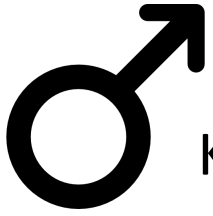


Caso clínico

Paola Riffo Ubilla

Nefróloga Pediátrica Hospital Base de Osorno



KMRS.

Healthy

2006 (2 years old)

Nephrotic syndrome
debut

- Creat → normal
- C3-C4 → normal
- VIH, hepatitis B y C (-)

Infrequently relapsing
nephrotic syndrome

+

Hypertension

(enalapril, losartan, furosemide)

2009 (years old)

Renal Biopsy

Steroid-sensitive NS

Steroid-sensitive
NS

2006

2011

Corticosteroid-dependent
NS

CDNS (Minimal change glomerulopathy)

Duration of disease → 5 years

7 years old

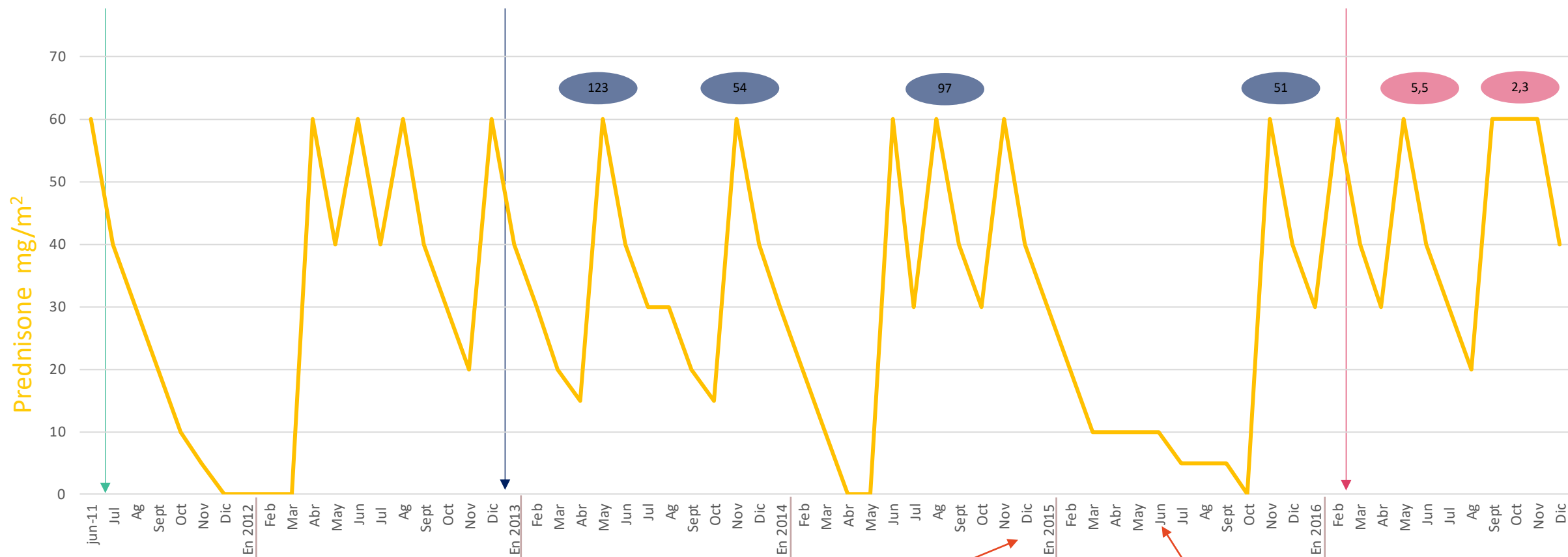


eGFR: 143 ml/min/1.73m²
Height Z score: -0.5 (119 cm)
BMI Z score: +2.2 (31 kg)

Cyclofosfamide
(8 weeks)

CsA

FK
CsA stop



Deep venous thrombosis right leg
(anticoagulation)

Scrotal and suprapubic edema
(discard DVT)

Primary lymphedema
(Manual drain KNT)

Several hospitalizations

Primary Lymphedema



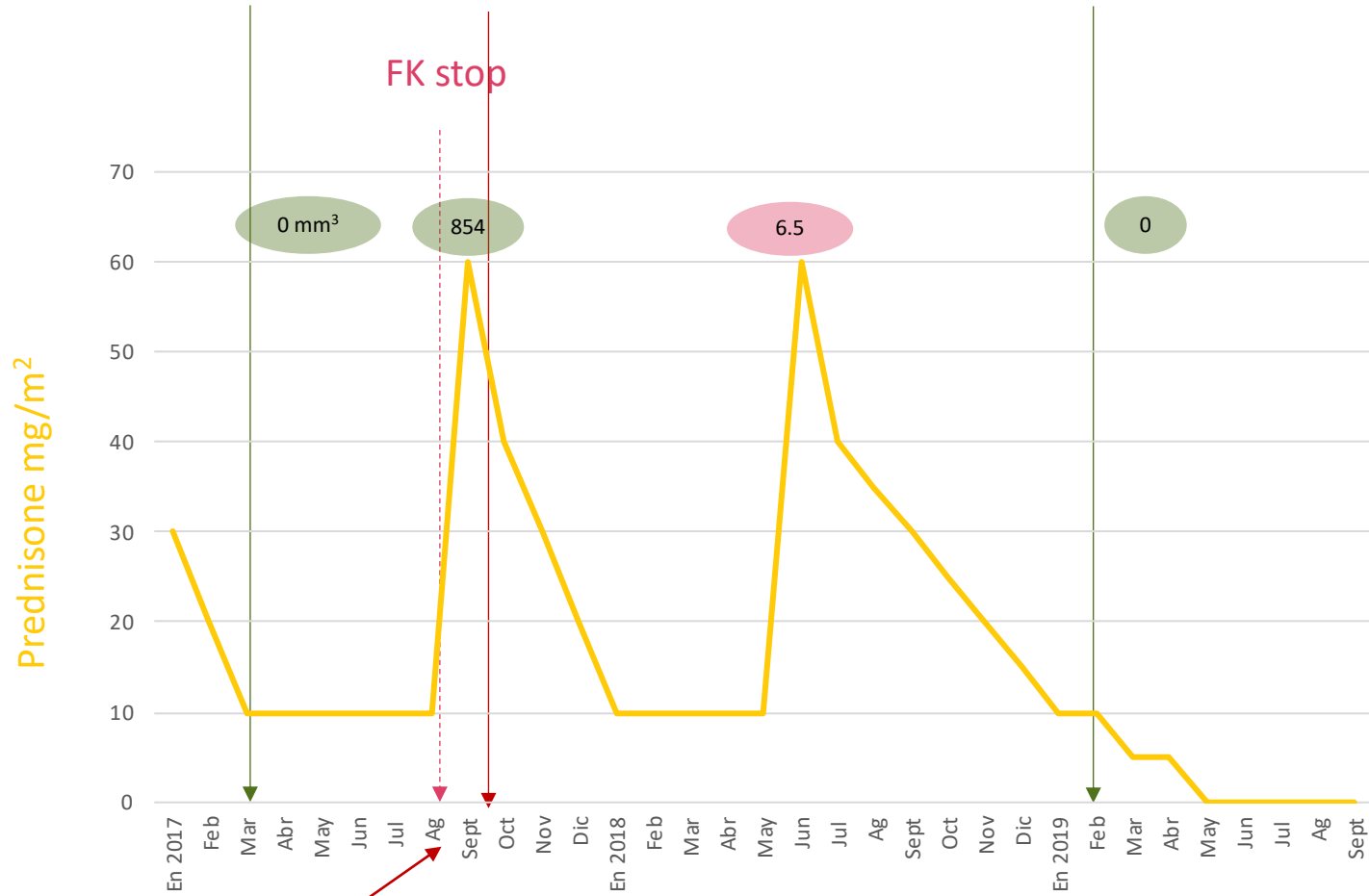
Legs, scrotal and suprapubic edema

Persistent (still in remission)

Rituximab 500mg x 2

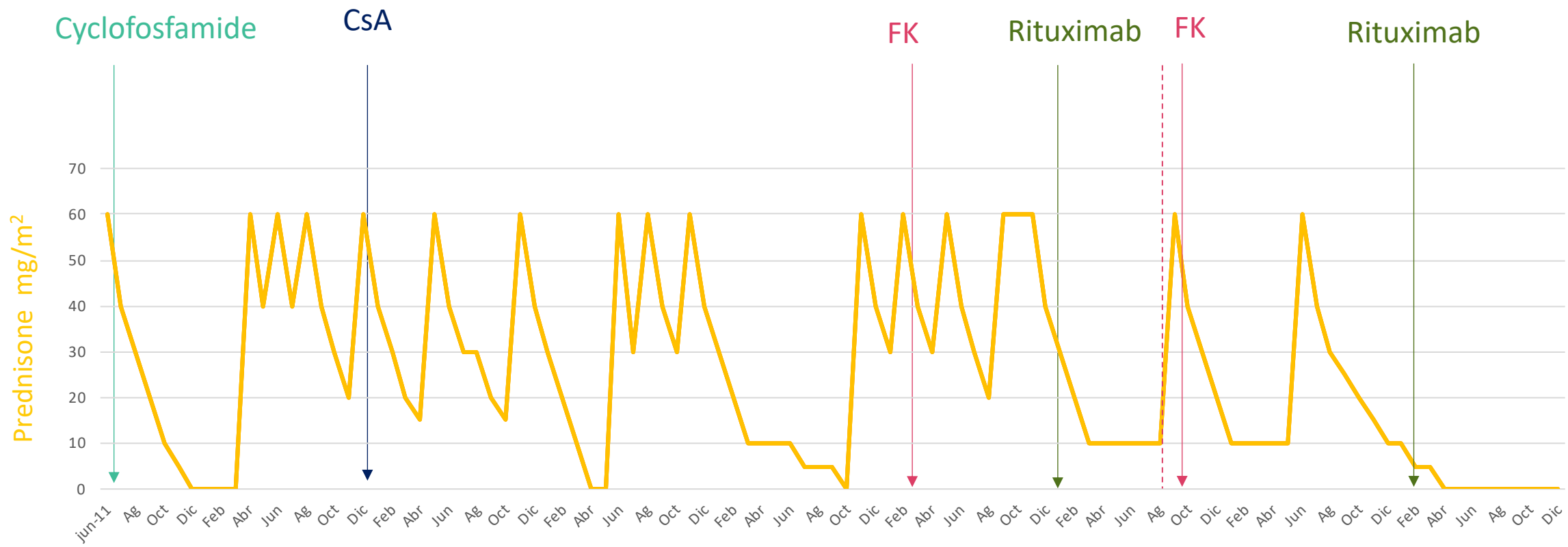
FK

Rituximab 500mg x 2

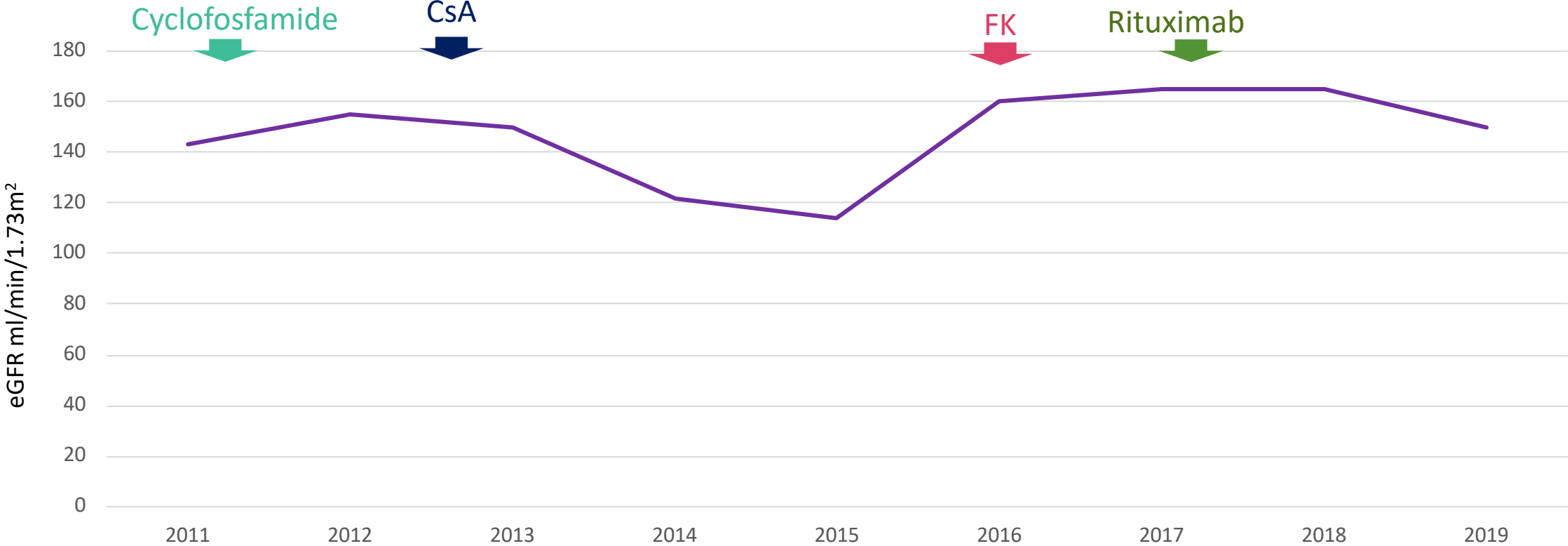


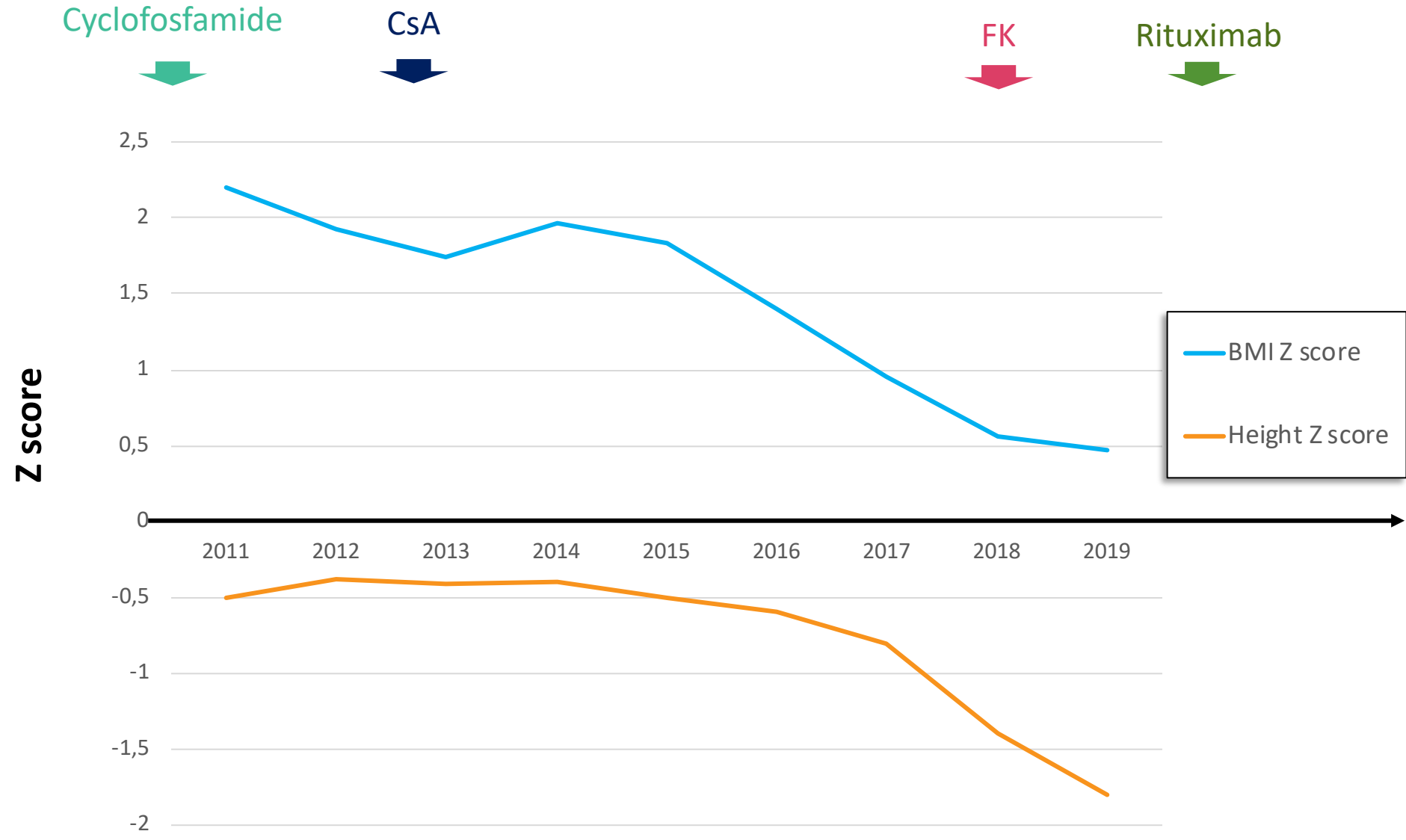
Skin infection
Sepsis

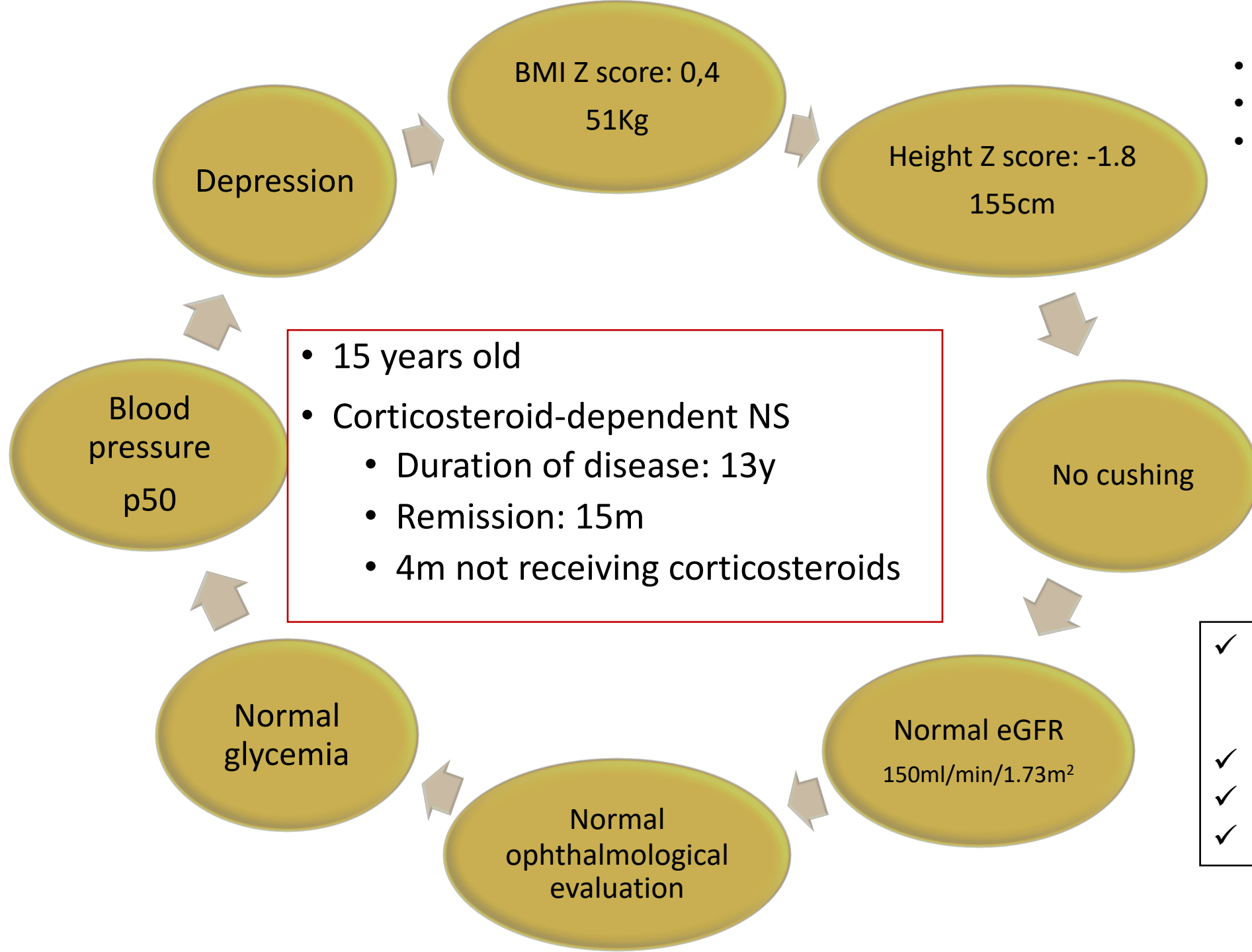
Moderate
depression



eGFR







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

- Ciclofosfamide (8 weeks)
- CsA (3.5 years)
- Tacrolimus (3.5 years).

- ✓ Tacrolimus 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

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

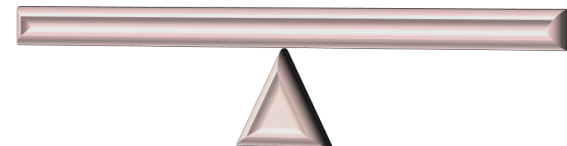
Long-term prognosis:
Complete resolution
Normal kidney function

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

Efficacy



Safety



Rituximab: B-lymphocyte– depleting monoclonal antibody

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???

Efficacy of Rituximab vs Tacrolimus in Pediatric Corticosteroid-Dependent Nephrotic Syndrome

A Randomized Clinical Trial

Biswanath Basu, MD; Anja Sander, PhD; Birendranath Roy, MD; Stella Preussler, MSc; Shilpita Barua, MD; T. K. S. Mahapatra, MD; Franz Schaefer, MD

- 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).

