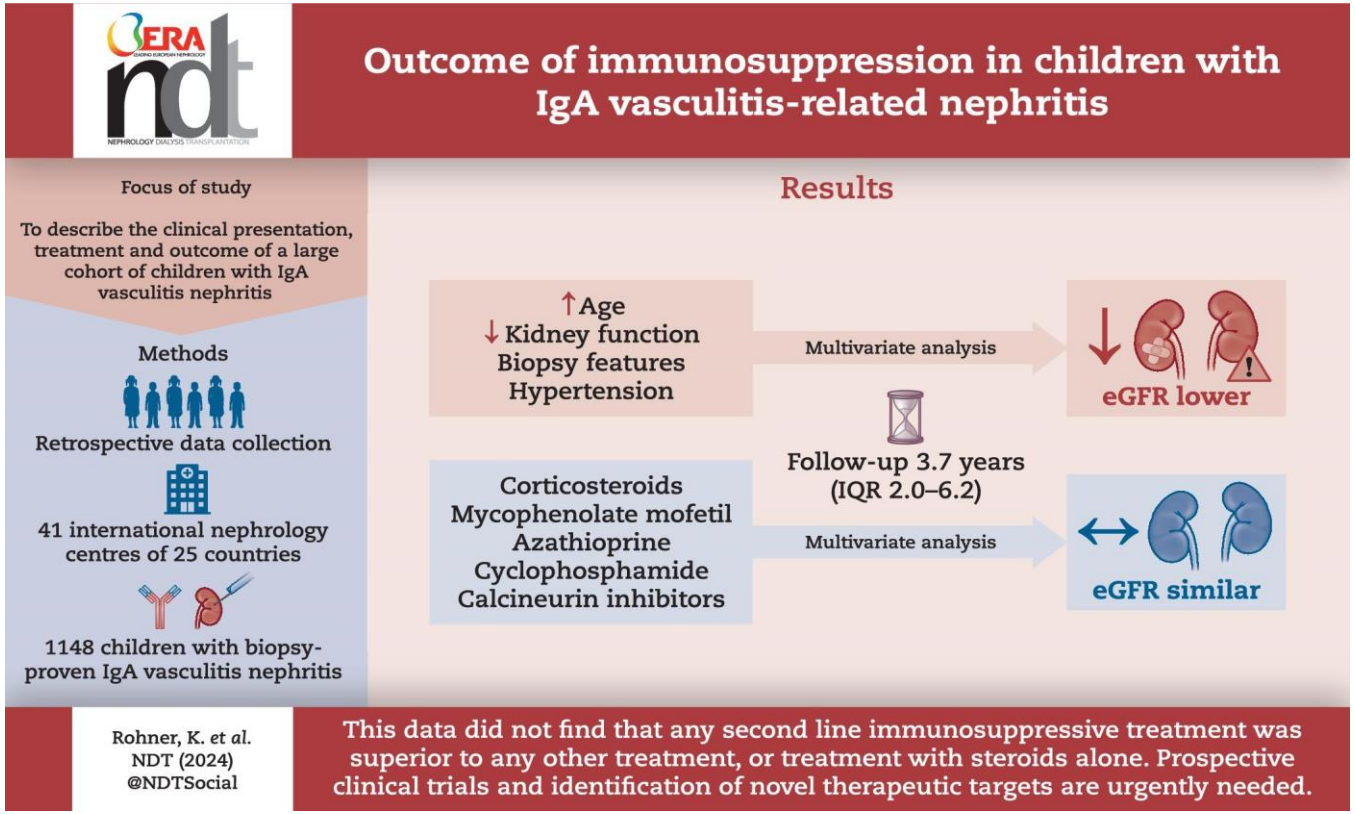
Seza Ozen, Stephen D Marks, Paul Brogan, Noortje Groot, Nienke de Graeff, Tadej Avcin, Brigitte Bader-Meunier, Pavla Dolezalova, Brian M Feldman, Isabelle Kone-Paut Pekka Lahdenne, Liza McCann, Clarissa Pilkington, Angelo Ravelli, Annet van Royen, Yosef Uziel, Bas Vastert, Nico Wulffraat, Sylvia Kamphuis, Michael W Beresford

Rheumatology, kez041, https://doi.org/10.1093/rheumatology/kez041
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KDIGO — Kidney Disease Improving Global Outcomes

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Treatment of biopsy proven nephritis



- Up to 80% steroids
- Complete response 36-57% at 6 months

Kidney outcomes:

- 36% persisting proteinuria
- 29% eGFR <90 mL/min/m^2
- 3% CKD stage 4–5.

Key milestones for IgA vasculitis

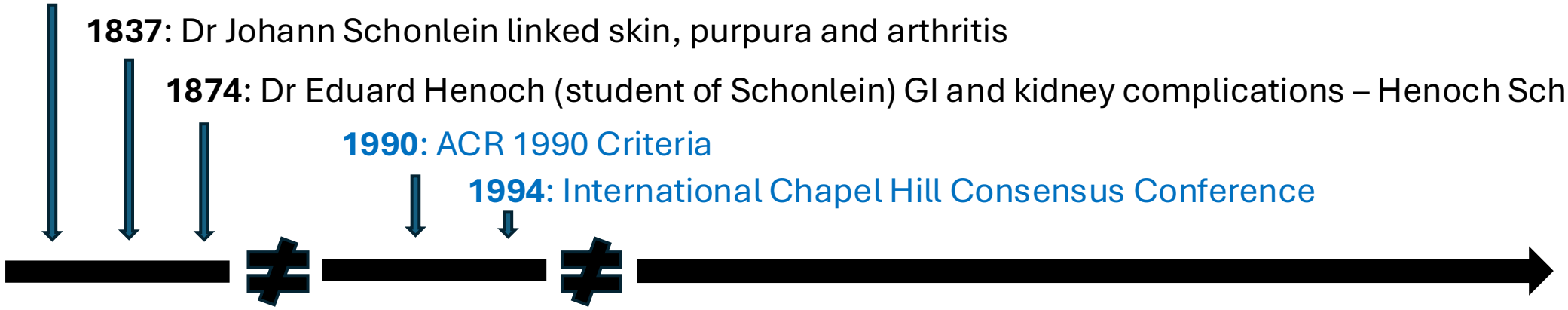
1801: Dr William Heberden described a 5-year old boy

1837: Dr Johann Schonlein linked skin, purpura and arthritis

1874: Dr Eduard Henoch (student of Schonlein) GI and kidney complications – Henoch Schonlein Purpura

1990: ACR 1990 Criteria

1994: International Chapel Hill Consensus Conference



2026:EULAR IgAV study group adult criteria

2025: International collaboration pathway to clinical trials

2024: Multi-centre global kidney outcome data in children n=1148

2024: Multi-centre global kidney histology scoring n=361

2019: SHARE diagnostic recommendations

2012: Disease renamed IgA vasculitis to reflect biology

2010: EULAR/PRINTO/PRES criteria for HSP in children

2006: EULAR/PRINTO/PRES proposed consensus for HSP in children

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Rare Kidney Diseases

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Rohner K, et al. NDT. 2024; Barbour SJ, et al. Clin J Am Soc Nephrol. 2024; Oni L, et al. Kidney Int Rep. 2025

Summary

- IgAV is commonly associated with nephritis
- Treatment relies on RAAS, immunosuppression
- Outcomes have not changed over time
- Unmet need in high quality evidence led by science

Acknowledgements

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