

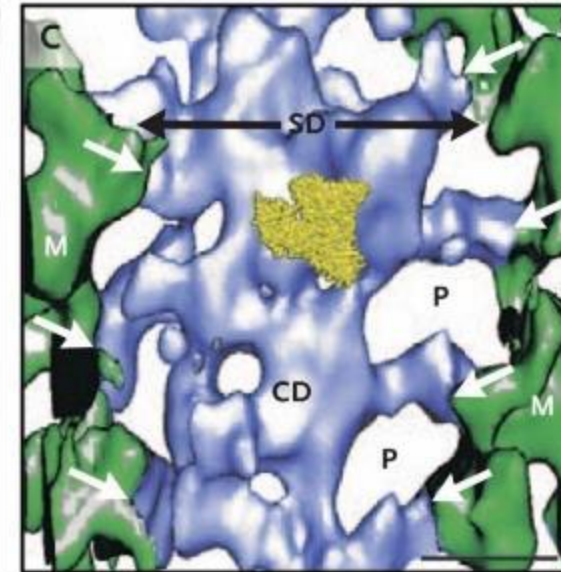
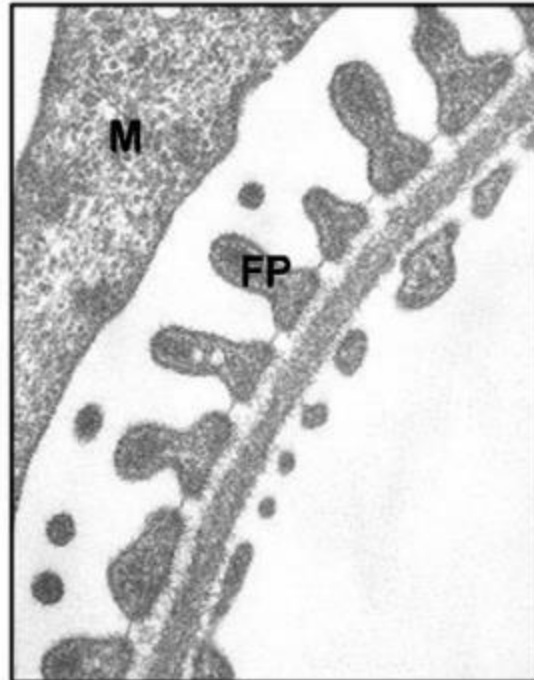
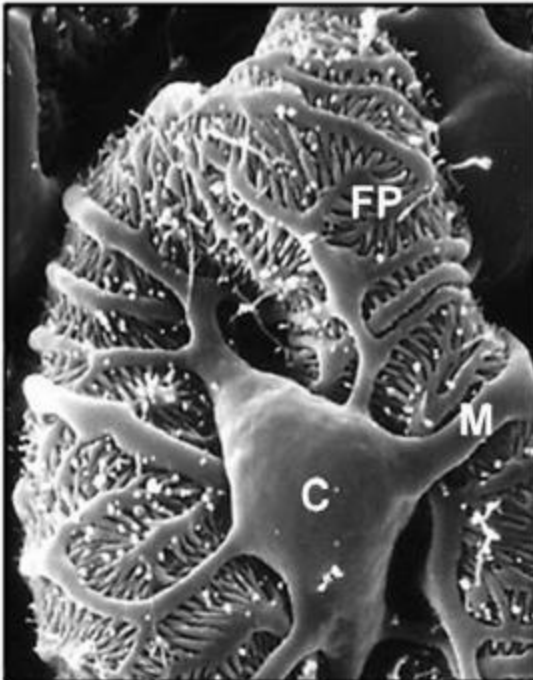
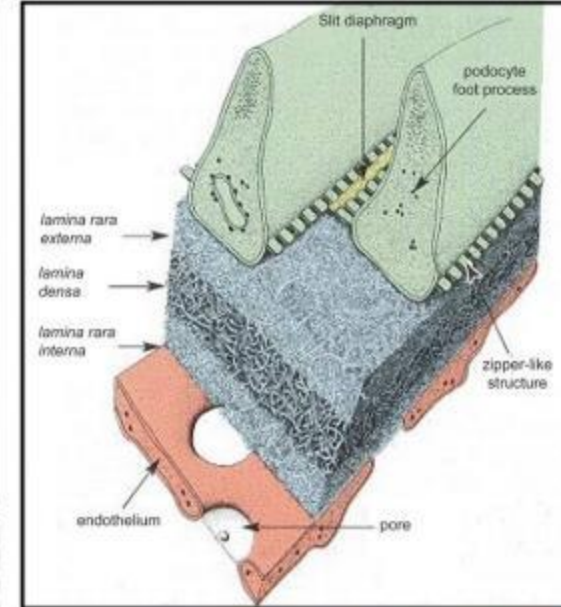
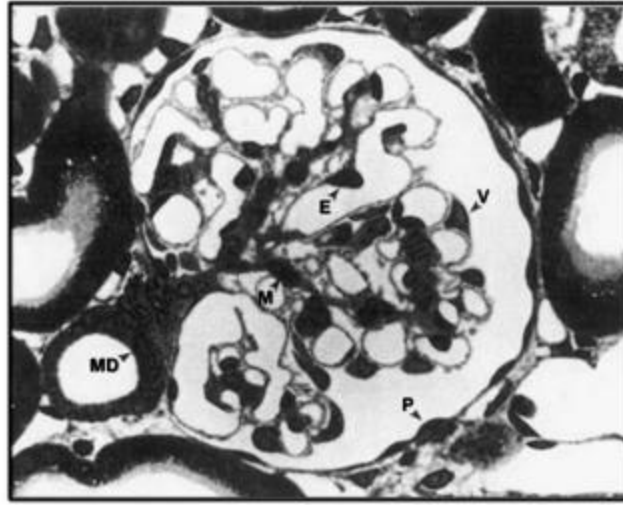
Sindrome nefrosica corticosensibile

Francesco Emma

*UOC di Nephrologia e Dialisi
Ospedale Pediatrico Bambino Gesù, Roma*

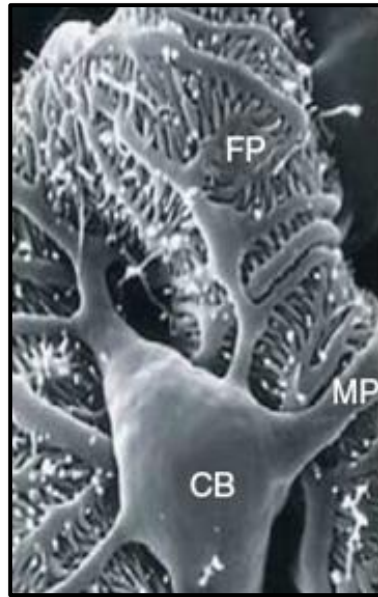
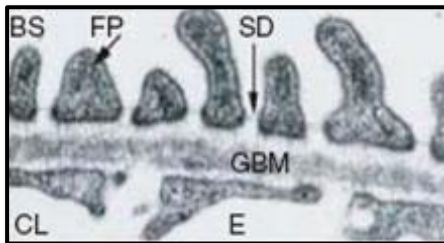
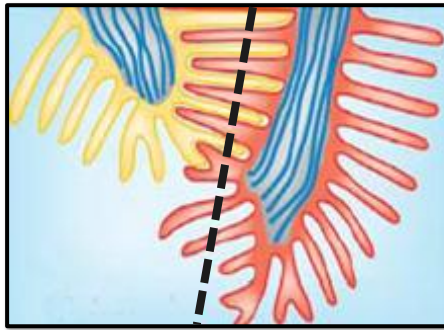
Roma 27 aprile 2019

Il glomerulo

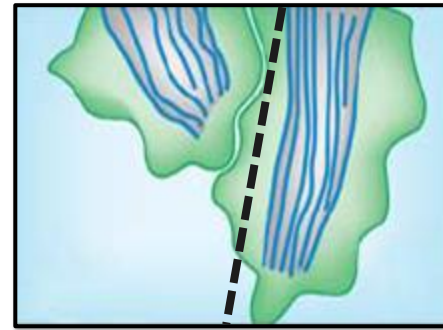


Fusione dei pedicelli

Normal



Nephrotic syndrome



Definizioni...

Steroid Sensitive Nephrotic Syndrome (SSNS)

Responsta al PDN (60mg/m²/24h) in 4-6 settimane

Steroid Resistant Nephrotic Syndrome (SRNS)

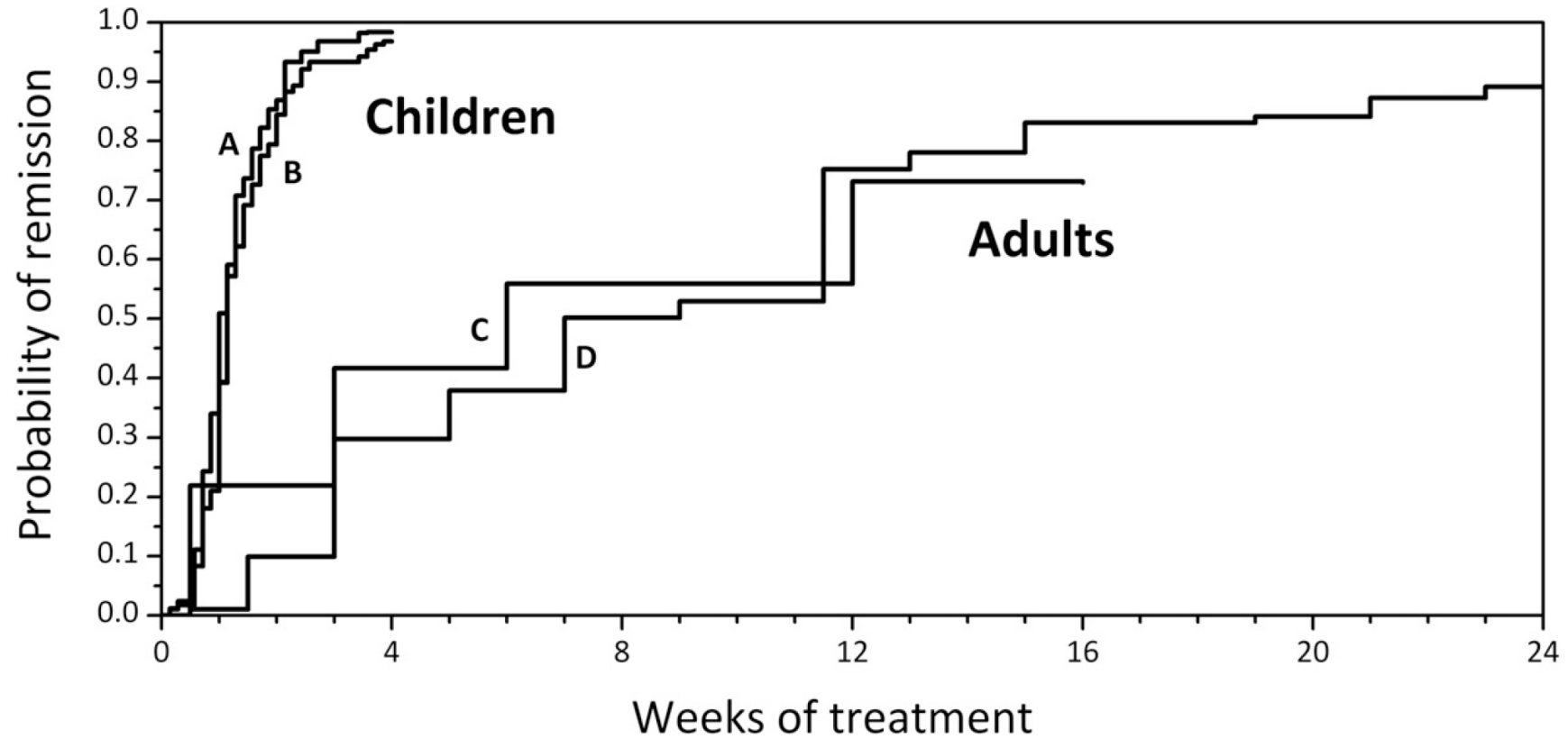
Assenza di responsta al PDN (60mg/m²/24h) in 4-6 settimane ± boli di MP

Multi-Drug Resistant Nephrotic Syndrome (MDRNS)

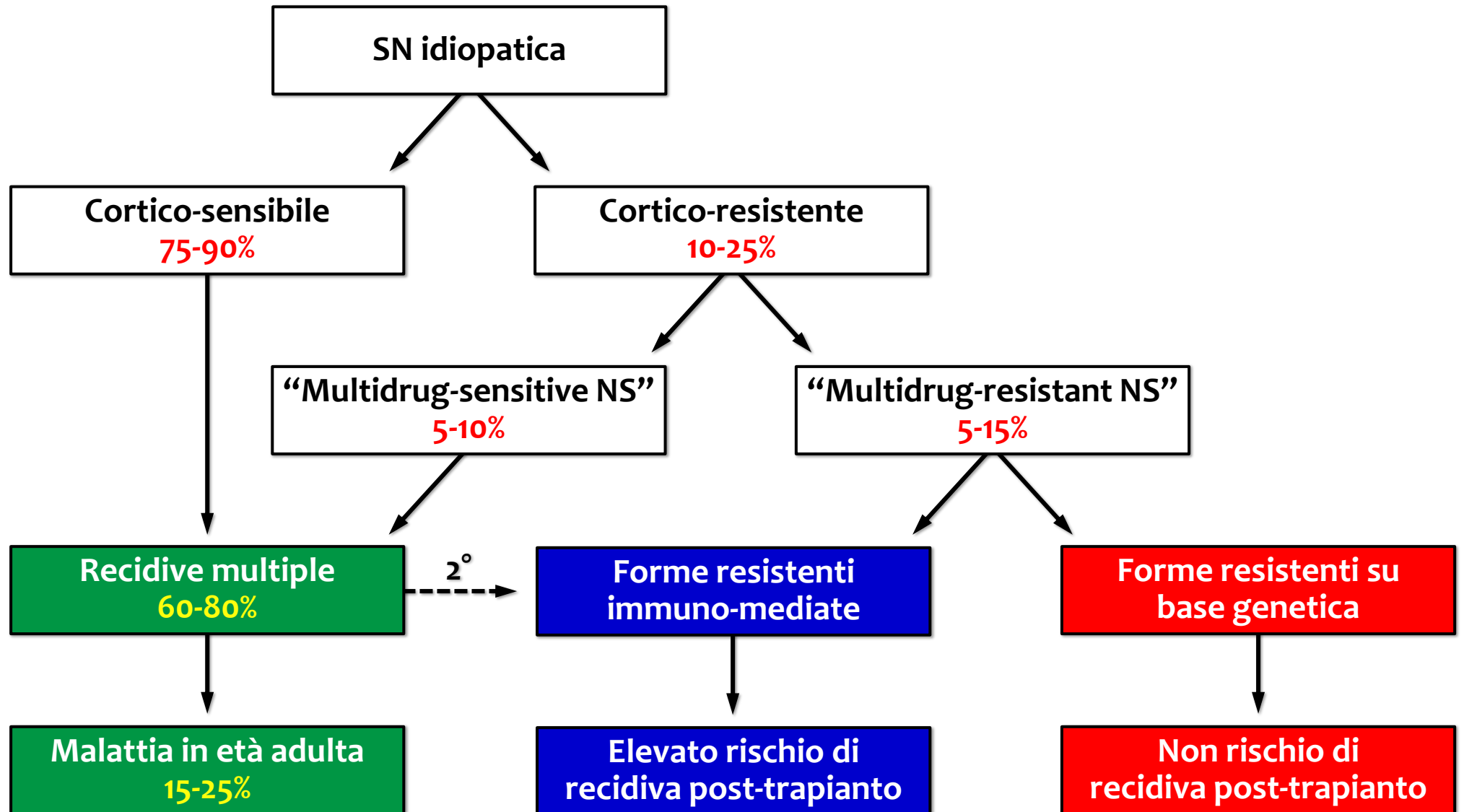
Mal definita, assenza di risposta a farmaci di seconda linea in:

- 6 mesi (remissione oarziale)**
- 24 mesi (remissione complete)**

Tempo alla remissione nelle forme corticosensibili



Sindrome nefrosica idiopatica del bambino



Rischi di bias negli studi clinici



- A. Randomizzazione casuale
- B. Allocazione in cieco
- C. Condotta dello studio in doppio cieco
- D. Analisi dei risultati in cieco
- E. Dati di esito incompleti
- F. Descrizione selettiva dei risultati
- G. Altri bias

Rischio di bias:



basso



incerto



elevato

Terapia iniziale per SNCS: confronto 3 vs 5-6 mesi di PDN

Study or Subgroup	Five - six months		Three months		Weight	Risk Ratio M-H, Random, 95% CI	Risk Ratio M-H, Random, 95% CI	Risk of Bias						
	Events	Total	Events	Total				A	B	C	D	E	F	G

2.8.2 High or unclear risk of bias for allocation concealment

Mishra 2012	1	37	1	37	2.5%	1.00 [0.06, 15.40]
Sharma 2002	8	70	24	70	18.3%	0.33 [0.16, 0.69]
Subtotal (95% CI)		107		107	20.8%	0.36 [0.18, 0.72]

Total events 9 25

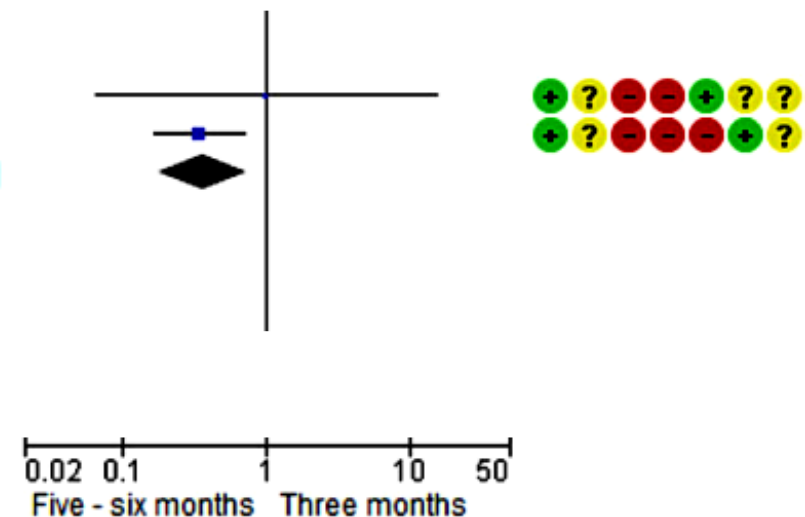
Heterogeneity: $\tau^2 = 0.00$; $\chi^2 = 0.58$, $df = 1$ ($P = 0.45$); $I^2 = 0\%$

Test for overall effect: $Z = 2.86$ ($P = 0.004$)

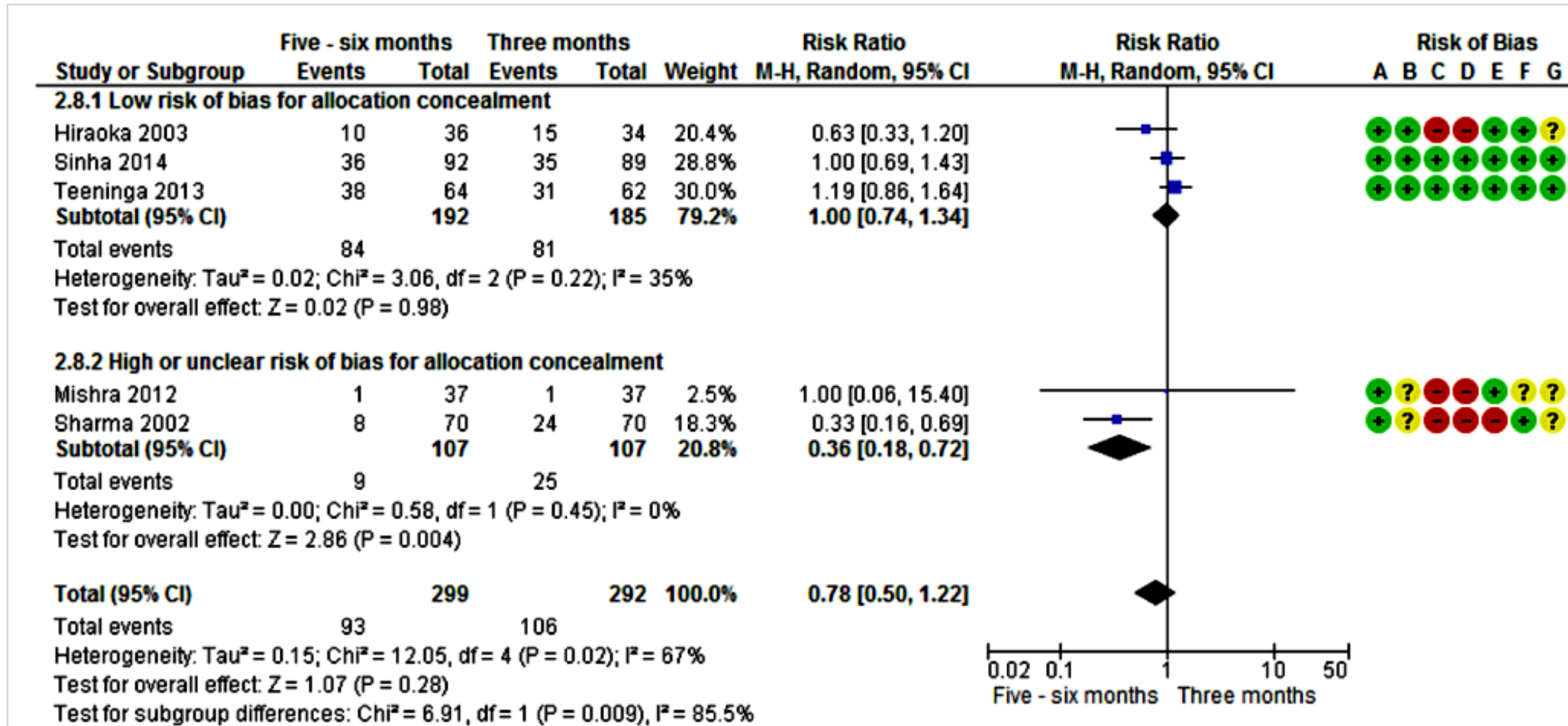
Heterogeneity: $\tau^2 = 0.15$; $\chi^2 = 12.05$, $df = 4$ ($P = 0.02$); $I^2 = 67\%$

Test for overall effect: $Z = 1.07$ ($P = 0.28$)

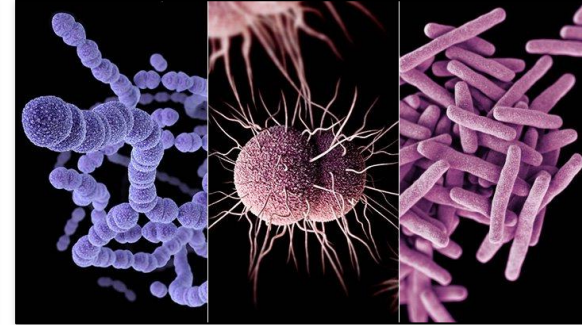
Test for subgroup differences: $\chi^2 = 6.91$, $df = 1$ ($P = 0.009$), $I^2 = 85.5\%$



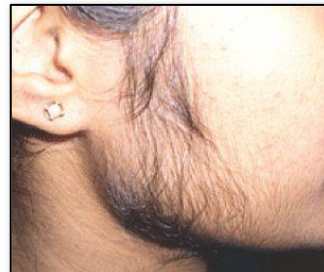
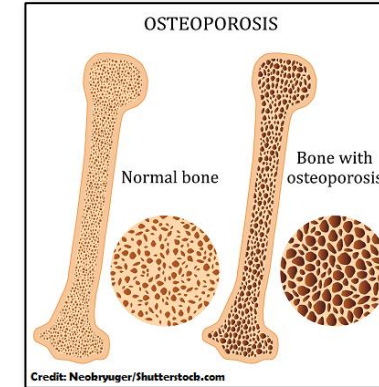
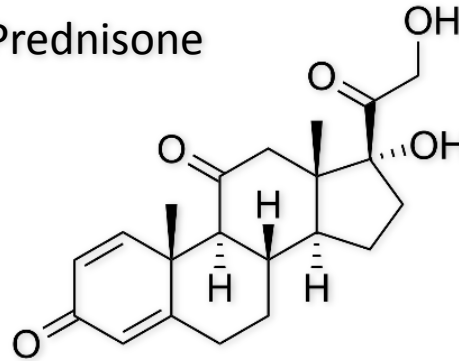
Terapia iniziale per SNCS: confronto 3 vs 5-6 mesi di PDN



Tossicità steroidea



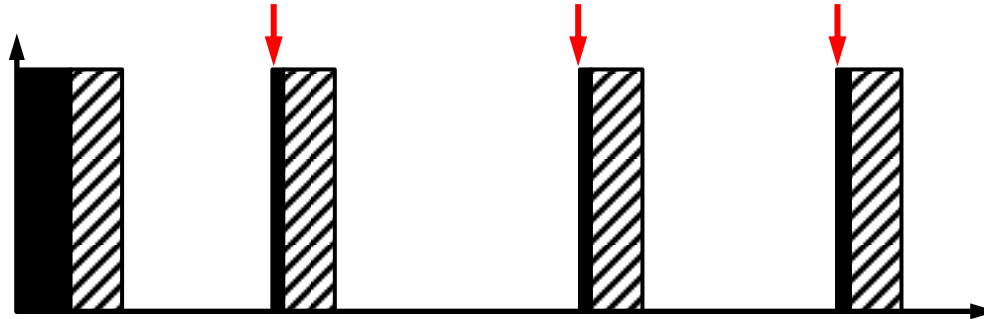
Prednisone



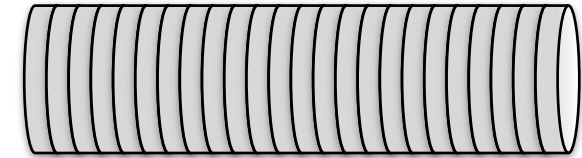
Principi del trattamento steroideo



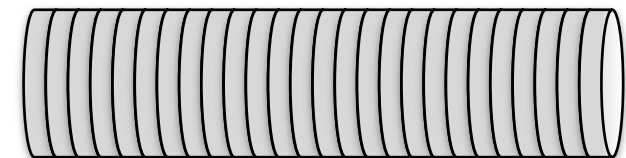
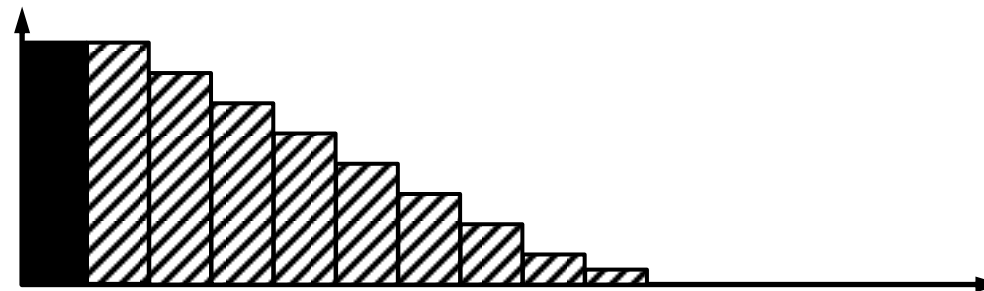
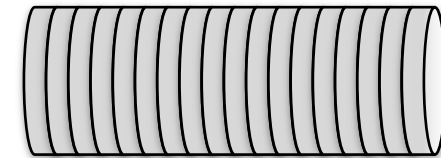
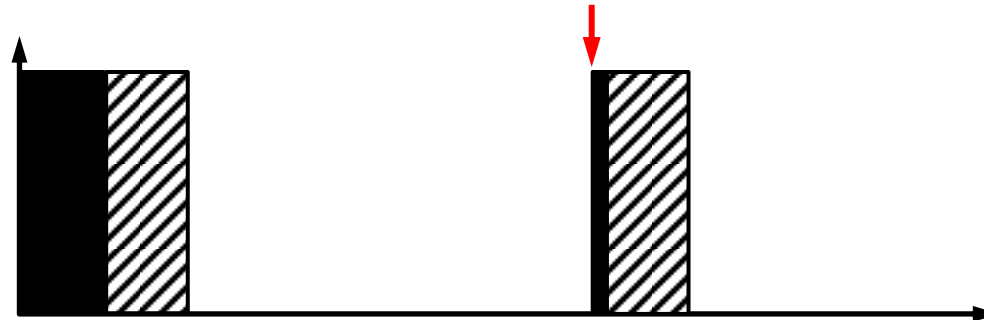
Ricaduta



Dose cumulative di PDN



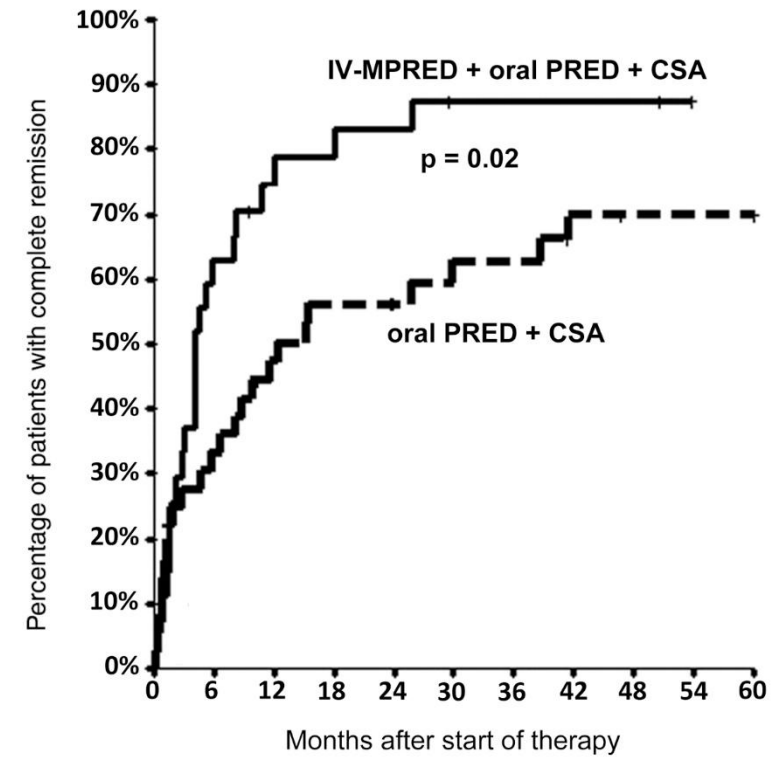
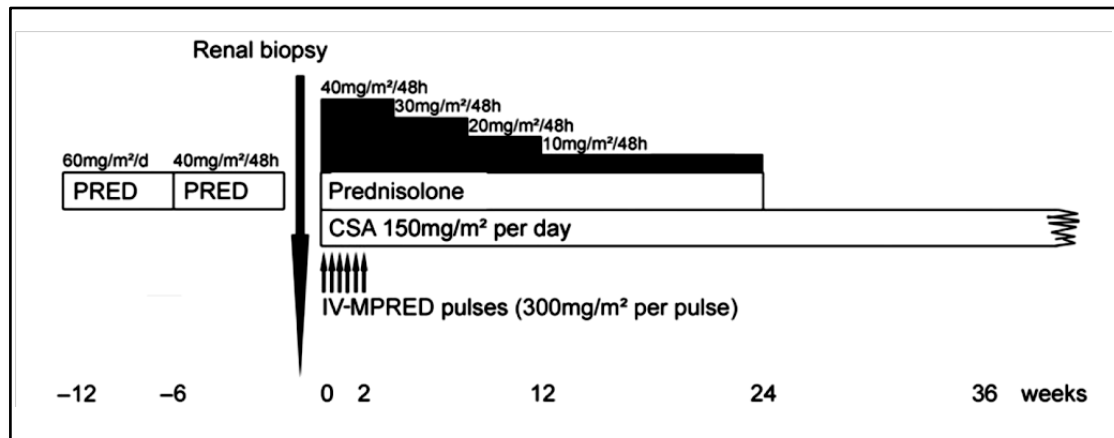
Trattamento migliore →



Farmaci di risparmio steroideo

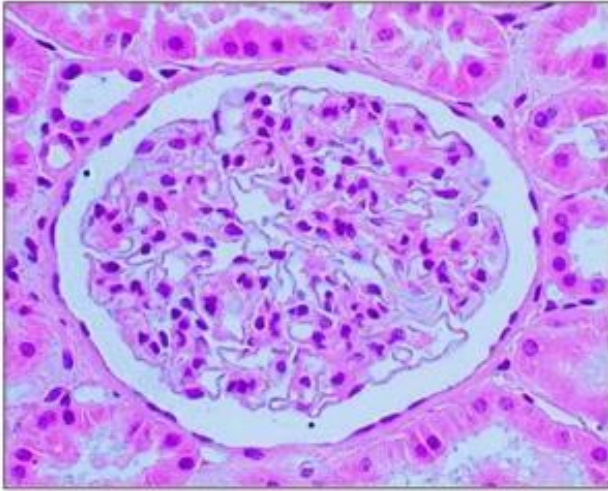
Farmaco	Efficacia	Indicazione	Commenti
Inibitori di calcineurina Ciclosporina A, tacrolimus	+++++	SDNS	Tossicità renale / ipertensione
Acido micofenolico	++++	FRNS/SDNS	Spesso efficace solo a dosaggi elevati (> 600 mg/m ²)
Levamisolo	+++	FRNS	Efficace nelle forme meno gravi Difficile da reperire
Ciclofosfamide	+++	?	Nei casi molto severi, spesso meno efficace
Rituximab, Ofatumumab	buona	?	Incertezze su: dosaggio ottimale, numero massimo di dosi, effetti secondari a lungo termine
Altre: immunoassorbimento, plasmaferesi, cellule staminali mesenchimali....	?	?	Probabilmente basso rischio di tossicità

La remissione completa nelle forme cortico-resistenti non genetiche è possibile

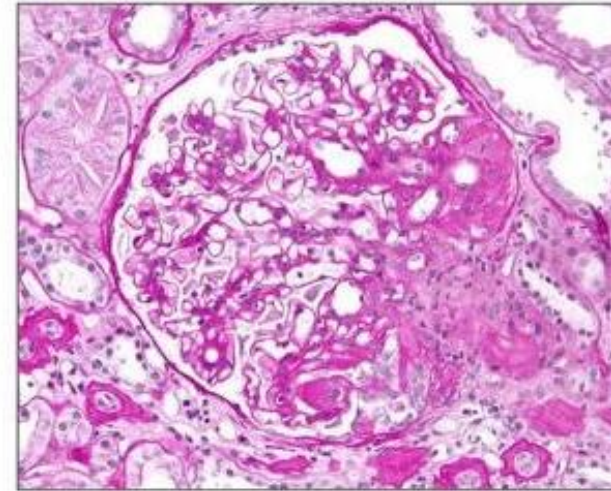


La diagnosi istologica dipende dal momento della biopsia

Minimal change disease
MCD

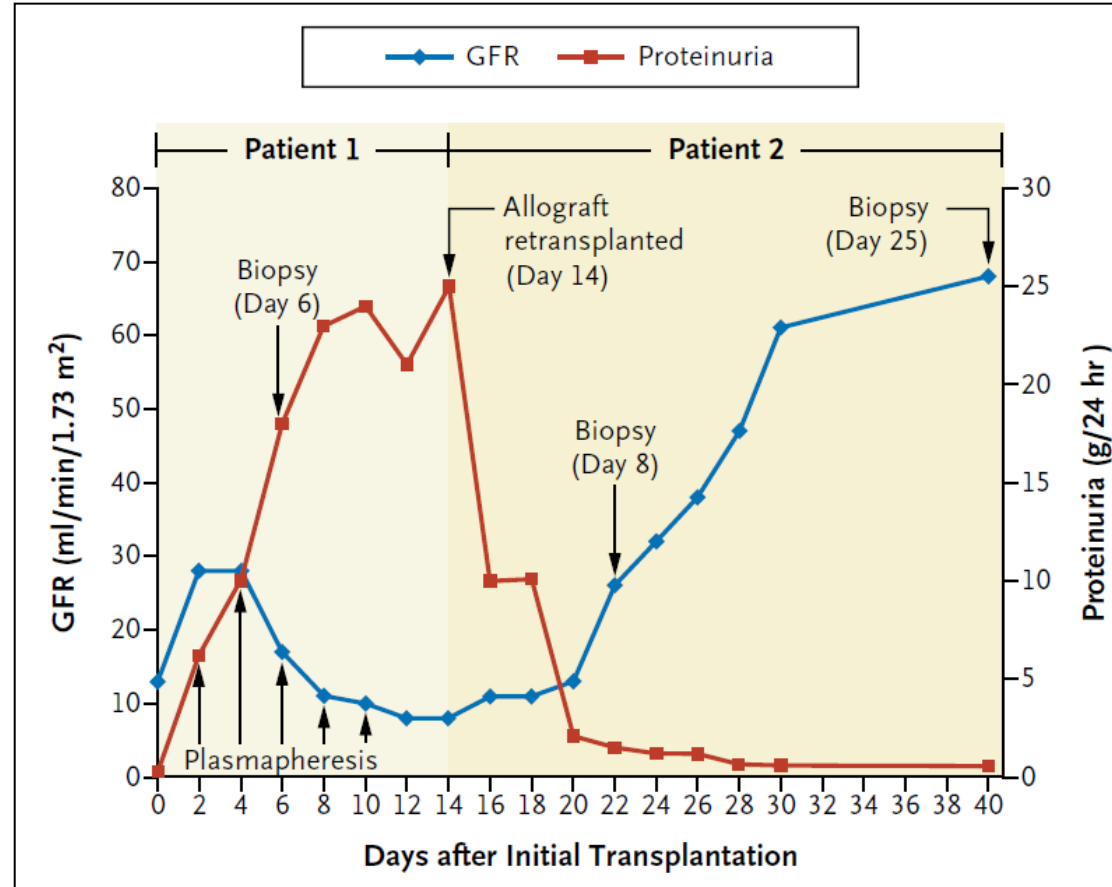


Focal segmental glomerulosclerosis
FSGS



Conta la risposta clinica!

Recidiva post-trapianto: un caso molto illustrativo...



Gallon et al, NEJM 2012

Non sempre si riesce a curare la recidiva post-trapianto

Grazie



francesco.emma@opbg.net