Anti-nephrin and other podocyte autoantibodies: toward an immunological stratification of idiopathic nephrotic syndrome

Actualités Néphrologiques Jean Hamburger
19-20 mai 2025

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Department of Nephrology and Transplantation
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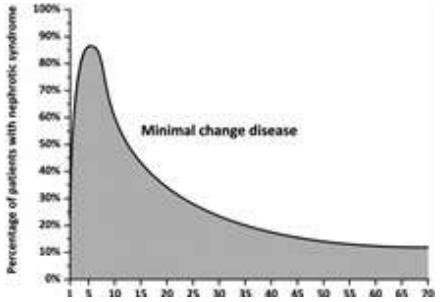
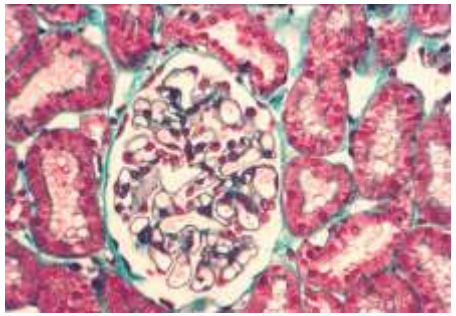
idiopathic Nephrotic Syndrome

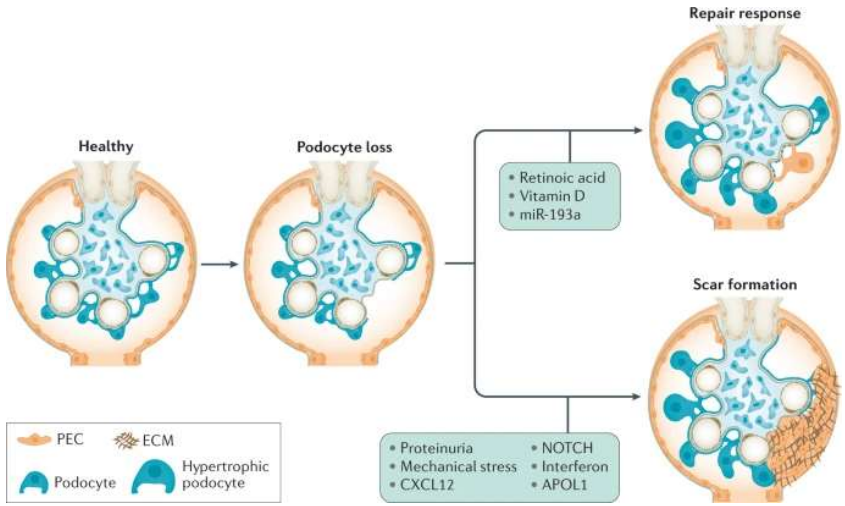
Minimal Change Disease



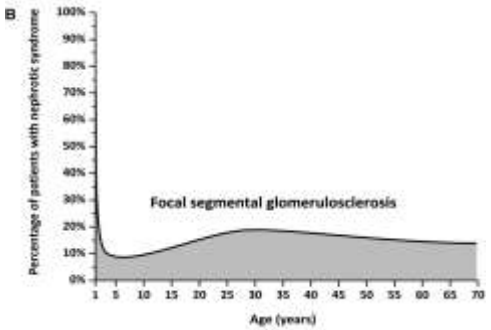


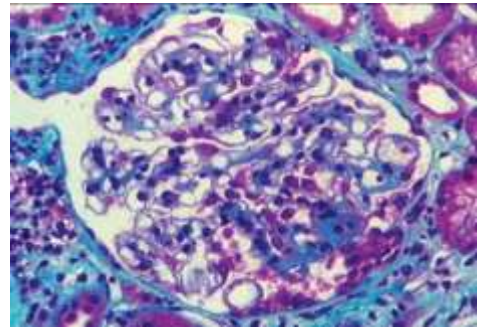
Negative IF

10–15% of adult NS
75% children



Primary Focal Segmental Glomerulosclerosis





IF deposits in sclerotic lesions

30% of adult NS
20% children

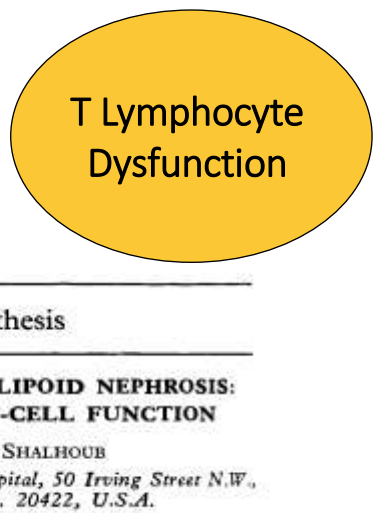
Podocyte injury
Podocytopathies

Vivarelli et al. CJASN 2017
Sahali et al Seminars Immunopathol 2014
Kopp et al. Nat. Disease primers 2020

Idiopathic Nephrotic Syndrome

An immunological related disorder

Pathophysiology of immunological nephrotic syndrome : From T-cell mediated glomerular disease….



Hypothesis

PATHOGENESIS OF LIPOID NEPHROSIS: A DISORDER OF T-CELL FUNCTION

ROBERT J. SHALHOUB
Veterans Administration Hospital, 50 Irving Street N.W., Washington D.C. 20422, U.S.A.

T Lymphocyte Dysfunction

Immune dysregulation

- sensitivity to steroids and immunosuppressive drugs
- HLA-risk loci involved in adaptive immunity (GWAS) [43-45]

B Lymphocyte Dysfunction

Circulating permeability factors [17-21]

?

Podocyte Alteration

- Atopy and T_H2 cell mediated immunity [28-36]
- Association to thymoma
- Treg dysfunction causes CD80 overexpression [41]
- Alterations to the distribution and/or function of T cell subsets [42]

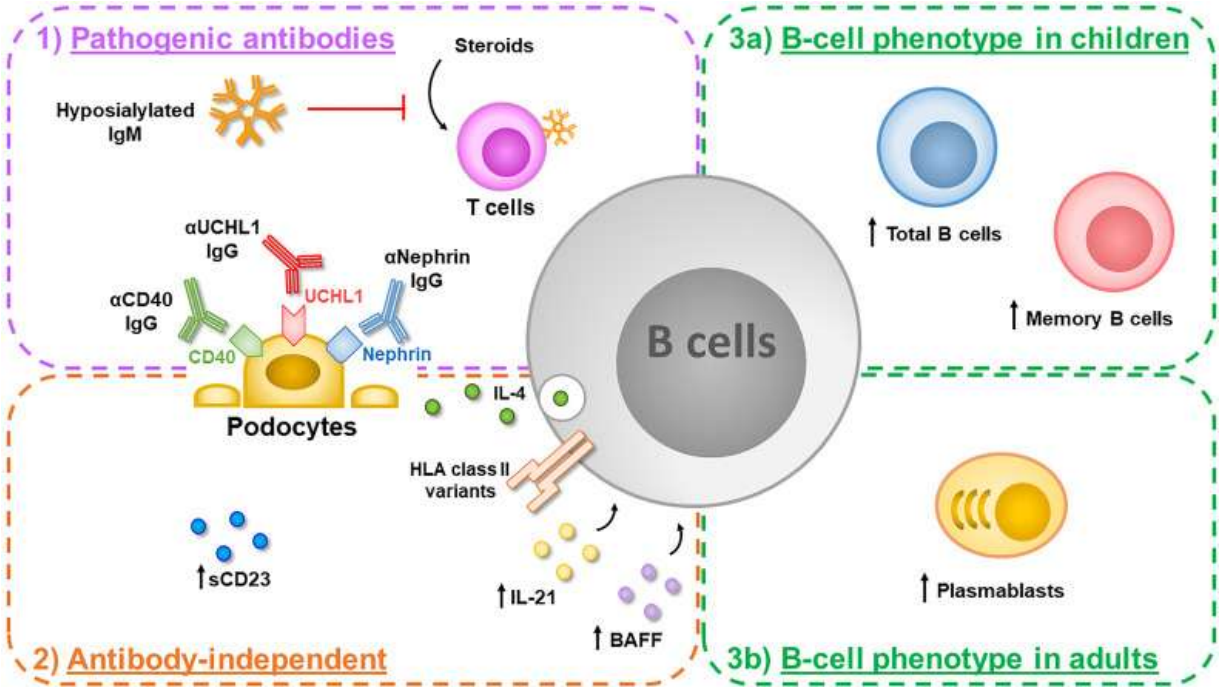
- Hodgkin lymphoma
- Successfull use of B cell-depleting agents
- Anti-UCHL1 IgG autoantibody in mice [37]
- B cell-induced local IL-4 induces foot process effacement & proteinuria [38, 39]
- Increase of sCD23 and IgE level

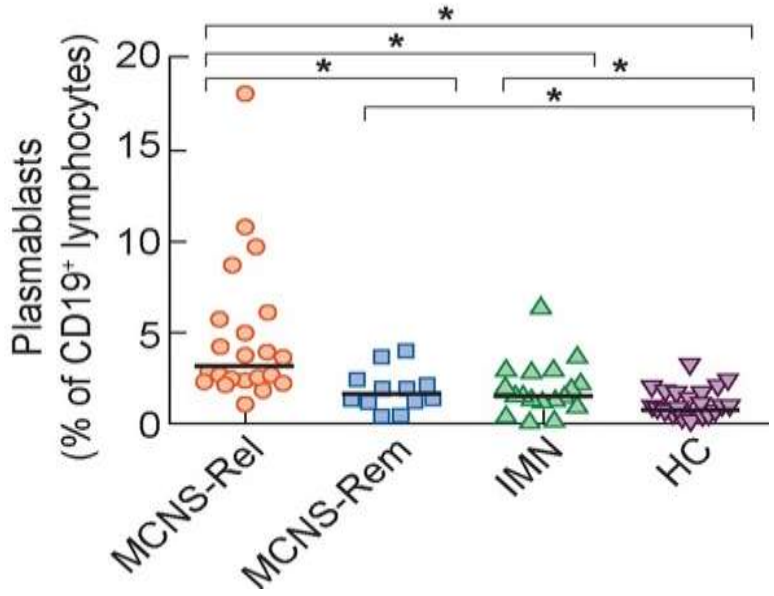
The Quest for the permeability Glomerular Permeability Factor...

- **Hemopexin**: *JASN 1999, Ki 2000, Ki 2005,*
- **c-MIP**: *J Exp Med 2003, Science signal. 2010*
- **Cytokines, IL13, CLCF-1**: *PN 2006, JASN 2007, JIR 2015*
- **TLR3-CD80 (B7-1)**: *JASN 2009*
- **CD34**: *JASN 2007*
- **Angiopoietin like-4**: *Nat Med 2011*
- **suPAR**: *Nat Med 2011*

Gauckler et al. Autoimmunity Reviews 2020

… Toward B-cell Immunity





Several studies:

- **Elevation of plasmablasts**
- **Extrafollicular response involvement**

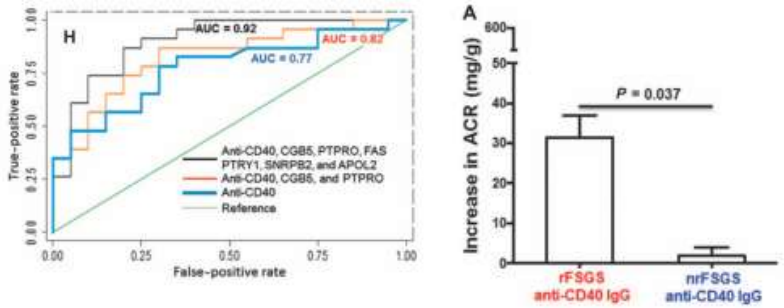
Vivarelli et al. Lancet 2023
Colucci et al. Front. Immunol. 2022
Oniszczuk et al. NDT 2022

Idiopathic Nephrotic Syndrome: Auto-immune glomerular disease ?

Several autoantibodies have been identified, based on recurrent FSGS after kidney transplantation and idiopathic NS

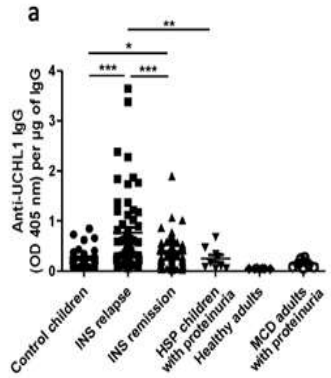
rFSGS

Anti CD40 Ab

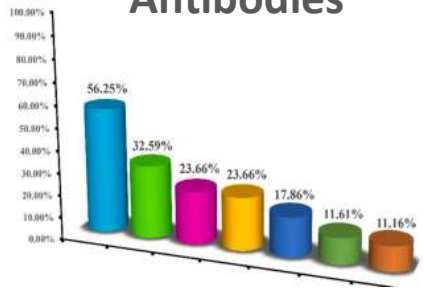


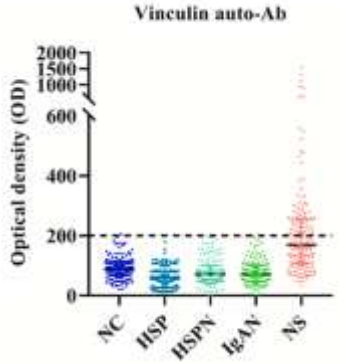
iNS

Anti UCHL1 Ab



Mass spectrometry :66% with auto-Antibodies





Define a sub-group of iNS :

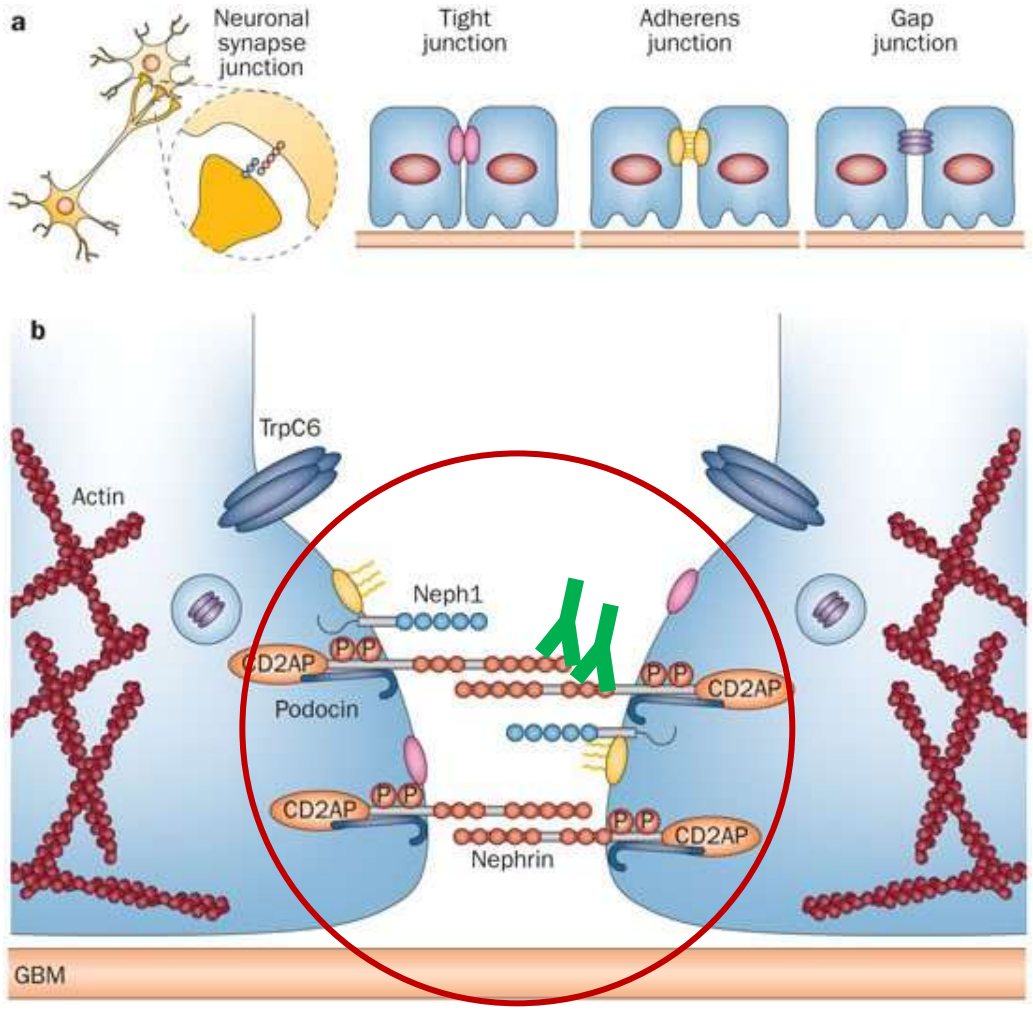
Autoimmune podocytopathies

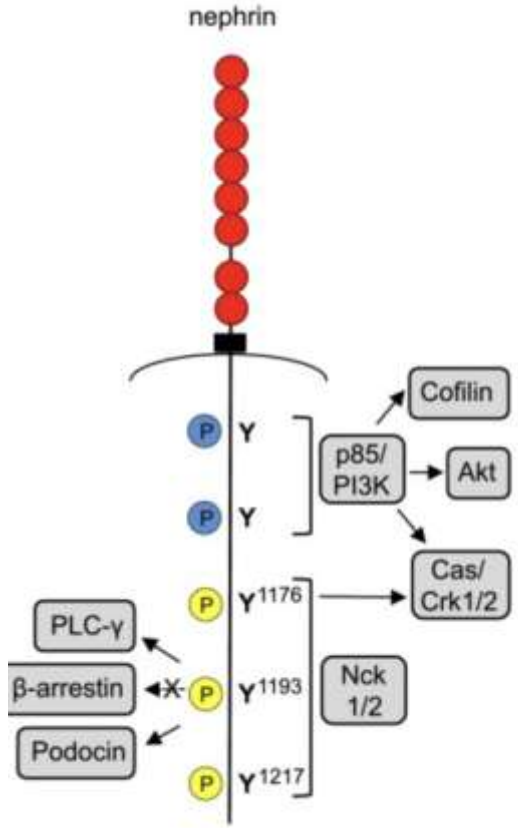
Qing et al. JASN 2022

Delville et al. ScienceTrans Med 2014
Jamin et al. J of Autoimmunity 2018
Qing et al. Clin. Immunology 2021

- **Key role of the humoral immune response ++**
- **Existence of a glomerular permeability factor**
- **Targets: podocytes and slit diaphragm**

Discovery of Anti-Nephrin Antibodies





Grahammer et al. Nat Review Nephrology 2013

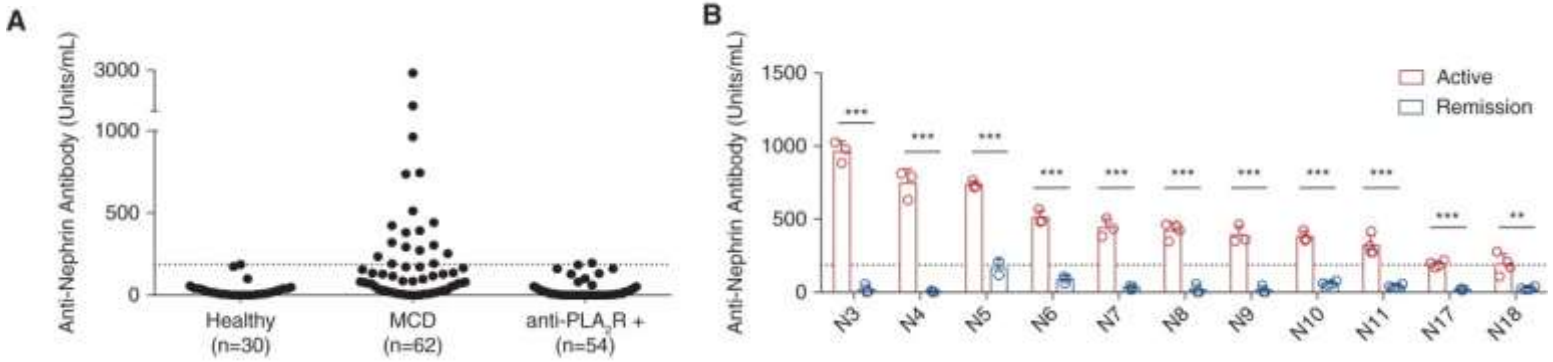
Discovery of Anti-Nephrin Antibodies

Study	Detection method of circulating anti-nephrin autoantibodies
Watts *et al.*, JASN 2022 [1]	Immunoprecipitation and signal-enhanced ELISA with lab-produced recombinant human nephrin ectodomain, 6xHIS-tagged, expressed in HEK293

Hengel et al. CKJ 2025

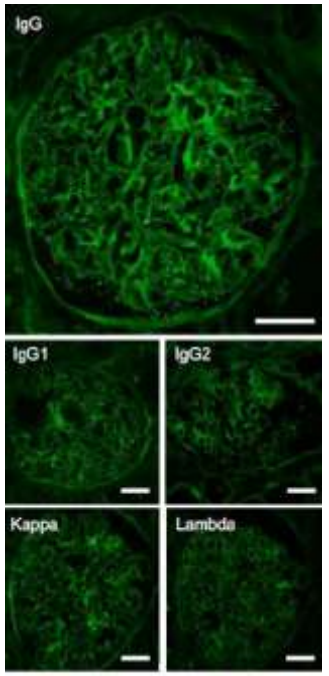
Neptune cohort: 41 children and 21 adults with MCD

- ***18/62 (29%) anti-nephrin Ab***
- ***1/54 MN***
- ***12/18 seropositive patients → Remission***



Disappearance of antibodies with treatment

positive IF on MCD kidney biopsy

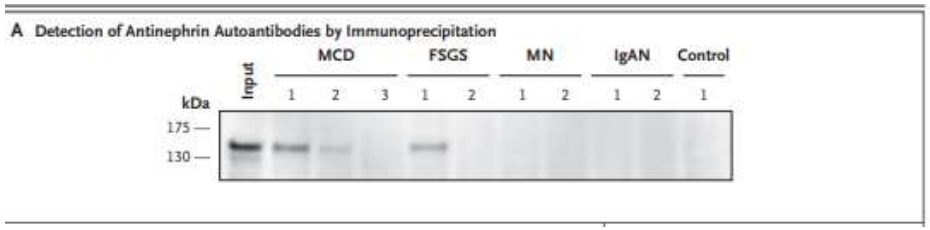


Watts et al. JASN 2022

Discovery of Anti-Nephrin Antibodies

Hengel *et al.*, *NEJM* 2024 [2]	Immunoprecipitation with lab-produced recombinant human nephrin ectodomain, 8xHIS- and twinstrep-tagged, or hybrid assay of immunoprecipitation followed by ELISA of eluted recombinant nephrin, expressed in HEK293	– 182 children with INS (NEPHROVIR cohort, cohorts from Bari and Rome) – 357 adults with biopsy-proven glomerular diseases (MCD, pFSGS, MN, IgAN, ANCA, SLE from Hamburg GN Registry and Bari cohort) – 117 healthy controls (50 children, 67 adults)

Hengel et al. CKJ 2025



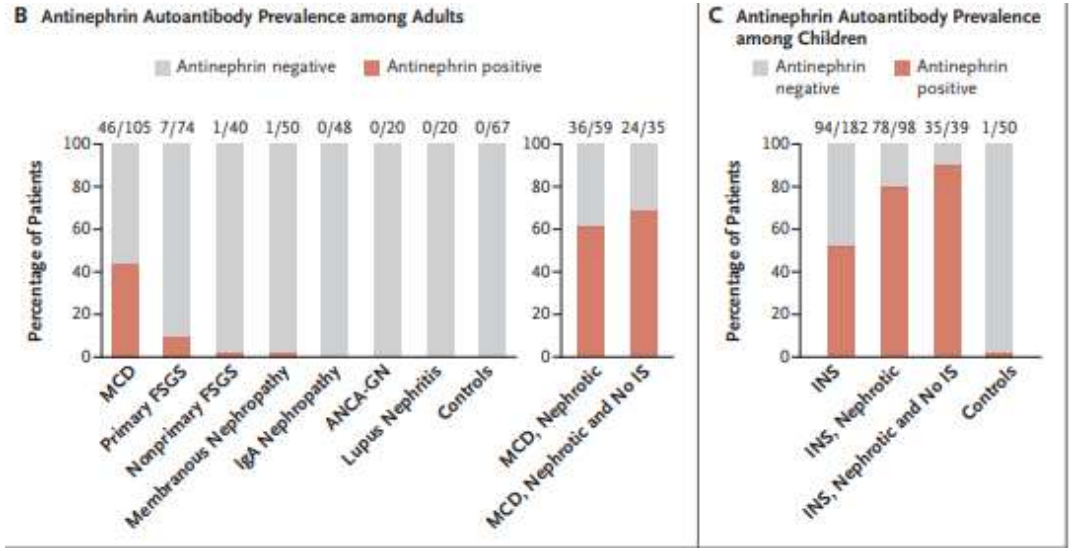


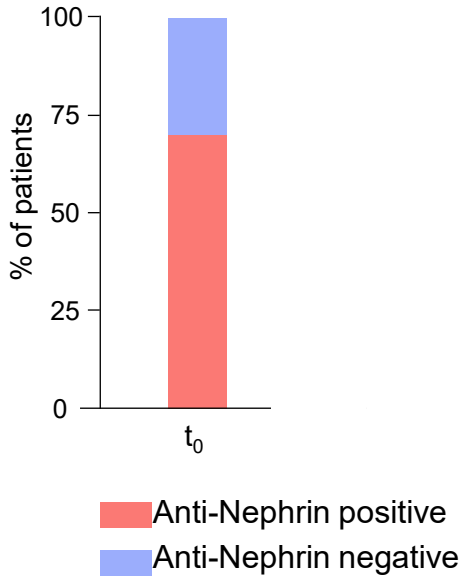
Table 1. Prevalence of Antinephrin Autoantibodies in Patients with Glomerular Diseases and Controls

Patient Classification	Antinephrin Autoantibody–Positive Ratio (%)
Idiopathic nephrotic syndrome	94/182 (52%)
Idiopathic nephrotic syndrome, nephrotic,[a] no IS[b]	35-39 (90%)
Minimal change disease	46/105 (44%)
Minimal change disease, nephrotic,[a] no IS[b]	24/35 (69%)
Primary FSGS	7/74 (9%)
Nonprimary FSGS	1/40 (2%)
Membranous nephropathy	1/50 (2%)
IgA nephropathy	0/48 (0%)
ANCA vasculitis	0/20 (0%)
Lupus nephritis	0/20 (0%)
Healthy adults	0/67 (%)
Healthy children	1/50 (2%)

Fischman et al. AJKD 2025

Hengel et al. NEJM 2024

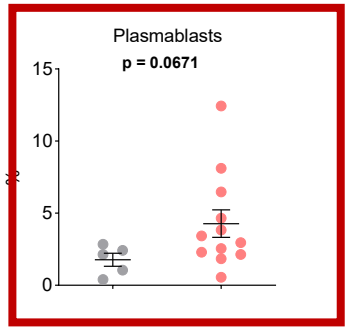
Discovery of Anti-Nephrin Antibodies



12/17 (71%) anti-nephrin positive
In *de novo* MCD

At Diagnosis

	Total (N=17)	Anti-Nephrin (-) (N=5)	Anti-Nephrin (+) (N=12)	p-value
Age (years), Median (IQR)	35.0 (25.0-47.0)	43.0 (25.0-52.0)	34.5 (26.0-43.3)	0.8
Sex, Male, n (%)	9 (53)	2 (40)	7 (58)	0.6
UPCR (mg/mmol), Median (IQR)	705.4 (461.6-864.1)	620.0 (512.5-697.69)	788.6 (466.64-878.8)	0.3
Albumin level (g/L), Median (IQR)	17.0 (11.0-20.0)	**20.0 (20.0-23.0)**	**13.5 (10.0-17.0)**	0.06
Creatinine level (µmol/l), Median (IQR)	86.0 (69.0-99.0)	85.0 (69.0-91.0)	88.0 (71.3-103)	0.6
eGFR at diagnosis (ml/min/1.73m²), Median (IQR)	89.0 (79.0-106)	101 (89.0-106)	85.5 (77.8-115)	0.8



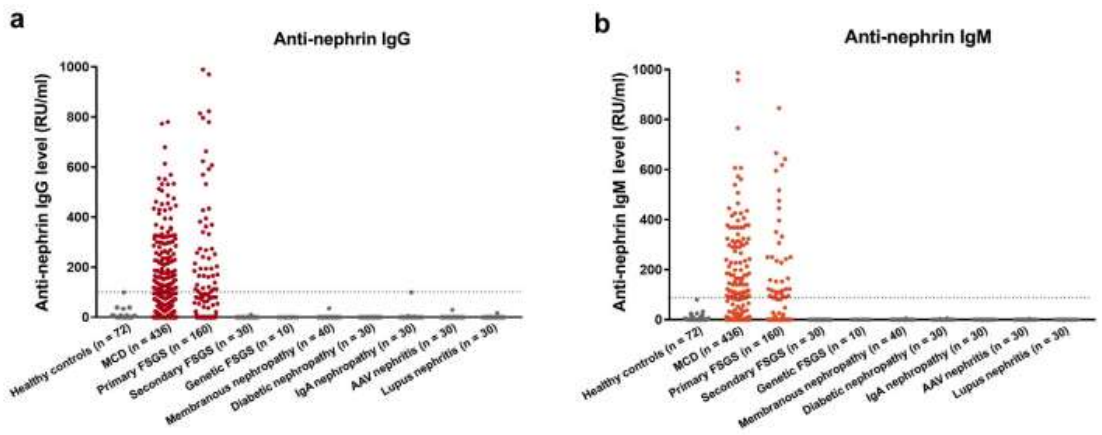
Plasmablasts

Anti nephrin Ab (+) : 4,3 % ± 3,3 (p=0,07)

Anti nephrin Ab (-) : 1,77 % ± 1

Hengel,Sakhi* et al. KI reports 2026*

Discovery of Anti-Nephrin Antibodies



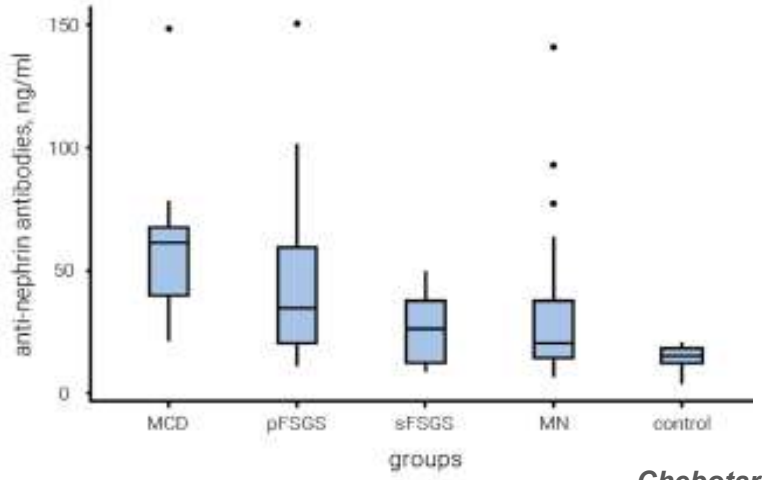
Shu et al. KI 2025

Anti-nephrin Ab : 43 %

- IgG : 30% and IgM : 26%
- 13,1 % both IgG & IgM

- **IgM : younger, more women, more nephrotic**
- **IgG & IgM :Similar prognosis in terms of response and relapse**

But having both IgG/IgM ➔ More severe forms



Chebotareva et al. Nephrology 2024

77 with glomerulopathies and 11 healthy subject

ELISA test

Anti nephrin level

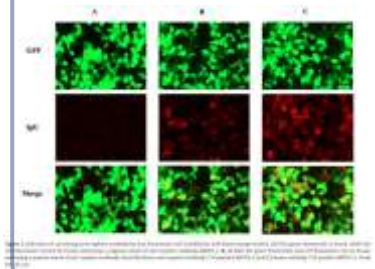
- MCD : 61,2 [28,9-66,3] ng/mL
- FSGS : 32,5 [17,2-58,4] ng/mL. (primary: 45,2[20-64,3] ng/mL)
- MN : 20,3 [14,4–38,4] ng/mL
- HC : 15,3 [12–18,9] ng/mL,

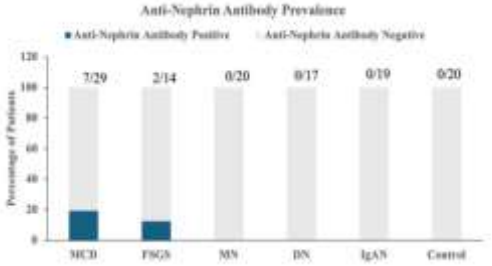
Anti-nephrin antibodies detected by other groups, but with heterogeneous assays

Can They Be Used as a Diagnostic Tool for INS?

Anti-Nephrin Antibodies : Diagnosis tool

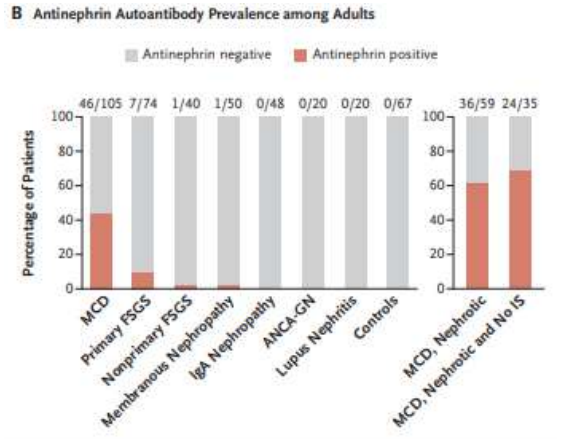
Indirect IF





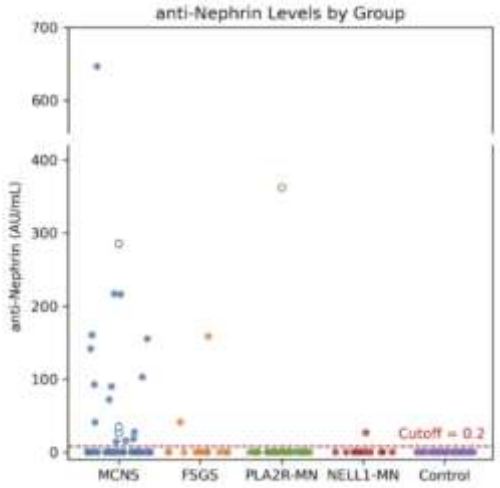
Chen et al .CKJ 2025

Western Blot



Hengel et al. NEJM 2024

ELISA



Hayashi et al. Scientific report 2026

Anti-nephrin Ab in iNS diagnosis

Specificity 98%

Variable Sensitivity

Type of Test

Histological sub-type

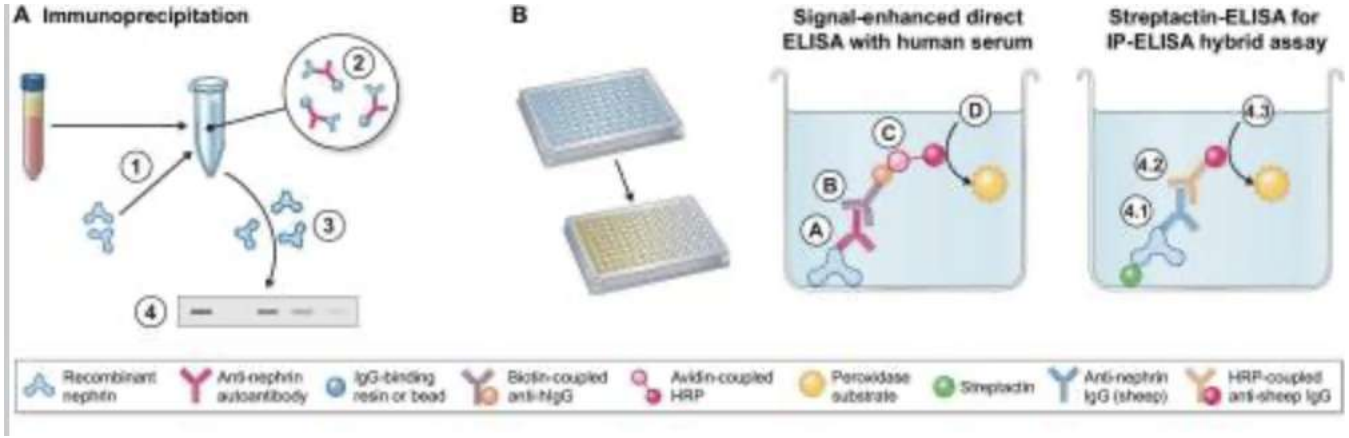
Disease-course dependent detection

A broad variety of technics

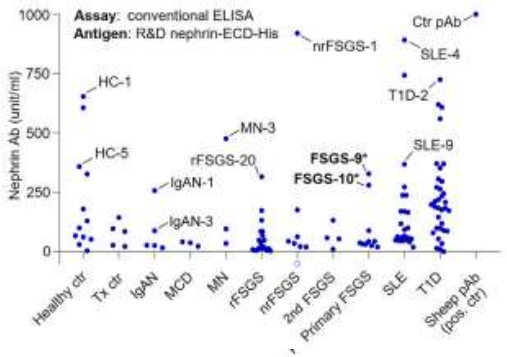
Limitations:

- Low antibody titer
- Complexe Ac-Ag complex rapidly internalized

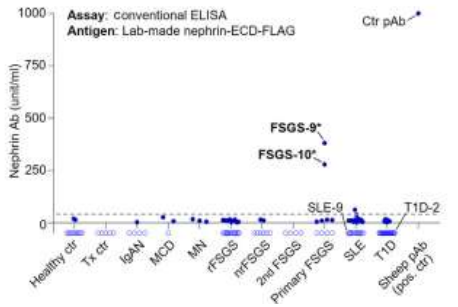
2 cohorts and plasma
127 patients and validation 21 × 2 FSGS/MCD



ELISA



Mouse cell produced nephrin
False positive ++



HEK cell produced nephrin

Method	Specificity	Sensitivity	Clinical feasibility
Conventional ELISA	**Low à moderate**	**Variable**	**High**
Immunoprecipitation – Western blot (IP–WB)	**Excellent**	**High**	**Low**
On-beads ELISA (IP-based)	**Good**	**Moderate**	**Moderate to high**
Immunofluorescence (cellular or tissue-based)	**Very low**	**Very low**	**Low**

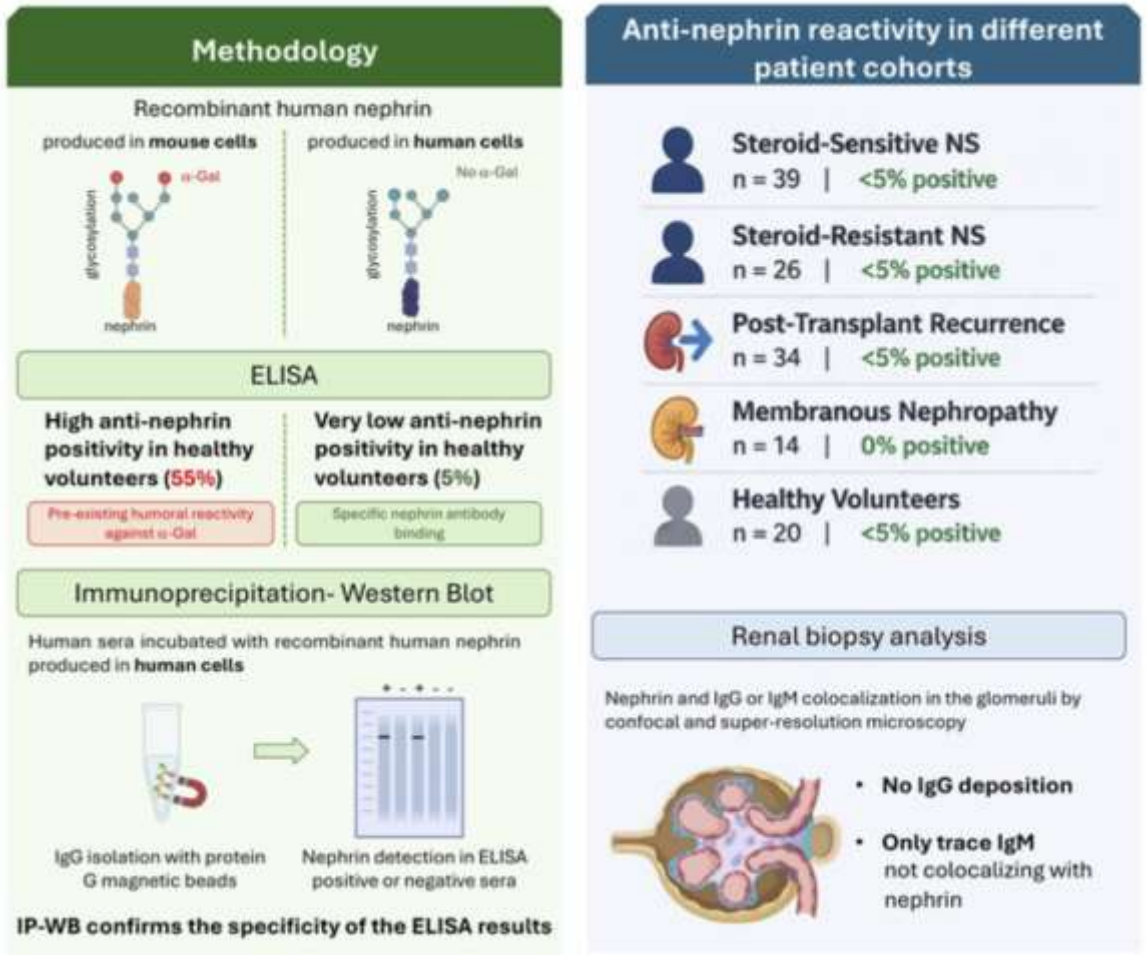
Hengel et al. CKJ 2025
Liu et al. KI 2025

A broad variety of technics

And sometimes non-reproducible results..

- **False positives already observed with murine recombinant nephrin**
- **Sensitivity issues**

To date: no validated French assay available

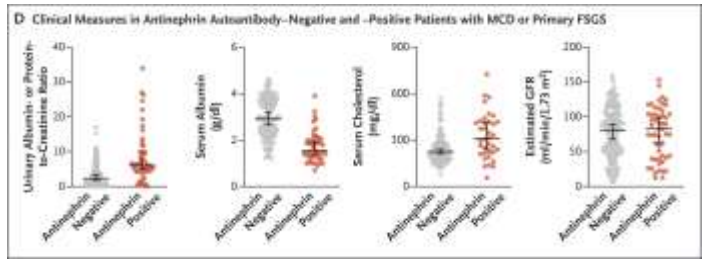


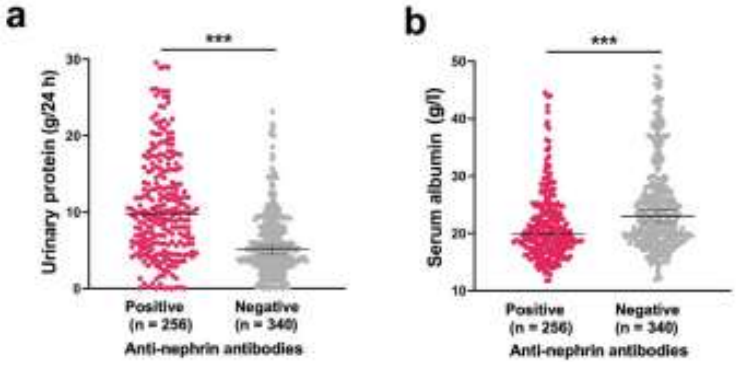
Pecoraro et al. JCI 2026

Can They Be Used for INS Stratification?
Impact on therapeutic management

Anti-nephrin Ab and Clinical Presentation

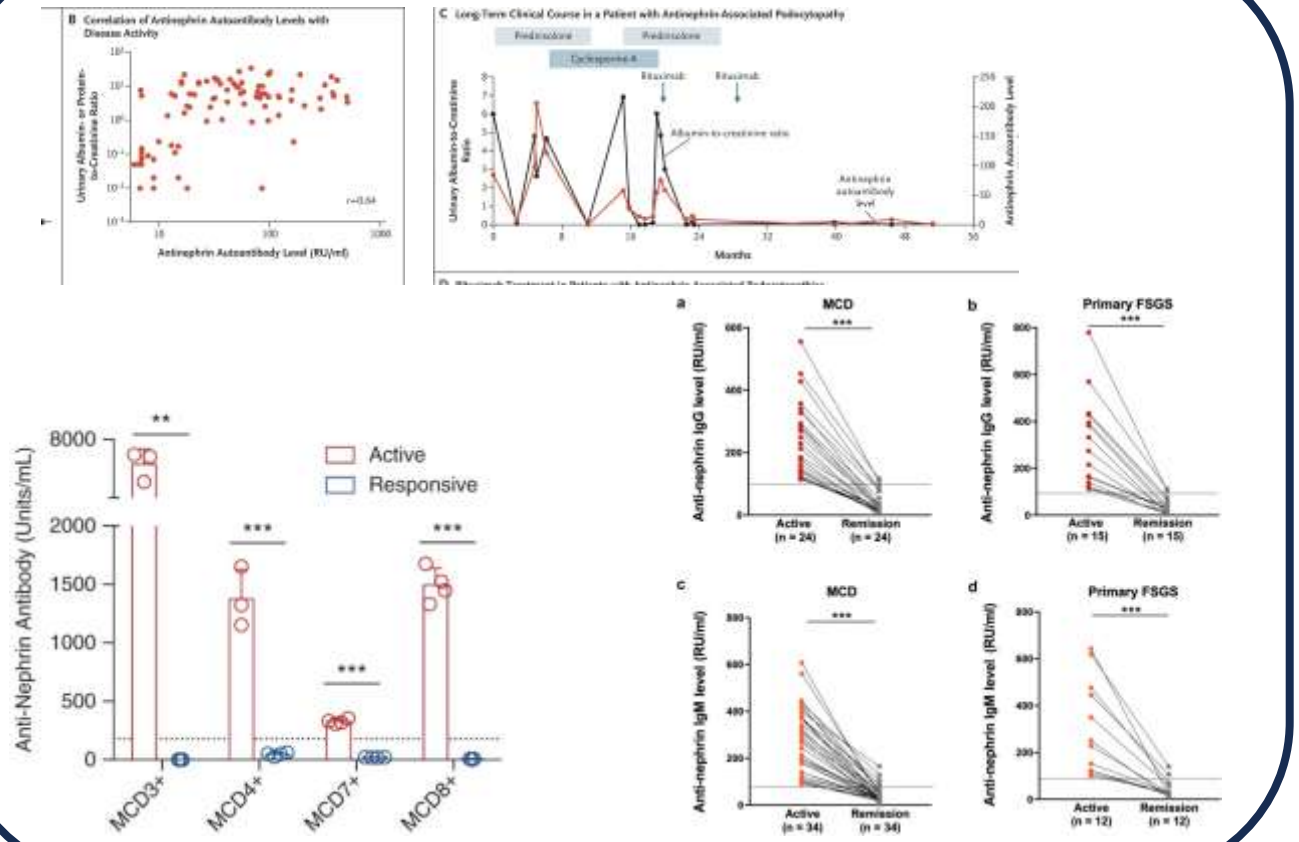
Compareason antinephrin (+) vs antinephrin (-)





Deeper nephrotic syndrome

Correlation anti-nephrin Ab and activity

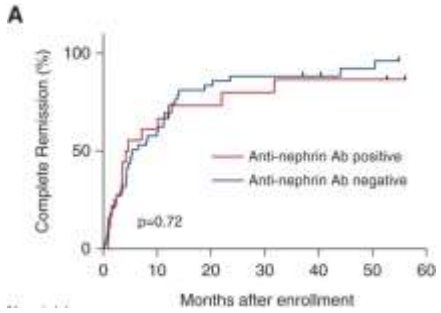


Patients Anti-nephrins (+) ➔ Deeper nephrotic syndrome
Correlation of antibodies with clinical activity

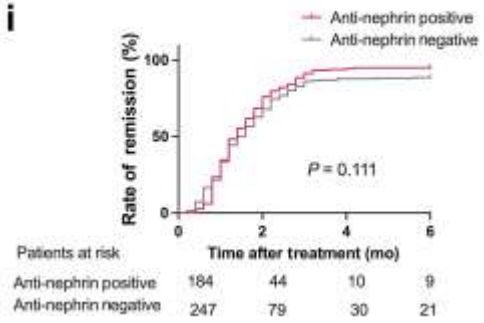
Watts et al. JASN 2022
Shu et al. KI 2025
Hengel et al. NEJM 2024

Anti-nephrin Ab : Prognosis

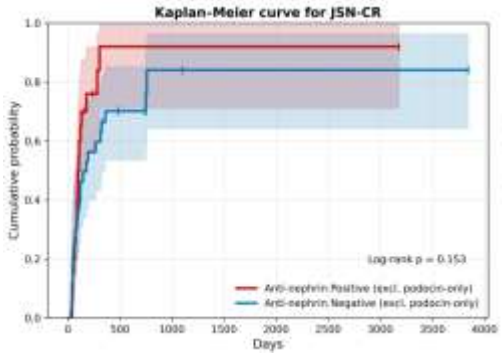
Compareason antinephrin (+) vs antinephrine (-)



Watts et al. JASN 2022

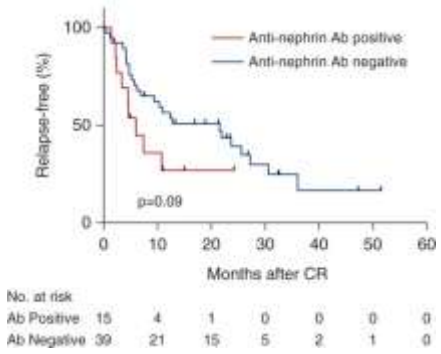


Shu et al. KI 2024

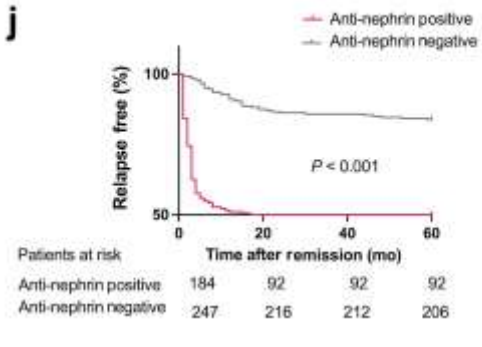


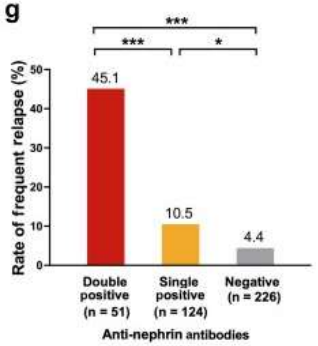
Hayashi Scientific report 2026

No difference in remission probability



Watts et al. JASN 2022





Shu et al. KI 2024

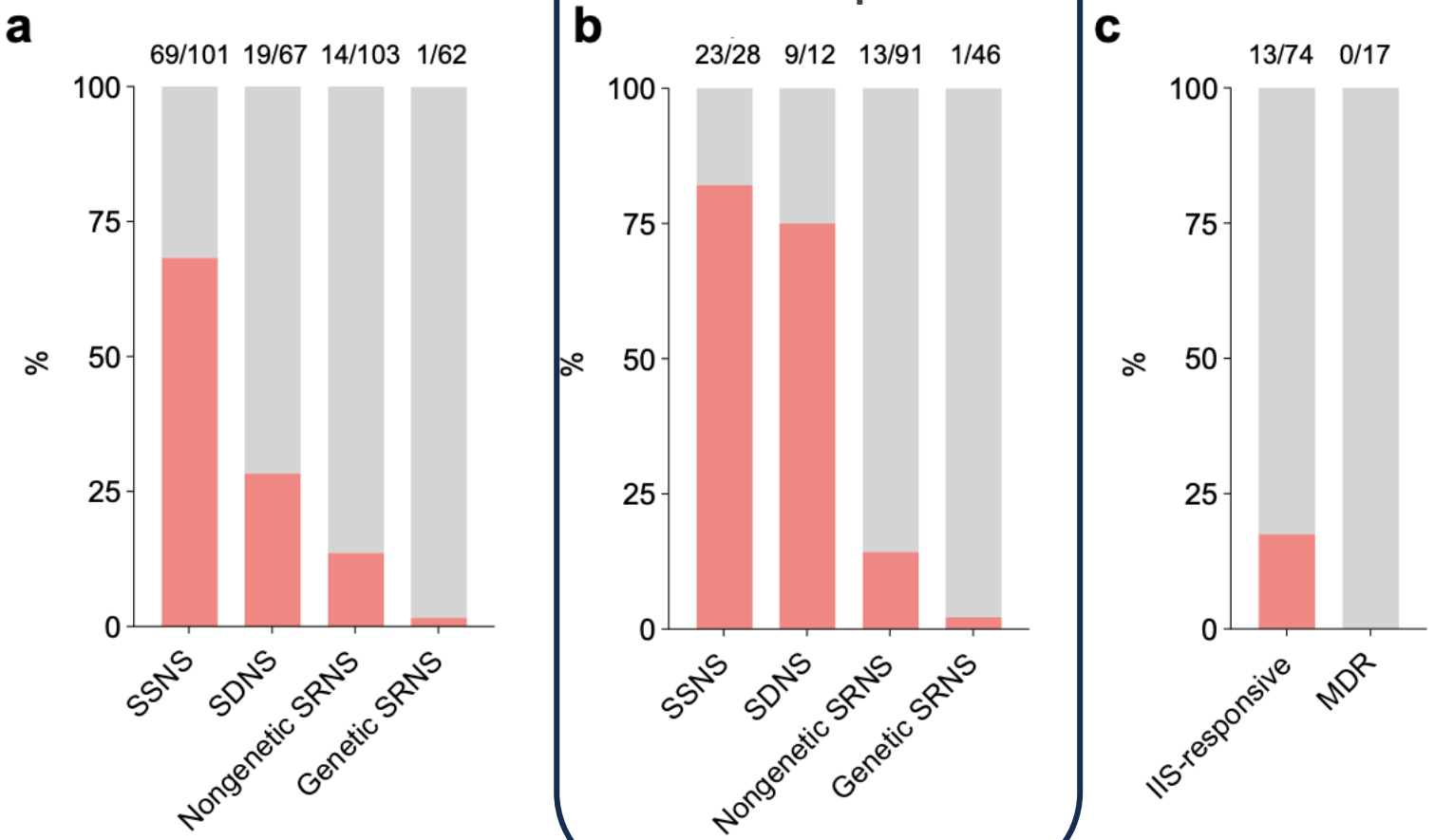
Steroid-Dependant

	Model A adjusted OR (95%CI)	*p* value	Model B adjusted OR (95%CI)	*p* value
Age (years)	1.05 (1.00–1.11)	0.052	1.05 (1.00–1.11)	0.054
Gender (Male)	0.23 (0.04–1.23)	0.087	0.16 (0.03–0.97)	0.046*
Serum creatinine (mg/dL)	0.62 (0.19–2.06)	0.439	0.67 (0.20–2.26)	0.520
Serum albumin (g/dL)	0.26 (0.05–1.36)	0.110	0.21 (0.04–1.13)	0.068
Urinary protein (uPCR, g/gCr)	0.90 (0.77–1.05)	0.164	0.86 (0.72–1.02)	0.090
Any antibody positive	9.88 (1.38–70.78)	0.023*	_	_
Anti-nephrin antibody positive	_	_	9.31 (1.22–70.98)	0.031*

Hayashi Scientific report 2026

Shorter relapse-free period

Interest in Steroid-Resistant Patients



Measurement during active NS phase

Ab Prevalence

- 70% of steroid-sensitive NS
- Genetic form negative
- 20% of Non genetic steroid-resistant forms
- 20% of IS-responders
- 0% of multi-drug resistant

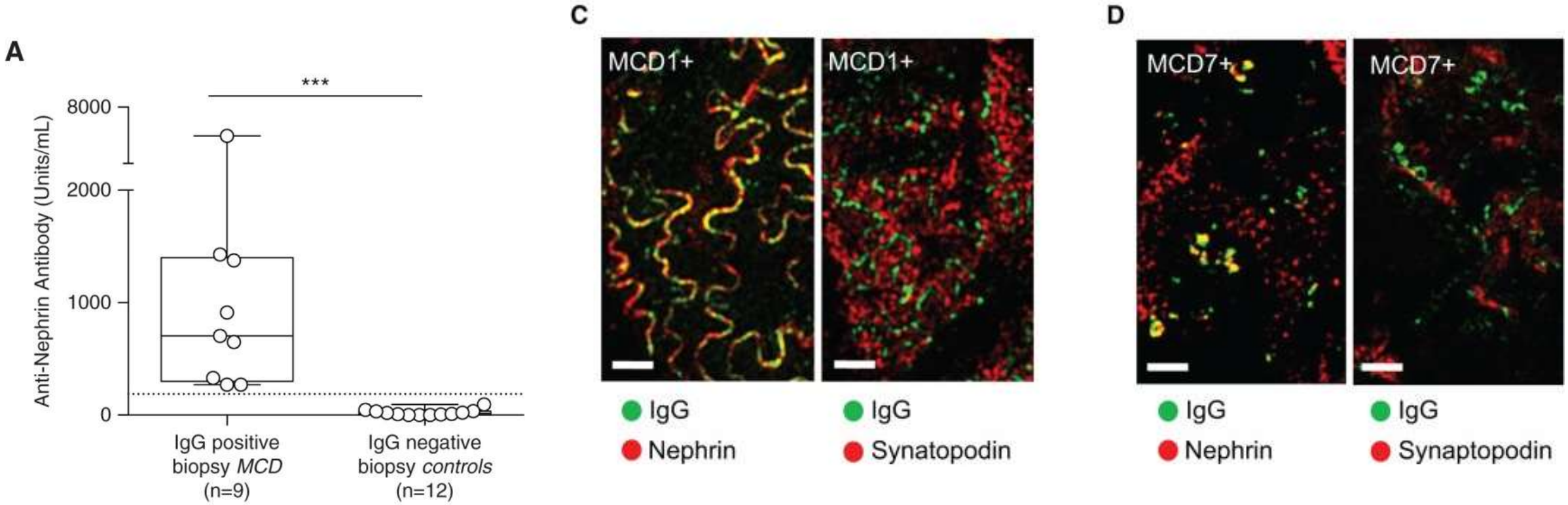
➔ **Therapeutic decision-making tool**

May facilitate therapeutic escalation decisions in steroid-resistant patients

Hengel et al. KI 2025

What About Histology?

New paradigm on histological pattern in idiopathic nephrotic syndrome ?

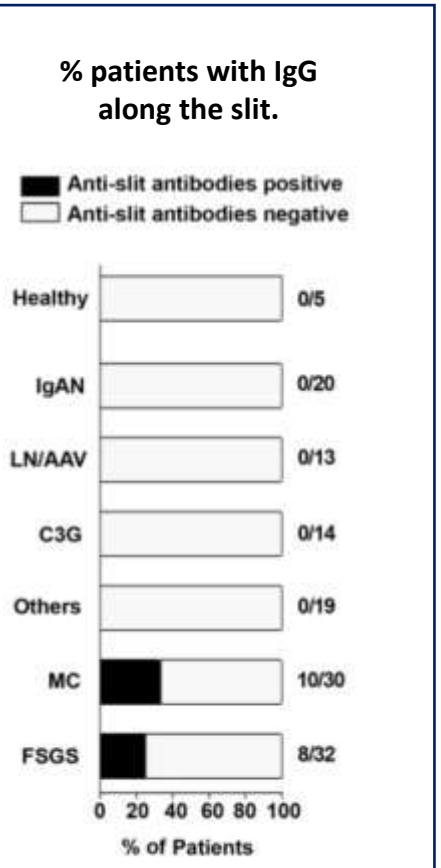


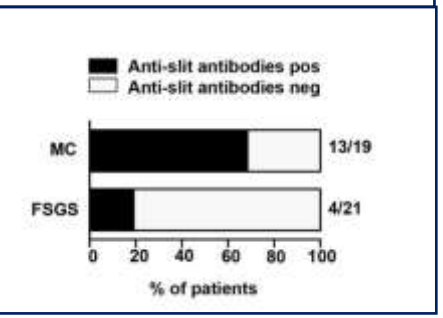
Anti-nephrin Ab (+) all have IgG deposits
IgG and nephrin colocalization

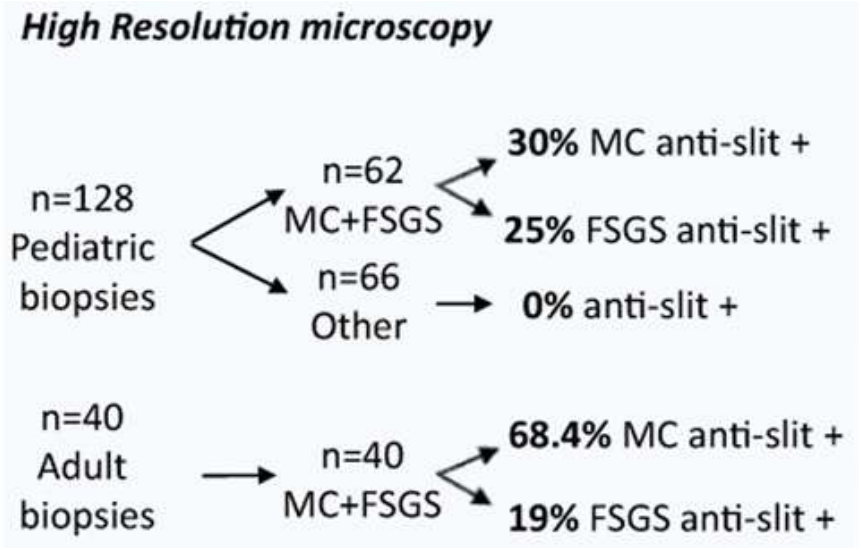
But some NS cases are IgG deposit (+) but without colocalization and antinephrin Ab (-)

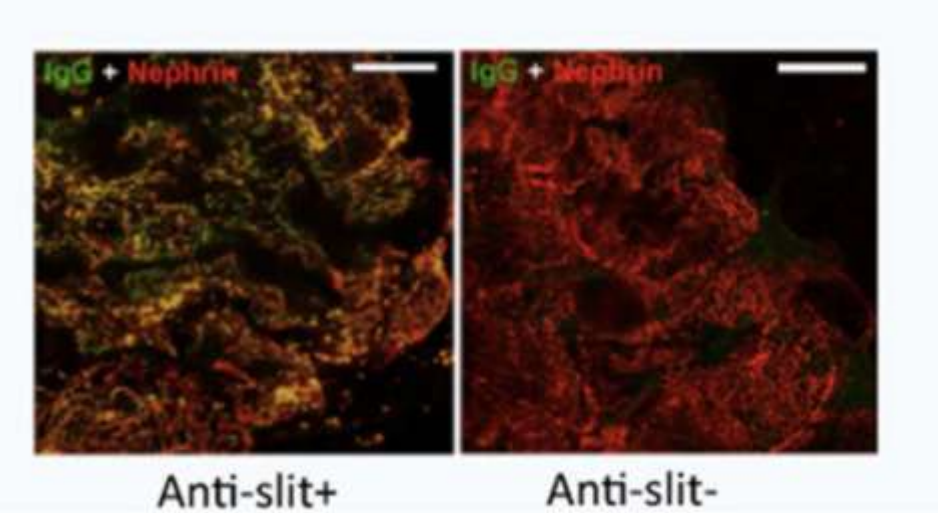
Watts et al. JASN 2022

Idiopathic nephrotic syndrome anti-slit antibody (+)





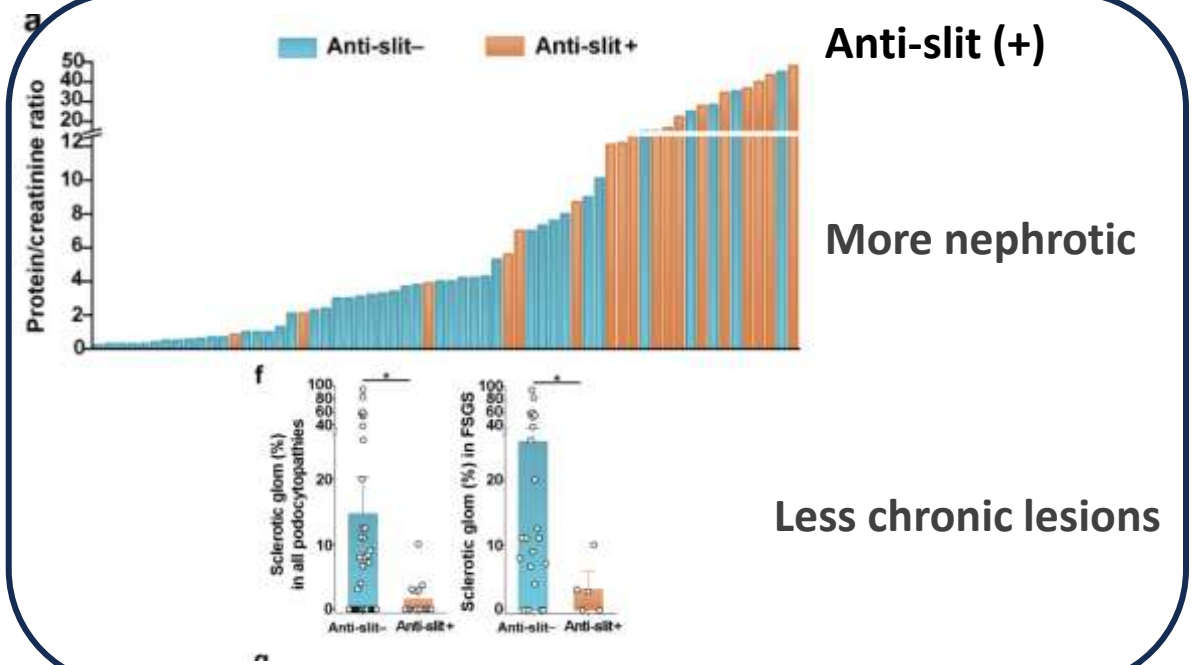




New sub-group
Patients anti-slit Ab (+)

Raglianti et al. KI 2024

Idiopathic Nephrotic syndrome anti-slit antibody (+) Characteristics

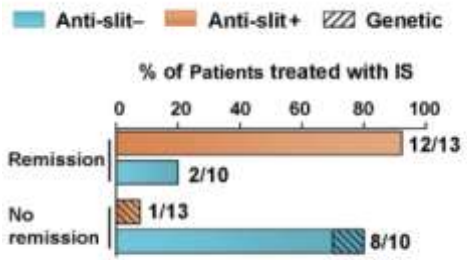


Anti-slit (+)

- More nephrotic
- Not associated with genetic forms
- Response to 2nd line of IS therapy

Spectrum of auto-immune néphrotic syndrome

Steroid Resistant Nephrotic Syndrome

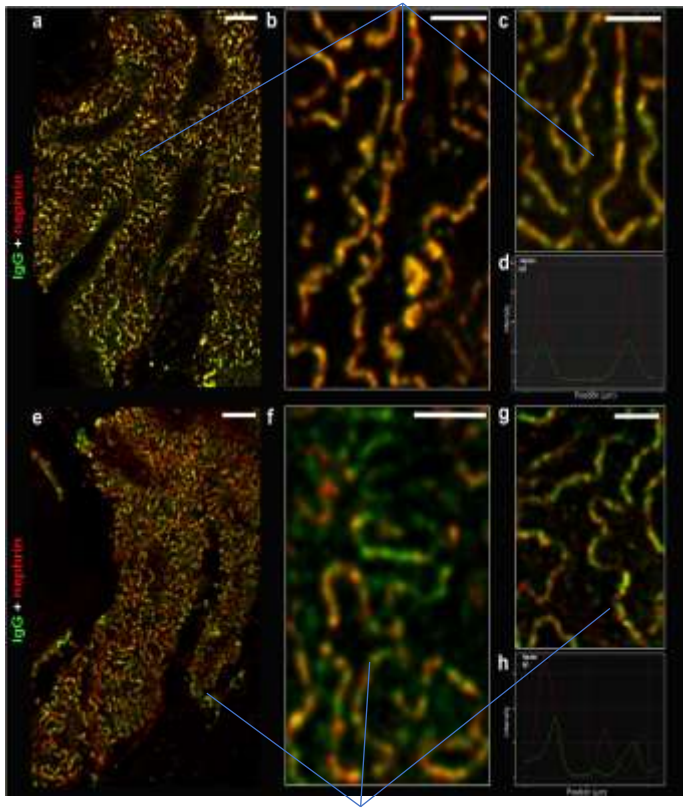


2nd line IS therapy
Anti slit (+) = 90% remission

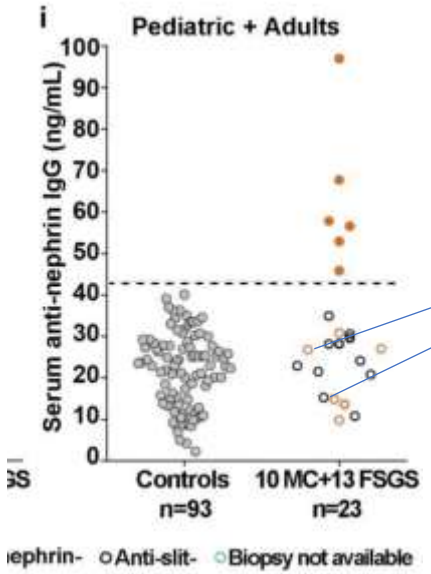
Ragliant et al. i KI 2024

Idiopathic nephrotic syndrome anti-slit antibody (+)

Colocalisation
IgG/nephrin



No Colocalisation
IgG/nephrin



Patient anti-slit(+)

- No cololcalization with nephrin
- ELISA : Anti-nephrin (-)

STED microscopy + ELISA

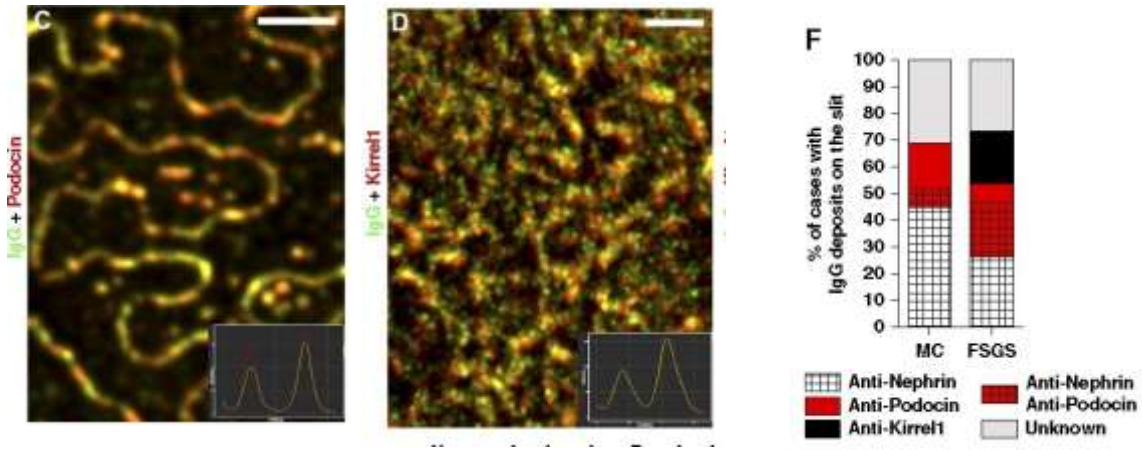
Pediatric patients → **78%** anti-slit + anti-nephrin +
Pediatric patients → **22%** anti-slit+ anti-nephrin -

Adult patients → **55%** anti-slit + anti-nephrin +
Adult patients → **45%** anti-slit+ anti-nephrin -

Other target ?

Raglianti et al. KI 2024

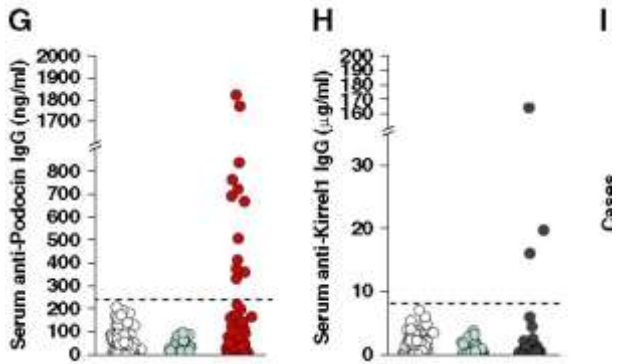
Idiopathic nephrotic syndrome anti-slit antibody (+)

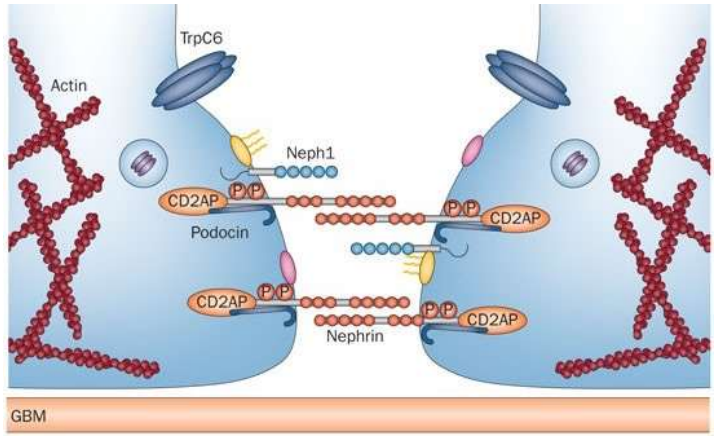


N= 116 biopsises

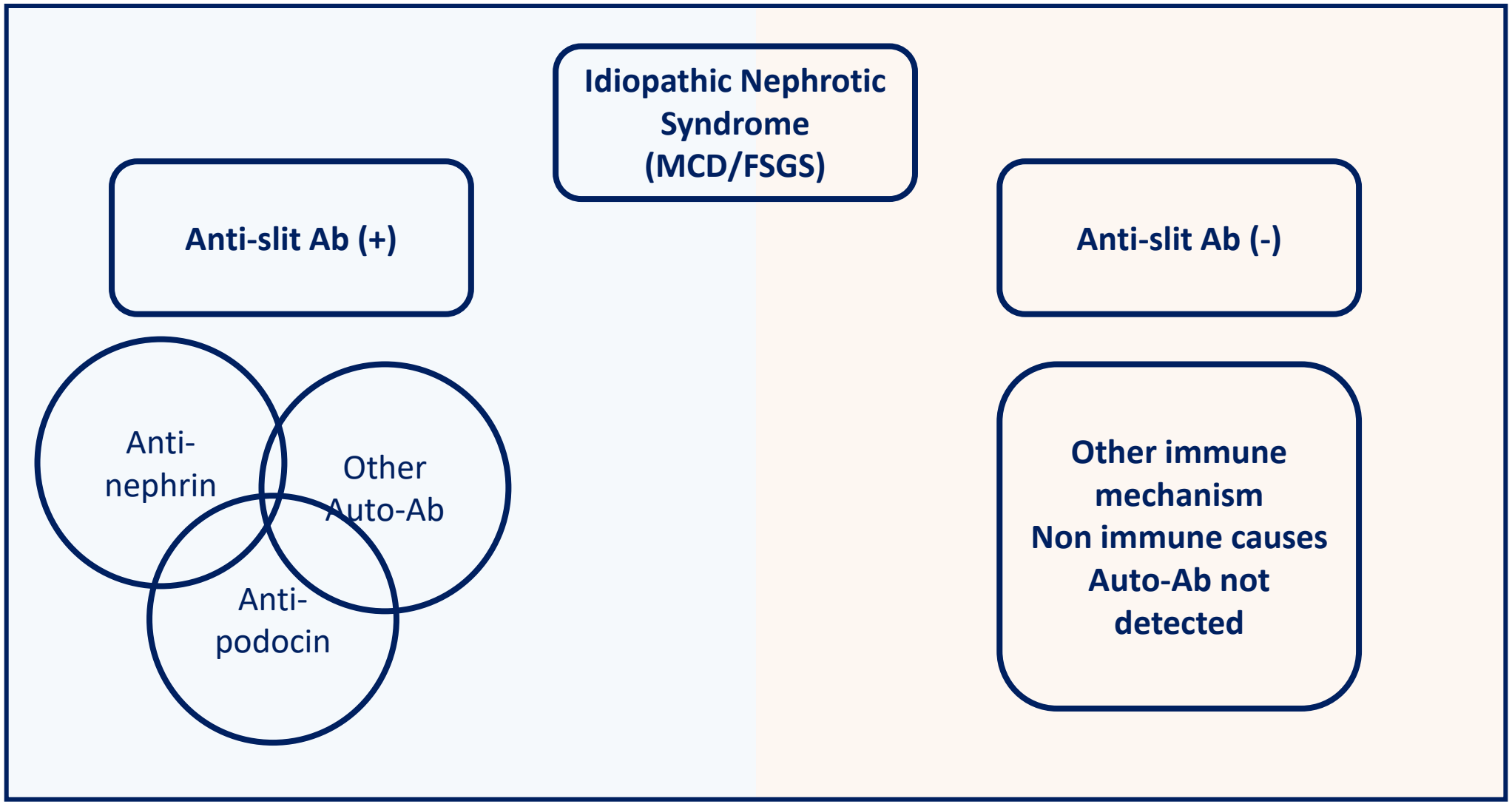
➔ ***Identification of anti-podocines 18%***

➔ ***Identification of anti-Kirrel 1 5%***





Raglianti et al. KI 2024
Raglianti et al. JASN 2025


Idiopathic Nephrotic Syndrome (MCD/FSGS)
Anti-slit Ab (+)
Anti-slit Ab (-)
Anti-nephrin
Other Auto-Ab
Anti-podocin
Other immune mechanism
Non immune causes
Auto-Ab not detected

A new biomarker of recurrent FSGS after kidney transplantation?

Anti-nephrin Ab and relapse risk in FSGS

Pre-transplant serum n = 39 KT recipients

- **Recurrence:** 22 recurent vs. 17 non-recurent FSGS

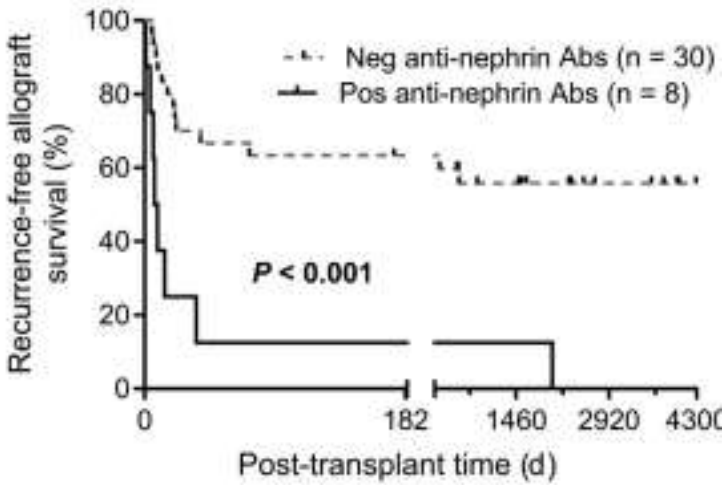
- **Anti-nephrin Ab (+): 38% in recurrent FSGS**

Recurrence risk: HR = 4.9

Allograft biopsy: IgG deposits colocalized with nephrin only in recurent cases.

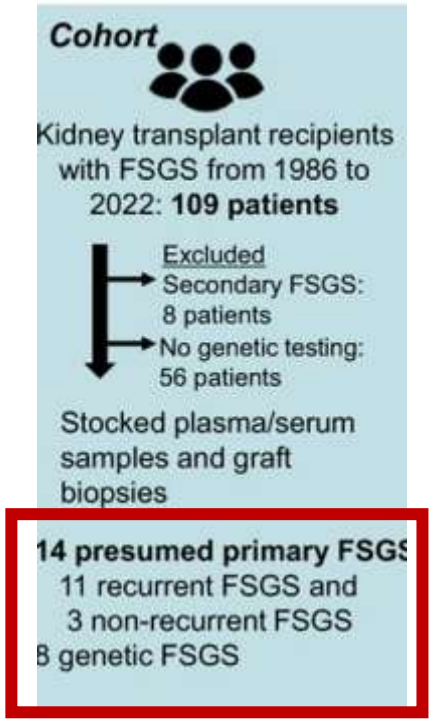
Relapse prediction

Se 8/21 = 38%
Sp: 17/17 = 100%
PPV: 8/8 = 100%
NPV17/30 = 57%



Batal et al. KI 2024

Anti-nephrin Ab at recurence



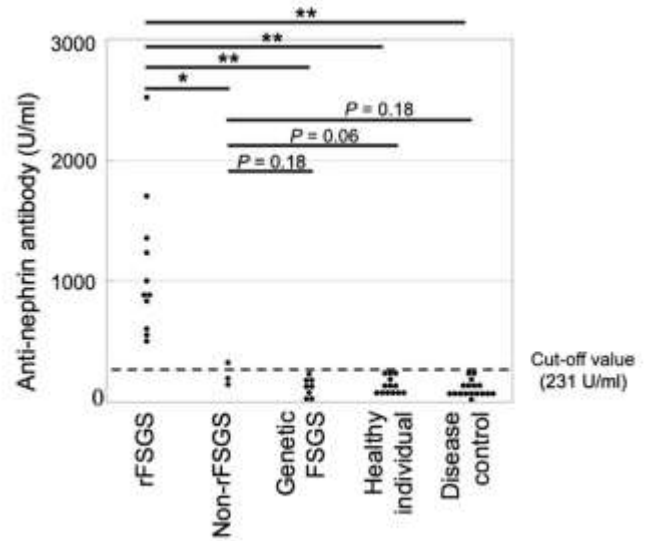


Figure 2| Anti-nephrin antibody levels during post-transplant focal segmental glomerulosclerosis (FSGS) recurrence. The cutoff

Shirai et al. KI 2024

Anti-nephrin Ab and relapse risk in FSGS : How to manage ?

Living donor case

Case

An adolescent with pFSGS and evidence of anti-nephrin antibodies detected prior to living-donor KTx

Pretransplant reduction of anti-nephrin antibody levels by RTX and TPE.

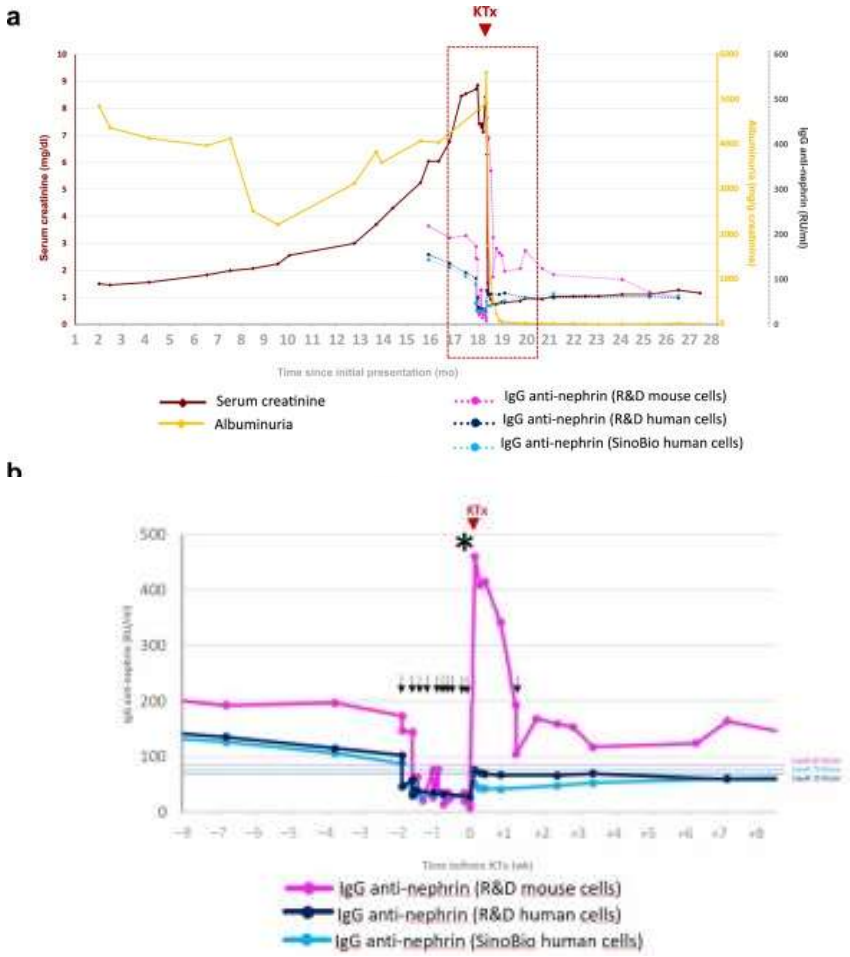
More than 1 year of follow-up without recurrence of proteinuria.

Methods

Monitoring of anti-nephrin antibody levels by ELISA, Immunoprecipitation and Western Blot using different nephrin proteins.

Glomerular co-staining of nephrin and IgG by immunofluorescence.

Characterization of cross-reactivity with the additional slit diaphragm protein NEPH3.



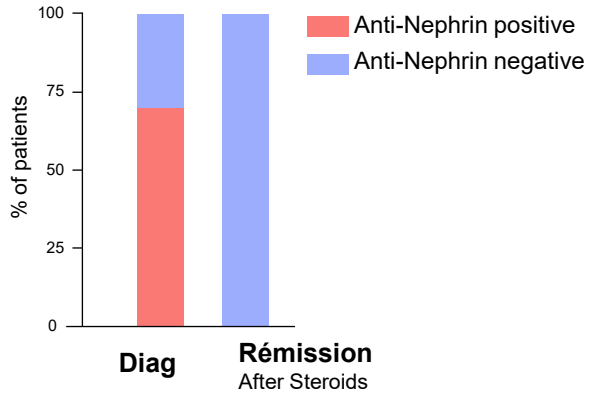
➢ **What about deceased donor management ?**

Habbig et al. KI 2025

Pathogenicity of anti-nephrin antibodies ?

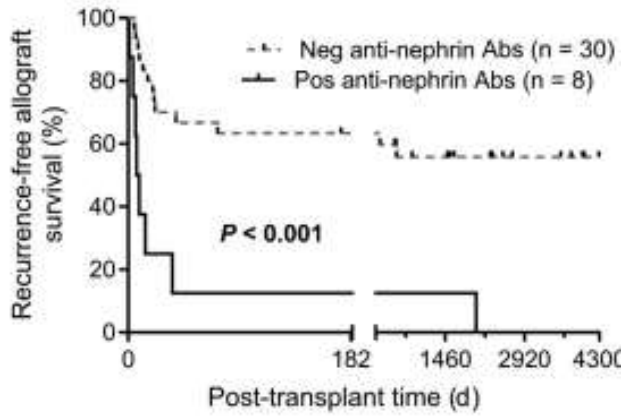
Anti-nephrin antibodies: are they pathogenic?

➢ **Correlation with disease activity**

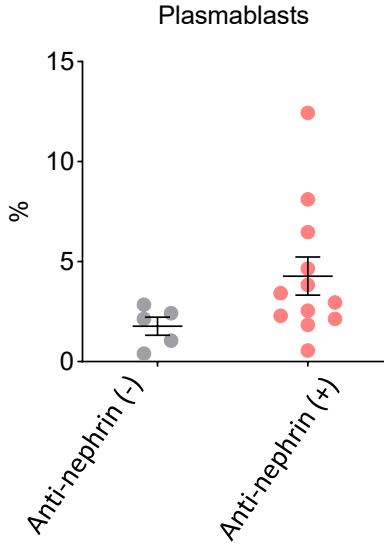


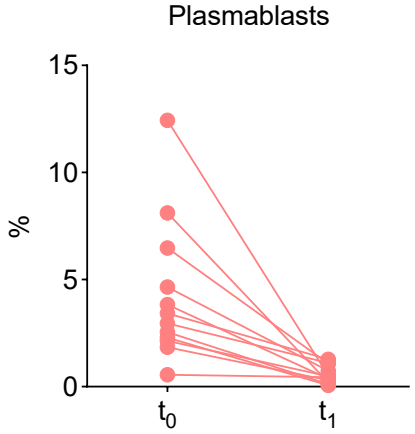
Negativation of Anti-nephrin Ab in less than 8 weeks
After steroid therapy initiation

Correlation with relapse



➢ **Correlation with B cell markers**





Hengel, Sakhi* et al. KIR 2026*
Batal et al. KI 2024

Anti-nephrin antibodies: are they pathogenic?

- **Correlation with disease activity**

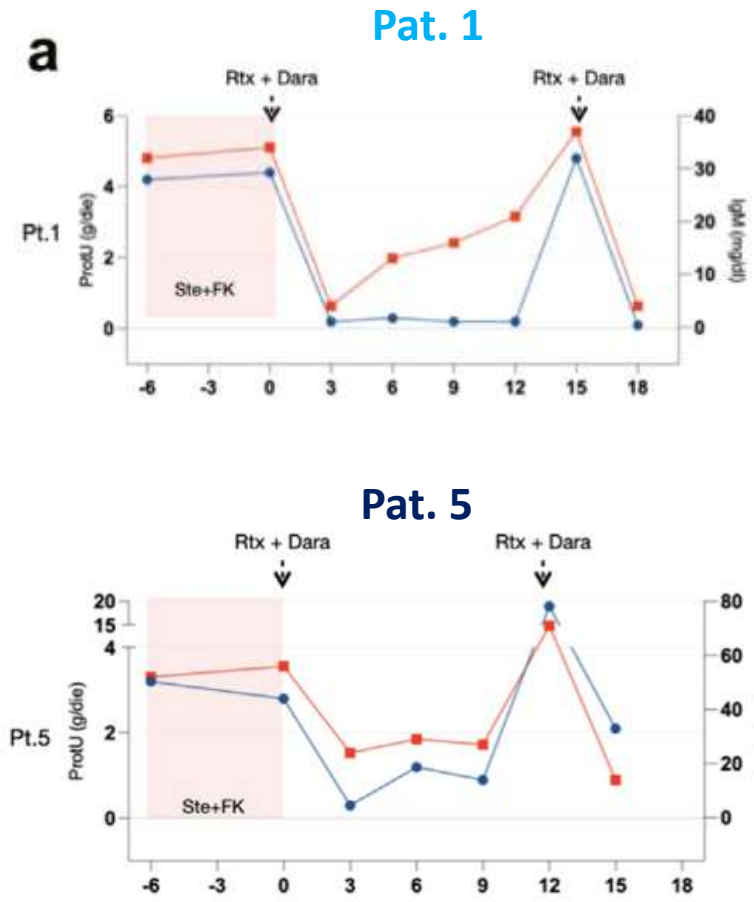
Anti-nephrin antibodies: are they pathogenic?

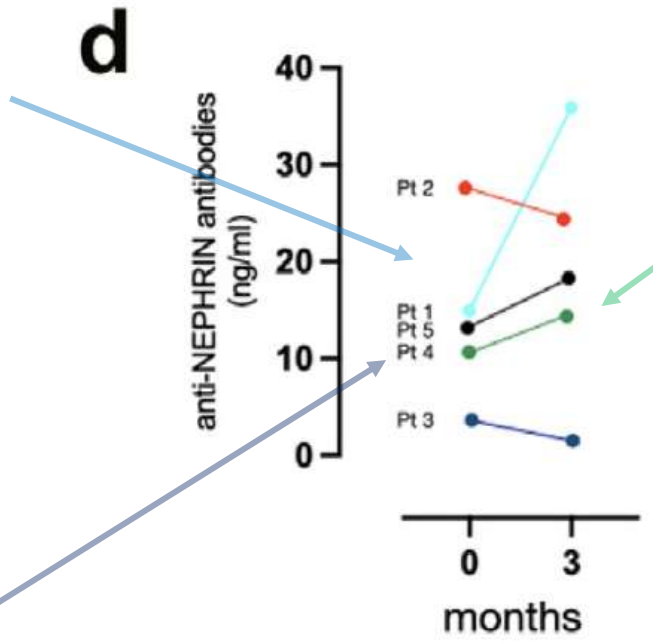
- **Correlation with disease activity** **Not always**

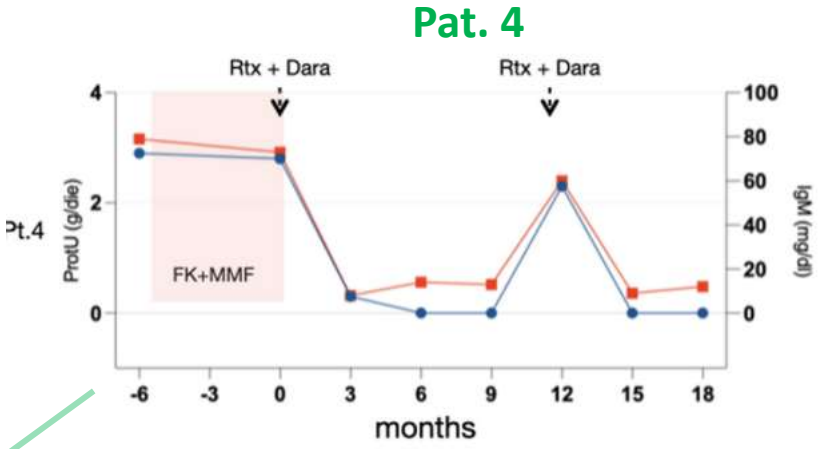
Anti-nephrin antibodies: are they pathogenic?

- Correlation with disease activity **Not always**

MDR-Nephrotic Syndrome







Clinical remission in patients 1, 4 and 5 without decrease of anti-nephrins Ab

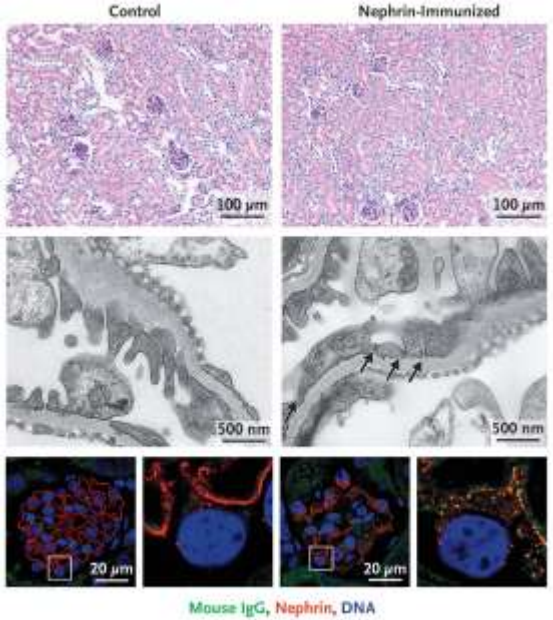
Month 0→ 3

Angeletti et al. KIR 2024

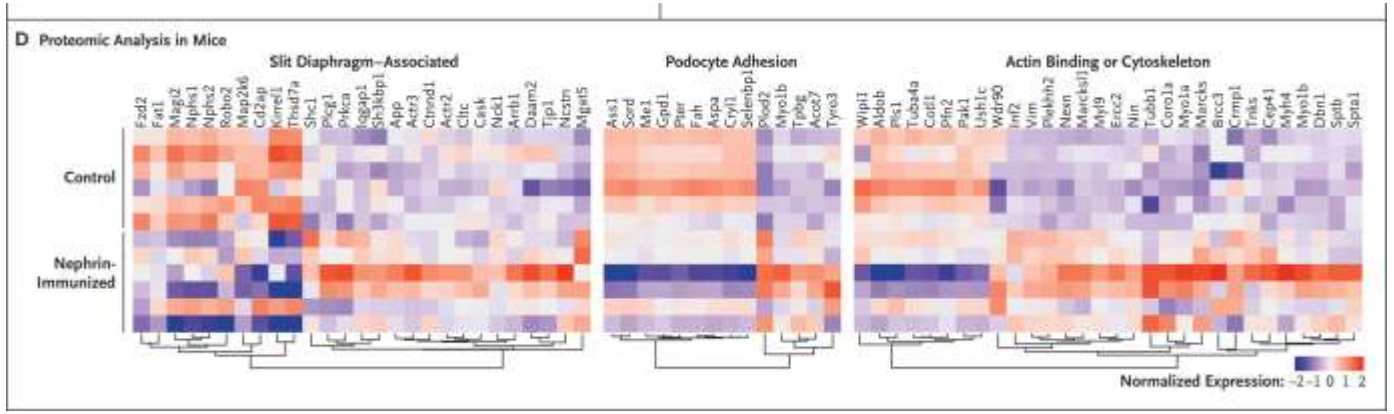
Anti-nephrin antibodies: proposed mechanism of action

immunization with rec nephrin + Freund adjuvant

Nephrotic Syndrome
Histological pattern MCD-Like
IgG deposit

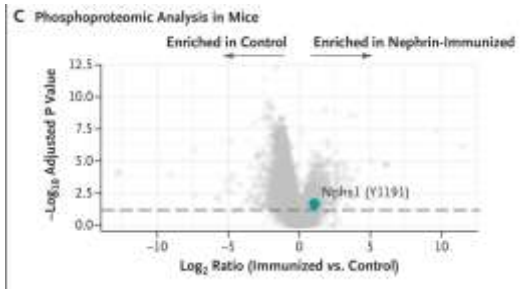


Glomerular Proteome



- ↑ Endocytic trafficking proteins
- ↓ Slit diaphragm proteins
- Altered podocyte adhesion proteins
- Cytoskeletal protein remodeling

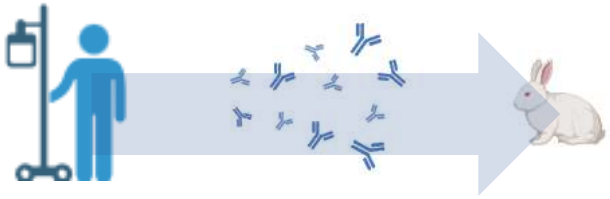
Nephrin phosphorylation



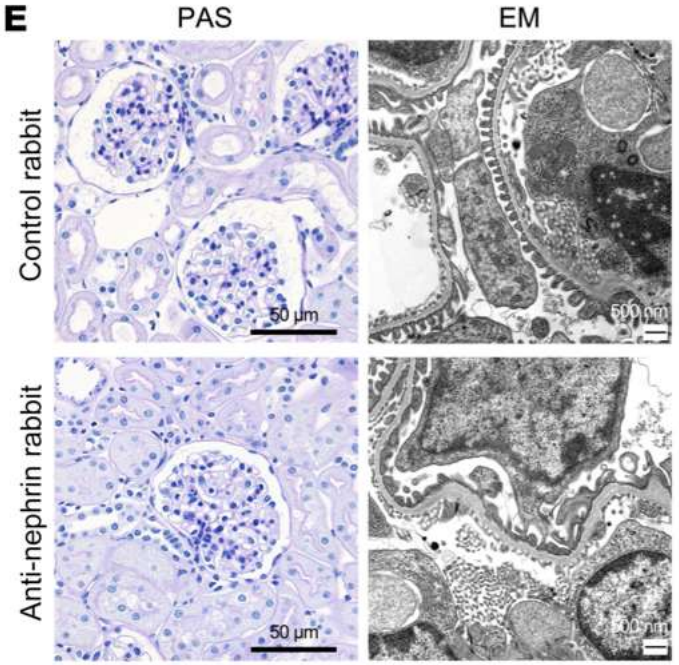
Hengel et al. NEJM 2024

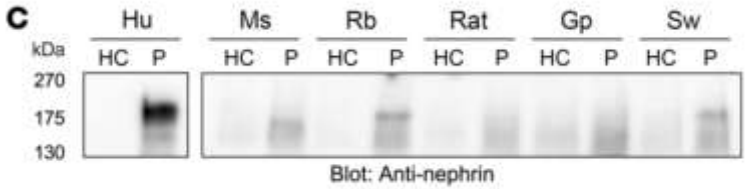
Anti-nephrin antibodies: proposed mechanism of action

Patient with NS-derived total IgG purified from PLEX



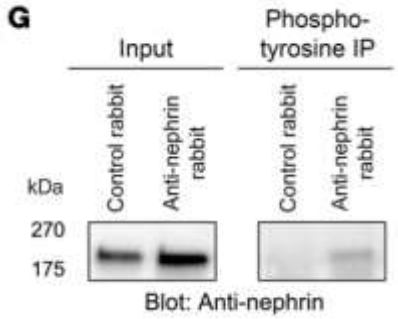
Nephrotic Syndrome
MCD-like pattern

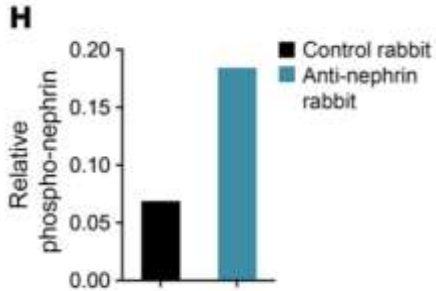




React to rabbit glomerular lysat++

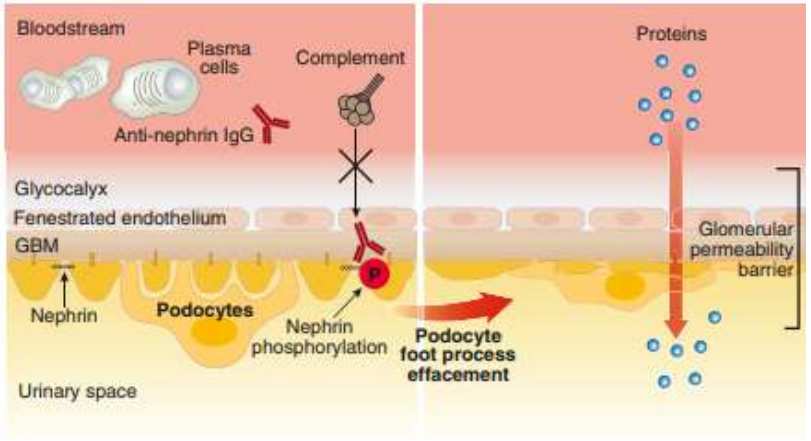
Nephrin phosphorylation





Hengel et al. JCI 2024

Anti-nephrin antibodies: proposed mechanism of action



Vivarelli et al. KI 2025

Anti-nephrin antibodies

Nephrin binding / immune complex formation

Nephrin phosphorylation

Nephrin internalization

Slit diaphragm and podocytes dysregulation

But some previous studies showed decreased nephrin phosphorylation in MCD

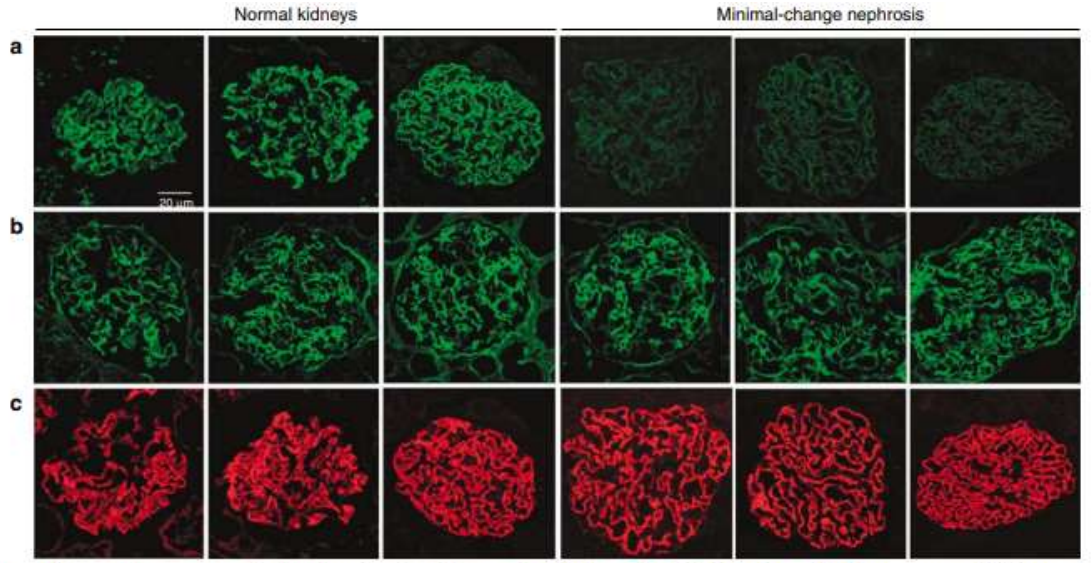
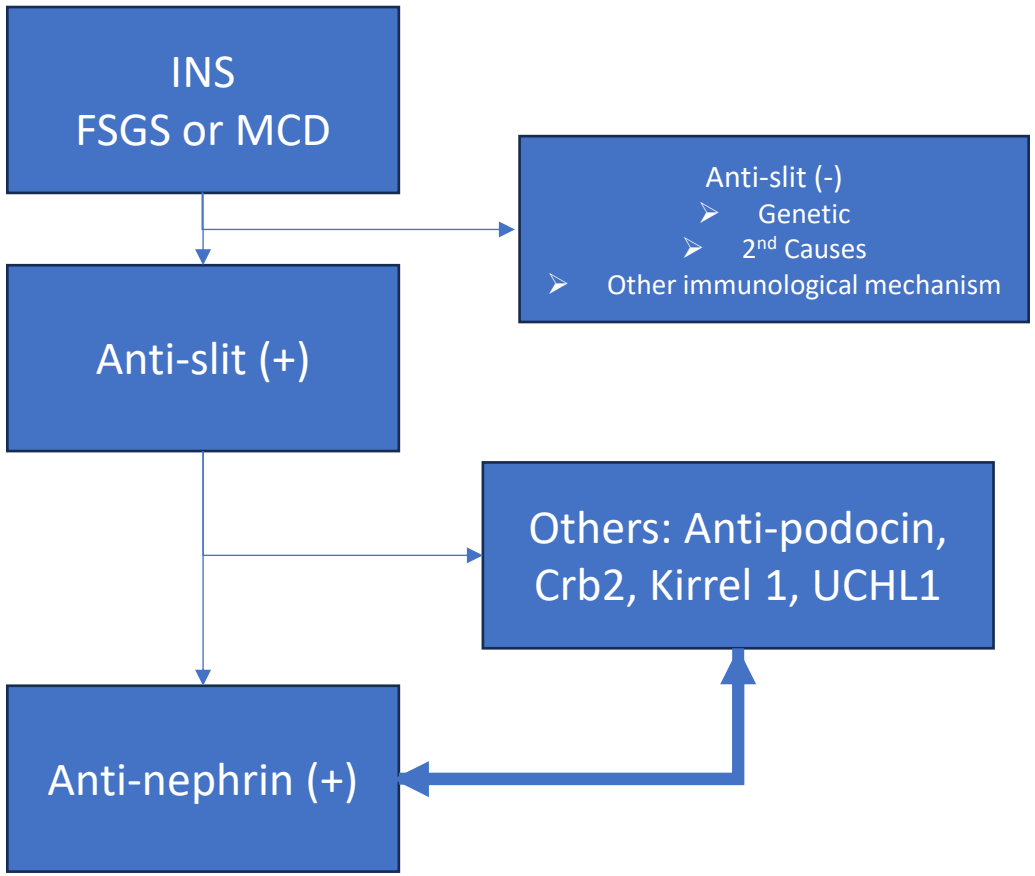


Figure 6 | Immunofluorescence of phosphorylated nephrin, total nephrin, and podocin in glomeruli of human normal kidneys and minimal change nephrosis. Biopsy specimens from donor kidneys and minimal change nephrosis were immunostained with anti-pY1228 (**a**), anti-total nephrin (**b**), and anti-podocin (**c**) antibodies. Three representative cases were shown. Signals were detected by using LSM510-Meta confocal microscopy under a constant laser and detector setting.

Uchida et al. KI 2008

Conclusion

Redefining the Nosological Framework



Circulating anti-nephrin antibodies

- **Prevalence 70% in IS-naive MCD**
- Primary FSGS <20%
  - ➔ Recurrent FSGS >70%

➔ **Prognostic and therapeutic management tool?**

- ➔ In steroid-resistant forms: **may facilitate intensification**
- ➔ Interest in monitoring and relapse prediction?
- ➔ **In pre-transplant setting ++**

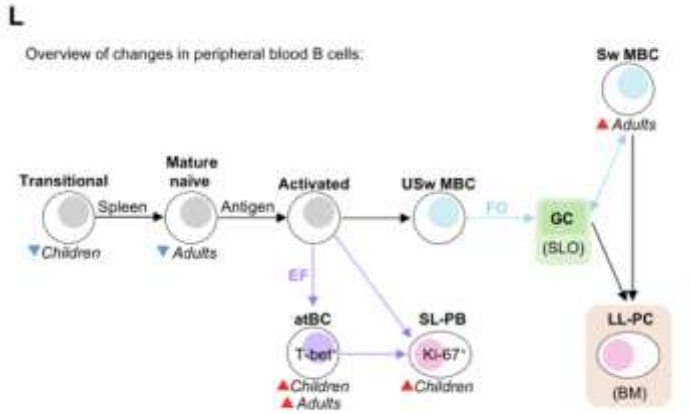
Blume et al. JASN 2025

ERKNet
The European
Rare Kidney Disease
Reference Network

Thank You

ORKiD
FILIÈRE ORPHAN KIDNEY DISEASES

Anti-nephrin nephrin : Mechanisms of action

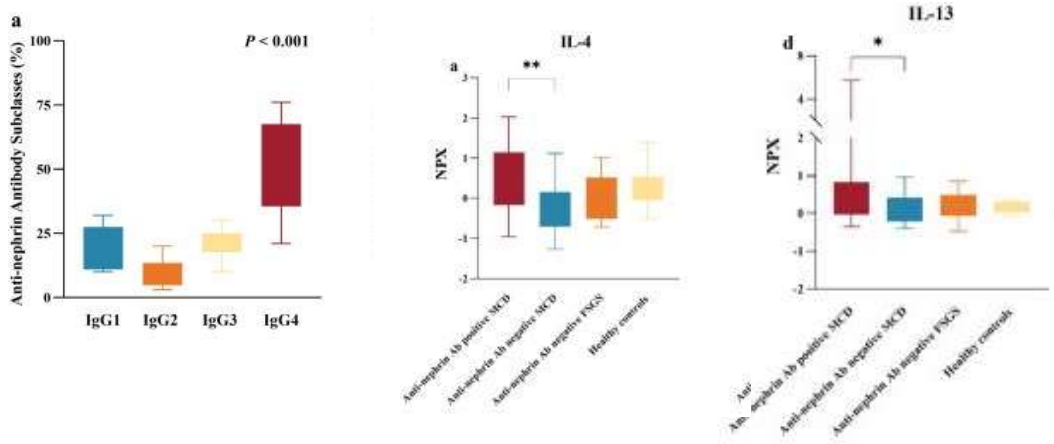


Réponse GC (>adulte)

Réponse EF (>enfant)

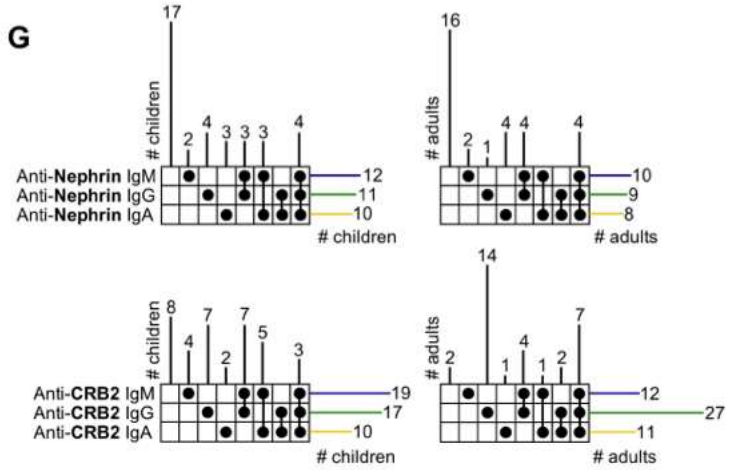
Al aubodah Bioxv 2026

60 MCD (14 enfants)

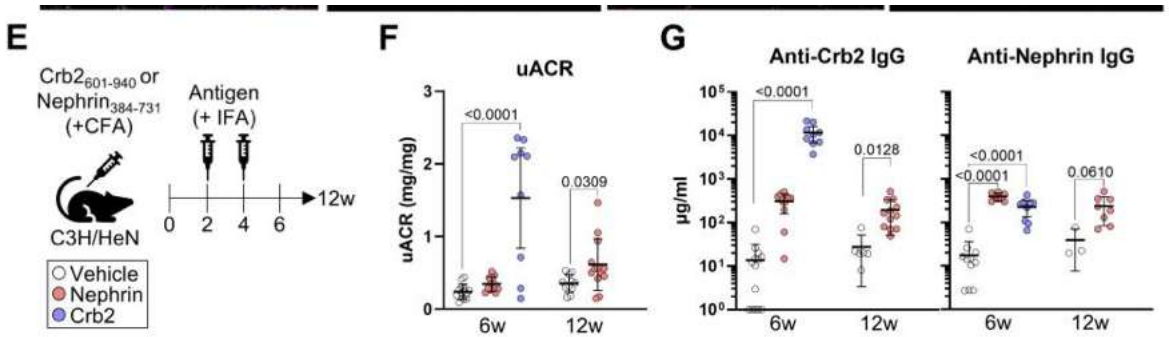


Augmentation Antinephrine-IgG4
Réponse Th2
Phase plus tardive ?

Guo Annals of medicine 2026



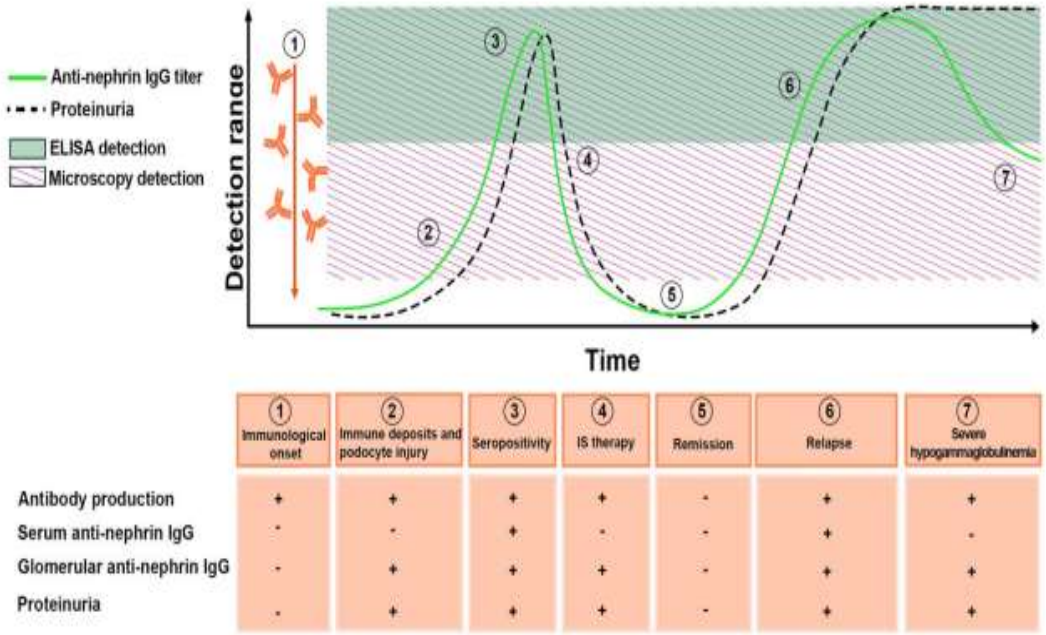
Enfant Plus d'auto anticorps IgM > Phase précoce ?



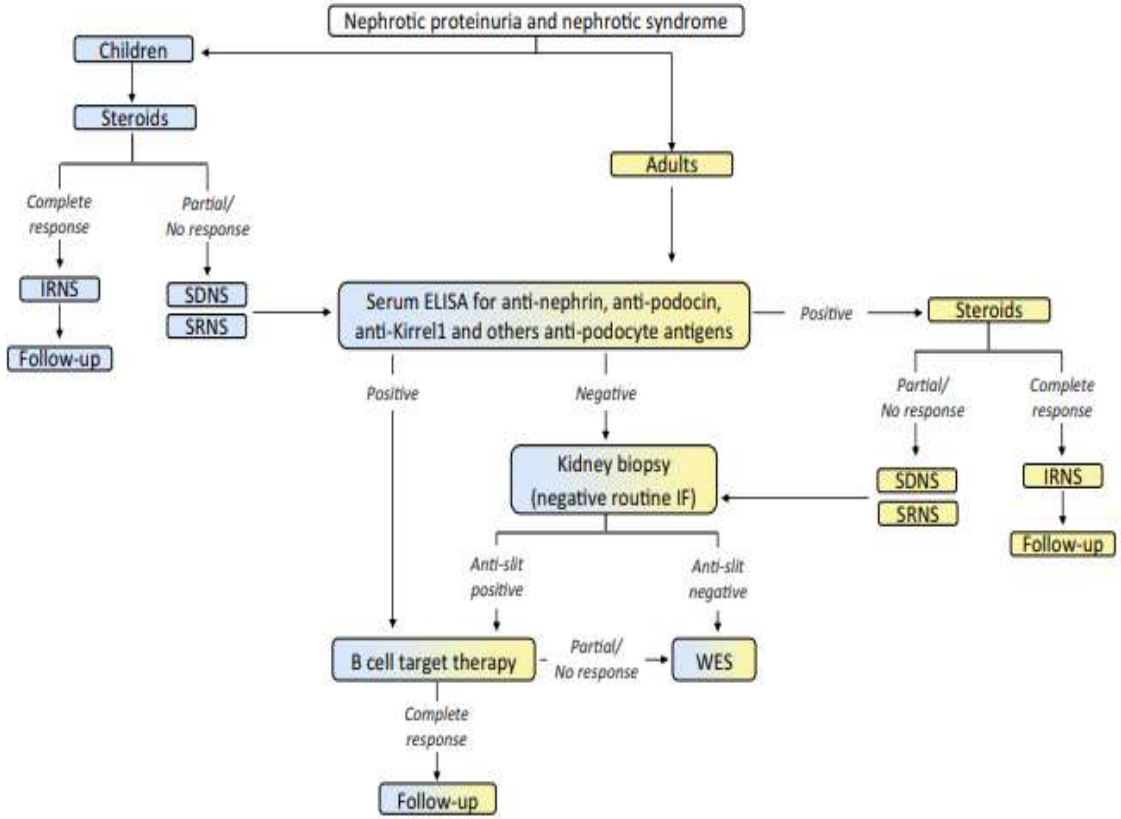
Intermolecular Spreading

Al aubodah Bioxv 2026

Conclusion



	1 Immunological onset	2 Immune deposits and podocyte injury	3 Seropositivity	4 IS therapy	5 Remission	6 Relapse	7 Severe hypogammaglobulinemia
Antibody production	+	+	+	+	-	+	+
Serum anti-nephrin IgG	-	-	+	-	-	+	-
Glomerular anti-nephrin IgG	-	+	+	+	-	+	+
Proteinuria	-	+	+	+	-	+	+

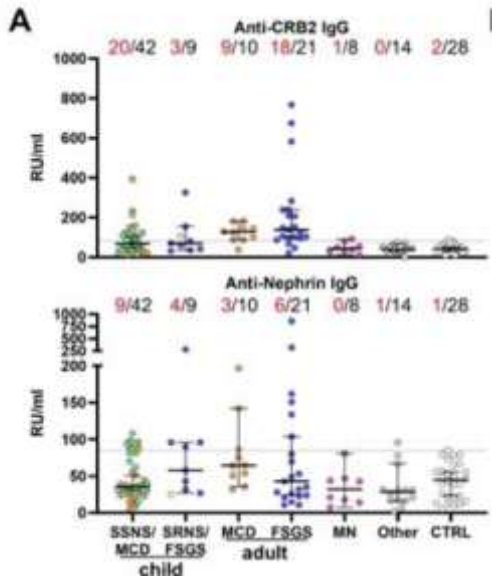


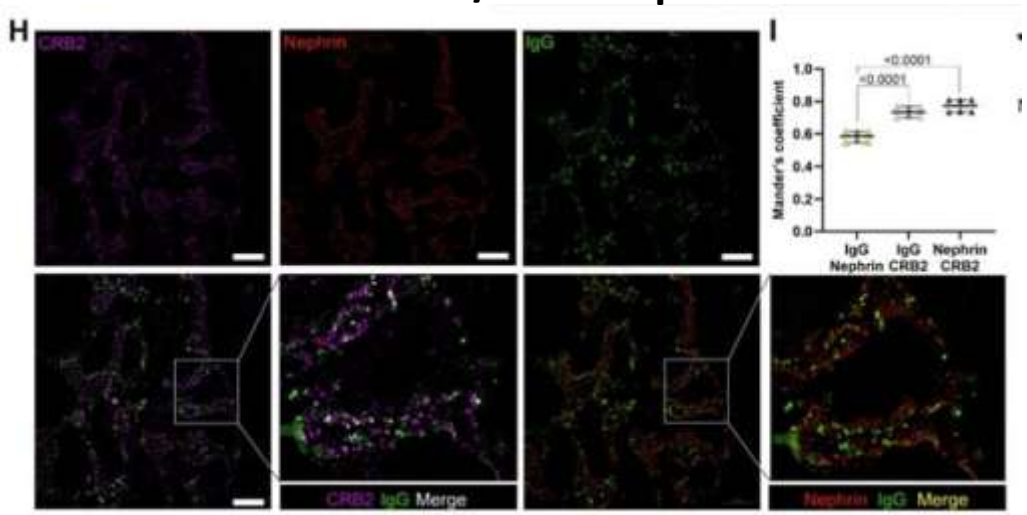
Raglinati NDT 2025

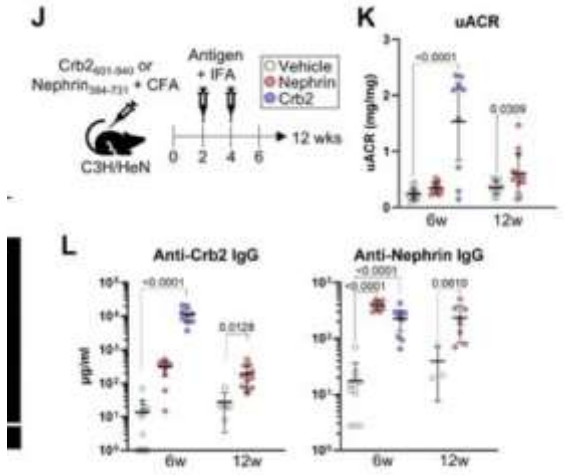
Anti-nephrin nephrin : Mechanisms of action

Epitope spreading ?

Some patients:
Anti CRB2 / Anti Nephrines





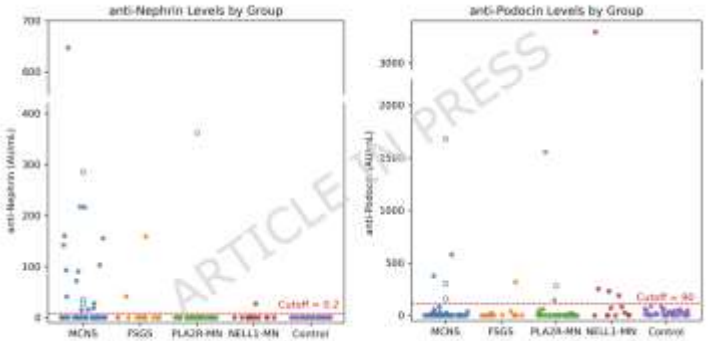


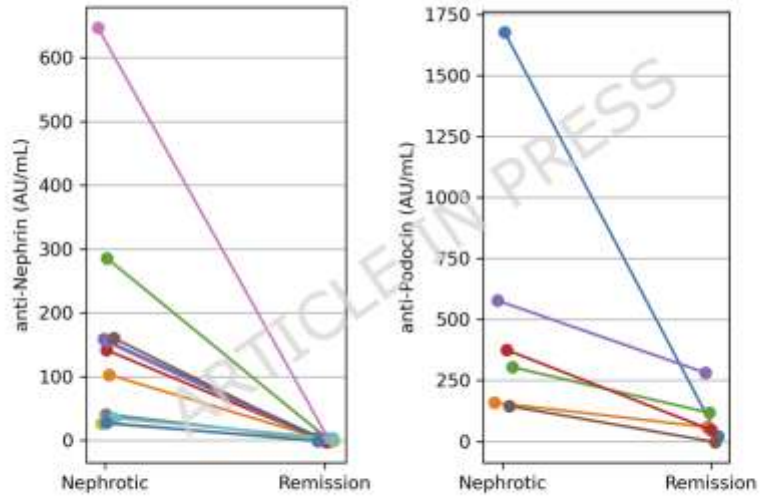
Mouse immunization contre Crb2

- **Develop anti-Crb2 autoantibodies**
- **Mais aussi of anticorps anti nephrines**

Study type
•Étude **Retrospective monocentric**
•Cohort of adult patients with biopsied nephrotic syndrome
Population (n)
•**n = 114 patients**
  •MCD: 47
  •FSGS: 14
  •MN (PLA2R/NELL1): 53
•Controls: n = 40

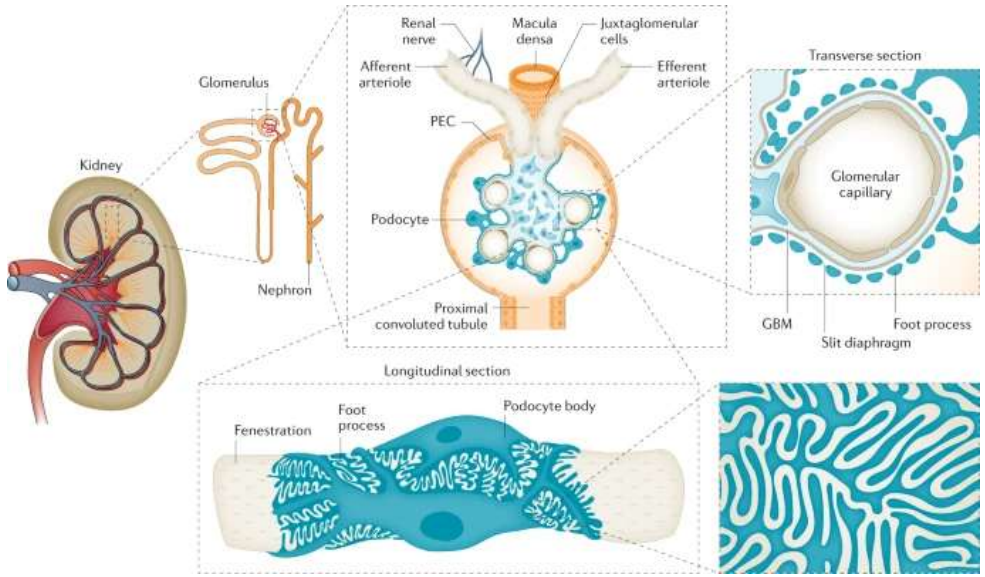
Main results
•**Distribution of Ac :**
  •Anti-nephrin : surtout **MCNS (~38%)**
  •Anti-podocine : **non-specific**, present in several GN (↑ NELL1-MN)
•**Initial severity:**
  •Patients Ac+ → **proteinuria ↑ and creatinine ↑**
•**Prognosis/outcome:**
  •Pas d'impact on la **initial remission**
  •Mais association forte avec **SDNS**
    •OR ≈ **9–10** (independent in multivariate)
•**Biological dynamics:**
  •Diminution/disparition of Ac in remission
•**Stratification:**
  •Double positivity → **maximum risk of relapse / dependency**

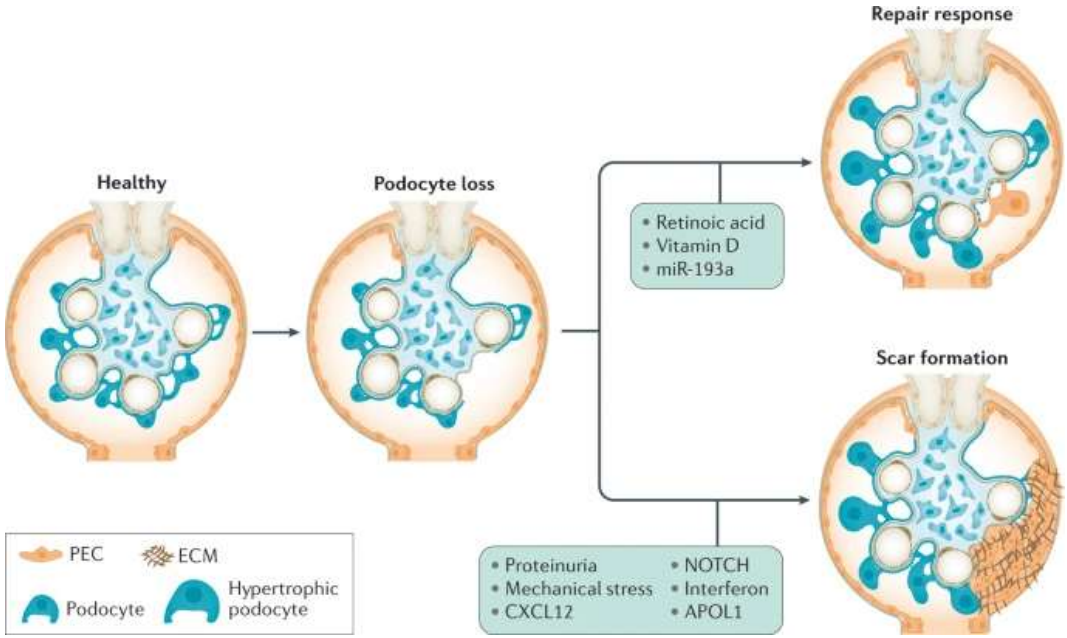




Hayashi scientific reports 2025

Podocyte = Primary Target



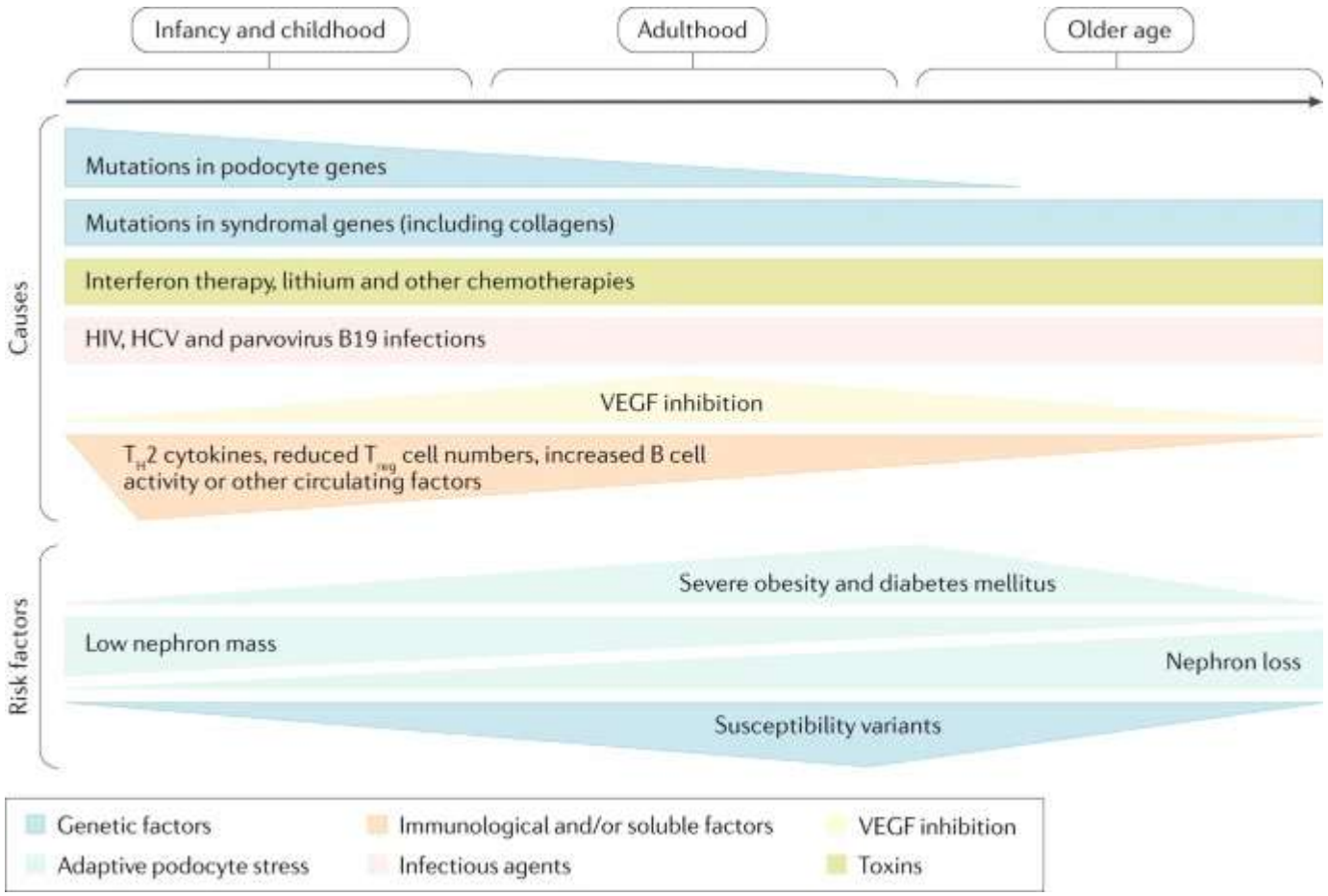


- Podocyte injury (ds le SNI : auto immune)
- Stress → remaniement cytosquelette → **foot process effacement,**, loss of adhesion and **detachment**.
- Disruption of filtration barrier → Proteinuria

➔ MCD
➔ FSGS

Kopp Nat. Disease primers 2020
Romagnani Natu disease primer 2025

Cause of podocytopathies



1. Genetic causes
2. Immunological causes
3. Causes infectieuses et toxiques.
4. Hemodynamic and metabolic causes
•Obesity, diabetes, hyperfiltration, nephron reduction

Kopp Nat. Disease primers 2020
Romagnani Natu disease primer 2025

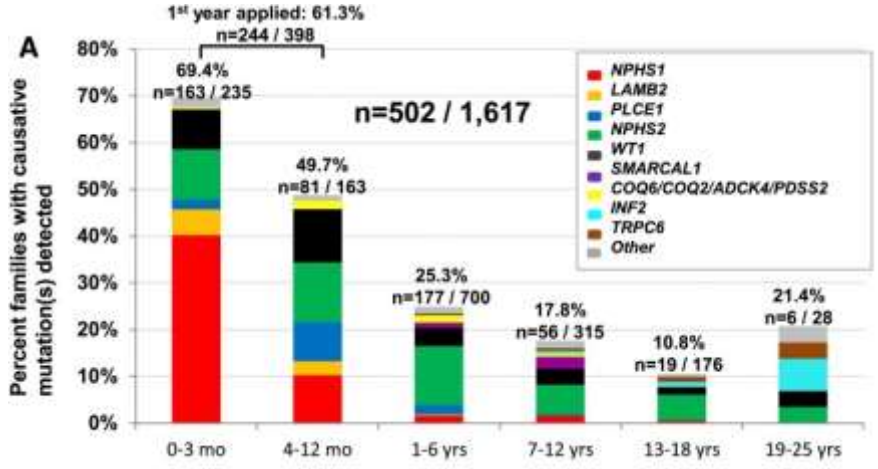
Idiopathic Nephrotic Syndrome

% pathogenic variant

MCD
- Steroid-sensitive = 80%
- Steroid-resistant = 20%

FSGS-NS
- Steroid-sensitive = 40%
- Steroid-resistant = 60%

30% of steroid-resistant NS = genetic



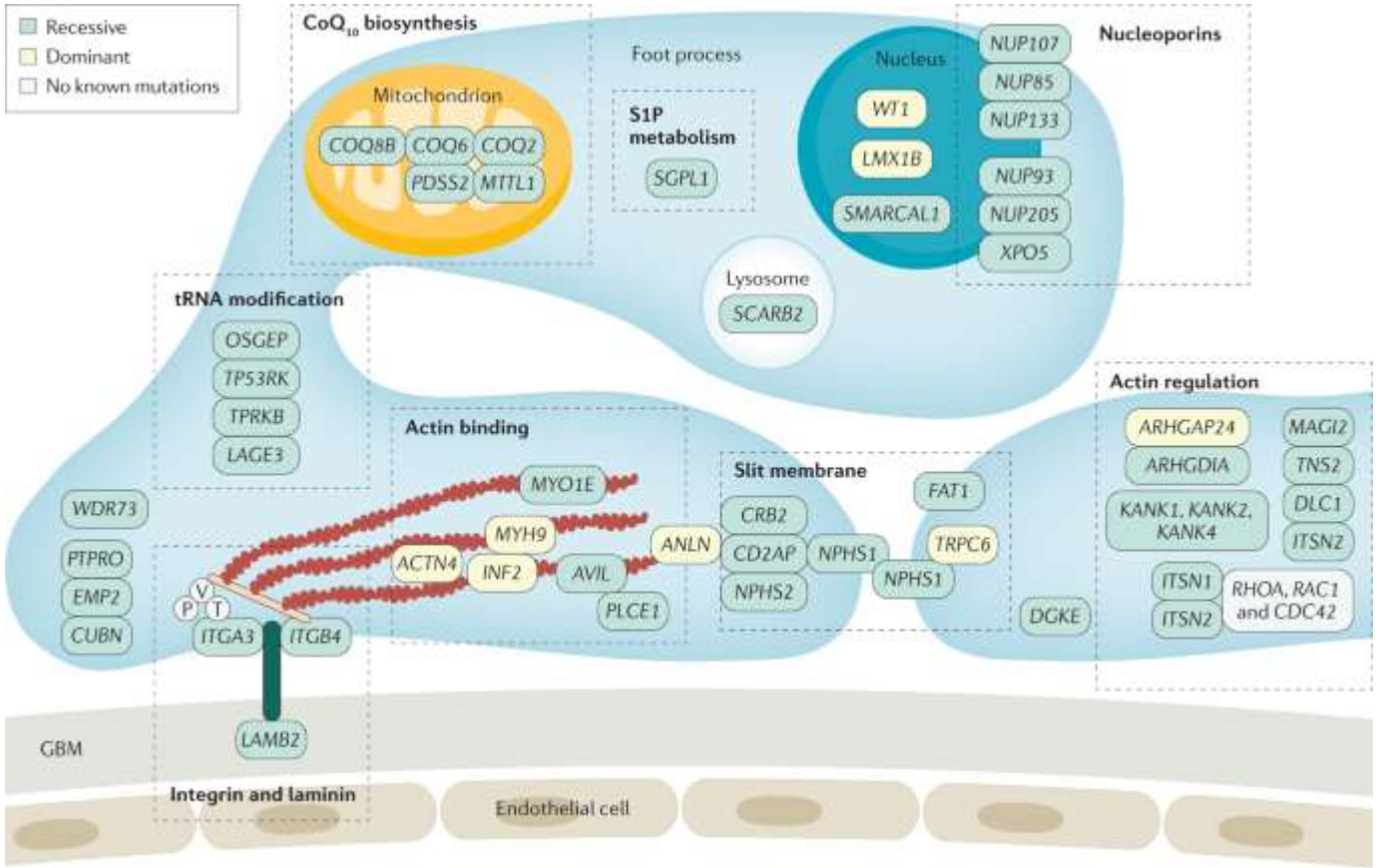
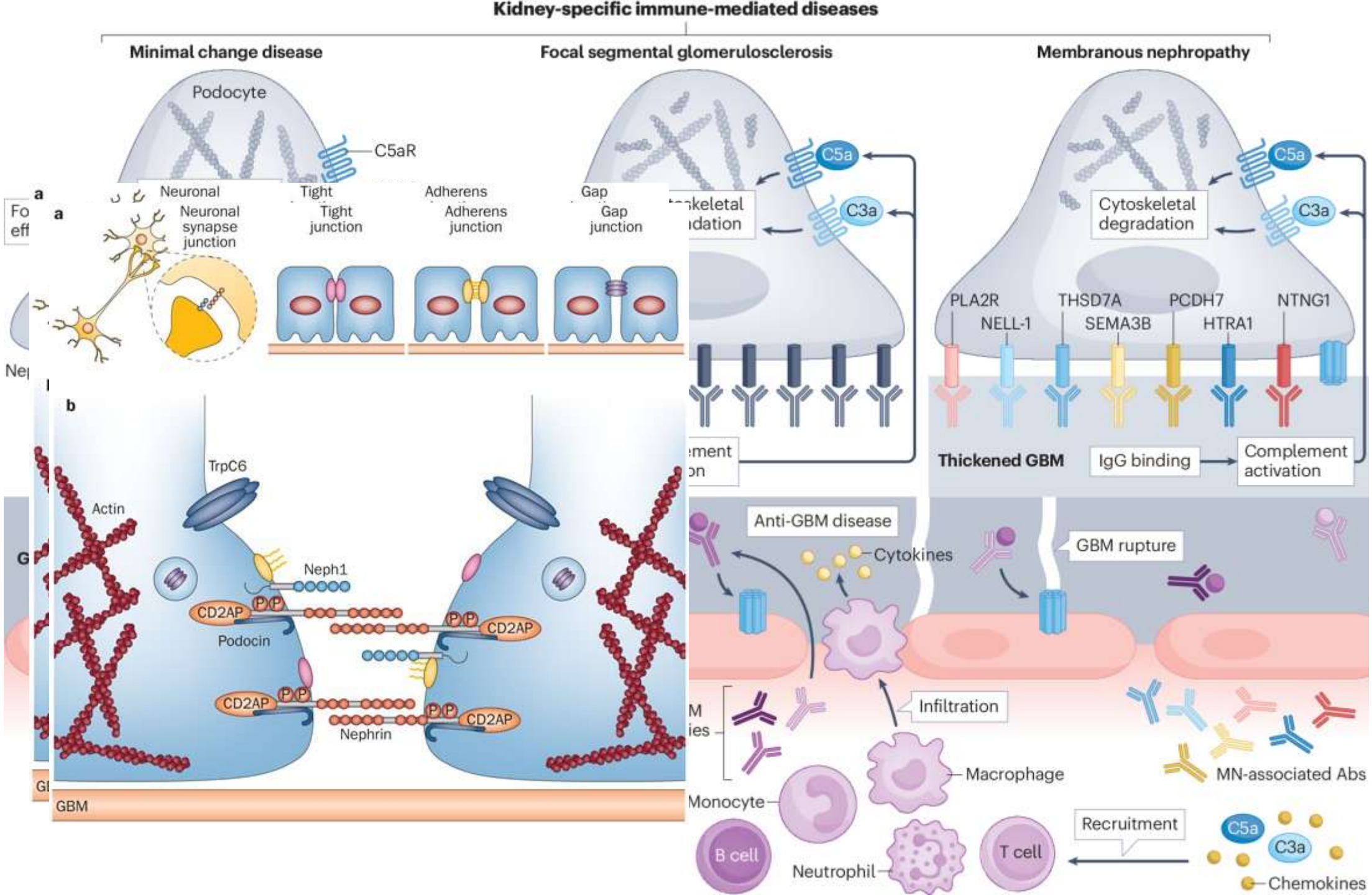


Table 1 | **Genes implicated in FSGS**

Function of the gene product	Gene
Slit diaphragm proteins	*NPHS1, NPHS2, CD2AP, CRB2, TRPC6, FAT1*
Actin binding	*PLCE1, ACTN4, MYO1E, MYH9, INF2, ANLN, AVIL*
Actin regulation	*ARHGDIA, ARHGAP24, KANK1, KANK2, KANK4, MAGI2, DLC1, ITSN1, ITSN2, DAAM2*
Nuclear transcription factors	*LMX1B, WT1, SMARCAL1, NXF5*
Nuclear pore complex proteins	*NUP93, NUP85, NUP107, NUP133, NUP160, NUP205, XPO5*
Mitochondrial proteins	*COQ2, COQ6, COQ8B (ADCK4), PDSS2, MTTL1*
KEOPS complex (tRNA modification)	*OSGEP, TP53RK, TPRKB, LAGE3*
Lysosomal proteins	*SCARB2*
Adhesion proteins	*ITGA3, ITGB4, LAMB2*
Glomerular basement membrane proteins	*COL4A3, COL4A4, COL4A5, COL4A6, LAMA5*
Other	*SGPL1, CUBN, PTPRO, WDR73, EMP2, DGKE, ALG1*

Sadowski et al, J Am Soc Nephrol 2015
Kopp Nat. Disease primers 2020


Kidney-specific immune-mediated diseases
Minimal change disease
Focal segmental glomerulosclerosis
Membranous nephropathy
Podocyte
C5aR
a
Neuronal
Neuronal synapse junction
Tight
Tight junction
Adherens
Adherens junction
Gap
Gap junction
b
TrpC6
Actin
Neph1
CD2AP
Podocin
Nephrin
GBM
C5a
C3a
Cytoskeletal degradation
PLA2R
NELL-1
THSD7A
SEMA3B
PCDH7
HTRA1
NTNG1
Thickened GBM
IgG binding
Complement activation
Anti-GBM disease
Cytokines
GBM rupture
Infiltration
Macrophage
MN-associated Abs
Monocyte
B cell
Neutrophil
T cell
Recruitment
Chemokines

The Role of Anti-B Cell Therapies

- Steroid-sparing aim
  - En entretien, 1st intetion (RIFIREIN)
  - CD/FR
    - SOC jusqu'à peu : tacrolimus/ MMF
    - Ritux
    - Obi
    - Dara
  - Steroid-R
    - Standard of Care
    - Data
    - Nephrin antin ?