

## *Parliamo insieme di sindrome nefrosica*



**SINePe**  
Società Italiana di  
Nefrologia Pediatrica

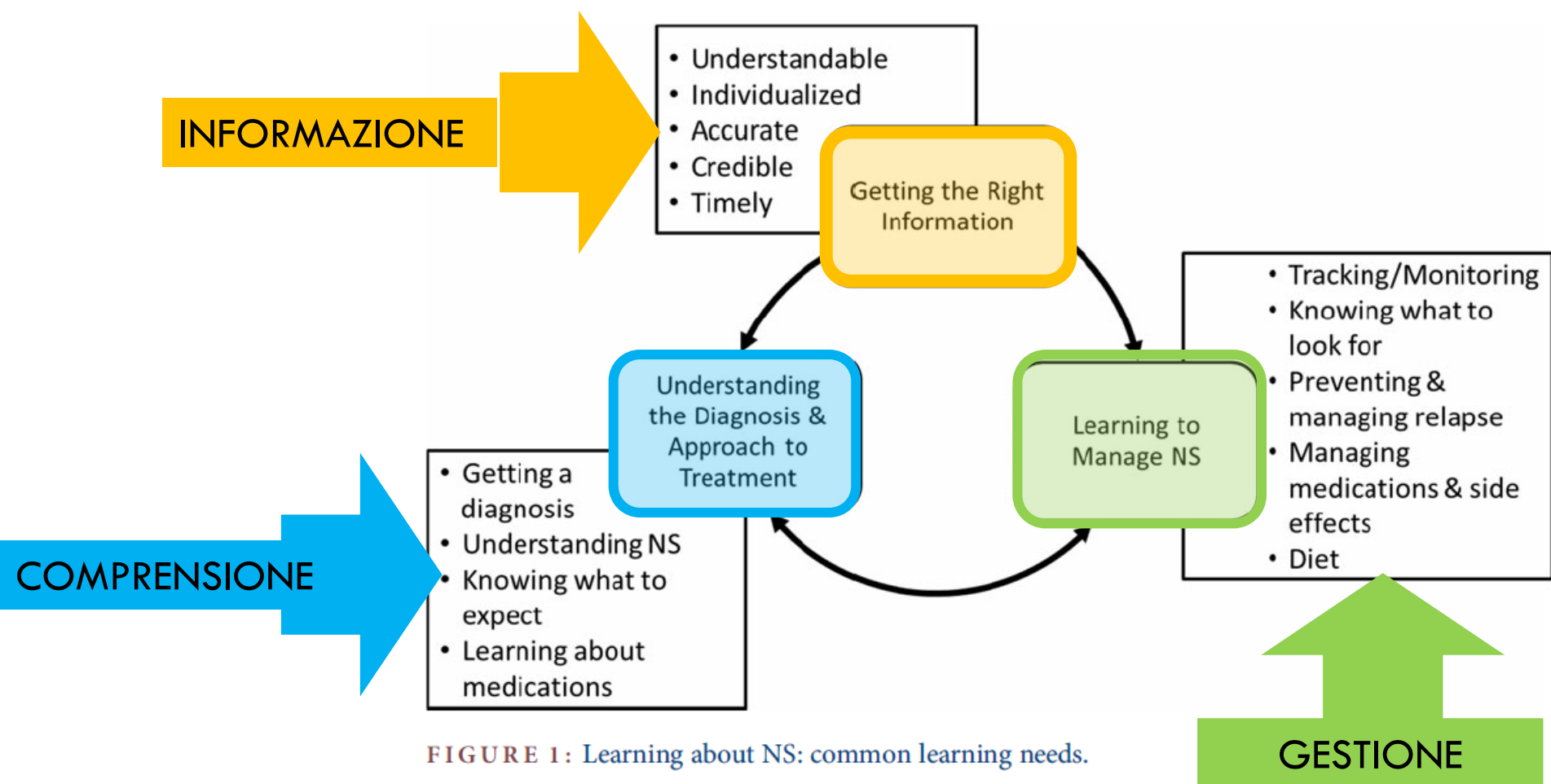


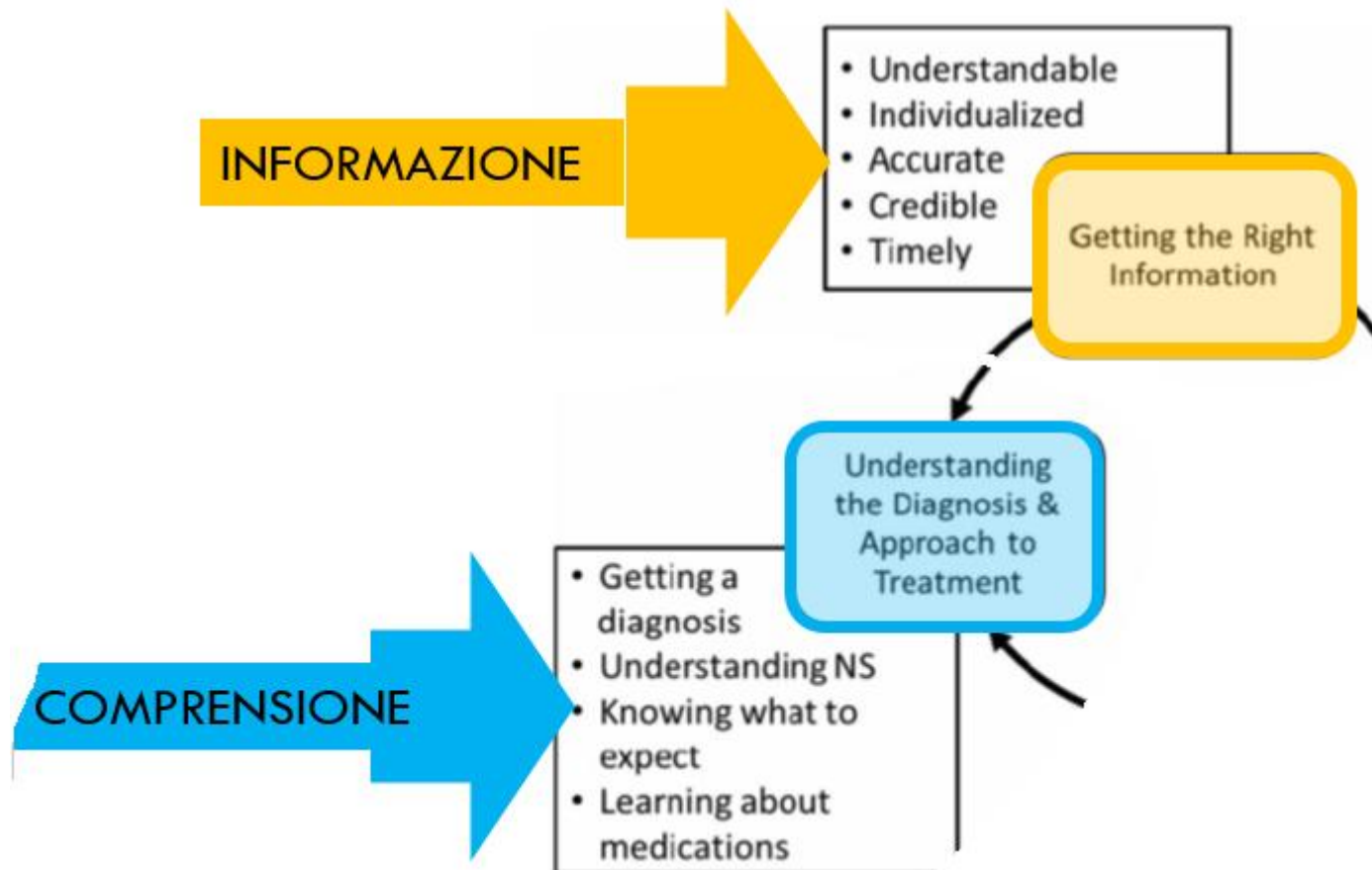
Licia Peruzzi

Ciro Corrado, Francesca Nuzzi, Fabrizio Pugliese, Mattia Parolin,  
Rosa Roperto, Giacomo Di Zazzo

*Original Article*

Learning to live with nephrotic syndrome: experiences of adult patients and parents of children with nephrotic syndrome



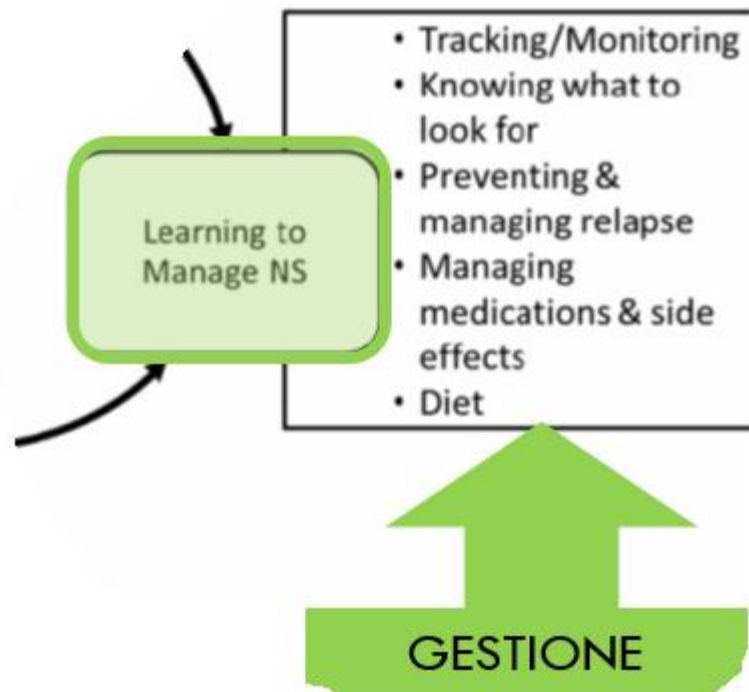


### **Gruppo A - Sala Cavour: Diagnosi e terapie**

#### **Moderatore Francesco Emma**

Dopo una breve presentazione introduttiva sull'argomento si aprirà la discussione sulle domande preparate dalle famiglie.

- Nuovi approcci terapeutici con cellule staminali e sparsentan. Giovanni Montini
- Quando fare la biopsia renale e cosa ci può dire: Luisa Murer
- Cosa ci può dire la genetica: Paola Romaganani
- Anticorpi Monoclonali: quando pensarci e che vantaggi e svantaggi: Gianmarco Ghiggeri



## **Gruppo B - Sala Campidoglio: Gestione del quotidiano** **Moderatore Carmine Pecoraro**

Presentazioni indirizzate a temi inerenti alimentazione, sport, vaccinazioni, scuola.  
 Dopo una breve presentazione introduttiva sull'argomento si aprirà la discussione sulle domande preparate dalle famiglie.

- Vaccinazioni: paure o pericoli. Carmine Pecoraro
- Lo sport: fa bene o fa male? Mario Giordano
- All'asilo e a scuola: si o no? Andrea Pasini
- Quali indicazioni sull'alimentazione: Alberto Edefonti

# SINDROME NEFROSICA: .....UNA MALATTIA CHE NON AVEVO MAI SENTITO



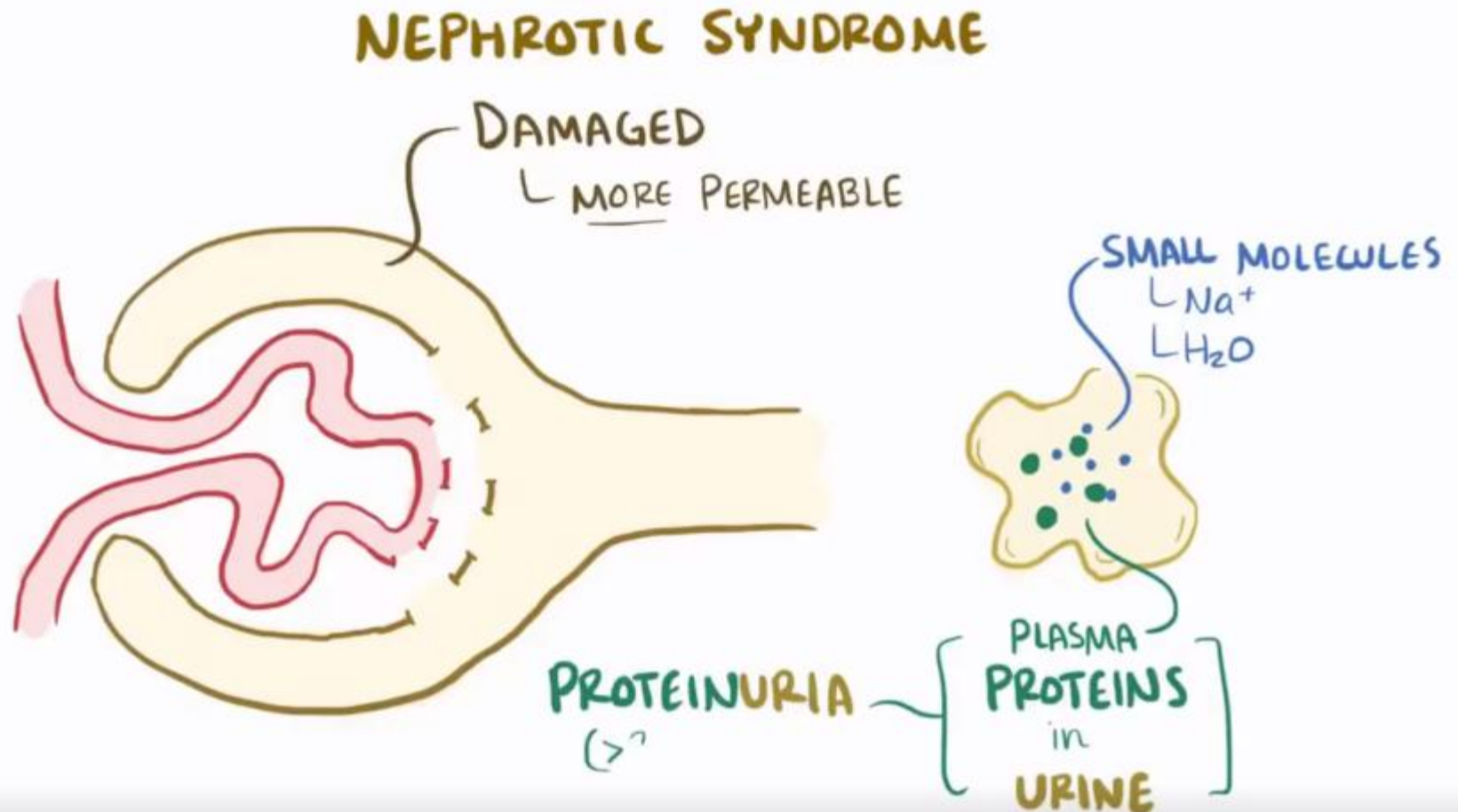
Pensavamo a una allergia.....  
E invece.....

- **Edema**
- **Proteinuria nefrosica**  
( $\geq 50$  mg/kg/die  $\rightarrow$   $40$  mg/m<sup>2</sup>/h - Upr/Ucr  $> 2$  g/mmol o 2 mg/mg)
- **Ipoprotidemia** ( $\leq 6$  g/dL)
- **Ipoalbuminemia** ( $\leq 2.5$  g/dL)

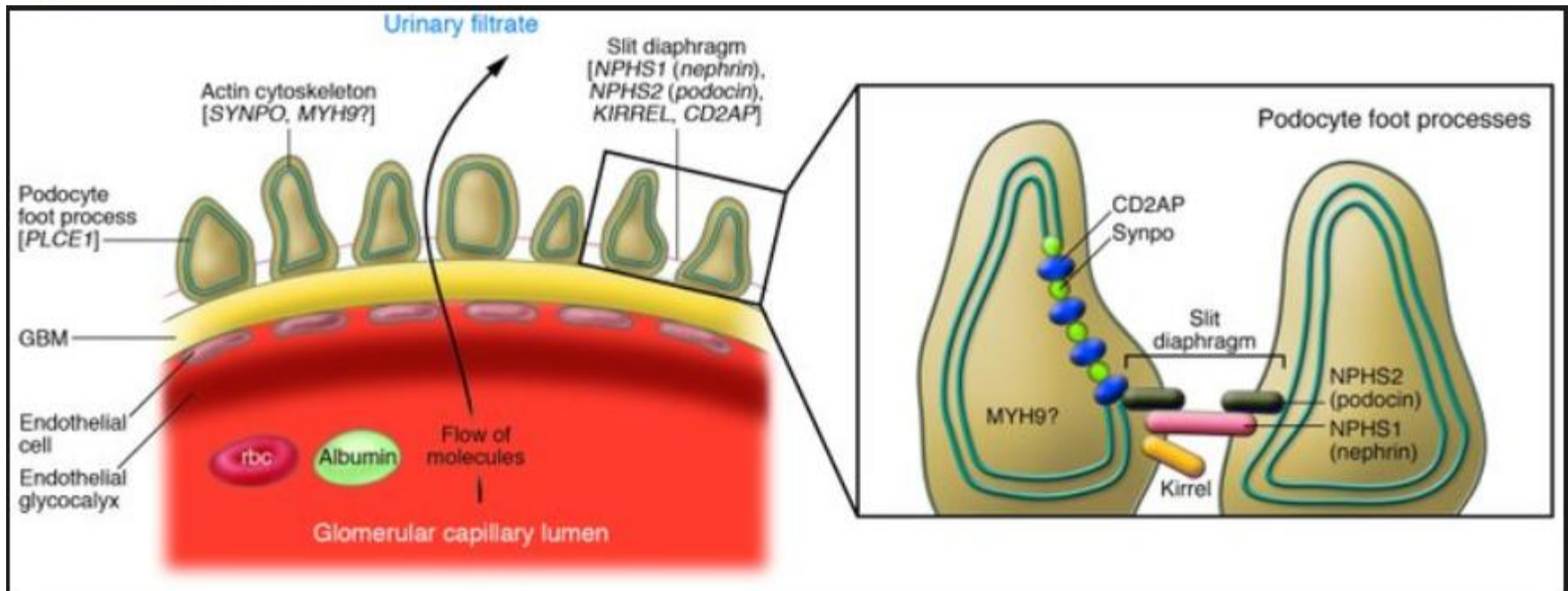
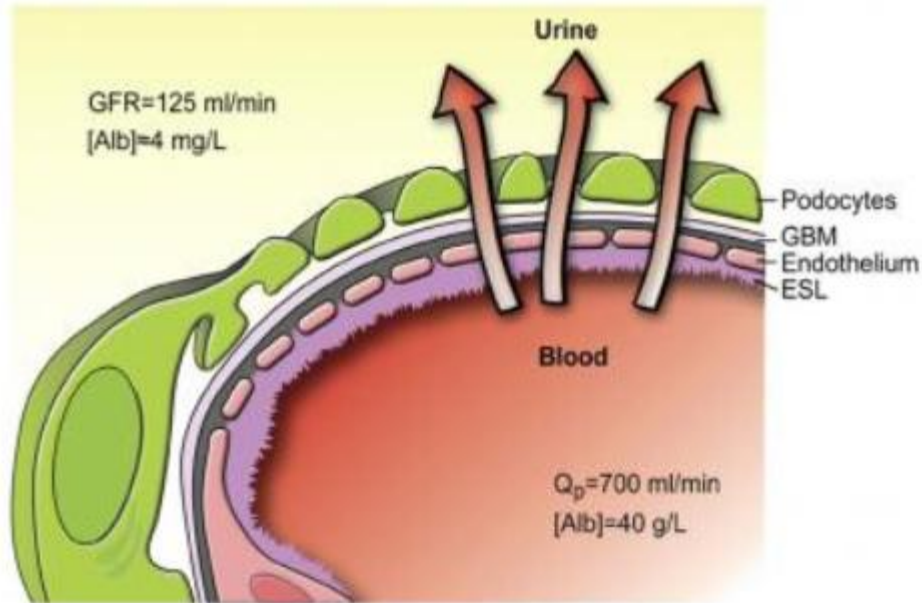


# UN MECCANISMO DIFFICILE DA CAPIRE

La bambina è gonfia..... ma cosa c'entrano le proteine nelle urine?

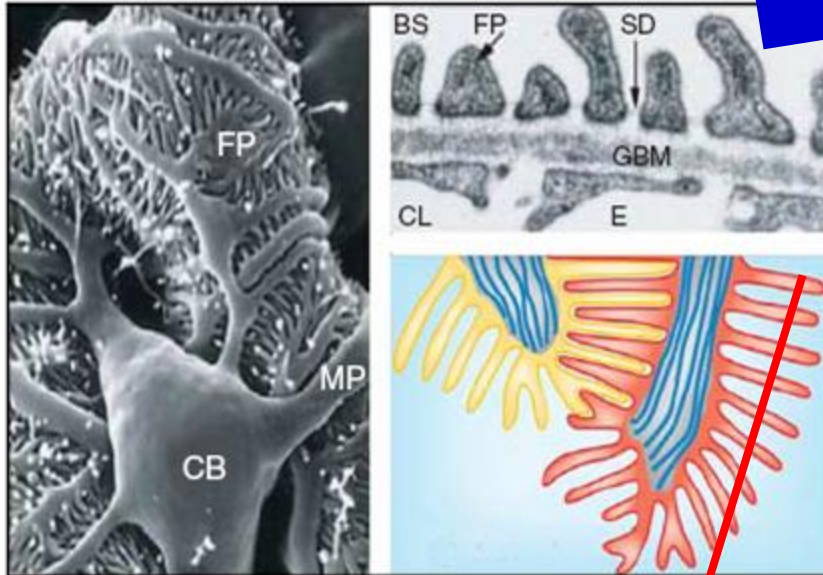


# IL GLOMERULO

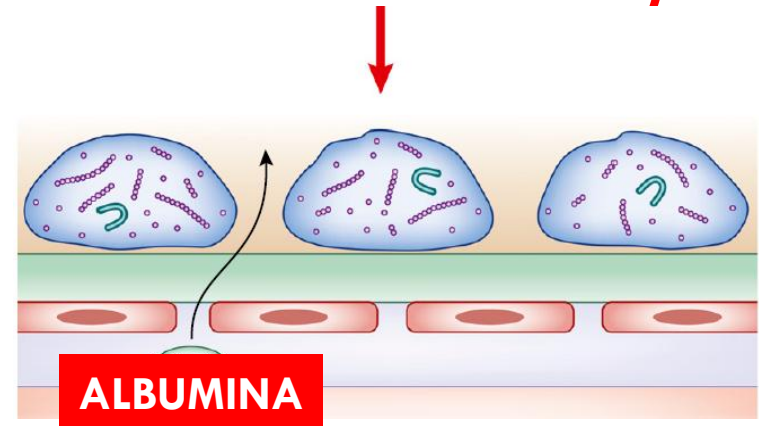
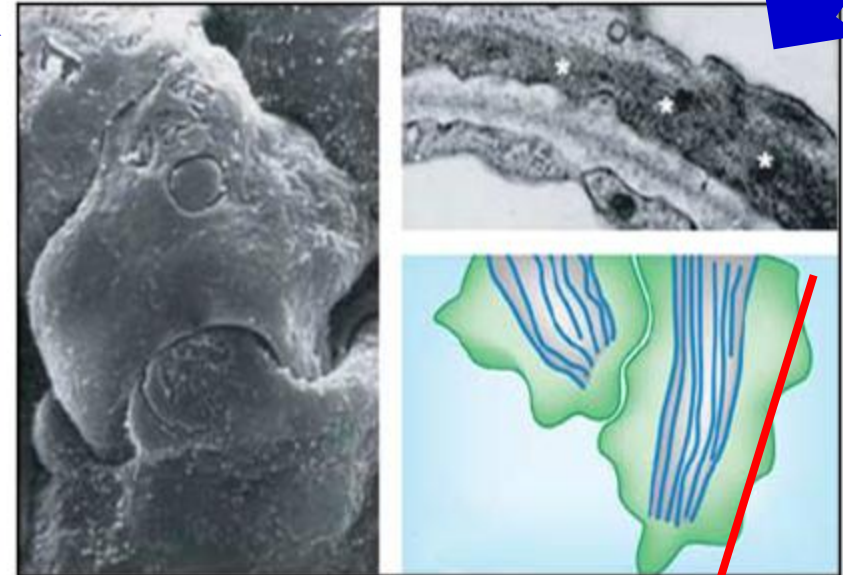


# LA BARRIERA DI FILTRAZIONE E' ALTERATA

Normale

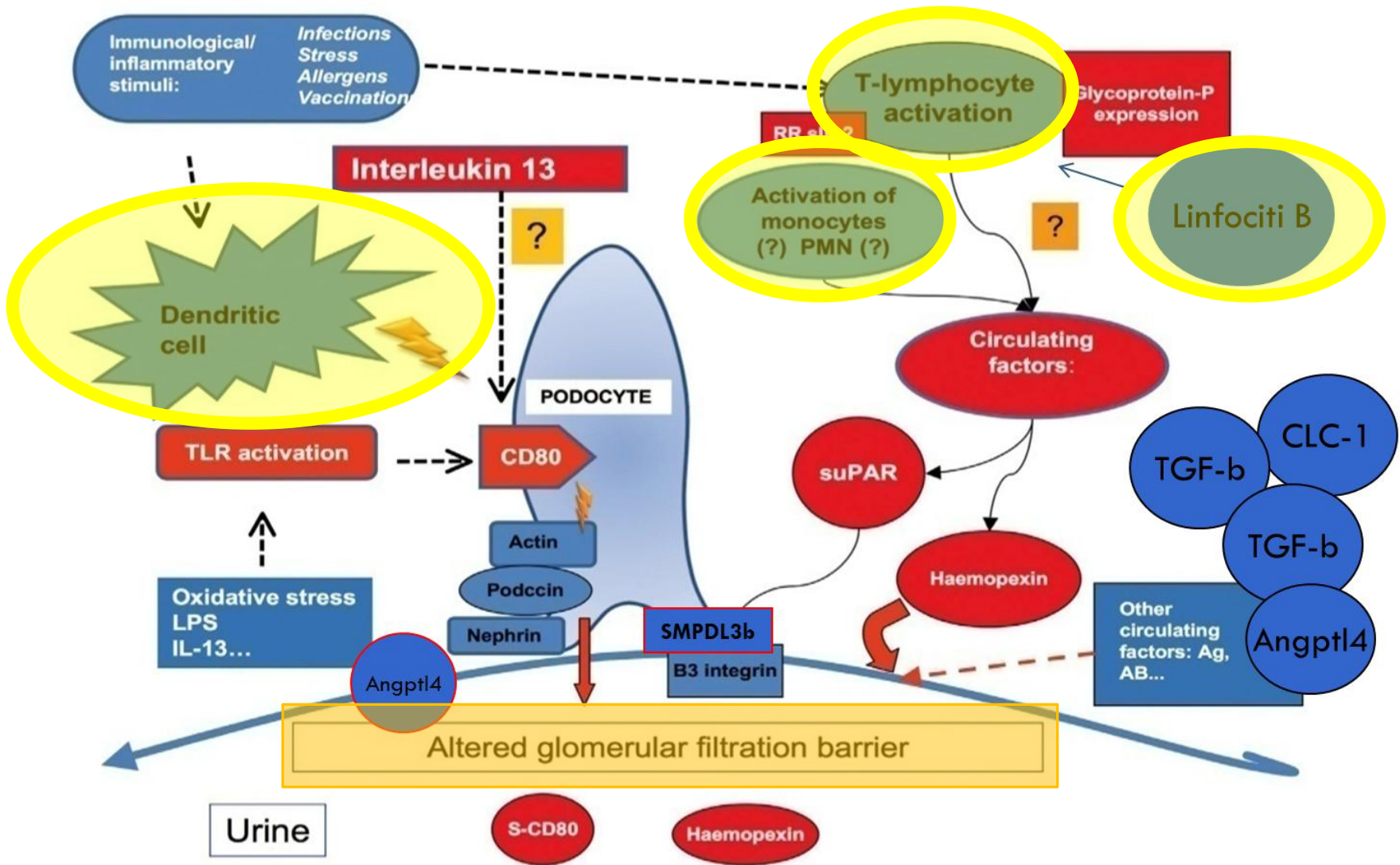


Sindrome nefrosica





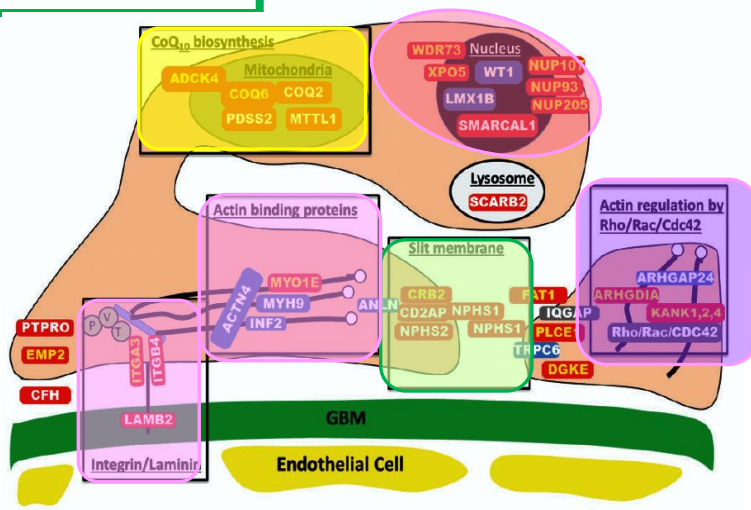
# LA BARRIERA DI FILTRAZIONE PUO' ESSERE IL BERSAGLIO DI UN ATTACCO «IMMUNOLOGICO»



**MA OGGI SAPPIAMO ANCHE CHE IL PROBLEMA PUO' ESSERE NELLE  
STRUTTURE CHE COMPONGONO LA BARRIERA DI FILTRAZIONE**

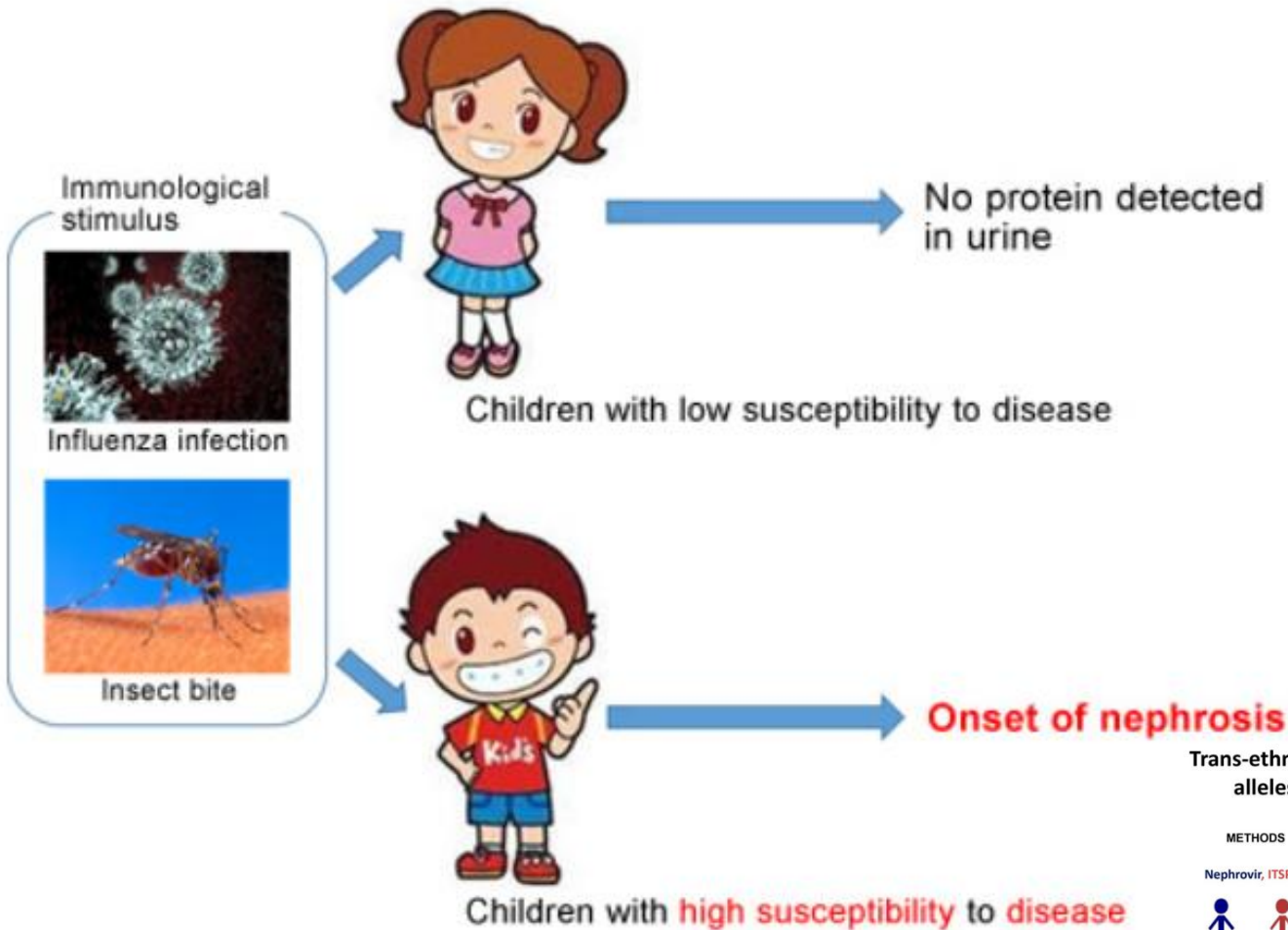
**Table 3. Genetic FSGS: Genetic variants with syndromes with Mendelian and mitochondrial inheritance**

Cell Matrix	Slit Diaphragm Complex	Cytoskeleton and Related	Mitochondria Function	DNA Repair, Transcription, Nuclear Transport	Cell Signaling	Lysosome	Cilia
<b>Nonsyndromic</b> <i>COL4A3</i>  <i>COL4A4</i> <i>COL4A5</i>	<i>NPHS</i> (nephrin)  <i>NPHS2</i> (podocin) <i>CD2AP</i> <i>PTPRO</i> (GLEPP1) <i>MYO1E</i>	<i>ACTN4</i>  <i>INF2</i> <i>AHRGP24</i> <i>AHRGDIA</i>	<i>INF2</i>	<i>WT 1</i> (Denys–Drash, Frasier) <i>NUP95</i> <i>NUP203</i> <i>XP05</i> (exportin 5) <i>NXF5</i> (nuclear export factor 5) <i>PAX2</i>	<i>PLCE1</i>  <i>TRPC6</i>		<i>TTC21B</i>
<b>Syndromic</b> <i>ITGB4</i> (epidermolysiss bullosa) <i>LAMB2</i> (Pearson)		<i>MYH9</i> (Esptein, Fechtner)	<i>INF2</i> (Charcot–Marie–Tooth) <i>MT-TL1</i> , <i>MT-TL2</i> tRNA leucine (MELAS) <sup>a</sup> <i>MT-TY</i> , tRNA tyrosine (MELAS) <sup>a</sup> <i>COQ2</i>  <i>COQ6</i> <i>PDSS2</i> (Leigh) <i>ADCK</i>	<i>WT1</i> (Denys–Drash, Frasier) <i>LMX1B</i> (Nail-patella)  <i>SMARCAL1</i> (Schimke immune-osseous dysplasia) <i>NXF5</i> <i>EYA1</i> (Branchio-oto-renal) <i>WDR73</i> (Galloway–Mowat, nephrocerebellar syndrome) <i>LMNA</i> (partial lipodystrophy)	<i>KANK4</i>	<i>SCARB2</i> (action myoclonus)	



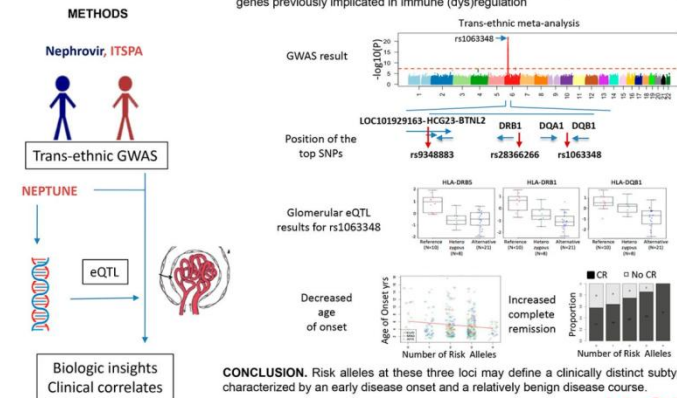
Clin J Am Soc Nephrol 12: 502–517, March, 2017

# O ENTRAMBI.....



## Trans-ethnic, genome-wide analysis reveals immune-related risk alleles and their phenotypic correlates in pediatric SSNS

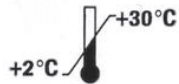
**RESULTS.** Three independent SNPs: 2 in *HLA-DR/DQ* region, 1 in *BTNL2* region; genes previously implicated in immune (dys)regulation



# PASSANO LE PROTEINE E COMPARE LA PROTEINURIA: LA SCHIUMA



IVD



2011-02

23054941

LOT

Specific Gravity Densidad Densidade 60 sec/seg.	1.000	1.005	1.010	1.015	1.020	1.025	1.030
pH 60 sec/seg.	5.0	6.0	6.5	7.0	8.0	9.0	
Leukocytes Leucocitos 60-120 sec/seg.	neg.	ca. 15	ca. 75	ca. 125	ca. 500	Leuko/ $\mu$ L	
Blood/Hemoglobin/ Sang(re)(ue)/Hemoglobina 60 sec/seg.	neg.	ca. 5-10	ca. 10	ca. 25	ca. 25	ca. 50	ca. 50
Nitrite/Nitrito/Nitritos 60 sec/seg.	neg.	+	++				ca. 250 Ery/ $\mu$ L
Ketones/ C.Cetónicos 60 sec/seg.	neg.	5 (0.5)	15 (1.5)	50 (5)	150 (15)	mg/dL (mmol/L)	
Bilirubin/Bilirrubina/ 60 sec/seg.	neg.	+	++	+++			
Urobilinogen(o)/ Urobilinogênio 60 sec/seg.	normal	1 (17)	4 (70)	8 (140)	12 (200)	mg/dL ( $\mu$ mol/L)	
Protein/Proteínas/ Proteínas 60 sec/seg.	neg.	15 (0.15)	30 (0.3)	100 (1)	300 (3)	1000 (10)	mg/dL (g/L)
Glucose/Glucosa/ Glicose 60 sec/seg.	normal	100 (5.5)	300 (17)	1000 (55)			mg/dL (mmol/L)

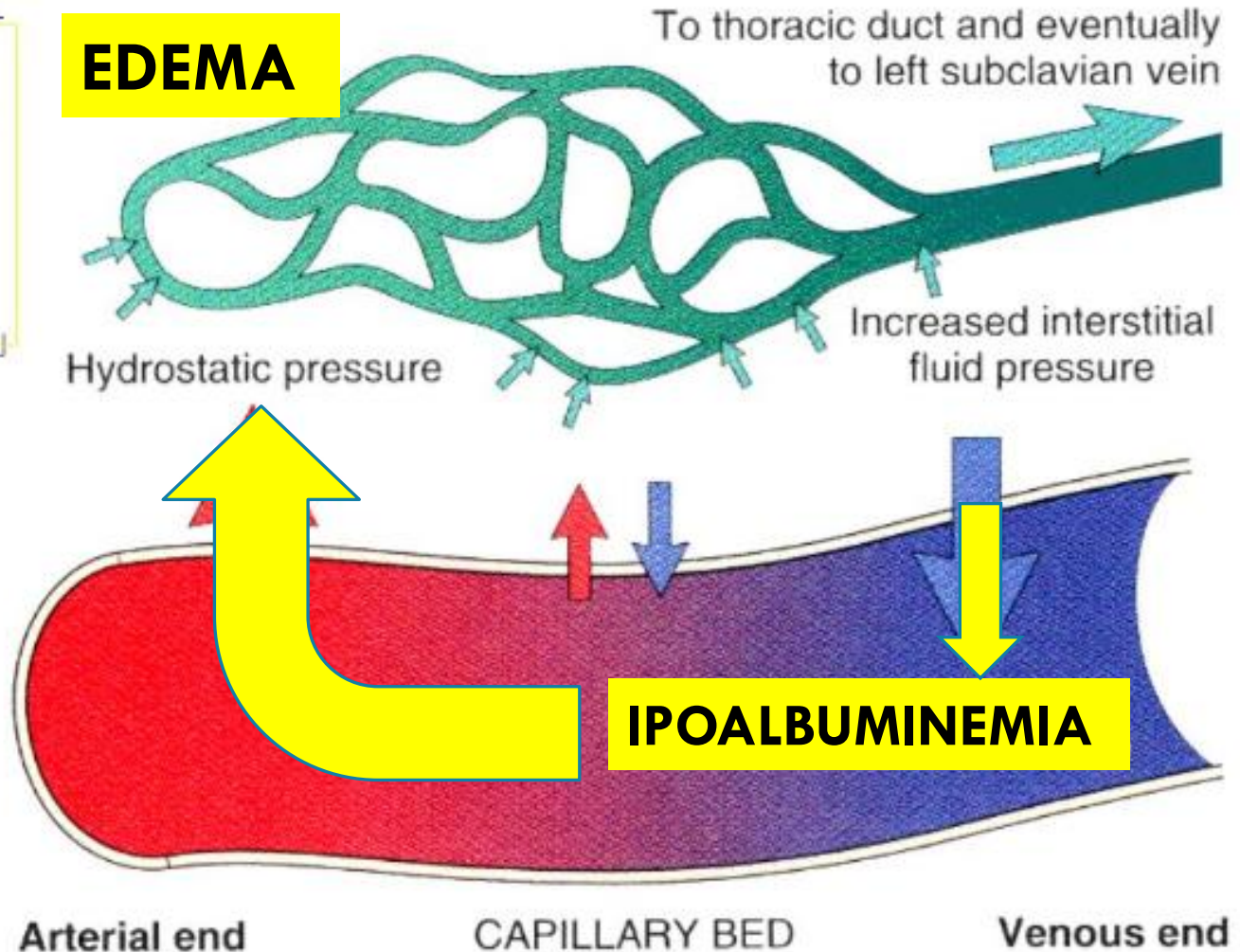




# MA PERCHE' GONFIA?

Viene mantenuta dagli effetti contrapposti tra la pressione idrostatica vascolare e la pressione colloidale osmotica del plasma

**EDEMA**



# SINDROME NEFROSICA NEL BAMBINO

**CONGENITA:** esordio entro i primi 3 mesi

**INFANTILE:** esordio tra i 4 e i 12 mesi

**DEL BAMBINO:** esordio oltre i 12 mesi

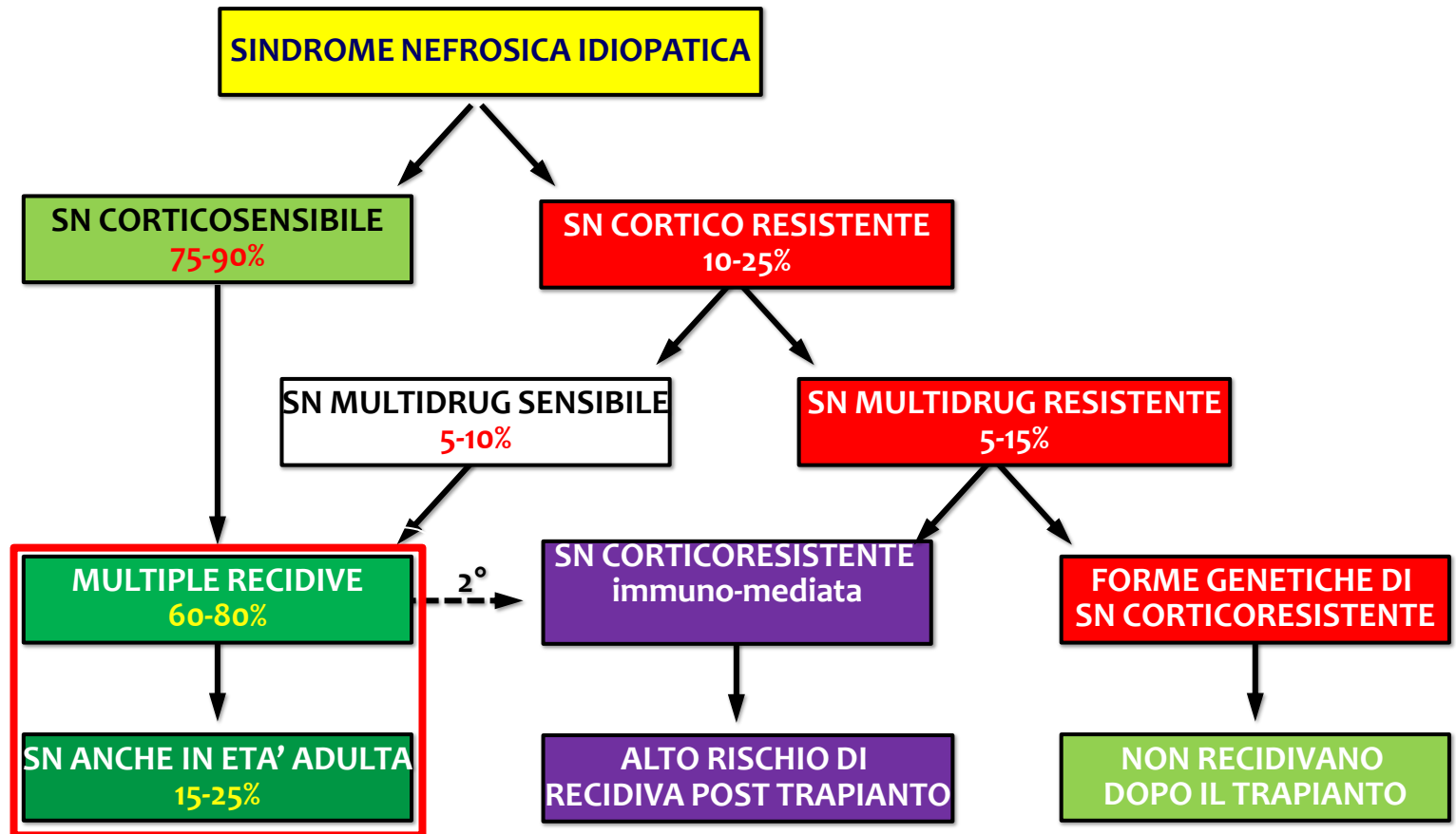
>90% STEROIDO RESISTENTI

**LA PROVA PRINCIPALE: LA  
RISPOSTA AL CORTISONE**

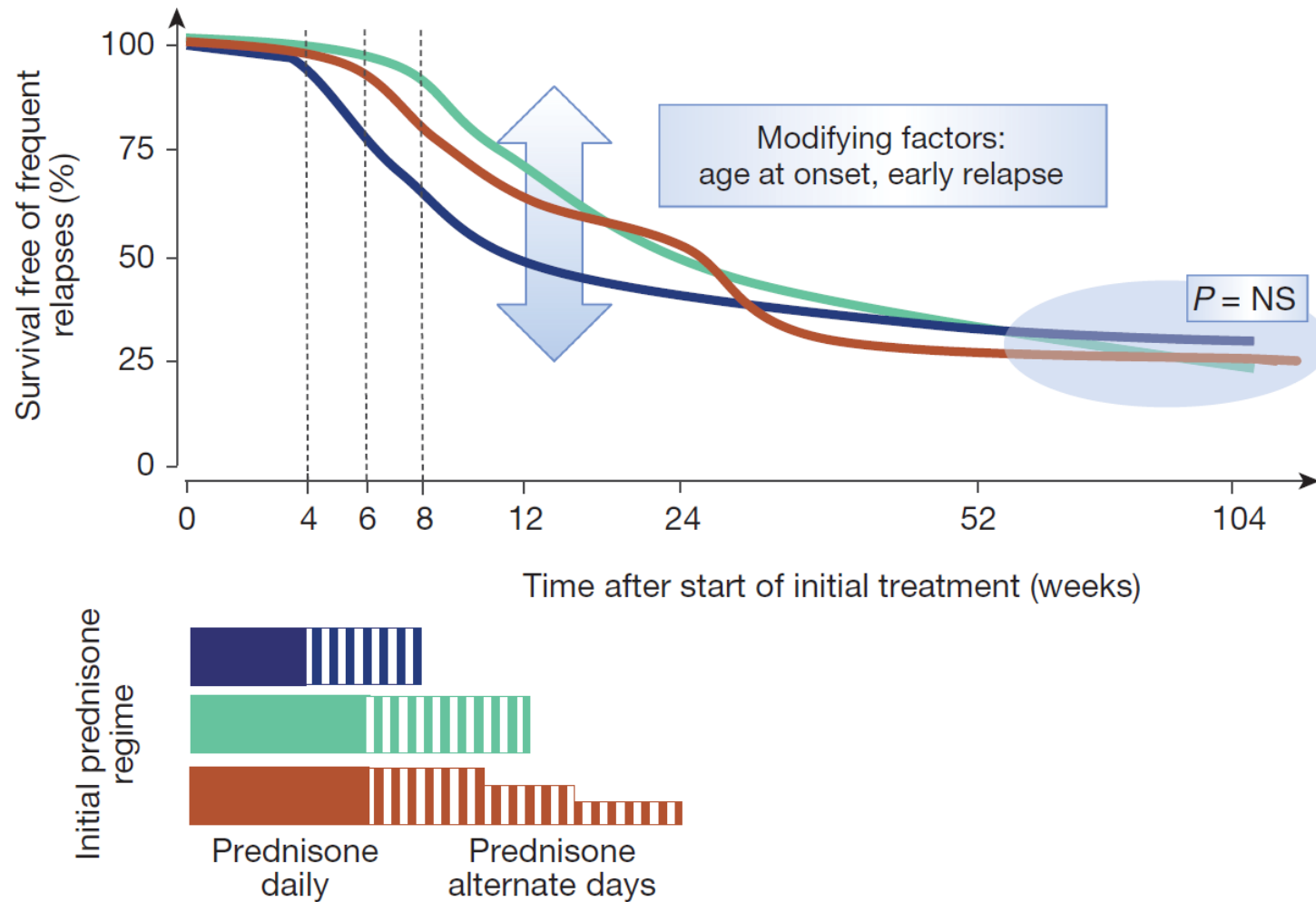
80% STEROIDO SENSIBILI

20% STEROIDO RESISTENTI

# SINDROME NEFROSICA NEL BAMBINO



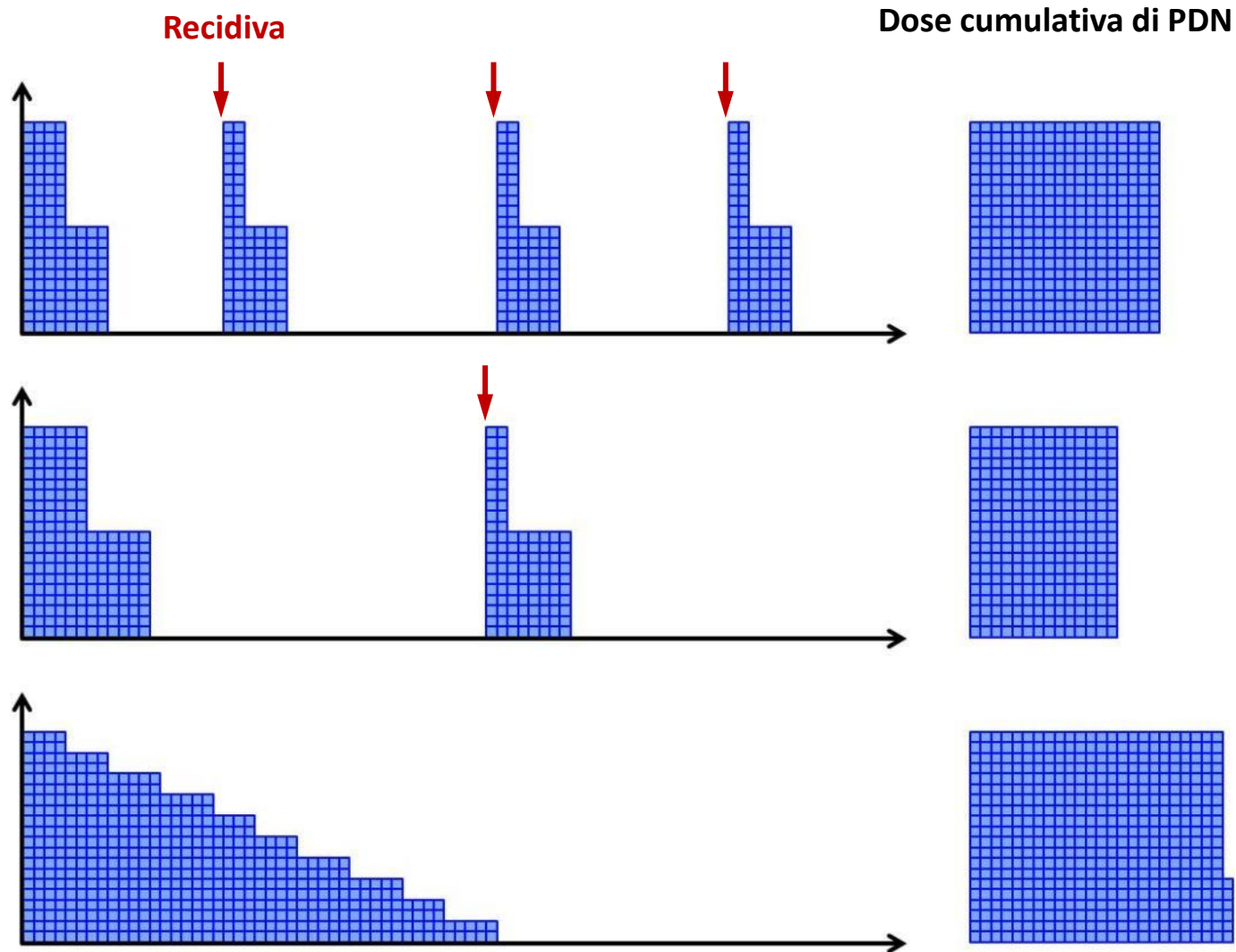
# IL CORTISONE: MOLTI SCHEMI...RISULTATI ANALOGHI



**Figure 1 | Lack of effect of extending initial prednisone treatment on long-term freedom from frequent relapses. NS, not significant.**



# IL CORTISONE: MOLTI SCHEMI.... QUELLO CHE CONTA E' LA DOSE CUMULATIVA



REVIEW

Open Access



# The Italian Society for Pediatric Nephrology (SINePe) consensus document on the management of nephrotic syndrome in children: Part I - Diagnosis and treatment of the first episode and the first relapse

Andrea Pasini<sup>1\*</sup>, Elisa Benetti<sup>2</sup>, Giovanni Conti<sup>3</sup>, Luciana Ghio<sup>4</sup>, Marta Lepore<sup>4</sup>, Laura Massella<sup>5</sup>, Daniela Molino<sup>6</sup>, Licia Peruzzi<sup>7</sup>, Francesco Emma<sup>5</sup>, Carmelo Fede<sup>3</sup>, Antonella Trivelli<sup>8</sup>, Silvio Maringhini<sup>9</sup>, Marco Materassi<sup>10</sup>, Giovanni Messina<sup>11</sup>, Giovanni Montini<sup>4</sup>, Luisa Murel<sup>2</sup>, Carmine Pecoraro<sup>6</sup> and Marco Pennesi<sup>12</sup>

**Table 5** Steroid protocol

Prednisone (PDN)	Dosage	Duration
Treatment of the first episode		
60 mg/m <sup>2</sup> (maximum 60 mg)	in single or 2 divided doses	6 weeks
40 mg/m <sup>2</sup> (maximum 40 mg)	on alternate days	6 weeks
Treatment of the first relapse		
60 mg/m <sup>2</sup> (maximum 60 mg)	in a single or 2 divided doses	Until urine protein is negative for 5 days
40 mg/m <sup>2</sup> (maximum 40 mg)	on alternate days	4 weeks

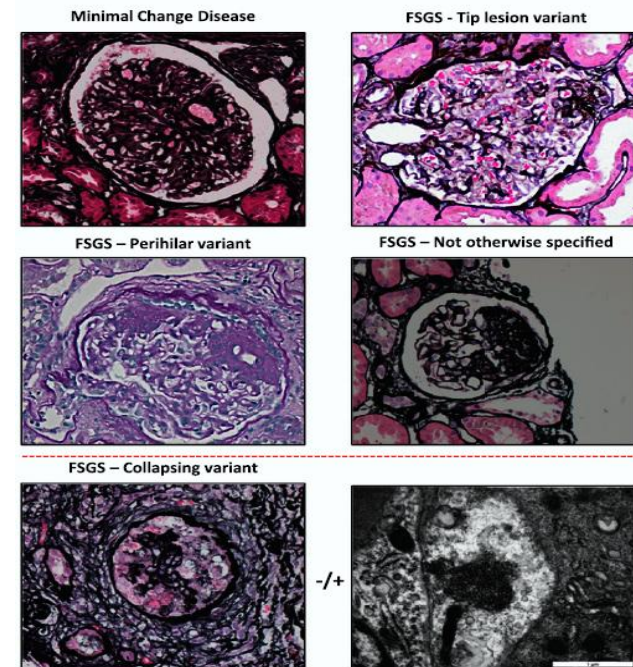
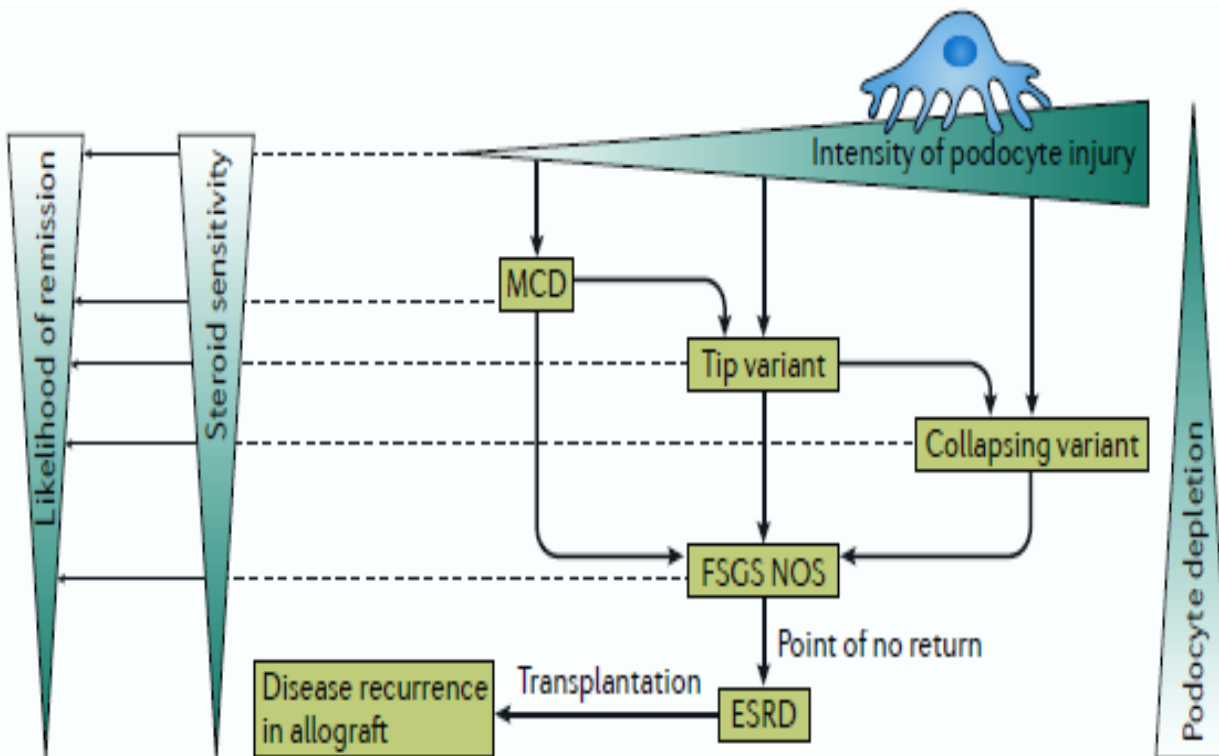
# L'OBIETTIVO: MANTENERE LA REMISSIONE E RISPARMIARE IL CORTISONE

FARMACO	Efficacia	Indicazione	Commenti
Ciclosporina e Tacrolimus	+++++	SDNS	Tossicità renale / ipertensione
Micofenolato mofetile	++++	FRNS/SDNS	Spesso necessarie alte dosi (> 600 mg/m <sup>2</sup> )
Levamisolo	+++	FRNS	Difficile da reperire
Ciclofosfamide	+++	?	Tossicità; talora nn protettivo
Rituximab	Buona	?	Può compromettere la memoria immunologica



*Altri farmaci innovativi*

# LA BIOPSIA RENALE





# LA QUALITA' DELLA VITA DEI BAMBINI E DELLE FAMIGLIE

23/4/2019

The effect of blended training on the quality of life of children with nephrotic syndrome



J Family Med Prim Care. 2018 Sep-Oct; 7(5): 921–926.

PMCID: PMC6259550

doi: 10.4103/jfmpc.jfmpc\_176\_18; 10.4103/jfmpc.jfmpc\_176\_18

PMID: [30598934](#)

## The effect of blended training on the quality of life of children with nephrotic syndrome

[Sedighe Khanjari](#),<sup>1</sup> [Sedighe Jahanian](#),<sup>2</sup> and [Hamid Haghani](#)<sup>3</sup>

## PROGRAMMI EDUCAZIONALI STRUTTURATI PER BAMBINI E FAMIGLIE

USO DI MULTIPLI STRUMENTI DIDATTICI PER MIGLIORARE LA  
FAMILIARIZZAZIONE CON  
**LA MALATTIA**  
**LA GESTIONE DELLA MALATTIA**



## MIGLIORAMENTO DELLE PERFORMANCE


- **FISICHE**
- **EMOTIVE**
- **SOCIALI**
- **SCOLASTICHE**
- **QUALITA' DI VITA GENERALE**

# UN IMPATTO IMPORTANTE PER LE FAMIGLIE



## ORIGINAL ARTICLE

### Multidimensional impact on families of children with steroid-sensitive nephrotic syndrome

Gurdeep S Dhooria <sup>1</sup>, Harmeet P Singh,<sup>2</sup> Deepak Bhat,<sup>1</sup> Harmesh S Bains,<sup>1</sup> Ravinder K Soni<sup>3</sup> and Mohit Kumar<sup>1</sup>

Departments of <sup>1</sup>Pediatrics, <sup>2</sup>Medicine, and <sup>3</sup>Statistics, Preventive Medicine, Dayanand Medical College and Hospital, Ludhiana, India

Journal of Paediatrics and Child Health 53 (2017) 354–357

**Table 2** Comparison of family impact score between nephrotic cases and controls

Family impact score	Nephrotic syndrome (n = 50); mean (SD)	Healthy control (n = 50); mean (SD)	Difference of means (95% CI)	P-value
Physical functioning	63.33 (22.66)	100 (0)	–36.66 (–43.02 to –30.30)	0.0001*
Emotional functioning	46.80 (24.61)	100 (0)	–53.20 (–60.10 to –46.29)	0.0001*
Social functioning	71.37 (26.09)	100 (0)	–28.63 (–35.95 to –21.30)	0.0001*
Cognitive functioning	58.50 (23.57)	100 (0)	–41.50 (–48.11 to –34.89)	0.0001*
Communication	72.67 (23.93)	100 (0)	–27.33 (–34.05 to –20.62)	0.0001*
Worry	46.20 (20.66)	100 (0)	–53.80 (–59.60 to –48.00)	0.0001*
Family daily activities	59.00 (30.16)	100 (0)	–41.00 (–49.46 to –32.53)	0.0001*
Family relationships	75.40 (18.87)	100 (0)	–24.60 (–29.90 to –19.31)	0.0001*
Total	60.76 (15.66)	100 (0)	–38.34 (–42.76 to –33.92)	0.0001*

\* $P < 0.05$  is significant. CI, confidence interval; SD, standard deviation.

### «PREOCCUPAZIONE»

- EFFETTI COLLATERALI DEI FARMACI A DISTANZA
- OUTCOME A DISTANZA

# LA QUALITA' DELLA VITA DEI BAMBINI

Saudi J Kidney Dis Transpl 2017;28(3):593-598  
© 2017 Saudi Center for Organ Transplantation

Saudi Journal  
of Kidney Diseases  
and Transplantation

## Renal Data from Asia–Africa

### Assessment of Quality of Life in Children with Nephrotic Syndrome at a Teaching Hospital in South India

Sonia Agrawal<sup>1</sup>, Sriram Krishnamurthy<sup>1</sup>, Bijaya Nanda Naik<sup>2</sup>

Table 3. PedsQL™ 4.0 Generic Core Scale Quality of Life scores in cases and controls.

Domains	Cases		Controls		P
	n	Median (interquartile range)	n	Median (interquartile range)	
Physical	50	71.87 (58.59–75)	50	62.5 (58.59–68.75)	0.004*
Emotional	50	65 (55–65)	50	60 (55–65)	0.029*
Social	50	70 (63.75–75)	50	65 (60–70)	0.010*
School	40	62.5 (56.25–65)	49	65 (52.5–65)	0.745
Total	40	65 (59.02–68.75)	49	62.19 (58.05–65.78)	0.012*

\*P value significant (<0.05).

DIVERSE DAI  
COETANEI  
SENZA SN

Table 4. Comparison of PedsQL 4.0 Generic Core Scale quality of life in different clinical phenotypes of nephrotic syndrome.

Parameters	Type of nephrotic syndrome			P
	Steroid dependent and frequently relapsing (n=24)	Steroid resistant (n=10)	Infrequently relapsing and first episode (n=16)	
Physical scores*	68.75 (56.25–75)	71.88 (65.63–75)	71.88 (65.63–75)	0.715
Emotional scores*	62.5 (51.25–70)	60 (60–71.25)	62.5 (56.25–65)	0.290
Social scores*	70 (65–75)	72.5 (63.75–75)	70 (60–75)	0.662
School scores*	60 (58.75–65)	65 (60–65)	60 (55–65)	0.529

\*Values depicted as median (interquartile range).

NON MOLTO  
DIVERSI NELLE  
FORME DIVERSE  
DI SN

# L'IMPATTO DELLA DURATA DELLA MALATTIA SULLA QUALITA' DELLA VITA

Pediatr Nephrol (2015) 30:1467–1476  
DOI 10.1007/s00467-015-3074-x

ORIGINAL ARTICLE

## The impact of disease duration on quality of life in children with nephrotic syndrome: a Midwest Pediatric Nephrology Consortium study

Table 6 Correlations between PROMIS and PedsQL measures

PROMIS domains	PedsQL™ 4.0 Generic Core Scales domains				
	Physical functioning	Emotional functioning	Social functioning	School functioning	Overall HRQOL
Mobility	0.57	0.29	0.35	0.41	0.52
Depression	−0.48	−0.69	−0.43	−0.49	−0.65
Anxiety	−0.49	−0.76	−0.36	−0.35	−0.62
Fatigue	−0.68	−0.62	−0.43	−0.58	−0.75
Peer relationships	0.20	0.27	0.41	0.31	0.34
Pain interference	−0.67	−0.64	−0.41	−0.62	−0.75



# ANCHE LO STRESS PUO' CAUSARE RECIDIVA?

ARTICLE IN PRESS

Journal of Psychosomatic Research xxx (xxxx) xxx–xxx



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Contents lists available at ScienceDirect

Journal of Psychosomatic Research

journal homepage: [www.elsevier.com/locate/jpsychores](http://www.elsevier.com/locate/jpsychores)



A longitudinal study on the effects of psychological stress on proteinuria in childhood steroid-sensitive nephrotic syndrome

Lianne Bakkum<sup>a,b,\*</sup>, Agnes Maresa Willemsen<sup>b</sup>, Lydia Zoetebier<sup>d</sup>, Antonia H. Bouts<sup>c</sup>

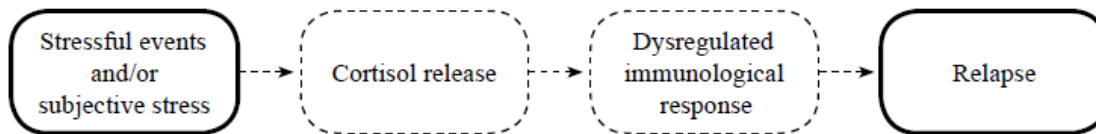


Table 4

Fixed effects results of the multilevel models for the effect of perceived stress on proteinuria.

Proteinuria	$\beta$	SE	95% CI
Same day	0.19**	0.04	[0.11, 0.27]
1 day later	0.19**	0.04	[0.10, 0.27]
2 days later	0.13*	0.04	[0.04, 0.21]
3 days later	0.14**	0.04	[0.05, 0.22]
4 days later	0.13*	0.04	[0.04, 0.21]
5 days later	0.12*	0.04	[0.03, 0.20]
6 days later	0.05	0.04	[−0.03, 0.14]
7 days later	0.05	0.04	[−0.04, 0.13]

EVENTO STRESSANTE ASSOCIATO A RECIDIVA NEI GIORNI SUCCESSIVI

**Conclusions:** Our findings suggest that psychological stress may trigger proteinuria in children with SSNS. Future research in larger samples is needed to support our findings.

Fondazione La Nuova Speranza onlus  
 Asnit onlus

**A.S.N.IT Onlus & LA NUOVA SPERANZA Onlus**

**organizzano:**

**IX INCONTRO NAZIONALE  
 MEDICI FAMIGLIE**

**DUE GIORNI PER PARLARE INSIEME DI  
 SINDROME NEFROSICA**

**CENTRO CONGRESSI CAVOUR  
 ROMA**

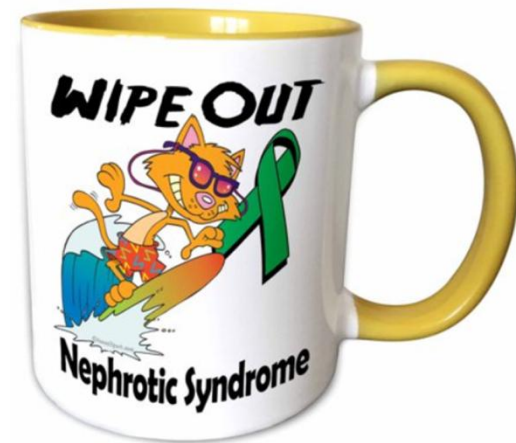
**27 E 28 APRILE 2019**

Con il patrocinio di:

SOCIETÀ ITALIANA  
 DI NEFROLOGIA  
 PEDIATRICA

REGIONE  
 LAZIO

ROMA



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 Kidney International  
 Saving Kidneys - Saving Lives

**SIGN UP  
 WITH NEPHCURE**

LIVING WITH KIDNEY DISEASE PATIENT CONNECTIONS EVENTS GET INVOLVED RESEARCH RECENT NEWS ABOUT US DONATE

**Understanding Nephrotic Syndrome and Glomerular Disease**

Nephrotic Syndrome 101 - NephCure Kidney Int...

Diseases that injure the glomeruli, the tiny filtering units within the kidney where blood is cleaned, are called Glomerular diseases.

Glomerular disease reduces the kidney's ability to remove a balance of poisons.

**Proteinuria and Hematuria**

**LIVING WITH KIDNEY DISEASE**

Just for Kids

Understanding Kidney Disease

Understanding Nephrotic Syndrome and Glomerular Disease

Nephrotic Syndrome

Focal Segmental Glomerulosclerosis (FSGS)

Minimal Change Disease (MCD)

Other Glomerular Diseases

Treatment Options

Diet and Nutrition

Managing Your Care

Educational Programs

End Stage Renal Disease

**THE Nephrotic Syndrome FOUNDATION**  
 SUPPORT EDUCATE FIGHT

Home For Patients Research and Updates Events Upcoming Events More

Working to support families, educate our community, and fight nephrotic syndrome.

*thank you*

In 2018 The Nephrotic Syndrome Foundation delivered 40 Backpacks of Hope, 14 hospital grade thermometers, hosted 25 patient families at Camp, offered 4 Finding Health sessions for