Childhood nephrotic syndrome: update 2026

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Childhood idiopathic nephrotic syndrome

- Acute onset of heavy proteinuria, hypoalbuminaemia and oedema
- 2-3/100,000 children per year in UK but 4-6 x increased incidence in Sri Lanka
- Modal age of presentation 2-3y; boys > girls
- Highly responsive to corticosteroid therapy
- 80% develop disease relapses
- 50% develop FRSSNS, needing use of more potent immunosuppressive therapies



What do we want for a child with newly-presenting nephrotic syndrome?

- Cure at first presentation
- Long-term remission
- Minimise relapse frequency
- Minimise drug toxicity

Overview

1. What's the best initial treatment?
2. How best to use steroids with relapsing disease?
3. Which steroid-sparing drug is best at preventing relapses?
4. How do we predict disease course early on?
5. What causes nephrotic syndrome?
6. What does the future hold for treatment options?

Pediatric Nephrology
https://doi.org/10.1007/s00467-022-05739-3

GUIDELINES

IPNA clinical practice recommendations for the diagnosis and management of children with steroid-sensitive nephrotic syndrome

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UKKA
UK Kidney Association

Clinical Practice Guideline:
The initial immunosuppressive treatment for children and young people with immune mediated glomerular diseases

Final version: September 2025
Review date: September 2028

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What’s the best initial treatment?

UKKA Harmonisation

Management of INS at disease presentation

Number	RECOMMENDATION	Grade
INS 1.1	We recommend that children with a 'typical' first presentation of nephrotic syndrome commence treatment with prednisolone 60 mg/m^2 per day or 2 mg/kg/day (maximum dose 60 mg) for 4 weeks.	**1A**
INS 1.2	We suggest that for children under the age of 5 years, prednisolone should be dosed according to body surface area, preferably using the Mosteller formula: $$BSA = \sqrt{\frac{height\ (cm) \times weight\ (kg)}{3600}}$$ This should be based on an estimated dry weight for the child rather than their oedematous weight.	**2C**
INS 1.3	We recommend that a kidney biopsy prior to treatment should be considered in children who present with atypical features such as macroscopic haematuria, abnormal kidney funtion (in the absence of hypovolaemia), hypertension with a euvolaemic state or low complement factor C3 or C4.	**1B**
INS 1.4	We recommend that children with typical newly-presenting nephrotic syndrome should not routinely be commenced on steroid-sparing drugs at the point of initial remission.	**1A**
INS 1.5	We suggest that in children aged >5 years prednisolone may be dosed by either body surface area or weight with a maximum dose of 60mg per day.	**2C**
INS 1.6	We suggest that prednisolone should be given as a single daily dose in the morning.	**2B**
INS 1.7	We recommend that children who have not entered remission after 4 weeks of prednisolone 60 mg/m^{2}per day are discussed with a paediatric nephrologist as they may require 3 consecutive daily doses of high dose intravenous methylprednisolone (500 mg/m^2, maximum dose 500 mg) to test steroid-responsiveness and may need a kidney biopsy.	**1C**
INS 1.8	We recommend that children who enter remission within 4 weeks of commencement of prednisolone at 60 mg/m^2 per day, continue to receive prednisolone at that dose for a total of 4 weeks, then reduce the dose to 40 mg/m^2 (maximum dose 40 mg) given on alternate days for a further 4 weeks, then stop.	**1A**
INS 1.9	We suggest that children who are late responders (i.e. only enter remission within 6 weeks of presentation) receive a more prolonged taper of prednisolone over at least 8 weeks and commence tacrolimus as a steroid-sparing agent.	**2D**
INS 1.10	We suggest consideration of a kidney biopsy and/or genetic testing in children who are late or partial responders.	**2D**

ISKDC regimen

- Prednisolone:
  - 60 mg/m^2 (max 80 mg) daily for 4 weeks
  - then 40 mg/m^2 (max 60 mg) on alternate days for 4 weeks

- ‘Gold standard’ regimen against which all others have been compared



No benefit to initial prolonged steroid course



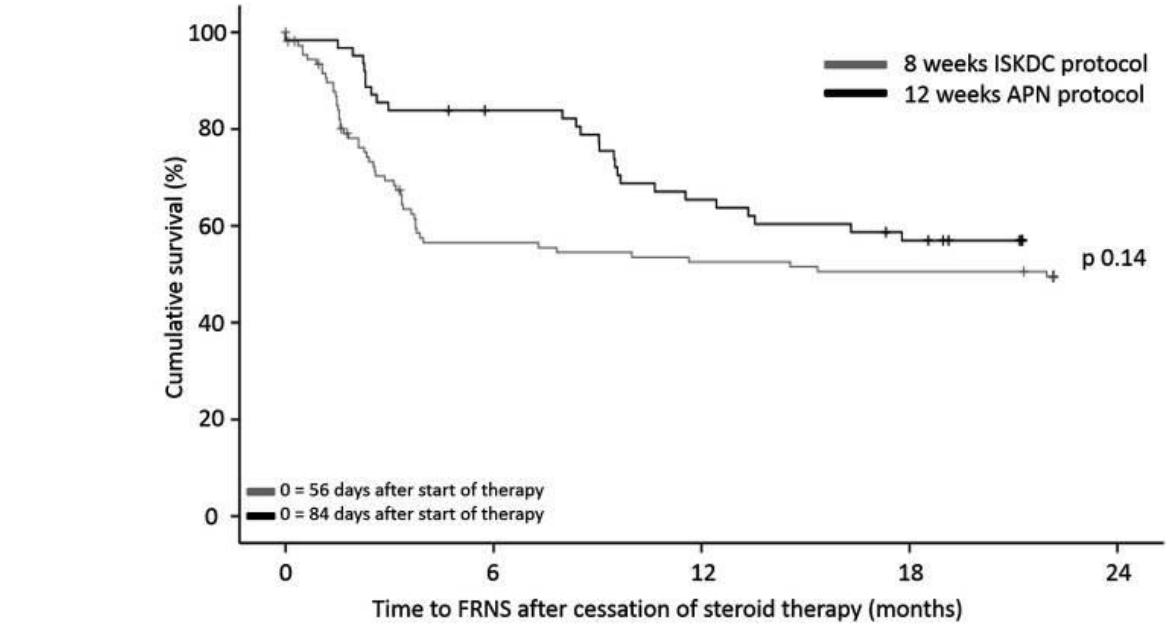
Authors' conclusions

There are now four well designed studies randomising 823 children which have clearly demonstrated that there is no benefit of prolonging prednisone therapy beyond two to three months in the first episode of SSNS.

Is 12 weeks better than 8?
Schijvens, 2021

- ISKDC = 4 weeks + 4 weeks
- APN = 6 weeks + 6 weeks

- IPD meta-analysis (PREDNOS vs Teeninga, 2013)
- Shorter time to first relapse in APN regimen
- But similar outcomes at 24 m

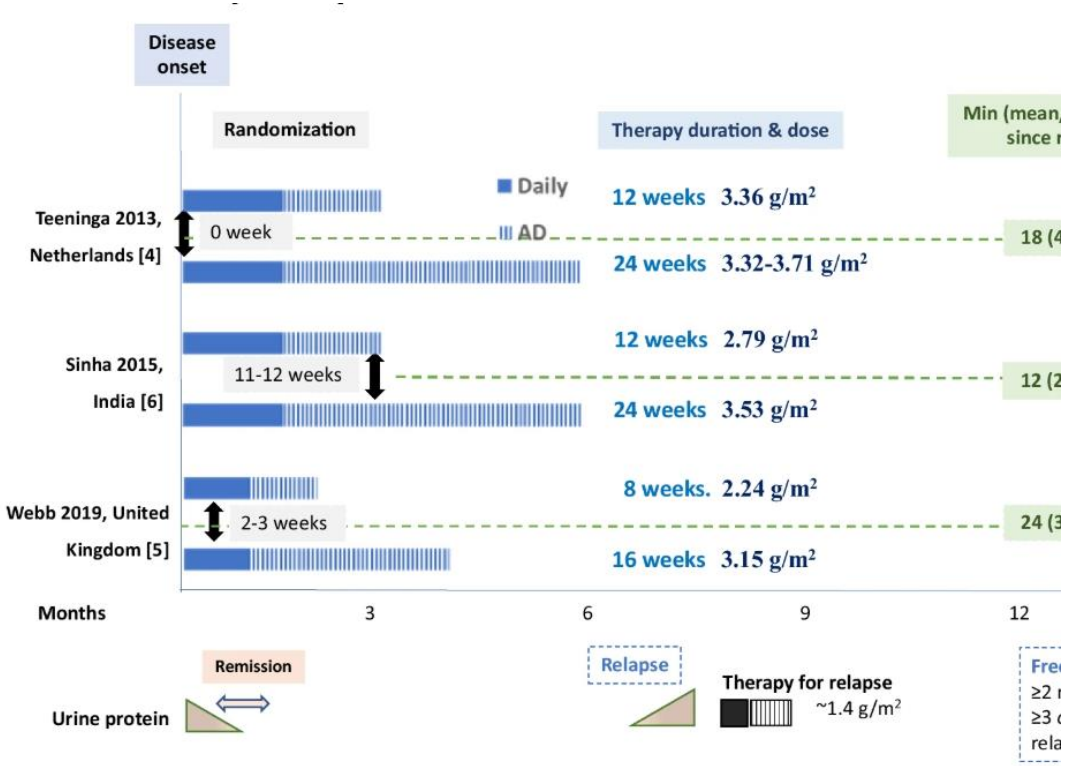


Are there demographic differences in steroid response?

- Teeninga, 2021: children <4 have shorter time to first relapse
- Sinha, 2014: children <3 may benefit from longer initial course (lower risk of relapse but not FRNS)
- PREDNOS: children ≤5 may have longer time to first relapse with extended course
- BSA versus weight-based dosing?

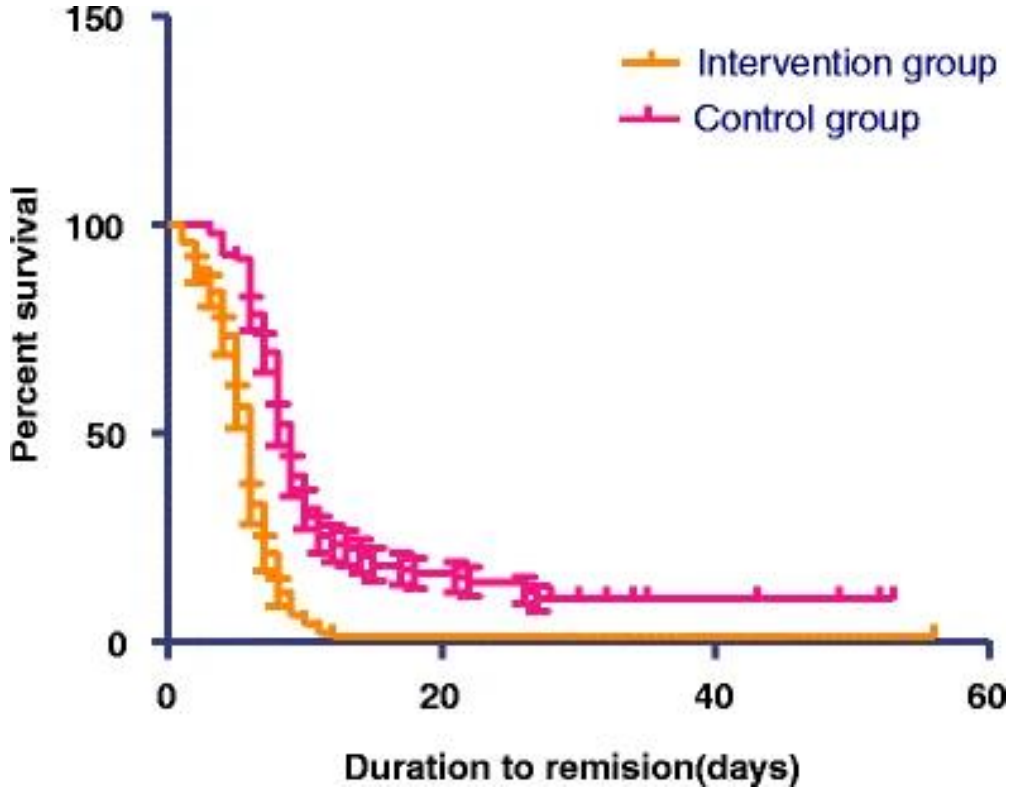
Sinha, 2026

- IPD meta-analysis of standard versus extended steroid
- Patient/disease variants as confounders
- Age >2.5 and initial cumulative prednisolone dose >3.135 g/m^2 reduced risks of the first relapse and frequent relapse
- Advises 12 week initial steroid course



Use of adjunctive non-steroid immunosuppression at first presentation

- Hoyer and Brodehl, 2006:
  - CsA on remission made no difference to disease course
- Zhang, 2014:
  - 211 children with new NS
  - Open label RCT
  - Standard treatment: 2 mg/kg daily 4 w +1.5 mg/kg alt days for 4 w + slow taper
  - IMP: std + azithromycin 10 mg/kg for 3 d
  - Shorter time to remission with AZM
  - No difference at 6 months



NEPHROVIR-3, 2022

- Levamisole for 6 months after initial remission
- 30% better relapse-free survival at 12 months
- LEARNS study (Netherlands) due to complete this year

Dossier, ESPN 2024

INTENT: Benz, 2026

272 children randomised within 5 days of initial remission

MMF for 12 w *and* Pred 40 mg/m^2 alt days for 2 w
n=136

Pred 60 mg/m^2 od for 6 w *then* Pred 40 mg/m^2 alt days for 6 w
n=136

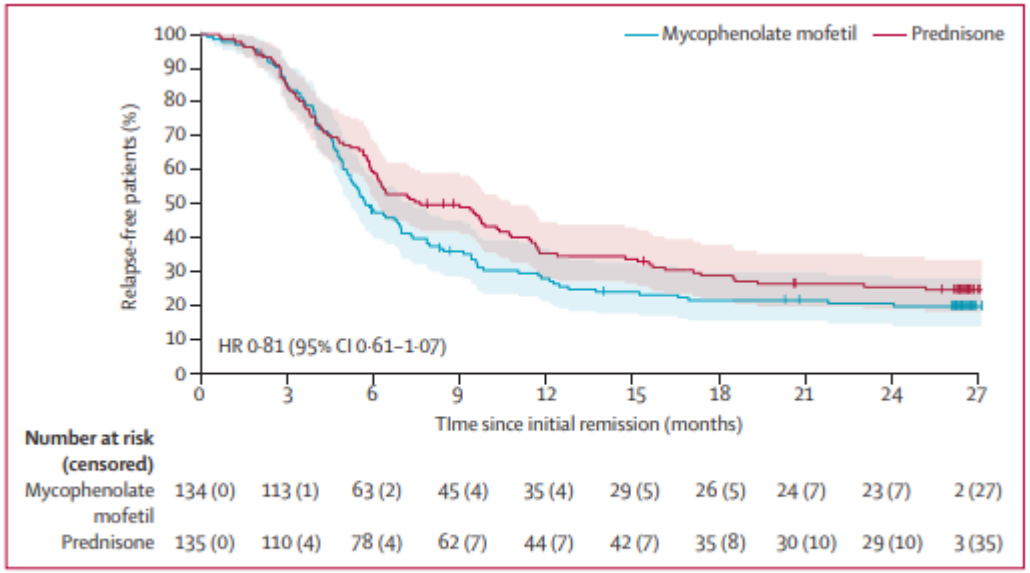
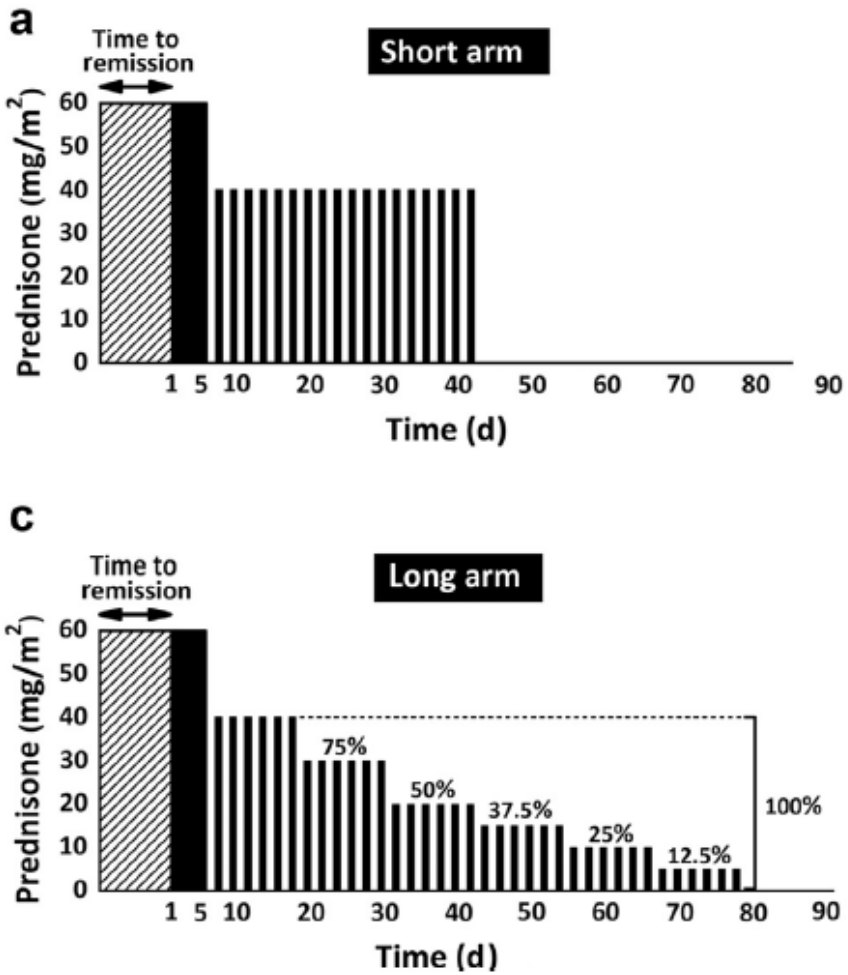
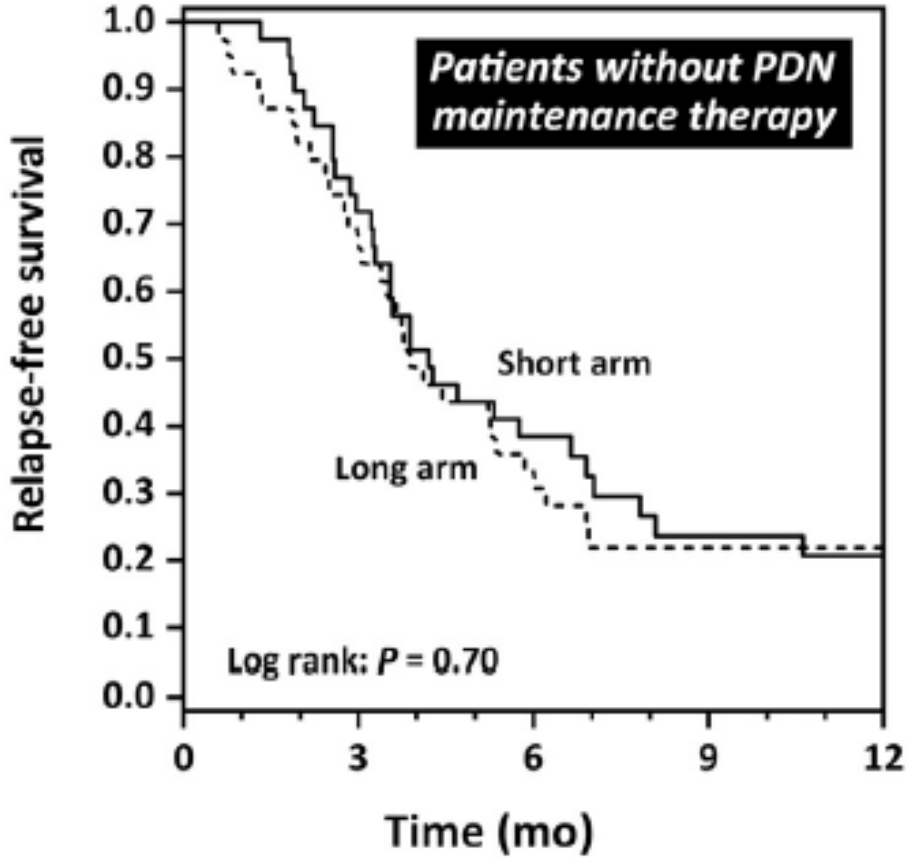


***Figure 3*: Treated relapse-free survival from initial remission to end of follow-up**
Shaded areas represent 95% point-by-point CIs. A post-hoc sensitivity analysis on the restricted mean survival time yielded consistent results (appendix p 31).

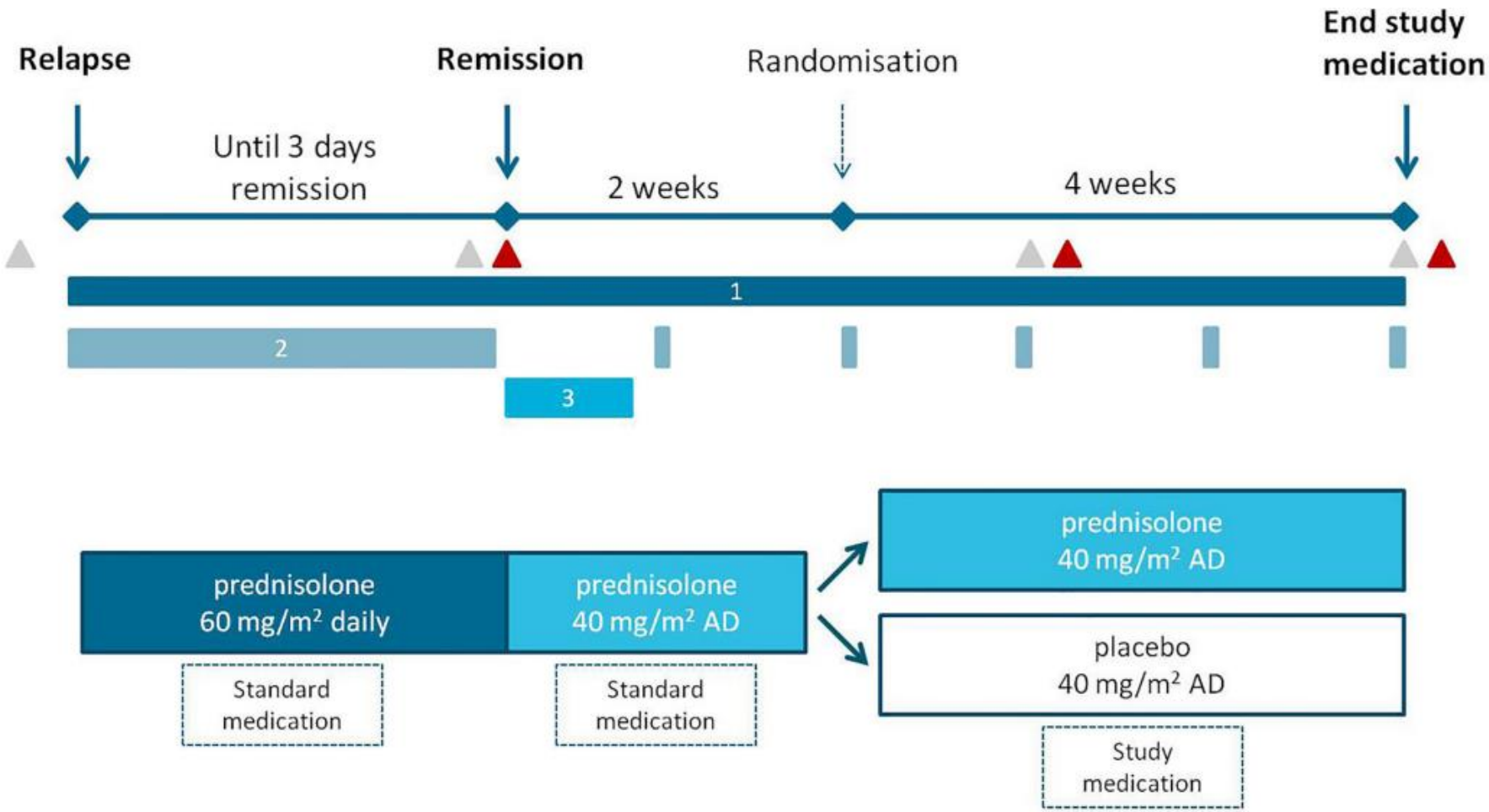
How best to use steroids with relapsing disease?

PROPINE (Gargiulo, 2021)



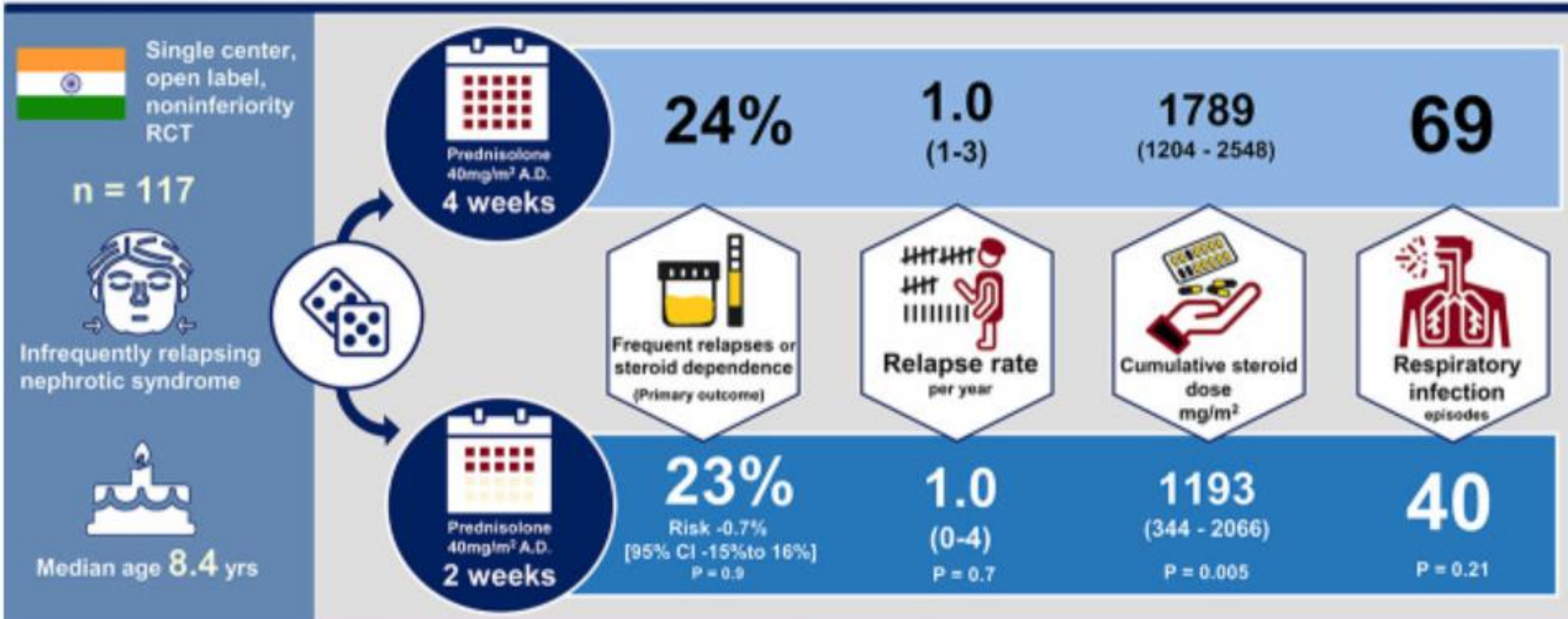


RESTERN (Schijvens, 2017)



Is short-duration prednisolone effective in treatment of nephrotic syndrome relapse in children?

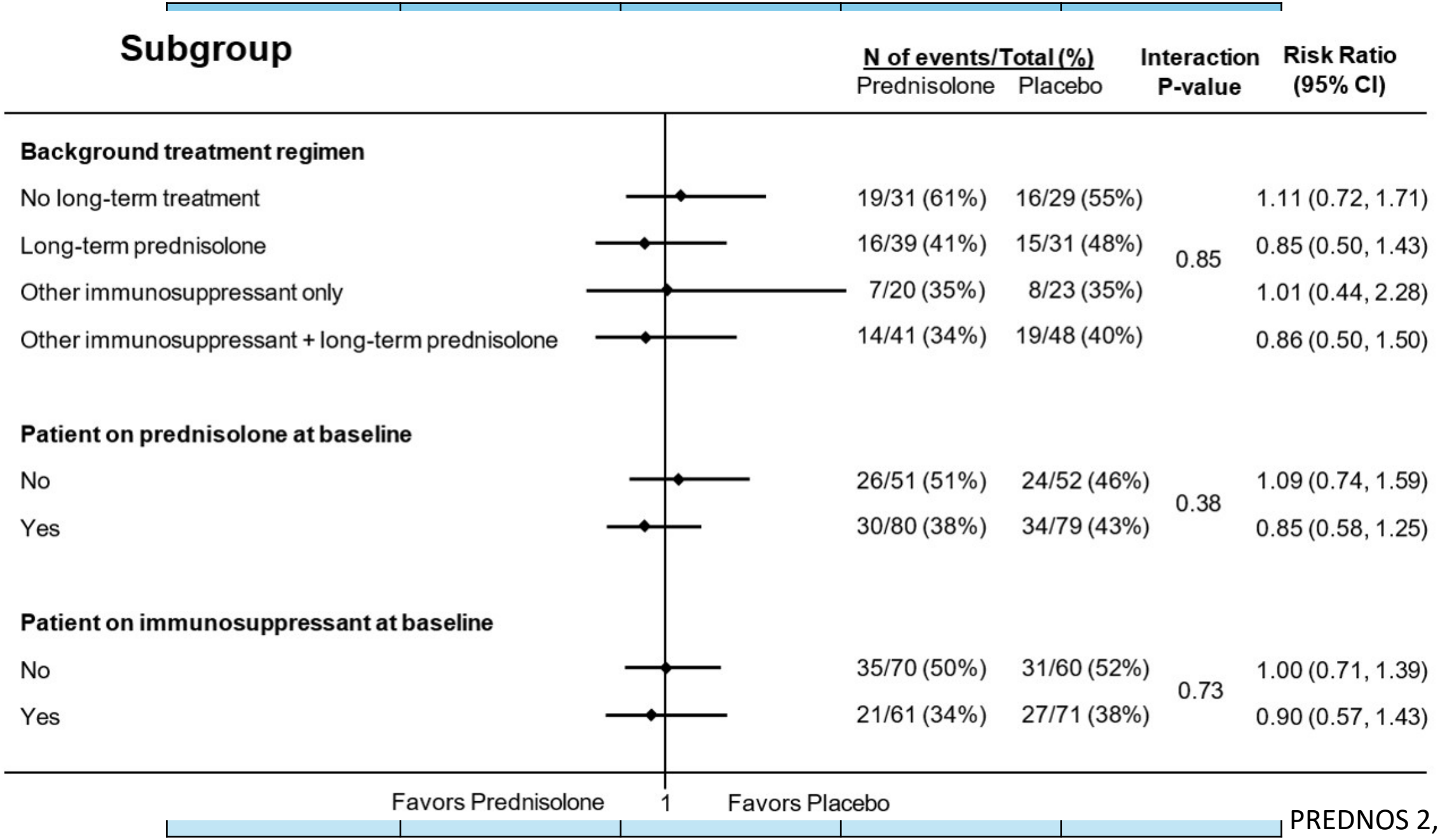
CJASN
Clinical Journal of the American Society of Nephrology



Conclusions: In children with infrequently relapsing nephrotic syndrome, a short steroid treatment for relapse led to a similar proportion of patients developing frequent relapses or steroid dependence; however, its non-inferiority could not be established.

Deepika Kainth, Pankaj Hari, Aditi Sinha, et al. ***Short-Duration Prednisolone in Children with Nephrotic Syndrome Relapse***. CJASN doi: 10.2215/CJN.06140420. **Visual Abstract by Divya Bajpai, MD, PhD**

Kainth, 2021



PREDNOS 2, 2022

Can a lower dose of prednisolone be used effectively to treat relapses?

- Fewer short-term effects and long-term steroid side-effects
- Doses <60 mg/m^2 commonly used *ad hoc* in the UK
- Previous uncontrolled studies have demonstrated effectiveness:
  - Choonara, 1989
  - Raja, 2017
- Current studies are addressing weaning regimes but equivalent cumulative prednisolone dose

European Journal of Pediatrics
https://doi.org/10.1007/s00431-019-03506-5

ORIGINAL ARTICLE

Lower prednisone dosing for steroid-sensitive nephrotic syndrome relapse: a prospective randomized pilot study

Yael Borovitz[1] · Hadas Alfandary[1,2] · Orly Haskin[1] · Shely Levi[1,2] · Shulamit Kaz[1] · Miriam Davidovits[1,2] · Amit Dagan[1,2]

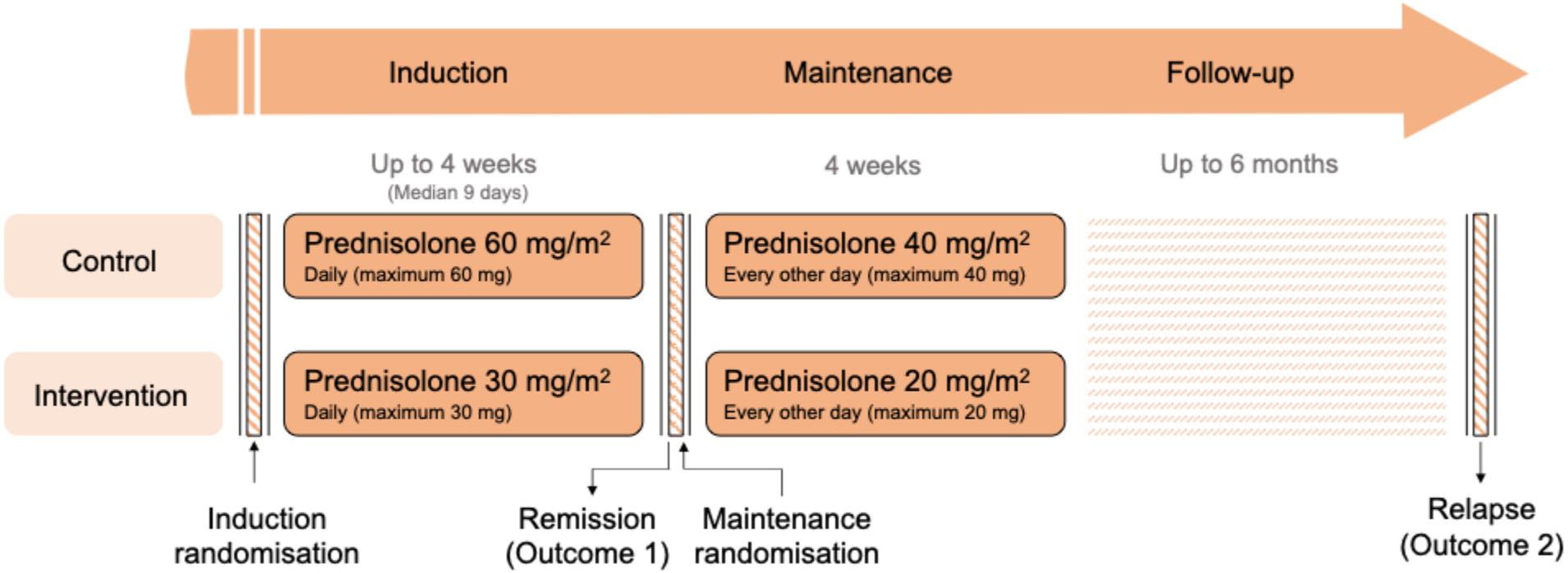
Received: 21 March 2019 / Revised: 10 October 2019 / Accepted: 14 October 2019
© Springer-Verlag GmbH Germany, part of Springer Nature 2019

- 30 children with relapsing SSNS
- Randomised to prednisone 2, 1.5, or 1 mg/kg/day for treatment of relapse
- Time to remission:
  - 7.2 ± 1.4, 10.2 ± 5.1, and 9 ± 3.3 days respectively
- Mean cumulative prednisolone doses:
  - 45.5 ± 3.4, 42.7 ± 25.9, and 24.9 ± 7.4 mg/kg, respectively ($P<0.05$)

Sheikh *et al*, 2019

- 60 children with SSNS
- Randomised to treat relapses with:
  - 1 mg/kg prednisolone (group A)
  - 2 mg/kg prednisolone (group B)
- Mean time to remission:
  - group A: 9.0 ± 2.2 days
  - group B: 8.6 ± 2.2 days
  - Mean difference (95%-CI) 0.4 (0.7-1.6) days; P=0.49]
- Median (IQR) cumulative dose of prednisolone:
  - group A: 12.5 (9-18) mg/kg
  - group B: 17 (14-21) mg/kg
  - P=0.002
- No difference in number of future relapses at 6 months

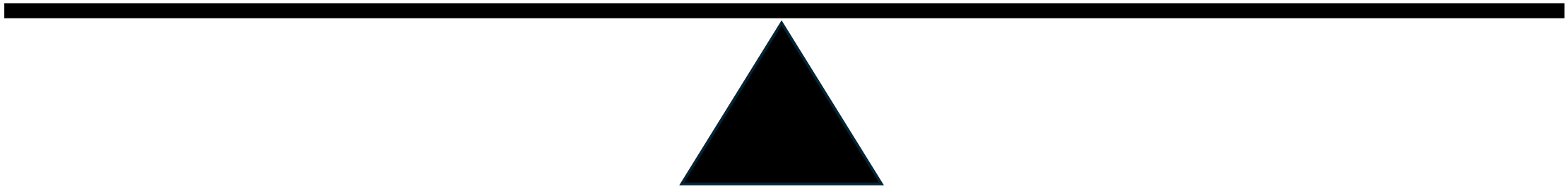
OPEN trial (Larkins et al, Australia)



Is it safe to stop steroids abruptly?

- PNNG advise not stopping steroids abruptly when given at NS doses (2021)
- NICE NG243 (2024): tapering steroids after high-dose for 3 weeks
- BSPED position statement (2026)

- ISKDC regimen used for >50 years
- Minimal reports of children with NS having symptomatic adrenal insufficiency (AI)
- Anecdotal reports of biochemical AI - ?significance



STARS study: Does **S**teroid **T**reatment cause **A**d**R**enal **S**uppression in children with idiopathic Nephrotic Syndrome treated with corticosteroids?

Cohort 1 – patients attending for synacthen tests for other indications (SST)

- Children aged 3-16 attending for a clinically indicated short synacthen test (SST) at one of the participating study hospitals

Cohort 2a – newly presenting with nephrotic syndrome (NS)

Cohort 2b – NS relapse (only if slow recruitment) (rNS)

- Children aged 3-16 diagnosed with iNS, planned to receive at least 4 weeks of corticosteroids at 60 mg/m^2/day as initial treatment
- Children aged 3-16 with RNS requiring repeated courses of high dose steroids

Study schema

Cohort 1

- SST and salivary cortisone (for validation)

Cohort 2a

- Salivary cortisone and symptom questionnaire at:
  - Diagnosis + 6 weeks
  - Diagnosis + 10 weeks
  - Diagnosis + 14 weeks
- Low salivary cortisone triggers SST

Which steroid-sparing drug is best at preventing relapses?

Steroid-sparing drugs: initial recommendations

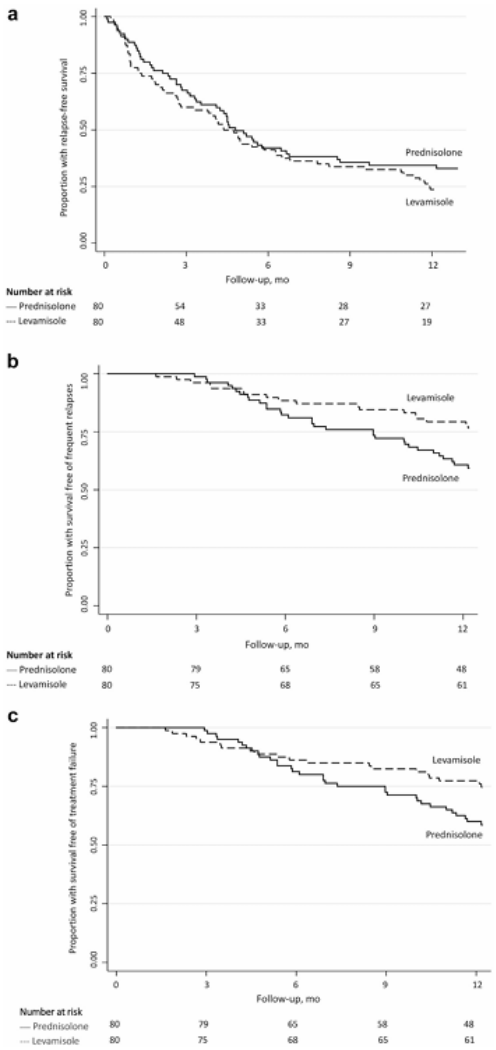
IPNA, 2021	KDOQI, 2025	Indian Soc Ped Neph	Japanese Soc Ped Neph
Low dose pred or any SSA for FRNS Lev/Cyc/CNI/MMF for SDNS	Cyc for FRNS CNI/MMF for SDNS	Cyc/MMF for FRNS and SDNS CNI for difficult to treat SDNS	No evidence to support one over another

UK harmonisation

INS 1.15	We suggest that for children with frequently relapsing, but not steroid dependent nephrotic syndrome, the maintenance treatment should use a personalised approach to conside one of the following treatments: 1. Levamisole 2. Mycophenolate mofetil 3. Tacrolimus (preferred calcineurin inhibitor)	**2C**
INS 1.16	We suggest that for children with steroid dependent nephrotic syndrome, where there are no contra-indications, the first choice of a steroid-sparing agent should either be mycophenolate mofetil or tacrolimus.	**2D**
INS 1.17	We suggest that for children with frequently relapsing INS or steroid dependent disease, a course of oral cyclophosphamide may be another therapeutic option with consideration of its side effect profile including the risk of infertility.	**2D**
INS 1.12	We suggest that children who experience a first relapse of nephrotic syndrome whilst taking the initial corticosteroid course or within 2 weeks of stopping could be treated with prednisolone 60 mg/m^2 per day (maximum dose 60 mg) once daily until remission followed by 40 mg/m^2 (maximum dose 40 mg) on alternate days for 4 weeks and then tapered over at least a further 8 weeks, and commence on either tacrolimus or mycophenolate mofetil as a steroid-sparing agent.	**2D**

Levamisole vs prednisolone Sinha, 2024

- Prednisolone given alternate day, increased to daily during infections
- Prednisolone arm non-inferior
- Levamisole use reduced cumulative prednisolone use



Levamisole-containing medicinal products - referral

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Current status
CMDh final position

Referral | Human

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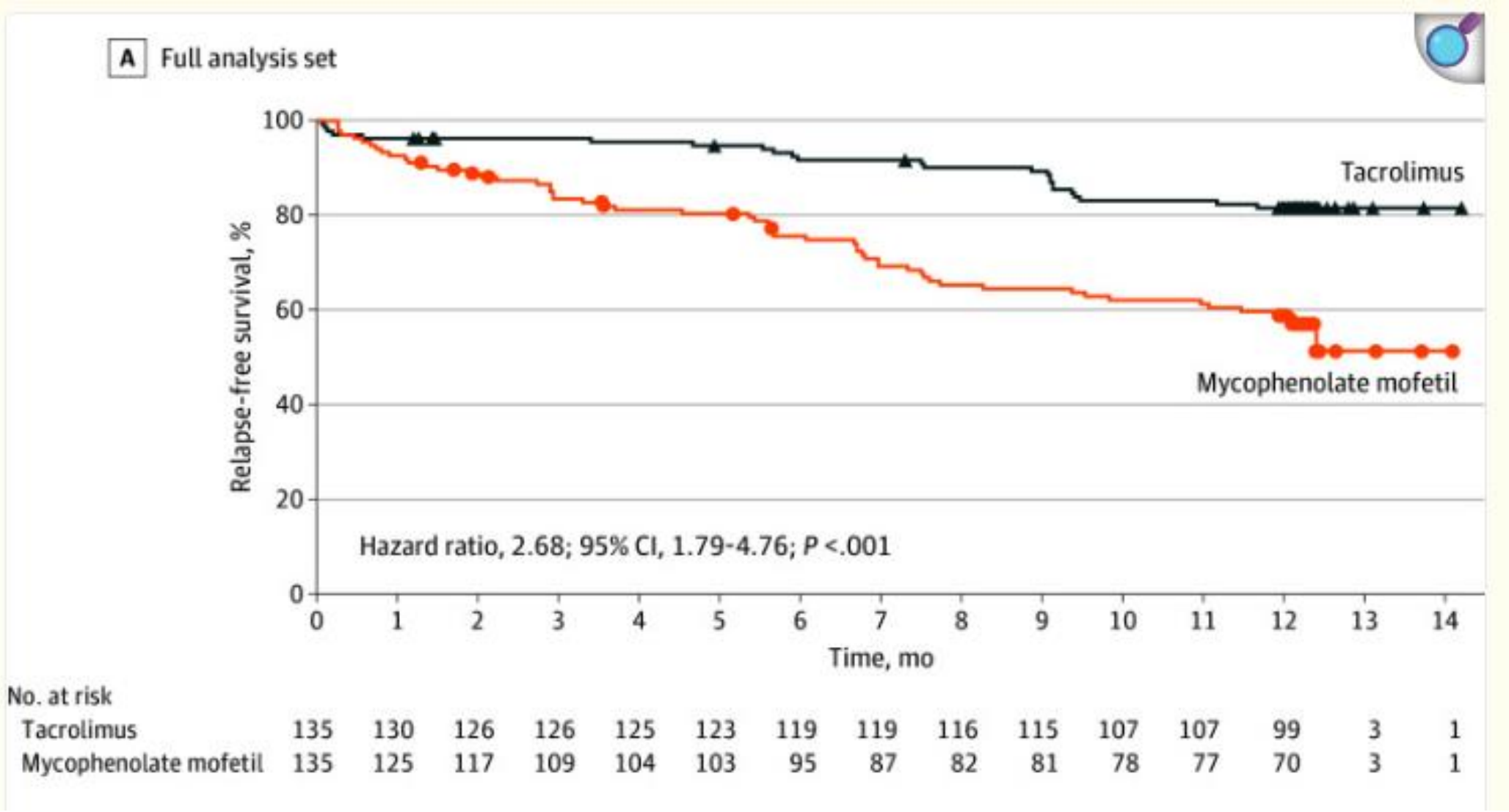
Procedure started → Under evaluation → Recommendation provided by Pharmacovigilance Risk Assessment Committee → **CMDh final position**

Overview

EMA recommends withdrawal of marketing authorisations for levamisole medicines.

Leukoencephalopathy confirmed as a serious side effect of levamisole.

Tacrolimus versus MMF



Wang, 2025

Target trial emulation in paediatric research: how can causal effects be estimated from observational data?

Cal H Robinson, Eddy Fan, Sonia M Grandi, Martin Urner, Rulan S Parekh

Conducting randomised controlled trials (RCTs) in rare paediatric diseases is often impractical or prohibitively expensive. Observational data from longitudinal cohort studies, disease registries, and population-based databases exist for children and adolescents, but standard observational analyses are typically limited by bias. Target trial emulation methods can improve the quality of observational analysis, address common sources of bias, and help fill evidence gaps in paediatric clinical practice. Applying target trial emulation methods in paediatric research creates unique opportunities, but also poses specific challenges. Tailored approaches are needed to address issues with small sample size, treatment-effect heterogeneity, longitudinal follow-up, and missing data. This Review aims to outline key concepts, such as what causal inference and target trial emulation are, justify the use of target trial emulation methods in paediatric observational research, discuss approaches to emulating key elements of an RCT protocol, and highlight unique paediatric applications of target trial emulation.

Lancet Child Adolesc Health 2025; 9: 663–72

Published Online July 11, 2025 https://doi.org/10.1016/S2352-4642(25)00131-2

Division of Nephrology, The Hospital for Sick Children, Toronto, ON, Canada (C H Robinson MD, Prof R S Parekh MD MS); **Child Health Evaluative Sciences, Research Institute, The Hospital for Sick Children, Toronto, ON, Canada** (C H Robinson, S M Grandi PhD, Prof R S Parekh); **Institute of Health Policy, Management and Evaluation, University of Toronto, Toronto, ON, Canada** (C H Robinson, E Fan MD PhD, M Urner MD PhD, Prof R S Parekh); **SickKids Research Institute, Peter Gilgan**

Introduction

Randomised controlled trials (RCTs) are the gold standard for evaluating comparative pharmacological treatment efficacy in a controlled study and provide the evidence used for regulatory approvals, health technology assessments, and insurer funding decisions. However, multiple barriers limit the conduct of traditional RCTs in rare paediatric conditions.[1–3] Thus, compared with adult populations, few paediatric and rare disease RCTs are conducted to evaluate vaccine effectiveness; medications to treat infections, cystic fibrosis, inflammatory bowel disease, and psychiatric conditions; neurodevelopmental outcomes of perinatal interventions; sports injury prevention; and public health initiatives.[22–32] However, paediatric applications of target trial emulation methods are not yet widely used. Given the unique considerations regarding clinical research in the paediatric population—such as smaller sample size, genetic causes of disease,

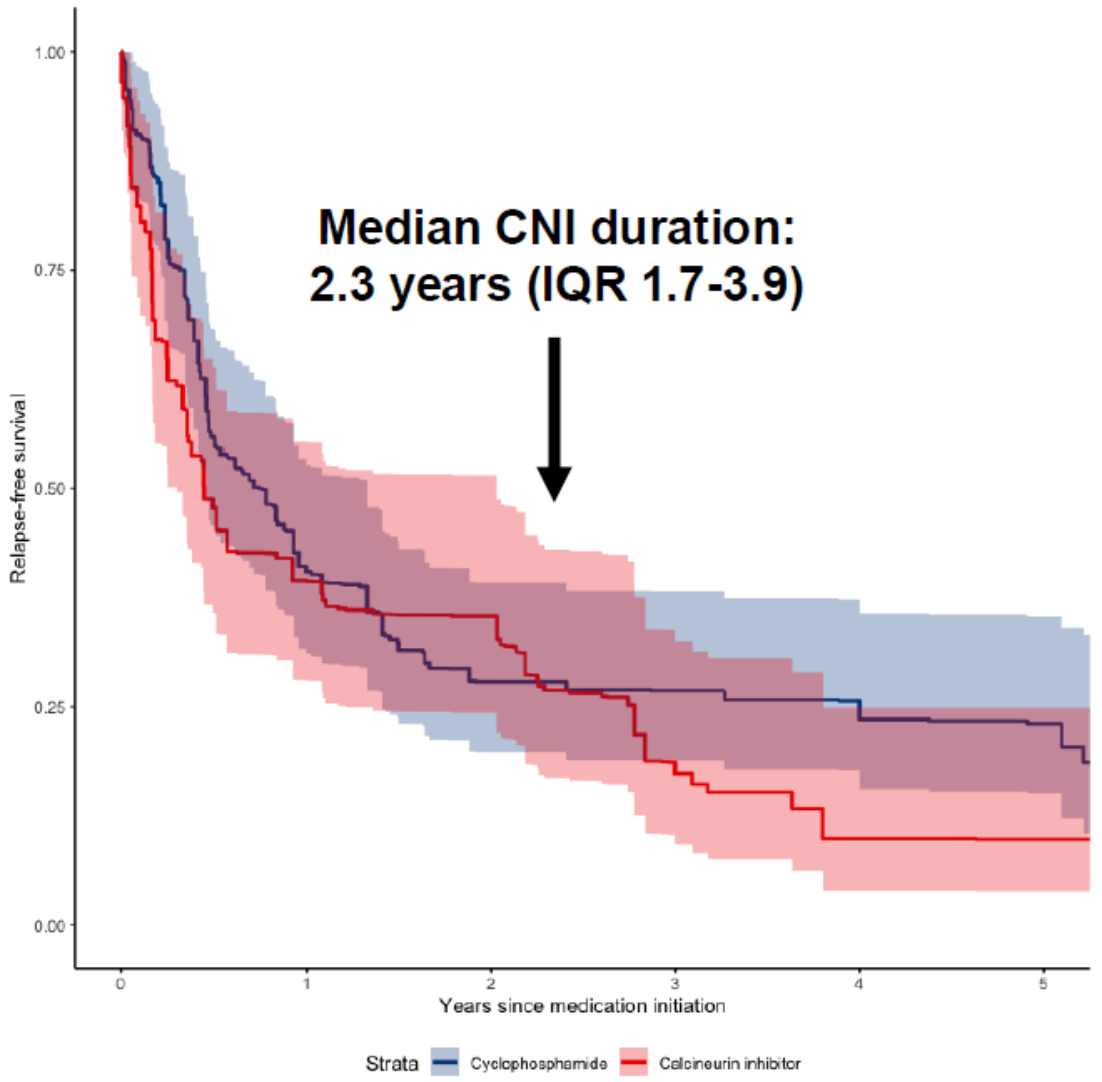
INSIGHT target trial emulation

Robinson et al. JAMA Peds (2025)

- 578 children with nephrotic syndrome
  - 252 initiated **cyclophosphamide**
  - 131 initiated **calcineurin inhibitors**
  - Median 5.5-year follow-up

- No difference in time-to-relapse

Outcome	Weighted effect estimate [95% CI]
Time-to-relapse	**HR 1.25 [0.84-1.87]**
Relapse count by 5-years	RR 1.04 [0.69-1.57]
Incident CKD	HR 1.29 [0.25-6.68]
Subsequent steroid-sparing med	HR 0.73 [0.45-1.18]
Hospitalization	HR 1.83 [1.14-2.92]
IV albumin use	HR 2.81 [1.65-4.81]



CONSIST – multiple interventions by trial emulation

Study	INSIGHT	CureGN	NEPTUNE	NEPHRO-VIR	H3 Africa	PodoNet	NURTuRE-INS	PREDNOS 2	KCL cohort	Israeli cohort	Hong Kong cohort	Rome cohort	Total cohort
Design	Pro	Pro	Pro	Pro	Pro	Pro	Pro	RCT	Retro	Pro	Retro	TBD	**Mixed**
Cohort	NS	Biopsy NS	NS ± biopsy	NS	NS	SRNS	NS	SSNS	NS	NS	NS	NS	**Mixed**
Region	6 centers; Canada	70 centers; mostly US	32 North American centers	39 centers; Paris, France	15 centers in 5 African countries	82 centers, 33 countries	23 centers; UK	125 centers; UK	Single center; UK	3 centers; Israel	Single center; Hong Kong	Single center; Italy	**Global**
Period	1993 - ongoing	2015 - ongoing	2009 - ongoing	2007 - 2010	2019 - ongoing	2009 - ongoing	2017 - 2023	2011-2014	2004 - ongoing	2014-ongoing	Ongoing	Ongoing	**1993-ongoing**
Study recruitment													
Total	860	494	402	273	626	1300	739	237	600	157	250	TBD	**>6000**
SSNS	830	390	320	250	595	0	518	237	550	144	250	TBD	**~4000**
SRNS	30	104	82	23	31	1300	221	0	50	13	0	TBD	**~2000**
Steroid-sparing immunosuppressive treatment													
CYP	307	67	19	15	TBD	95	TBD	60	TBD	TBD	TBD	TBD	**~800**
CNI	254	416	76	30	TBD	700	TBD	36	TBD	TBD	TBD	TBD	**~2400**
MMF	185	203	33	170	TBD	200	TBD	28	TBD	TBD	TBD	TBD	**~1300**
RTX	100	224	3	70	TBD	70	TBD	6	TBD	TBD	TBD	TBD	**~700**
LEVA	23	0	0	200	TBD	0	TBD	69	TBD	TBD	TBD	TBD	**~400**

Rituximab

- Serendipitous discovery in 2004
- Reorientated pathophysiology of NS
- MoA involves multiple cytotoxic pathways, including:
  - complement-dependent cytotoxicity
  - antibody-dependent cellular cytotoxicity
  - induction of phagocytosis and apoptosis
- Depletes naïve and memory B-cells, but not plasma cells

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rituximab and nephrotic and child

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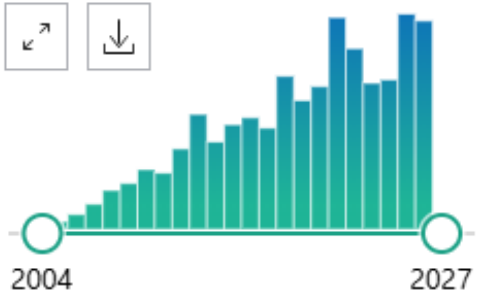
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552 results

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RESULTS BY YEAR



PUBLICATION DATE

- 1 year
- 5 years

9 articles found by citation matching

Rituximab-induced serum sickness with idiopathic nephrotic syndrome in a child.
Majima H, et al. Pediatr Int. 2022. PMID: 34657352 No abstract available.

Rituximab-induced serum sickness and anaphylaxis in a child with nephrotic syndrome.
Bayram MT, et al. Turk J Pediatr. 2020. PMID: 33108096

Rituximab-induced Crohn's disease in a child with refractory nephrotic syndrome.
Machida S, et al. Pediatr Int. 2022. PMID: 36271771 No abstract available.

Show all

Rituximab is effective in nephrotic syndrome

Reference	N	F/U (mo)	Study design	Dose	CR (%)
SDNS/FRNS					
Ravani et al. [53]	46	12	Off–on trial	375 mg/m^2 (1–5 doses)	48
Iijima et al. [18]	48	12	RCT	375 mg/m^2 (4 doses) vs. placebo	81
Ruggenenti et al. [19]	30	12	Off–on trial	375 mg/m^2 (1–2 doses)	100
Ravani et al. [54]	30	24	RCT	375 mg/m^2 vs. steroid	66 at 12mo, 34 at 24mo
Chan et al. [20]	346	24	Multicenter, retrospective cohort study	375 mg/m^2 or 750 mg/m^2. Multiple courses (2 to more than 7)	-
Gauckler et al. [21]	183	36	Retrospective	1000 mg (1 or 2 doses) or 375 mg/m^2 (1 to 4 doses)	Initial CR/PR at 6 mo: 82%; long-term relapse-free survival among responders: 55% at 3 years
Laslandes et al. [22]	134	36	Multicenter retrospective cohort	Variable regimens; maintenance RTX at 6–12 mo	Relapse rate reduced from 1.02 to 0.20 per pt-year; maintenance RTX associated with lower relapse risk (HR 0.35)
SRNS					
Magnasco et al. [25]	31	12	RCT	375 mg/m^2 (2 doses) vs. prednisone and calcineurin inhibitors	6 delayed-resistant patients achieved remission (3 RTX, 3 control)
Chan et al. [26]	246	32.4 (mean)	Retrospective	From 375 to 1500 mg/m^2	-
Gauckler et al. [21]	183	36	Retrospective	1000 mg (1 or 2 doses) or 375 mg/m^2 (1 to 4 doses)	-

Bigatti et al,B-cell-targeting therapies in podocytopathies, *Pediatr Nephrol*, 2026

Clinical and immunological factors that determine the treatment outcomes after rituximab in children with SDNS

- Older age at rituximab[51–54]
- White children[51]
- Use of maintenance immunosuppression after rituximab, e.g., MMF[69]
- Repeated courses of rituximab[50]
- Lower mitogen-stimulated T-cell subsets at baseline[25]

Lower risk of relapse



Histology[7,41,48] and high-dose rituximab[50] do not alter relapse risk



Higher risk of relapse

- East Asian, South Asian, and Black children[51]
- History of steroid resistance[48,51]
- Multidrug dependence[50,51]
- Low-dose rituximab without maintenance therapy[50]
- Repopulation of total memory B cells, especially switched memory B cells[14–16]

Chan *et al*, cJASN, 2023

Rituximab toxicity

- Infusion reactions
- Anti-drug antibodies
- Late-onset neutropenia
- Attenuated vaccine responses
- Hypogammaglobulinaemia

OPTIMIZING PATIENT CARE… PDATED DELPHI CONSENSUS RECO… OF SECONDARY HYPOGAMMAGLOBULINA… DISEASES

Mohammed Yousuf Karim,[1] Ramsay L Fuleihan,[2] Silvi… [7] Hirokazu Kanegane,[8] Samia J Khoury,[9] Sorena Kiani-Alikhan,[10] Viviana Moschese,[11] Christian… Chetan B Mukhtyar[16]

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TABLE 1. FINAL CONSENSUS STATEMENTS FOR THE MANAGEMENT OF SECONDARY HYPOGAMMAGLOBULINAEMIA IN PATIENTS WITH AUTOIMMUNE DISEASES

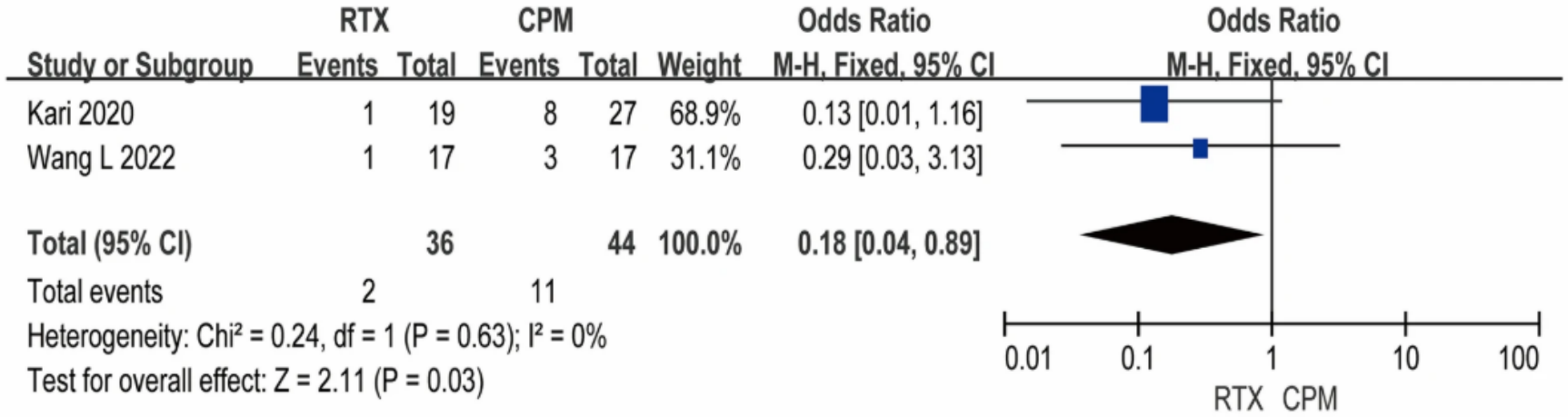
Theme	Statement	Agreement	Participation
Risk factors for, and susceptibility of, developing hypogammaglobulinaemia	All patients who receive BCTT should have immunoglobulin (i.e. IgG, IgA, IgM) levels assessed before and during treatment, either before each infusion or every 6 months (whichever is earlier), and after completion of treatment (every 6 months for a minimum of 1 year after or until IgG returns to normal)	95%	92%
	All patients commencing BCTT should have evidence-based, patient-specific, disease-specific and treatment-specific risk factors assessed, to allow stratification of hypogammaglobulinaemia risk and inform monitoring schedules	94%	92%
Risk of hypogammaglobulinaemia with newer BCTT drugs and biosimilars	In the absence of robust evidence, it is reasonable to extrapolate the extensive data available relating to rituximab's impact on hypogammaglobulinaemia to rituximab biosimilars and other, newer anti-CD20 therapeutics	92%	92%
	Early studies on novel approaches – such as CAR T-cell therapies, tyrosine kinase inhibitors and plasma-cell therapies – indicate that they may cause hypogammaglobulinaemia in patients with autoimmune disorders. However, more evidence is needed to confirm the causative effect and to establish the prevalence and clinical impact of hypogammaglobulinaemia in patients receiving these novel therapies	97%	92%
Monitoring hypogammaglobulinaemia	Hypogammaglobulinaemia is defined as an IgG level below the local laboratory age-specific range, with or without low IgA and/or IgM	84%	92%
	Patients receiving BCTT should have immunoglobulin (i.e. IgG, IgA, IgM) levels checked, alongside a detailed history and examination for severe, recurrent, atypical and/or sinopulmonary infections both at baseline and prior to each cycle of treatment, or at least every 6 months, whichever is earlier	98%	87%
Immunological assessment besides hypogammaglobulinaemia	Patients should be assessed for immunological abnormalities beyond hypogammaglobulinaemia as often as possible, but no less than twice a year	88%	87%
	Biannual assessment should include measurement of lymphocyte subsets (including B- and T-cell enumeration) and neutrophil counts to consider late-onset neutropenia. Additional assessments may be performed where clinically indicated and may include naïve/memory (CD27^{-}/$^{+}$) B-cell numbers	75%	87%
	Prior to commencement of BCTT, patients should undergo a detailed clinical and laboratory evaluation, including detailed immune assessments. Immune assessments should include immunoglobulin (i.e. IgG, IgA, IgM) levels and lymphocyte subsets. If there is baseline hypogammaglobulinaemia, or the patient is at high risk of developing hypogammaglobulinaemia, a specialist opinion should be considered	92%	87%

IgRT initiation, dosing and route of administration	IgRT should be initiated in patients with hypogammaglobulinaemia, considering patient preferences for method of administration. Guideline initiation parameters are serum IgG levels: • < 1.5 g/L, regardless of clinical features • 1.5–4 g/L with hypogammaglobulinaemia-associated severe infections[b] OR repeated sinopulmonary infections OR failure to respond to vaccines • 4–6 g/L with hypogammaglobulinaemia-associated severe infections OR repeated sinopulmonary infections AND failure to respond to vaccines • 1.5–6 g/L with bronchiectasis	83%	87%

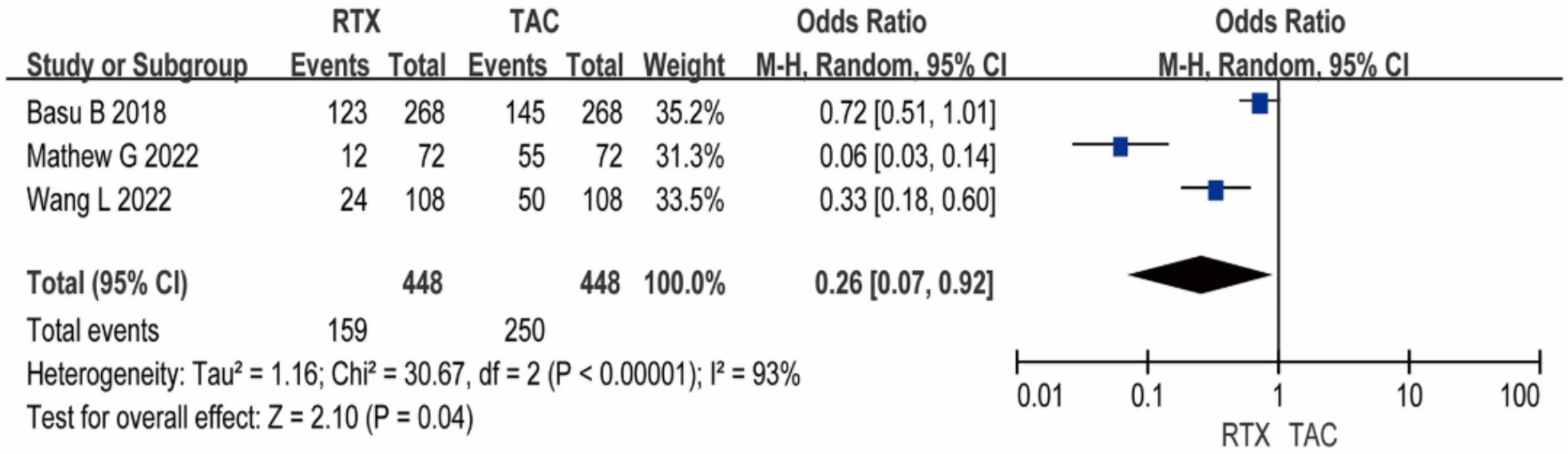
When to stop IgRT	Following discontinuation of IgRT, patients should be closely monitored for infections, and immunoglobulins should be checked every six months for a minimum of one year after. If required, IgRT reinstatement should be considered in line with the recommendations for initiation in these guidelines	95%	87%
Types of vaccines, timing of administration, age at vaccination, and timing after diagnosis	Prior to commencing BCTT, patients should have an up-to-date vaccination record according to the patient's history and routine age-appropriate local vaccination schedule, where possible	95%	87%
	Where possible, we recommend that patients with hypogammaglobulinaemia related to BCTT undergo vaccine challenges[a] to determine the level of functional humoral immunosuppression and reduce the risk of severe infections. The utility of each challenge should take into account the timing of BCTT doses, as these are known to blunt antibody responses for several months	88%	87%
	Vaccine challenges should include an assessment of the degree of antibody deficiency and a minimum of tetanus and polysaccharide pneumococcal vaccines where appropriate, with interpretation by an appropriate specialist.[a] The risk/benefit profile should be considered on an individual basis, according to previous vaccine and infection history (e.g. whether commencement should be delayed to enable vaccination challenge; or if IgG < 1.5 g/L, vaccination as a means of assessing response may not be needed)	88%	87%
Role of antimicrobial prophylaxis	Antimicrobial prophylaxis may be considered to prevent specific infections in patients with autoimmune diseases treated with immunosuppressive therapy, but there are few data in autoimmune patients with hypogammaglobulinaemia related to BCTT to compare its utility with IgRT. Antimicrobial prophylaxis may be used on a short-term basis while a patient with hypogammaglobulinaemia is being evaluated	77%	98%
	Local epidemiology, adverse events, antimicrobial resistance and gut dysbiosis, when available, should be considered in the management of patients being treated with antimicrobial prophylaxis	84%	98%
	In hypogammaglobulinaemia related to BCTT, antimicrobial prophylaxis for bacterial infections may be considered where IgRT is not available or contraindicated for any reason[a]	90%	84%
Monitoring patients for infections	As clinically significant hypogammaglobulinaemia and previous/long-term immunosuppression are risk factors predictive of severe infections, patients should be closely monitored for infections, particularly sinopulmonary infections, during and following treatment with BCTT, especially if they remain hypogammaglobulinaemic	100%	87%
Management of pregnant patients and considerations for the management of neonates	As there is little clinical evidence in this patient population, patients with autoimmune disorders planning for pregnancy should be evaluated by a multidisciplinary team of clinicians and undergo close immunological assessment, with BCTT only initiated or continued following a careful risk/benefit assessment	100%	87%
	Babies born to mothers with autoimmune disorders who have received BCTT, especially during the second or third trimester, should be evaluated for hypogammaglobulinaemia and B-cell deficiency, and expert advice from specialist physician[a] should be sought regarding immunologic evaluation, preventative treatment and timing of childhood vaccines	95%	87%

From: Rituximab in pediatric refractory nephrotic syndrome: a systematic review and meta-analysis evaluating therapeutic efficacy and adverse event profiles

a



b



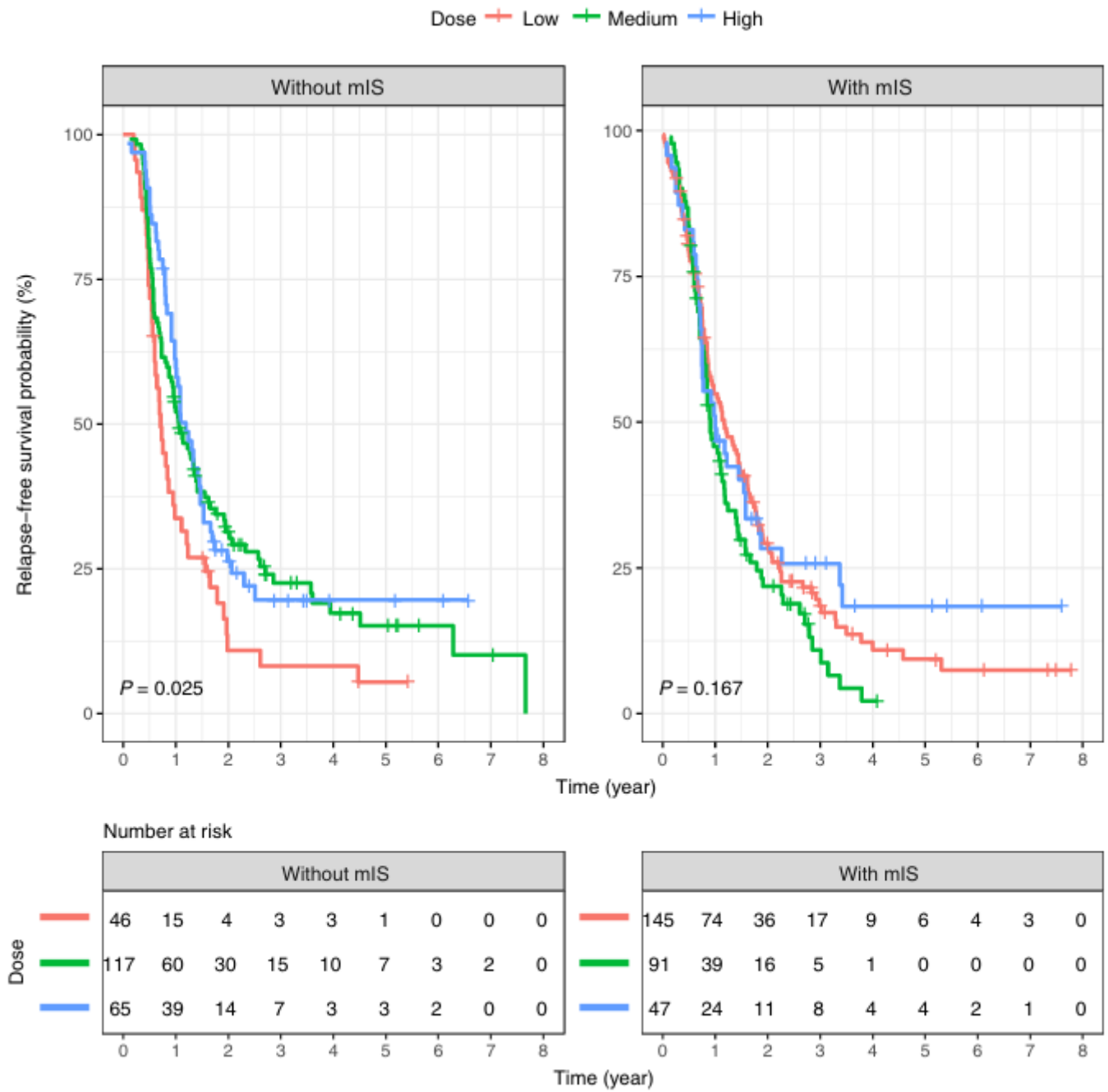
Meta-analysis results of the incidence of adverse events for RTX in RNS. **a** Adverse event incidence rate of RTX and CPM. **b** Adverse event incidence rate of RTX and TAC

outcomes for RTX versus TAC

Deng, 2026

Outstanding questions with rituximab

- What's the optimal dose?
- To use with or without concomitant immunosuppression?
- Where should it fit in the algorithm?
- Are repeat doses safe?
- Is it equally effective/safe in all ages?



Low: 375 mg/m^2
Medium: 750 mg/m^2
High: 1125-1500 mg/m^2

Chan, Kidney Int, 2020

Long-term efficacy and safety of repeated rituximab to maintain remission in idiopathic childhood nephrotic syndrome (NS): an international study

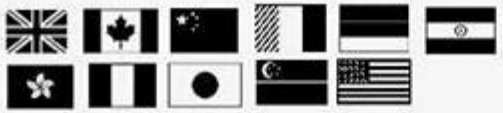
JASN
JOURNAL OF THE AMERICAN SOCIETY OF NEPHROLOGY

METHODS

Retrospective cohort study

346 children with NS
With ≥2 rituximab courses

16 Paediatric Nephrology centres



1149 rituximab courses

Follow-up 5.9 years (IQR 4.3-7.7)

OUTCOMES

	Rituximab course 1st	2nd	3rd	4th	
Relapse-free period, m	10 [9.0-10.7]	13.5 [12.3-16.0]	14.3 [12.3-16]	16.0 [13.3-19.7]	p<0.001
Relapse risk, HR_{adj}	1 [REF]	0.13 [0.10-0.18]	0.06 [0.04-0.09]	0.03 [0.02-0.06]	p<0.001
Time to B-cell repletion, m	6.1 [6.0-6.3], NS				
Significant hypo-IgG (11%) Infection (5%) Neutropenia (4%)	NS				

Conclusion: Children receiving repeated courses of rituximab experience an improving clinical response. Side effects appear acceptable but significant complications can occur. These findings support repeated rituximab use in childhood nephrotic syndrome.

Chan, 2022
doi: 10.1681/ASN.2021111472

Long Term Efficacy and Safety of Rituximab vs Tacrolimus in Children With Steroid Dependent Nephrotic Syndrome: The RITURNS Follow-Up Study

RITURNS trial

Children with SDNS N= 117

1st year relapse free survival

RTX 90%

TAC 63%

p<0.001

2 year follow-up

Original arm	Time to first relapse (weeks)	2nd line treatment*	Relapse free survival (weeks)	Overall 2-yr relapse-free survival
RTX	63 (60-71)	RTX + MMF <52 weeks**	1st line RTX 63 (60-71)	RTX 9%
		RTX + MMF 52-80 weeks**		
		RTX >80 weeks**		
	p=0.49		p<0.01	p<0.0001
TAC	59 (54-69)	RTX 1st relapse	55 (51-59) 2nd line RTX	RTX + MMF 67%
		RTX + MMF 2nd relapse		

*after relapse **time when the 1st relapse ocurred

SDNS, steroid dependent nephrotic sindrome; RTX, Rituximab; TAC, tacrolimus; MMF, Mycophenolate mofetil

KI REPORTS
Kidney International Reports

Basu B et al, 2023

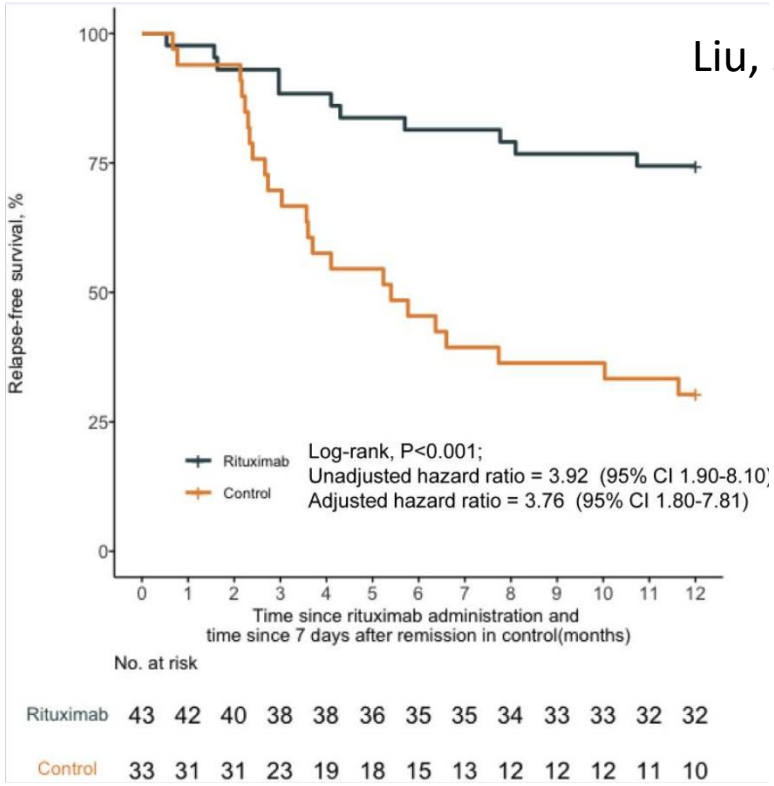
Visual abstract by:
Denisse Arellano, MD
@deniise_am

Conclusion The superior therapeutic effect of Rituximab in SDNS vanishes during the second year postexposure. Rituximab appears to yield longer remission when applied as 1st line as compared to 2nd line therapy. Maintenance MMF following Rituximab induces long-term disease remission.

Rituximab at first remission?

- 44 children with new NS
- Median age 4.3 y
- Prospective open-label, single arm
- Single dose 375 mg/m^2 given one week after remission
- Historic controls
- RFS at 12 m: 74 v 30%
- No significant toxicity

Liu, 2024



Should rituximab be the first steroid-sparing drug?

NO

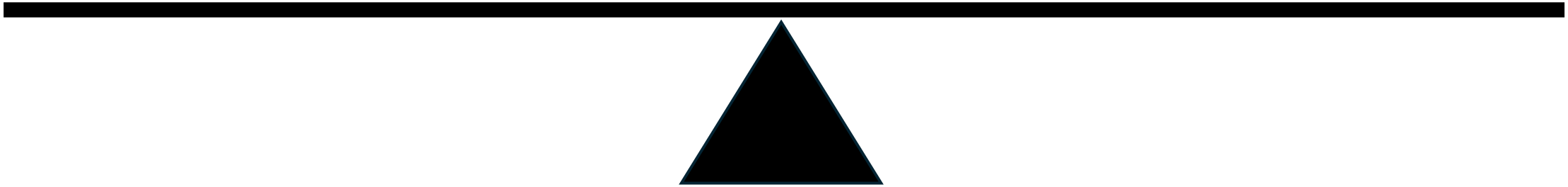
- Effect at 2 years is no better than other SSDs
- Memory B-cell repertoire takes until 10 years to reach stable repertoire
- Potential reduction in vaccine competency

Vivarelli & Colucci, 2025

YES

- Good safety profile compared to other SSDs
- Efficacy likely better when given early
- Hypogammaglobulinaemia risk may relate more to repeat doses and concomitant i/s

Dossier & Hogan, 2025



Efficacy and Safety of Rituximab Therapy to Sustain Remission in Young Children with Childhood Nephrotic Syndrome

Retrospective multicentre cohort
16 centres, 10 countries

Primary analysis
n=101

Matched comparison
<6years vs 6-21years
n=70 each

Efficacy

Median relapse-free survival: 8.8 months (95% CI 7.5-11.0)

Each month of concurrent immunosuppression reduced relapse risk by 8% (HR 0.92)

Annual relapse rate: 3→1
p<0.0001
84% needed repeat dosing

Safety

Hypogammaglobulinemia **57%**

Neutropenia **9%**

Infection **8%**

Low baseline IgG predicted hypogammaglobulinemia post-rituximab

Vs. older children

Earlier relapse
8.3 vs 11.3 months; *p=0.14*

More:

Hypogammaglobulinemia
63% vs 35%; *p=0.01*

Neutropenia 13% vs 7%

Infection 9% vs 4%

KI REPORTS
Kidney International Reports®

Conclusion: Reasonable efficacy with shorter relapse-free survival and higher complication burden versus older children. Reserve for refractory cases; monitor IgG closely.

Chan EY et al., 2026. VA by Cristina Popa, MD.

Obinutuzumab

- Second-generation, fully humanised anti-CD20
- Desgined to overcome rituximab failure in B-cell malignancies
- Glycoengineered to enhance interaction with Fc-gamma receptors to resist internalisation resulting in more potent B-cell depletion
- Increased efficacy over rituximab demonstrated in lupus
- Paediatric NS cohort data suggest low-dose obinutuzumab may be more efficacious than rituximab in preventing relapses
- Some UK experience – do we need to write up?

INShore

- Randomised (1:1) open label trial of obinutuzumab vs MMF
- 85 individuals with iNS aged 2 to 25 with SDNS or FRNS
- Primary outcome: % in each arm with sustained CR at 1 year
- Obi 1g x 2 doses week 0 & 2 and repeated week 24 & 26 (20 mg/kg in <45 kg)
- MMF 600 mg/m^2 bd for 52 weeks
- Roche reported in Oct 2025 that primary outcome was met: https://www.roche.com/media/releases/med-cor-2025-10-28

OBIRINS

Dossier *et al* (protocol: BMJ Open, 2025)

- French multi-centre study
- DBRCT, 1:1
- Children aged 3 to 17 with SDNS or FRNS
- Ritux 375 mg/m^2 single dose vs obi 300 mg/m^2 single dose
- Primary outcome: occurrence of first relapse within 12 m
- Recruitment from October 2023, aiming to close in October 2025 and for study completion in October 2027

What do we want for a child with newly-presenting nephrotic syndrome?

- Cure at first presentation
- Long-term remission
- Minimise relapse frequency
- Minimise drug toxicity

How do we predict disease course early on?

Clinical features that predict disease outcome

- Initial steroid sensitivity in children with steroid-resistant nephrotic syndrome predicts post-transplant recurrence (Ding, 2014)
- Factors predicting risk of frequent relapses in 277 children followed for one year from diagnosis (Dakshayani, 2018):
  - Young age at onset (under 5)
  - First relapse within 6 months
  - Relapses associated with infection
- Time to initial remission predicts relapsing course (Sinha, 2026)

Biomarkers with prognostic potential

Biomarker	Sample	Key Finding/Performance	Clinical Role	Ref(s)
Anti-nephrin IgG	Serum	Seroprevalence 68/28/14/2% (SSNS/SDNS/non-genetic SRNS/genetic SRNS); positivity in non-genetic SRNS predicts response to intensified immunosuppression	Diagnosis; predicts IS response	[7,14,15,16]
Other podocyte autoantibodies (e.g., anti-vinculin)	Serum	≥1 autoantibody in 66%; titres track disease activity	Diagnosis	[17]
Urinary NGAL (±VDBP)	Urine	AUC ≈ 0.90 for SRNS; uNGAL < 10 ng/mL = 100% NPV for SRNS at first episode	Predicts steroid resistance at onset	[18,19]
Urinary CD80	Urine	Elevated in MCD/SSNS relapse; low in FSGS/SRNS	Marks steroid-sensitive/MCD phenotype	[20,21]
Podocyte microparticles (nephrin, podocin)	Urine	AUC up to 0.90 vs. controls; correlate with proteinuria	Marks active podocyte injury	[22]
Urinary suPAR	Urine	AUC ≈ 0.70; higher in SRNS relapse	Associated with steroid resistance	[23]
Cytokine panel (IL-7/IL-9/MCP-1)	Plasma	AUC 0.85 for SRNS	Predicts steroid resistance	[24]
Serum TNF-α	Serum	Falls with remission in SSNS, highest in SDNS; higher baseline → longer time to remission	Disease-activity monitoring	[25]
Unswitched memory B cells	Blood	OR 3.6; >1.5% cut-off, AUC 0.79	Predicts 12-month relapse in SSNS	[26]
Class-switched memory B cells	Blood	More abundant in relapse after B-cell depletion	Predicts relapse post-rituximab (SDNS)	[27]
$CD19^+$ B cells/$CD8^+$ T:$CD19^+$ B ratio	Blood	AUC 0.72 (SRNS); ratio AUC 0.81 (SDNS)	Phenotype discrimination	[28]
Exploratory: metabolomics, miRNA, EVs, ANGPTL3, HSP70, fatty acids	Serum/Urine	Investigational; some high AUCs (e.g., uANGPTL3 AUC 0.98)	Hypothesis-generating	[29,30,31,32,33,34,35,36,37]
Anti-PSMA1 autoantibodies	Serum	AUC 0.904 for INS vs. controls; tracks proteinuria; validated in independent cohort	Diagnosis; autoimmune podocytopathy	[38]
Extrafollicular/atypical B cells	Blood	Expanded in active INS; ablated by rituximab; MZ-like resurgence marks post-RTX relapse	Mechanism; relapse monitoring	[39]
Plasma proteomic panel (VDB/ADIPOQ/MMP-2)	Plasma	AUC 0.78 for SRNS at presentation; validation cohort $n = 37$	Predicts steroid resistance	[40]

Lazaruc, Med Sci (Basel), 2026

Higher circulating levels of memory B-cells prior to rituximab predict risk of relapse

Table 1 | Baseline characteristics of patients

Parameter	Overall (n = 102)	Nonrelapsers (n = 40)	Relapsers (n = 62)	P
Demographics				
Age, yr (range)	10.5 [6.4–15.8] (2.7–24.0)	14.2 [10.2–17.9] (2.7–23.8)	8.6 [5.7–13.7] (3.1–24.0)	0.002
Sex, male	70 (71.4)	24 (60.0)	46 (74.2)	0.19
Age at onset, yr (range)	3.0 [2.0–5.0] (1.0–13.7)	3.7 [2.1–5.1] (1.0–13.7)	3.0 [2.0–4.0] (1.0–12.5)	0.11
Previous immunosuppressive drugs				
Anti-CD20	28 (27.5)	15 (37.5)	13 (21.0)	0.07
Prednisone	102 (100)	40 (100)	62 (100)	—
Mycophenolate mophetil	38 (37.2)	16 (40.0)	22 (35.5)	0.68
Calcineurin inibitors	88 (86.2)	33 (82.5)	57 (91.9)	0.21
3 drugs	26 (25.5)	9 (22.5)	17 (27.4)	0.65
B-cell subsets (% of total lymphocytes)				
$CD19^+$ B cells	8.2 [6.0–12.9]	8.5 [4.8–10.9]	7.9 [6.1–13.1]	0.46
Transitional	0.6 [0.2–1.0]	0.7 [0.2–1.1]	0.4 [0.1–0.9]	0.24
Mature-naïve	7.7 [4.8–12.4]	8.3 [4.9–12.8]	7.3 [4.7–11.5]	0.49
Total memory	3.4 [1.9–5.3]	2.2 [1.5–4.3]	3.8 [2.3–5.7]	0.003
IgM memory	1.3 [0.9–2.6]	1.0 [0.6–1.8]	1.8 [1.0–2.9]	0.004
Switched memory	1.4 [0.8–2.6]	1.0 [0.7–2.1]	2.0 [1.1–2.7]	0.009

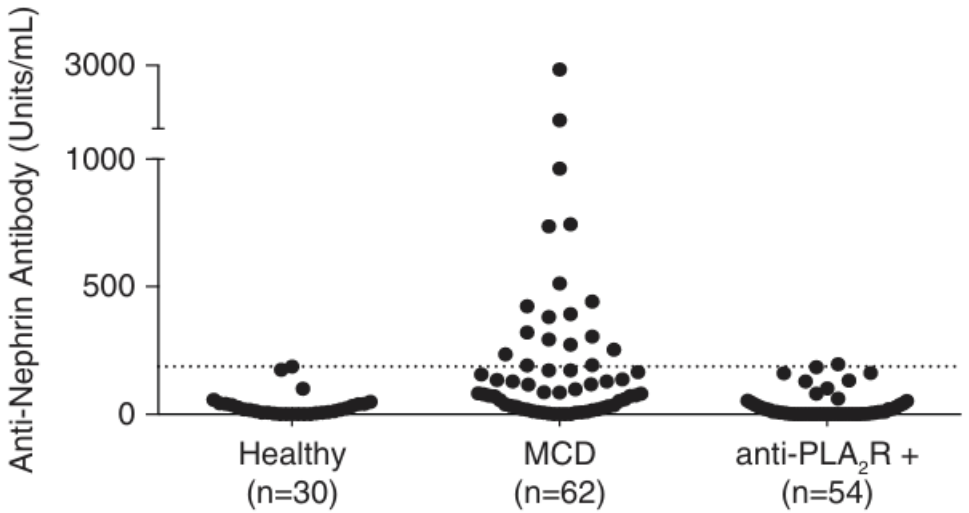
Values are n (%) or median [interquartile range], unless otherwise indicated.
P values were calculated by Mann–Whitney U test by comparing relapsers versus nonrelapsers.

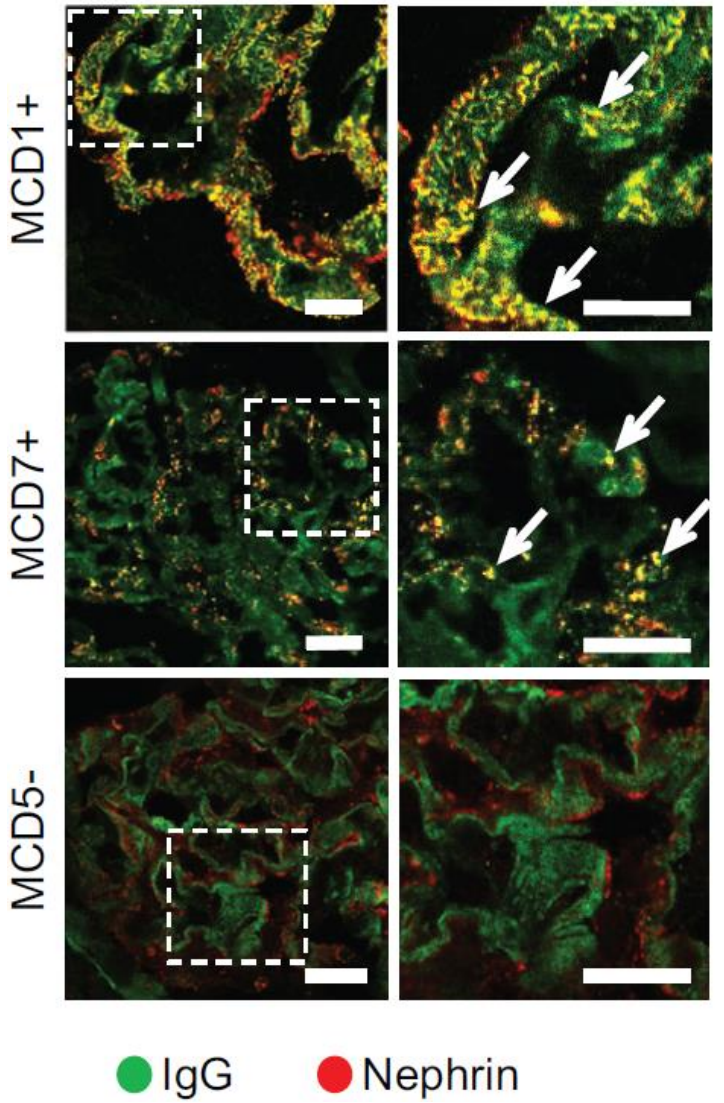
Colucci, Kid Int 2023

The anti-nephrin antibody (ANA) story

- Circulating ANA are present during active disease in some children with MCD
- Circulating autoantibodies to nephrin are present in patients with MCD and punctate IgG on biopsy
- Pretransplant ANA are associated with post-transplant proteinuria

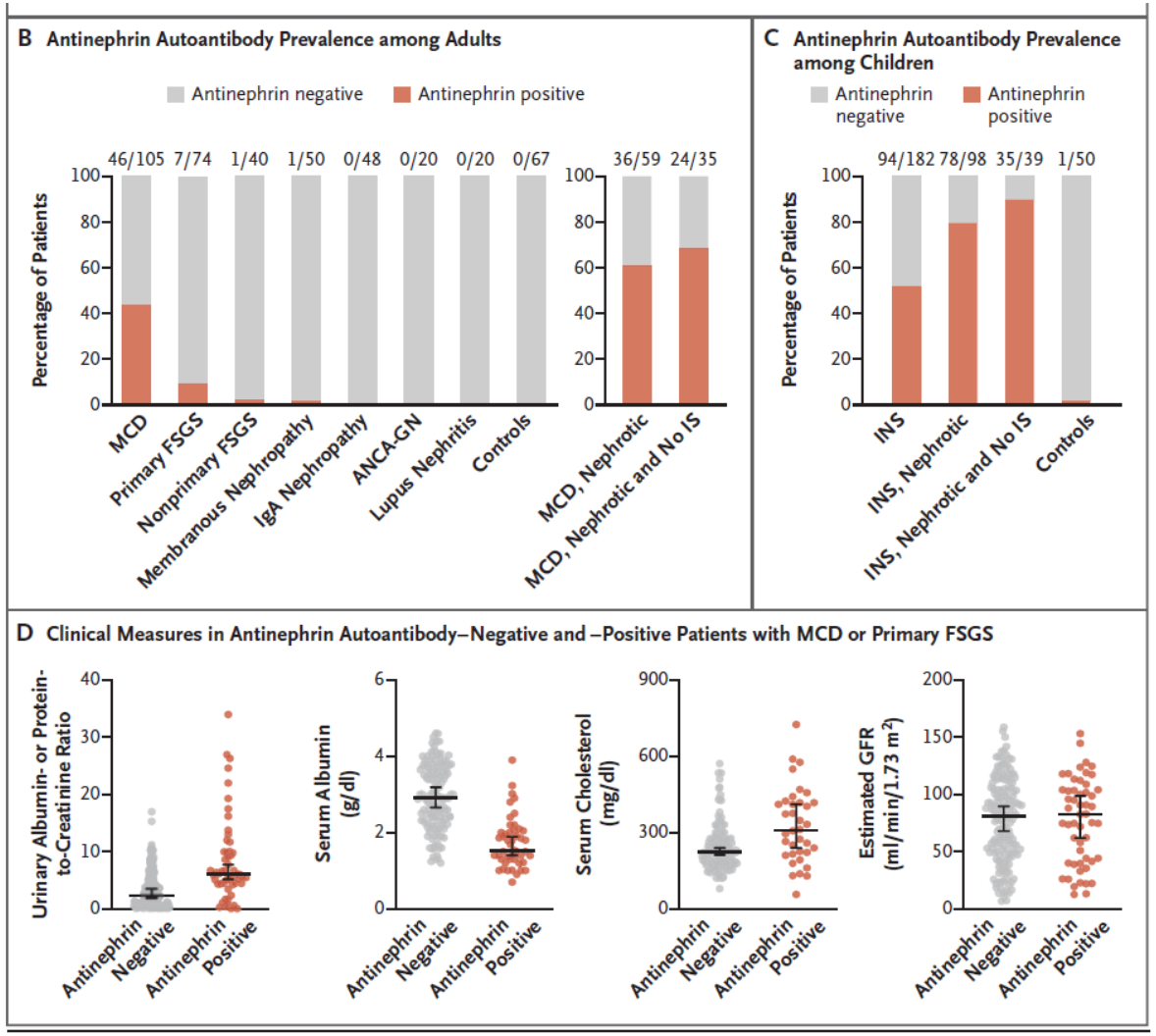
Watts, JASN, 2022



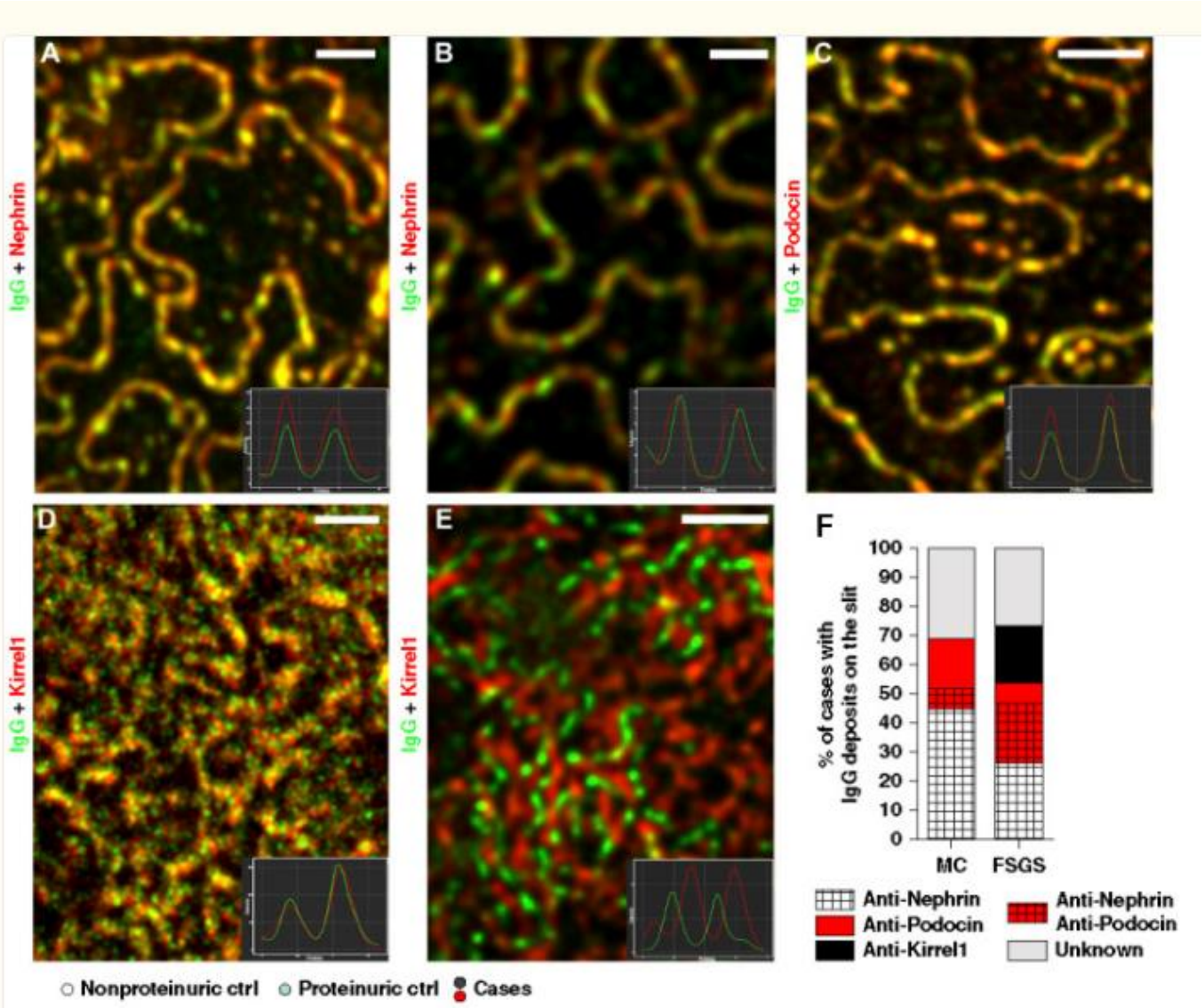


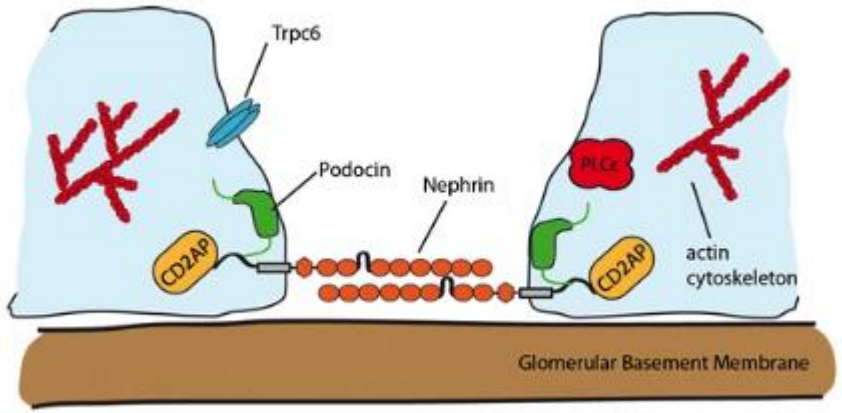
Hengel, 2024, NEJM

- Replicated presence of ANA in large multi-centre cohort of adults with MCD/FSGS and children with iNS
- Quantitative assessment of ANAs
- Correlation with disease activity
- Correlation with immunosuppression



Other anti-slit diaphragm antibodies exist and may explain more complex NS





Grahammer, 2012

Raglianti, 2026

B-cell depleting therapies

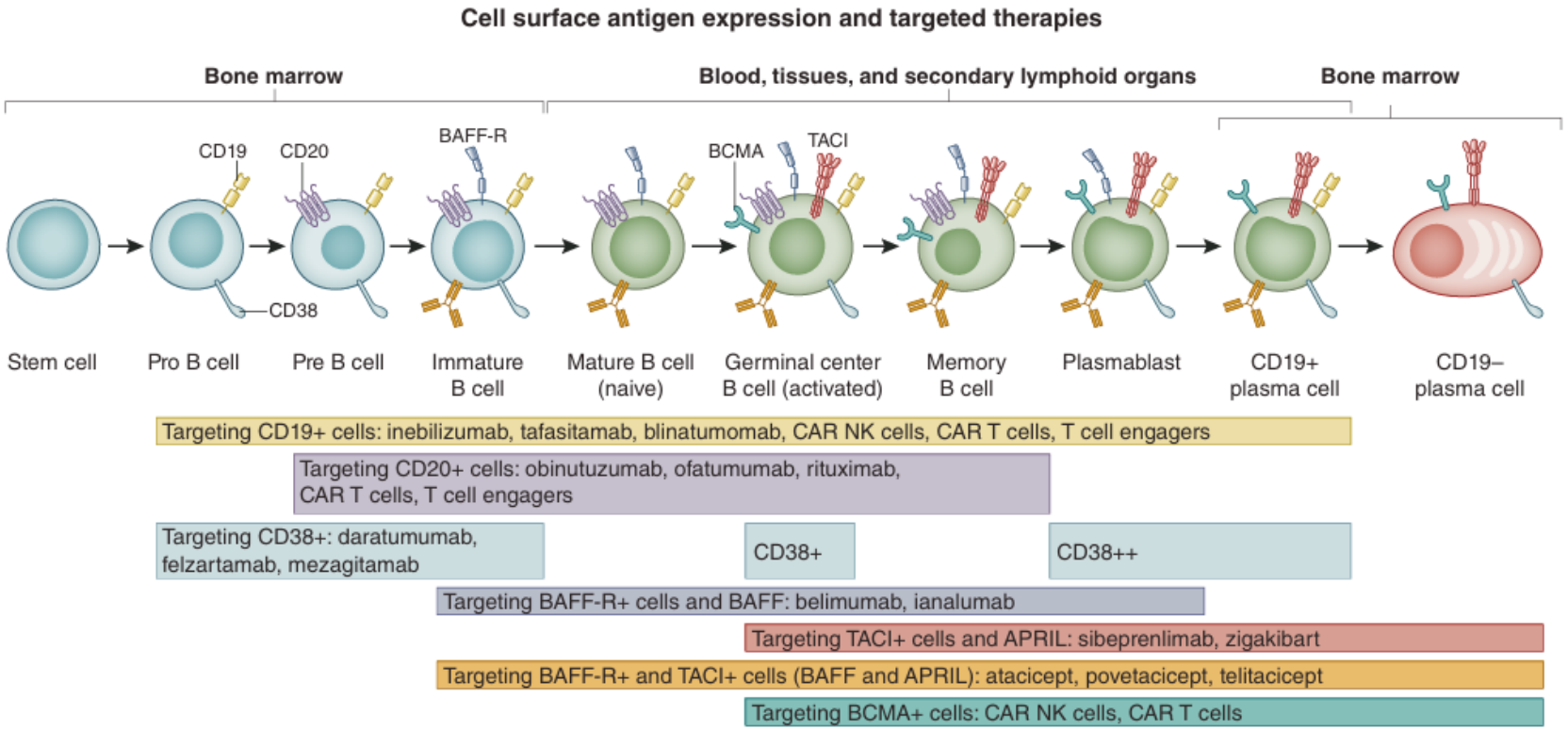


Figure 2 | B cell maturation, treatment targets, and therapies. APRIL, a proliferation-inducing ligand; BAFF, B cell activating factor; BAFF-R, B cell activating factor receptor; BCMA, B cell maturation antigen; CAR, chimeric antigen receptor; CAR NK, chimeric antigen receptor natural killer; Pro, progenitor; TACI, transmembrane activator and calcium modulator and cyclophilin ligand interactor.

- Anti-CD20 MABs
- Anti-CD38
- Anti-BAFF
- Anti-APRIL
- CAR T cells

Floege, 2026

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Challenge Your Thinking

Evaluation of pegcetacoplan in adults and adolescents with focal segmental glomerulosclerosis: Rationale and design of a sequential phase 2/3 study

J. BARRATT[1], C. RIELLA[2], Y. CHEN[2], J. MIN[2], AND J. MELNICK[2]
[1]University of Leicester, Leicester, England, UK; [2]Apellis Pharmaceuticals, Inc, Waltham, MA, USA

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BACKGROUND

- Focal segmental glomerulosclerosis (FSGS) is a rare, lesion of glomerular scarring, classified into primary, secondary, genetic, and of undetermined cause[1–3]
  - Often results in nephrotic syndrome, chronic kidney disease, and kidney failure in 40%–50% children and adults 5–10 years after initial diagnosis[4–6]
  - Current standard of care: glucocorticoids, calcineurin inhibitors, angiotensin-converting enzyme inhibitors, angiotensin receptor blockers, mycophenolate mofetil[6–8]
  - Targeted therapies for prevention or reversal of FSGS lesions are limited; prognosis remains poor[6,9–11]
- Evidence of complement activation in FSGS:[12–22]
  - Low serum C3 and C3 staining on kidney biopsies were associated with worse kidney outcomes[16,17]
  - Complement activation from initial podocyte injury results in further podocyte damage, tubulointerstitial and glomerular injury, and kidney fibrosis[13,18–22]
- **Hypothesis: complement inhibition may reduce proteinuria and interstitial inflammation while preventing disease progression in FSGS[23,24]**

AIM

Evaluate the efficacy and safety of pegcetacoplan added to standard of care in adults and adolescents with primary, genetic, or undetermined FSGS

METHODS

Study design: sequential phase 2/3 (**Figure**)

- Phase 2, single-arm, open-label portion will enrol ~15 US adults (≥18 years) before phase 3 randomisation
- Phase 3, randomised, placebo-controlled, double-blind, multicentre portion (dependent on phase 2 findings) will enrol ~250 patients, initially adults and then adolescents (12–17 years) after independent review of data collected from adults (enrolment from North America, Europe, Middle East, Africa, Latin America, and Asia-Pacific)

Study design

Trial registered on ClinicalTrials.gov, NCT07213960

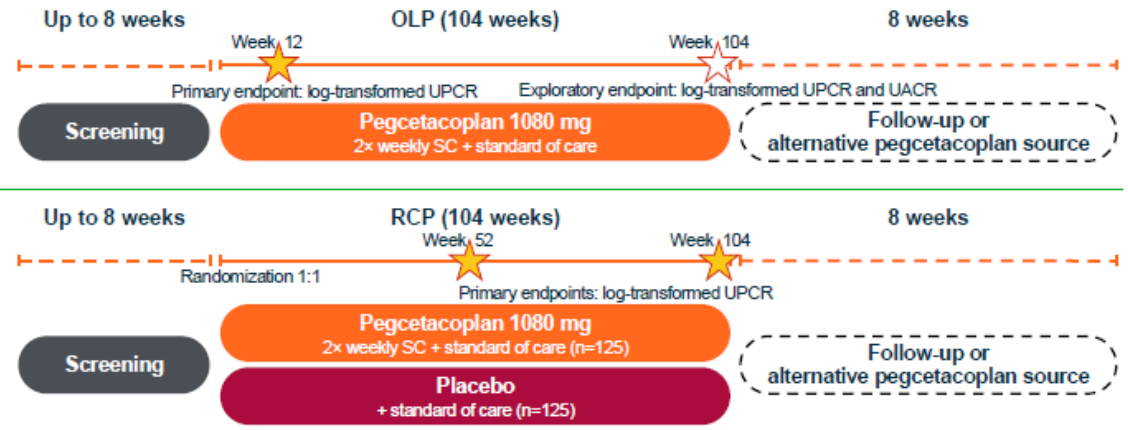


Table 1. Patient population

Inclusion criteria	
Age	• Phase 2: adults ≥18 years • Phase 3: adults ≥18 years; then adolescents 12–17 years
Body weight	≥30–≤100 kg
Diagnosis	Primary, genetic, or undetermined FSGS (excluding secondary)
Proteinuria	≥1.5 g/day on screening 24-hour urine collection and UPCR ≥1.5 g/g in ≥2 first morning urine samples
eGFR	≥25 mL/min/1.73 m²
Treatment	Stable regimen (for ≥12 weeks) of standard-of-care medications, including immunosuppressants
Vaccinations	Completed series against *Streptococcus pneumoniae* and *Neisseria meningitidis* (types A, C, W, Y, and B)
Administration	Willingness and ability to self administer pegcetacoplan
Pregnancy	Negative urine pregnancy test (women); contraception ≥90 days after last pegcetacoplan dose (men and women)
Consent	Willingness and ability to provide informed consent
Exclusion criteria	
Diagnosis	FSGS secondary to another condition
Coexisting conditions	• Type 1 or uncontrolled (A1C ≥8%) type 2 diabetes • History of kidney transplant

Table 2. Key study endpoints

Primary efficacy: Proteinuria reduction[25,26]	
Phase 2	Log-transformed UPCR reduction from baseline at week 12 (and exploratory endpoint at week 104)
Phase 3	Log-transformed UPCR reduction from baseline at week 52 and at week 104
Secondary key efficacy	
Phase 2	Log-transformed UACR reduction from baseline at week 12 (and exploratory endpoint at week 104)
Phase 3	• Percentage of patients achieving complete remission[5,24] (UPCR <0.3 g/g) at week 104 • eGFR slope through week 104
Safety	
Phases 2+3	Incidence and severity of adverse events
Pharmacodynamics and pharmacokinetics	
Phases 2+3	• Change in serum pegcetacoplan concentration over time • Change in serum C3 concentration over time

METHODS *(CONTINUED)*

Population: primary, genetic, or undetermined FSGS (**Table 1**)

Study endpoints: primary efficacy is proteinuria reduction at week 12 (phase 2) or weeks 52 and 104 (phase 3) (**Table 2**)

Phase 3 sample size calculations: assumptions based on data from the VALIANT (NCT05067127) phase 3 trial in patients with C3G and IC-MPGN[25]

- Week-52 urine protein-to-creatinine ratio (UPCR) reduction: pegcetacoplan, 40% vs placebo, 10%, with mean log-transformed UPCR −0.51 vs −0.11, respectively, and 0.8 standard deviation
- 250 participants will provide ~94% power at week 52 (0.0125 1-sided significance level) and ~90% power at week 104 (assuming similar UPCR reduction and 20% attrition at week 104)

CONCLUSIONS

By assessing the efficacy and safety of pegcetacoplan plus standard of care in adults and adolescents with FSGS, findings from this study will inform potential future treatment options for FSGS

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ABBREVIATIONS

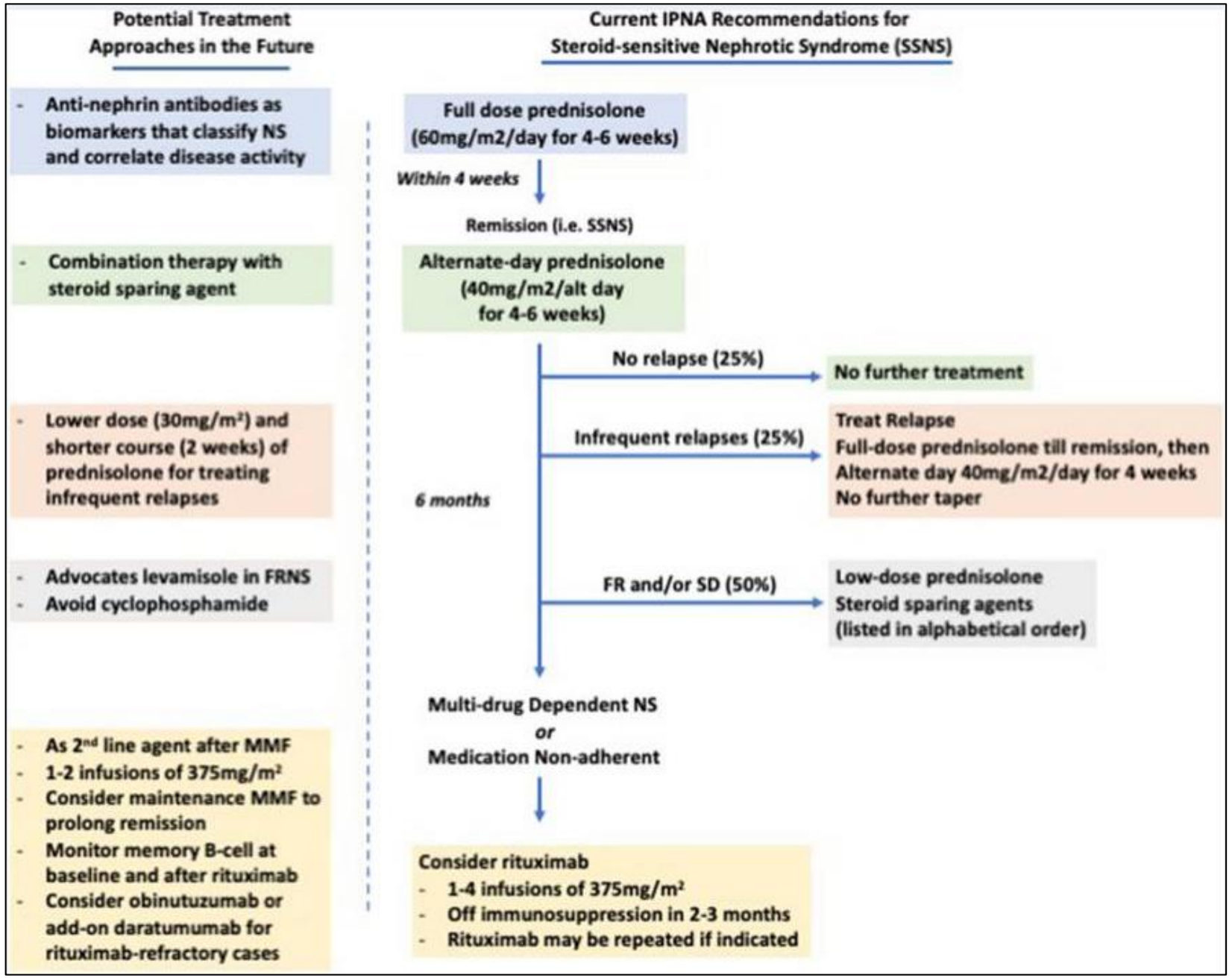
C3G, C3 glomerulopathy; eGFR, estimated glomerular filtration rate; FSGS, focal segmental glomerulosclerosis; IC-MPGN, immune-complex membranoproliferative glomerulonephritis; OLP, open-label period; RCP, randomised controlled period; SC, subcutaneous; UACR, urine albumin-to-creatinine ratio; UPCR, urine protein-to-creatinine ratio.

DISCLOSURES

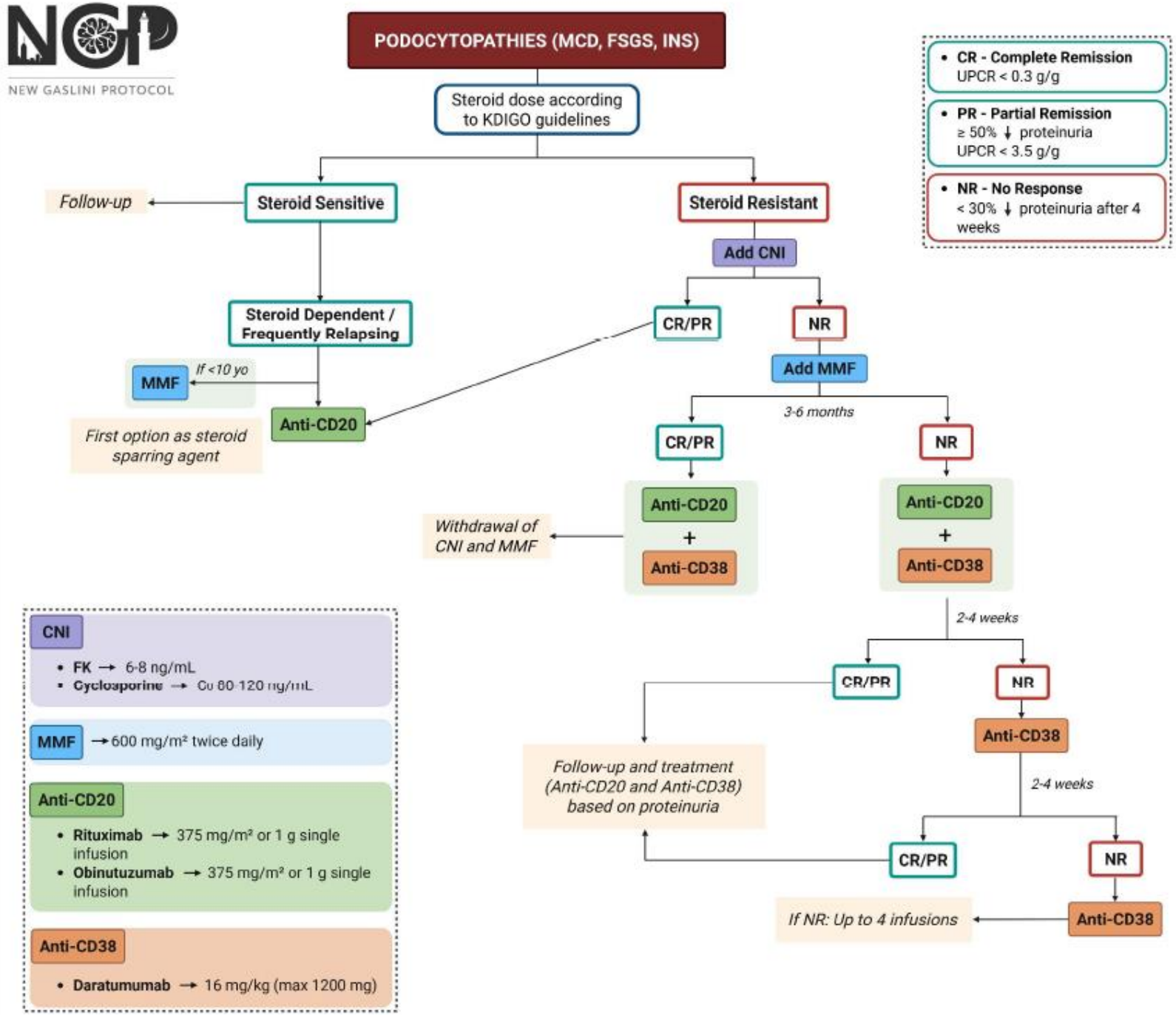
JB has received consulting fees from Argenx, Biohaven, Calliditas, Dimerix, Novartis, Travere Therapeutics, Vera Therapeutics, Otsuka , Travere, Biogen, Takeda, Vertex, Roche, and Alexion; he also received grant support from Argenx, Calliditas, Novartis, Travere Therapeutics, Visterra, and Alexion. CR, JMe, YC, and JMi are employees of Apellis (now BioGen) and may hold stock or stock options.

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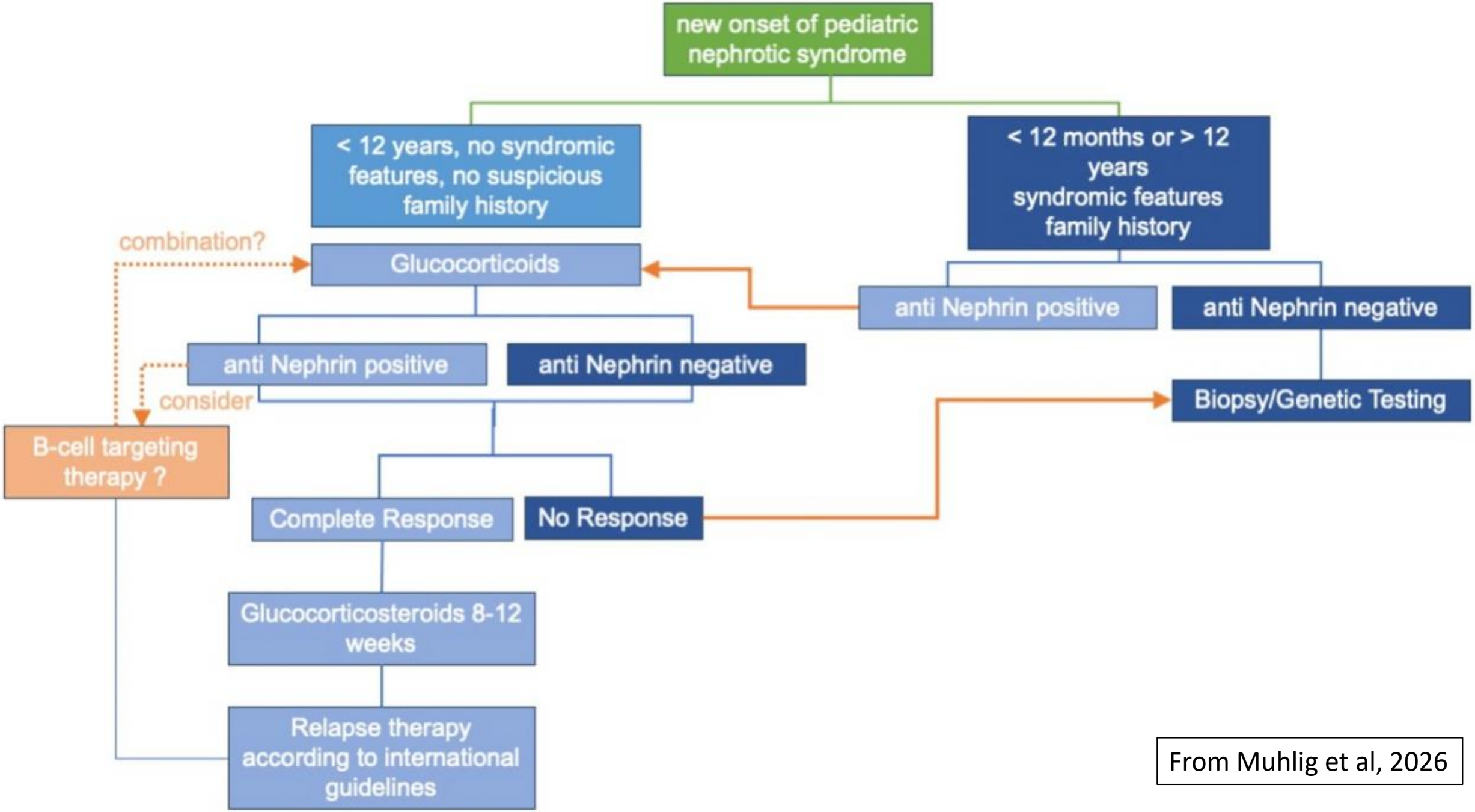
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Chan and Boyer, 2025



Angeletti, 2026



From Muhlig et al, 2026

Summary

- Childhood iNS treatment is still primarily steroids after 50 years
- More steroid for initial course but less for relapses?
- Anti-CD20 monoclonal antibodies increasingly important as steroid-sparing agents
- Obinutuzumab looks more promising than rituximab
- Newer B-cell depleting therapies offer still further hope
- Biomarkers offer the potential for more directed therapies
- Recent discovery of anti-nephrin antibodies may lead to a paradigm shift in iNS management

Questions?

Pediatric Nephrology (2026) 41:1909–1912
https://doi.org/10.1007/s00467-025-07115-3

EDITORIAL COMMENTARY

Optimal selection and timing of steroid-sparing therapy in childhood nephrotic syndrome: what determines long-term outcomes of adult survivors?

Shuichiro Fujinaga[1] · Hiroki Miyano[2] · Mayu Nakagawa[3] · Koji Sakuraya[1] · Amane Endo[3] · Yoshiyuki Ohtomo[2] · Hiromichi Shoji[3]

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- Decisions based on LT prognosis; no RCT >2 years duration
- Early studies of Cyc show better LT remission than for CNI-era
- Early use of rituximab may give better LT remission

Adult prognosis for childhood iNS

	n	Population	Relapses into adulthood
Trompeter, 1985	152	SSNS	5.5%
Fakhouri, 2003	102	SSNS, 1970-1975	42%
Rüth, 2005	42	SSNS	33%
Skrzypczyk, 2014	61	SSNS	16%
Aydin, 2019	50	SDNS and SRNS	9%
Fujinaga, 209	44	SDNS, MMF after CNI	66%
Korsgaard, 2019	39	SSNS	31%
Takemasa, 2024	81	SDNS and SRNS	54%