Caso clínico
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Healthy

2006 (2 years old)
Nephrotic syndrome debut
- Creatinine normal
- C3-C4 normal
- VIH, hepatitis B and C (-)

Infrequently relapsing nephrotic syndrome
+ Hypertension
(enalapril, losartan, furosemide)

2009 (years old)
Renal Biopsy

KMRS.

Steroid-sensitive NS
CDNS (Minimal change glomerulopathy)
Duration of disease: 5 years

2006

Steroid-sensitive NS

2011

Corticosteroid-dependent NS

7 years old

- eGFR: 143 ml/min/1.73 m²
- Height Z score: -0.5 (119 cm)
- BMI Z score: +2.2 (31 kg)
Deep venous thrombosis right leg (anticoagulation)

Scrotal and suprapubic edema (discard DVT)

Primary lymphedema (Manual drain KNT)

Several hospitalizations

Cyclofosfamide
(8 weeks)

CsA

FK
CsA stop

Prednisone mg/m²
Primary Lymphedema

Legs, scrotal and suprapubic edema
Persistent (still in remission)
Skin infection
Sepsis
Moderate depression

Rituximab 500mg x 2
FK stop
Rituximab 500mg x 2

Prednisone mg/m²

En 2017
Feb
Mar
Abr
May
Jun
Jul
Aug
Sep
Oct
Nov
Dic
En 2018
Feb
Mar
Abr
May
Jun
Jul
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Oct
Nov
Dic
En 2019
Feb
Mar
Abr
May
Jun
Jul
Aug
Sep
Cyclofosfamide  
CsA  
FK  
Rituximab

Prednisone mg/m²

Cyclofosfamide  
CsA  
FK  
Rituximab

Prednisone mg/m²

Cyclofosfamide  
CsA  
FK  
Rituximab

Prednisone mg/m²

Cyclofosfamide  
CsA  
FK  
Rituximab

Prednisone mg/m²
Cyclofosfamide
CsA
FK
Rituximab

eGFR


eGFR ml/min/1.73m²
BMI Z score: 0.4
51Kg
Height Z score: -1.8
155cm

- 15 years old
- Corticosteroid-dependent NS
  - Duration of disease: 13y
  - Remission: 15m
  - 4m not receiving corticosteroids

- Normal ophthalmological evaluation
- Normal glycemia
- Normal eGFR 150ml/min/1.73m²
- Blood pressure p50

- No cushing
- Normal eGFR 150ml/min/1.73m²
- Tacrolimus (3.5 years)
- CsA (3.5 years)
- Ciclofosfamide (8 weeks)
- Prograf XL® 9mg/d (0.15mg/kg/d)
- Losartan 50mg/día
- Cotrimoxazole F-S-S
- El cal D forte®
What do we do now???...continue with tacrolimus?

- YES
- NO
- Biopsy and decide according to result
If he relapses again...what immunosuppression should you use?

A. Rituximab
B. Mycophenolate
C. Biopsy and decide according to result
D. Only prednisone
Conclusions

Idiopathic NS is the most common disorder of glomerular function in children

Long-term prognosis:
Complete resolution
Normal kidney function

- 80-90% steroid-sensitive
- 40-50% CDNS/FRNS: chronic glucocorticoid treatment

- Limiting the long term adverse effects
- Steroid-sparing treatment
Guidelines KDIGO

Treatment option for childhood onset complicated FRNS/CDNS

Efficacy and safety established

Rituximab could be a first-line treatment for uncomplicated FRNS/CDNS

- A single course of rituximab reliably retains disease remission for 6-12 months (relative)
- Minimize corticosteroid exposure
- Good tolerability and lack of nephrotoxic effects
- Is the most efficacious in maintaining relapse-free survival in children with CDNS???
• N:120, 3-16y
• A single course of rituximab vs tacrolimus
• Relapse-free survival rate: 90% vs 63.3%
• The relative risk of developing a relapse: 5 times >tacrolimus vs rituximab
• Median time to first relapse: 40 w vs 29m
• The mean cumulative corticosteroid dose during the 12-month study period was lower with rituximab compared with tacrolimus (25.8 vs 86.3 mg/kg).