

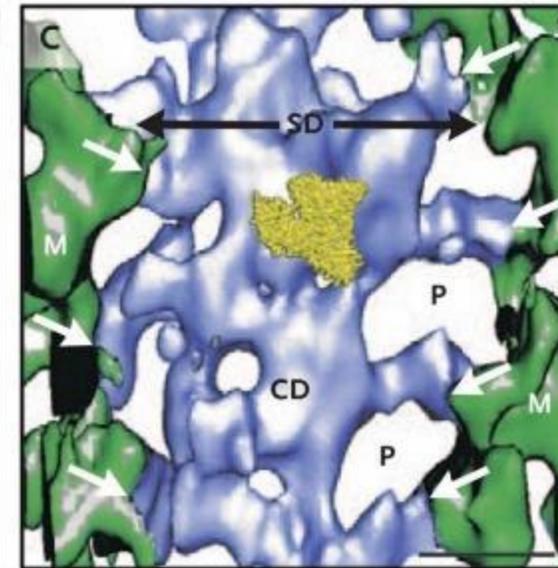
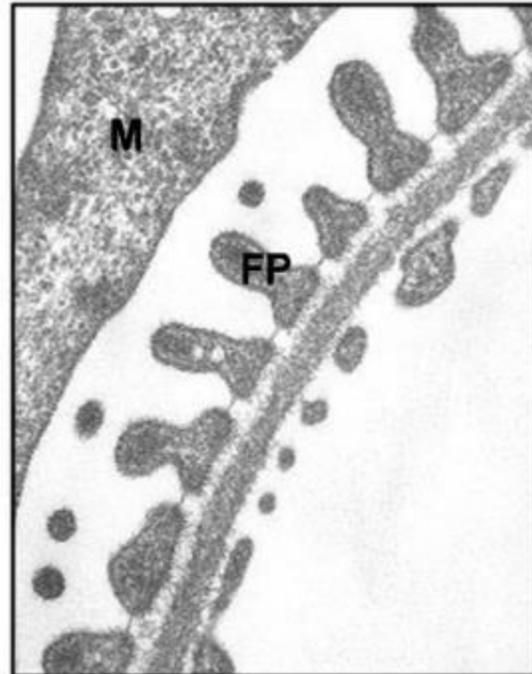
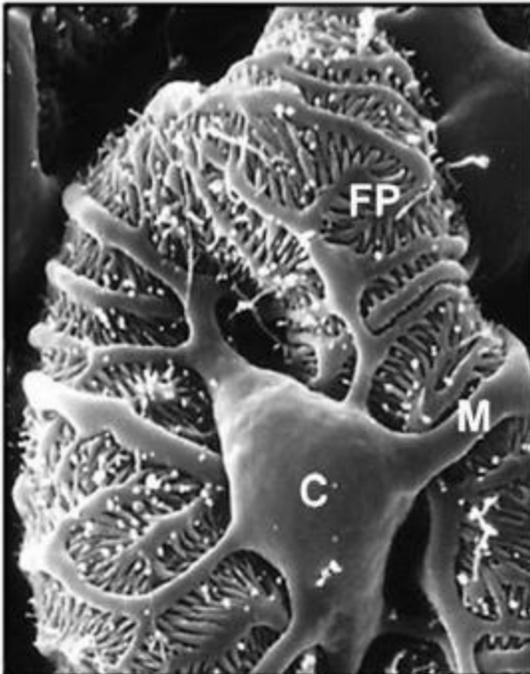
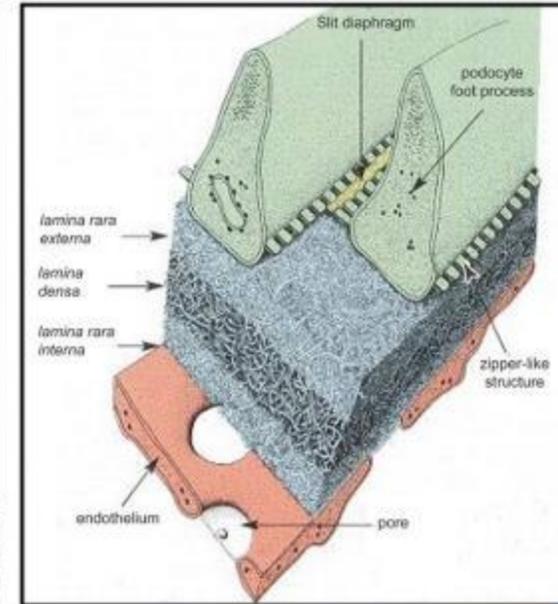
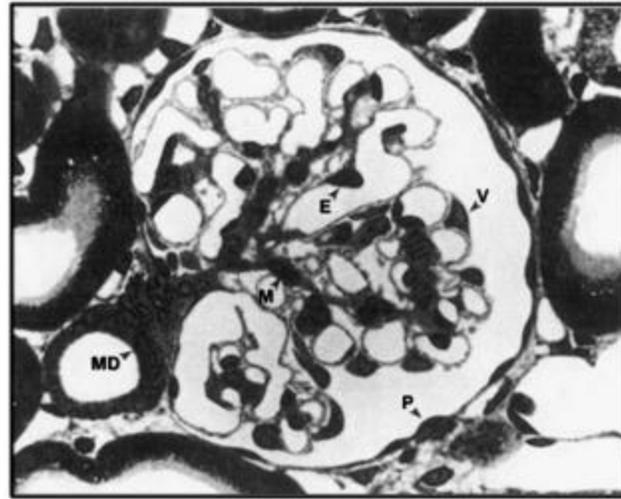
Sindrome nefrosica corticosensibile

Francesco Emma

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

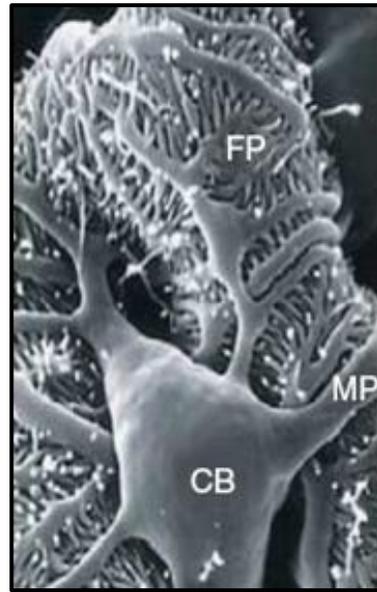
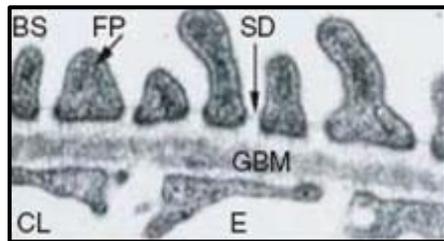
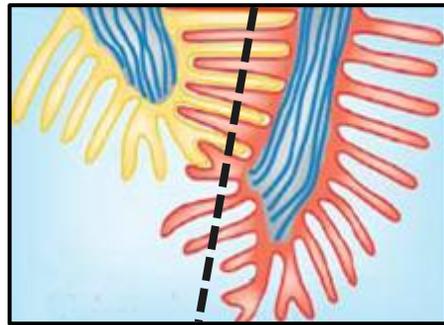
Roma 27 aprile 2019

Il glomerulo

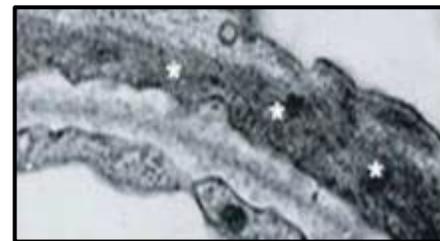
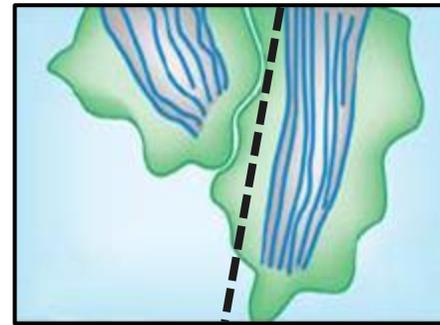


Fusione dei pedicelli

Normal



Nephrotic syndrome



Definizioni...

Steroid Sensitive Nephrotic Syndrome (SSNS)

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

Steroid Resistant Nephrotic Syndrome (SRNS)

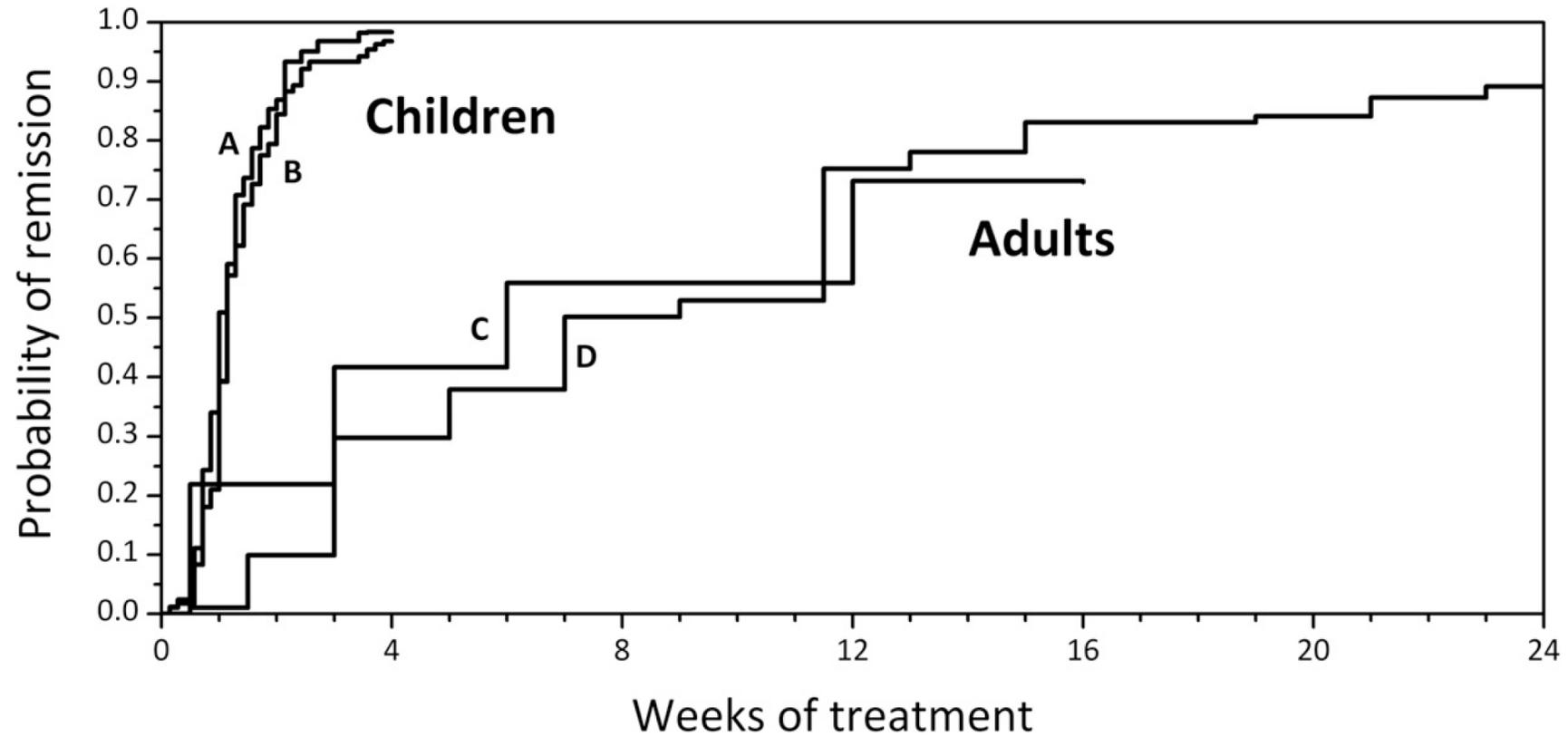
Assenza di responsta al PDN (60mg/m²/24h) in 4-6 settimane ± boli di MP

Multi-Drug Resistant Nephrotic Syndrome (MDRNS)

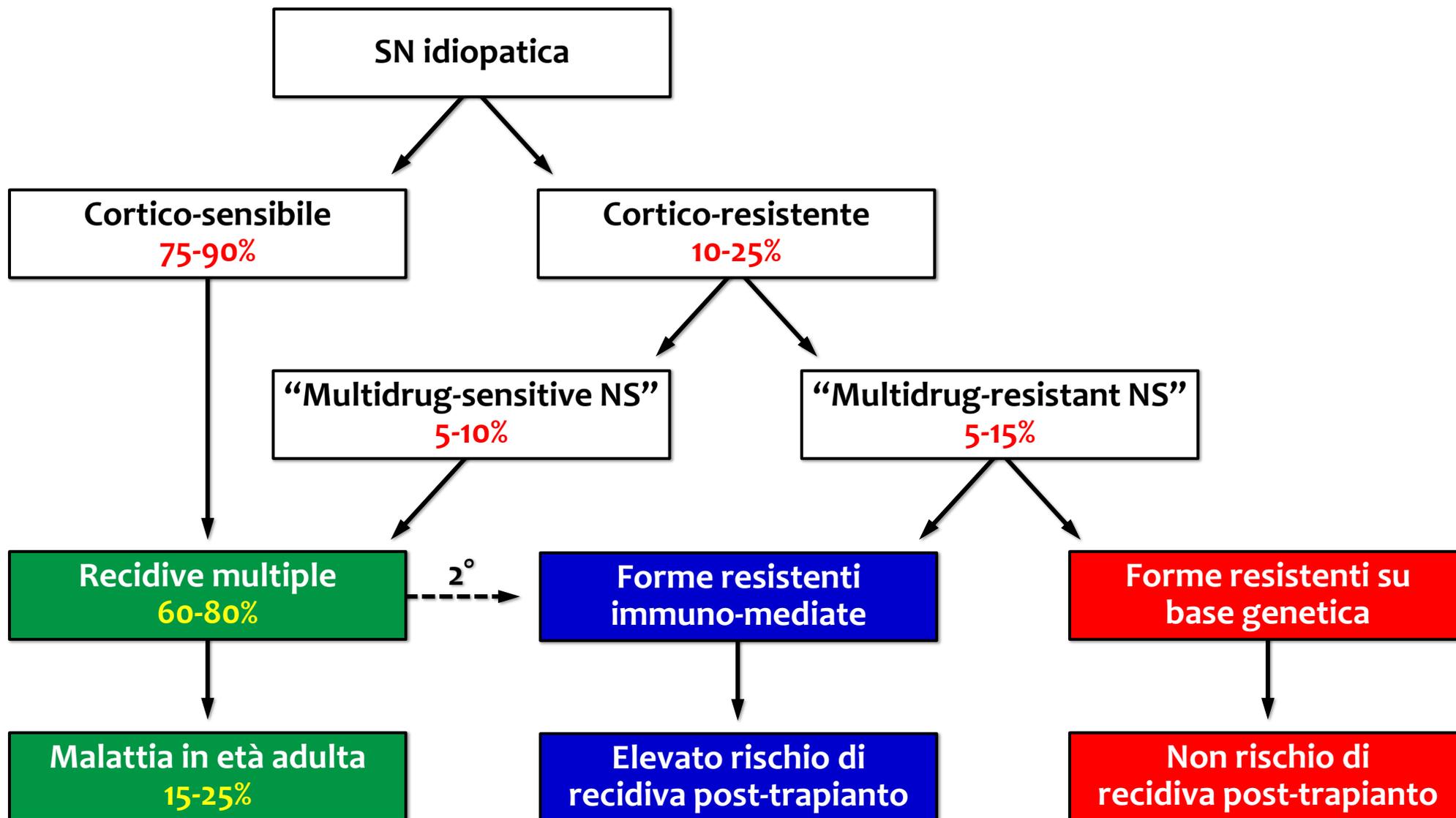
Mal definita, assenza di risposta a farmaci di seconda linea in:

- 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

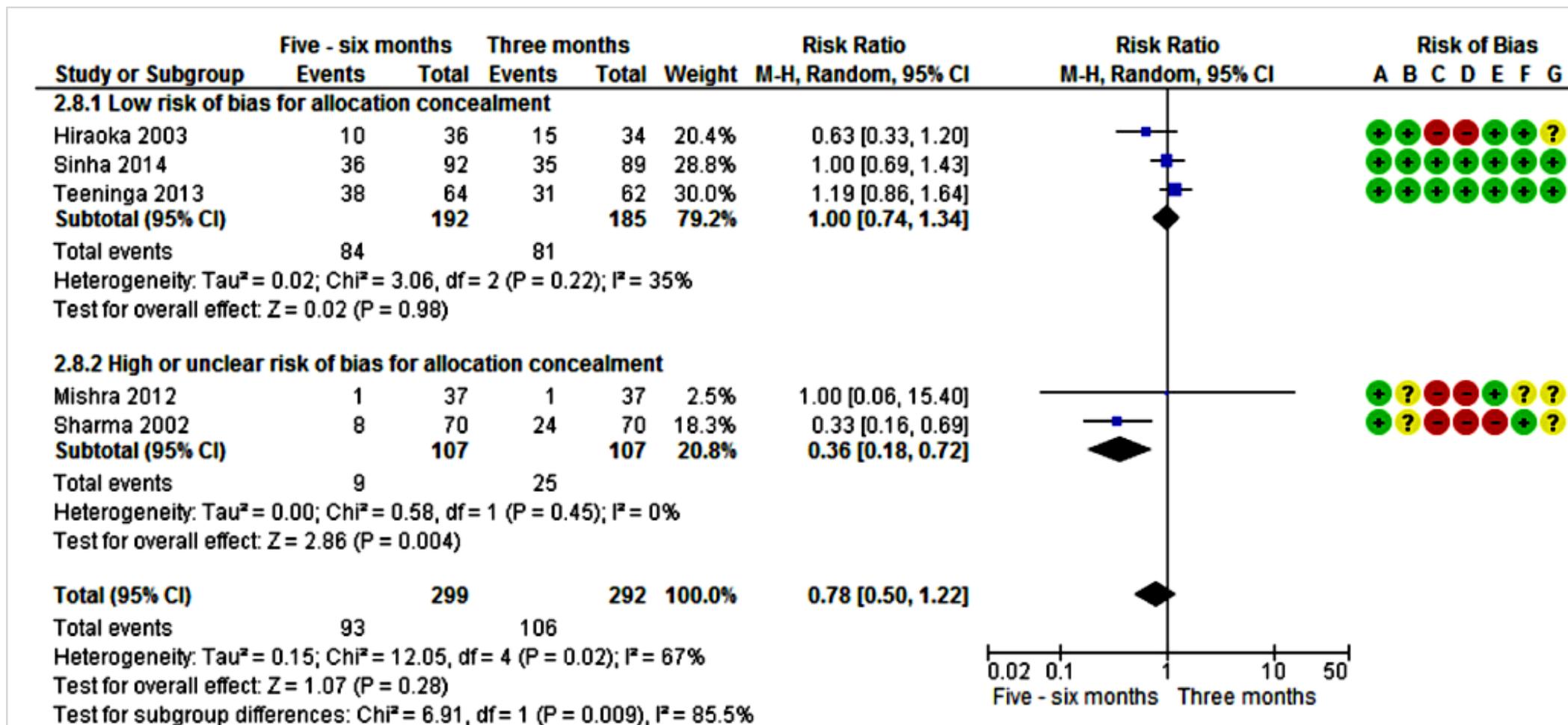


- A. Randomizzazione casuale
- B. Allocazione in cieco
- C. Condotta dello studio in doppio cieco
- D. Analisi dei risultati in cieco
- E. Dati di esito incompleti
- F. Descrizione selettiva dei risultati
- G. Altri bias

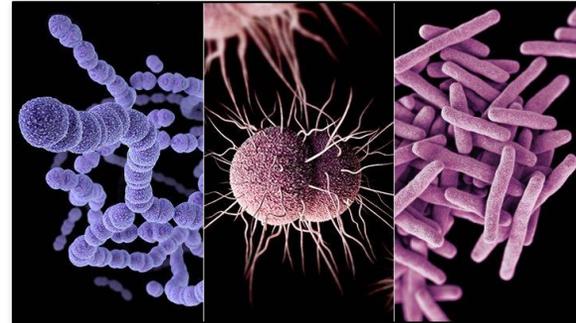
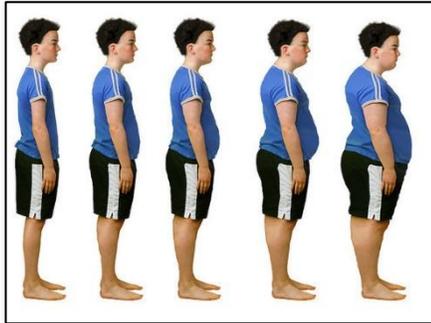
Rischio di bias:

-  basso
-  incerto
-  elevato

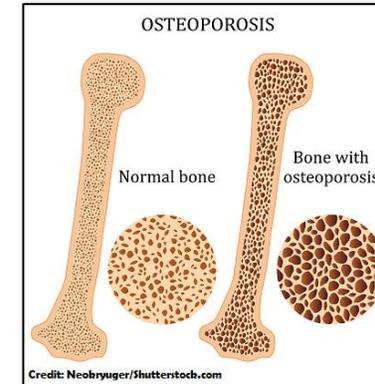
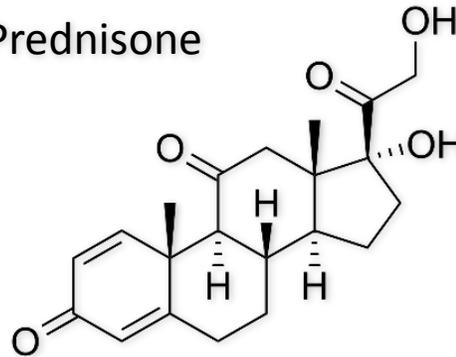
Terapia iniziale per SNCS: confronto 3 vs 5-6 mesi di PDN



Tossicità steroidea



Prednisone

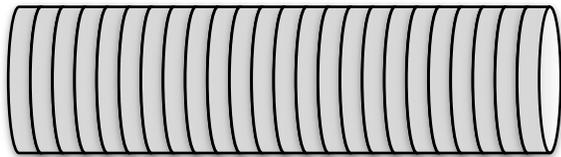
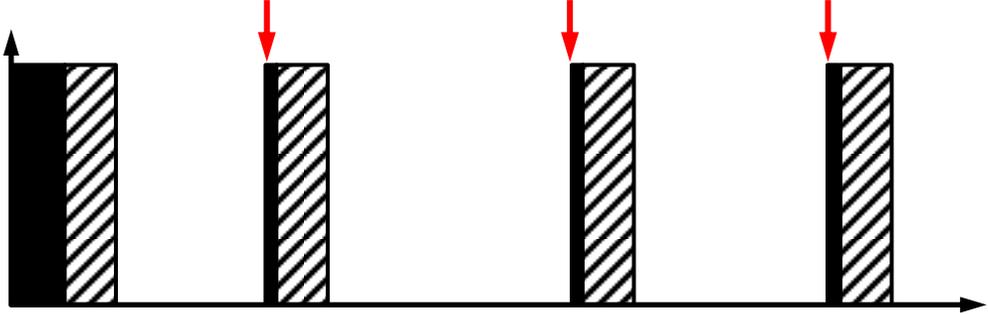


Principi del trattamento steroideo

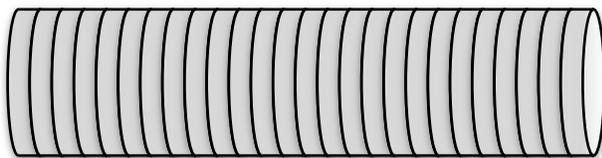
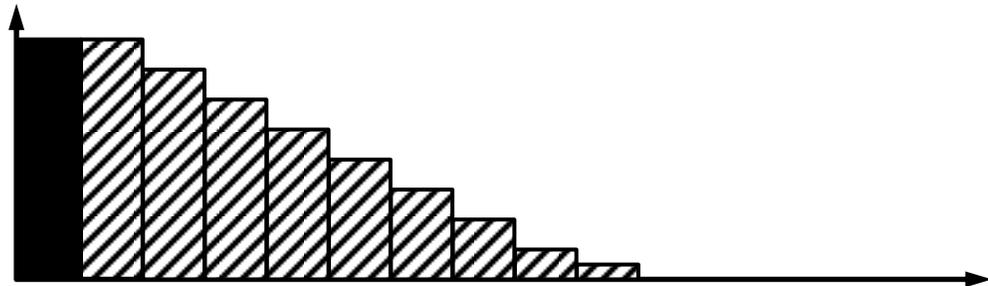
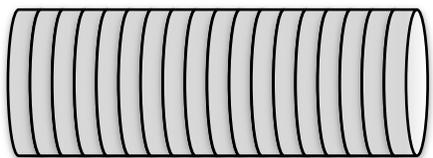
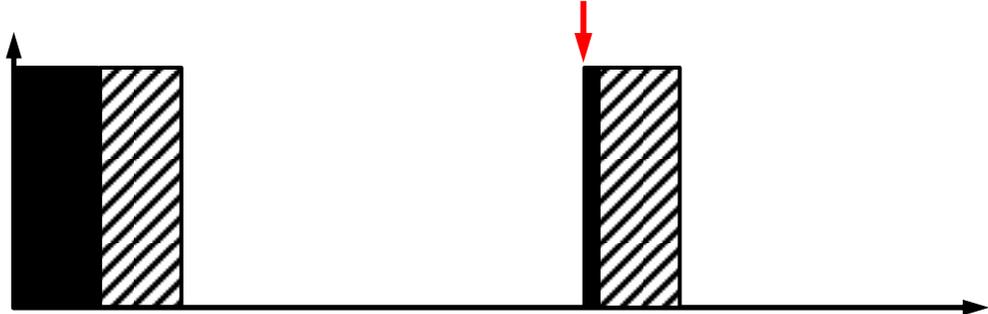
■ Giornaliero
▨ A gg alterni

Ricaduta

Dose cumulative di PDN



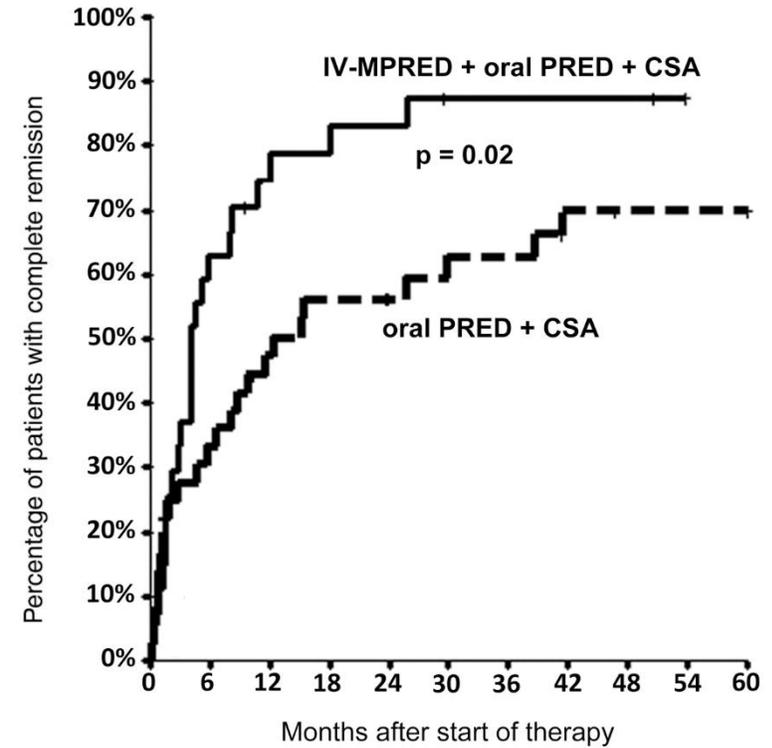
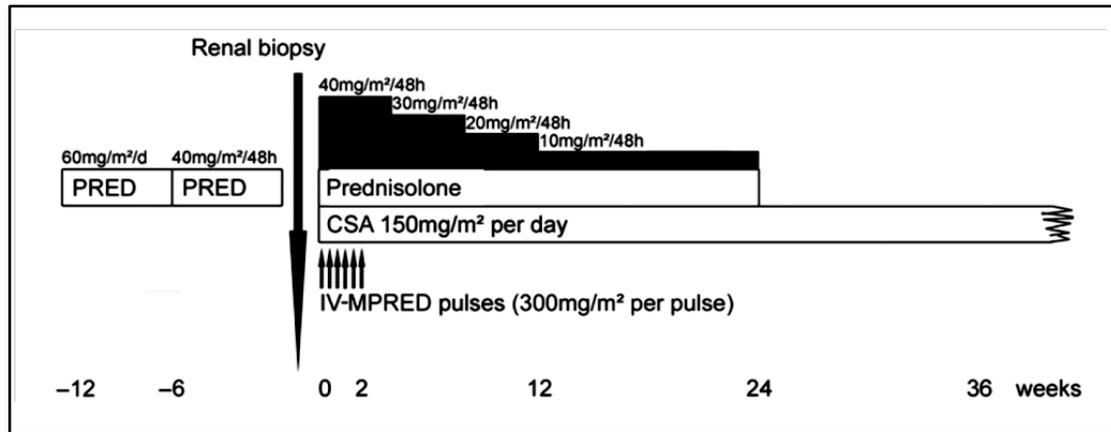
Trattamento migliore →



Farmaci di risparmio steroideo

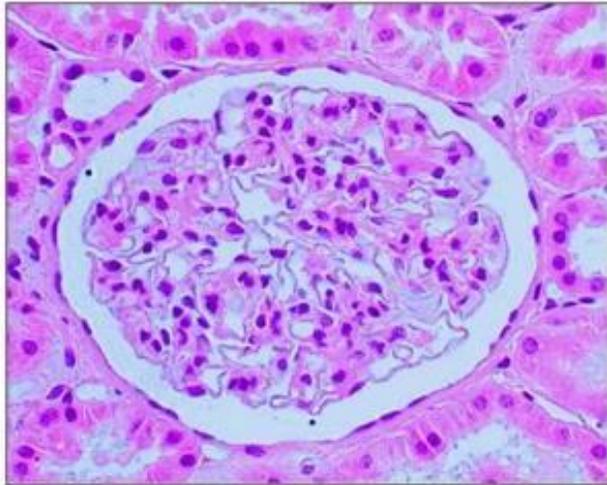
Farmaco	Efficacia	Indicazione	Commenti
Inibitori di calcineurina Ciclosporina A, tacrolimus	+++++	SDNS	Tossicità renale / ipertensione
Acido micofenolico	++++	FRNS/SDNS	Spesso efficace solo a dosaggi elevati (> 600 mg/m ²)
Levamisolo	+++	FRNS	Efficace nelle forme meno gravi Difficile da reperire
Ciclofosfamide	+++	?	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 complete nelle forme cortico-resistenti non genetiche è possibile

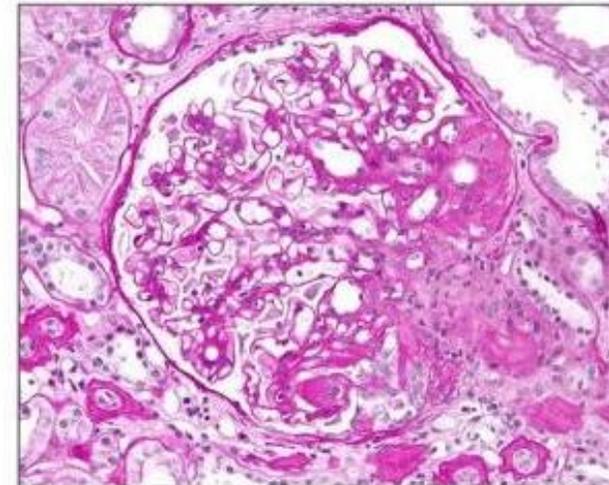


La diagnosi istologica dipende dal momento della biopsia

Minimal change disease
MCD

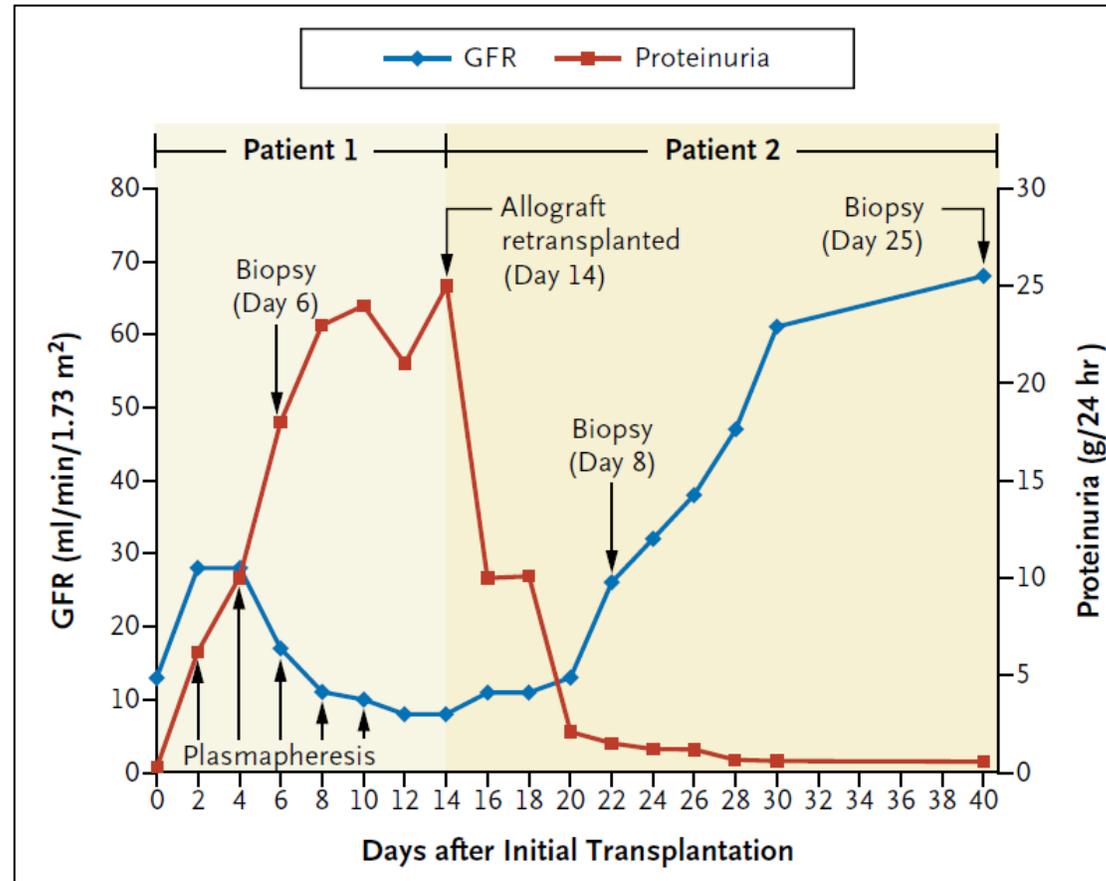


Focal segmental glomerulosclerosis
FSGS



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