

4.2 Prognosis

Practice Point 4.2.1: The prognosis for childhood nephrotic syndrome is best predicted by the patient's response to initial treatment and frequency of relapse during the first year after treatment. Therefore, a kidney biopsy is not usually needed at initial presentation, and instead is reserved for children with resistance to therapy or an atypical clinical course.

4.3 Treatment

4.3.1 Initial treatment of NS in children

Recommendation 4.3.1.1: We recommend that oral glucocorticoids be given for 8 weeks (4 weeks of daily glucocorticoids followed by 4 weeks of alternate-day glucocorticoids) or 12 weeks (6 weeks of daily glucocorticoids followed by 6 weeks of alternate-day glucocorticoids) (1B).

Practice Point 4.3.1.1: The standard dosing regimen for the initial treatment of nephrotic syndrome is daily oral prednisone/prednisolone 60 mg/m²/d or 2 mg/kg/d (maximum 60 mg/d) for 4 weeks followed by alternate day prednisone/prednisolone, 40 mg/m², or 1.5 mg/kg (maximum of 50 mg) for other 4 weeks, or prednisone/prednisolone 60 mg/m²/d (maximum 60 mg/d) for 6 weeks followed by alternate day prednisone/prednisolone, 40 mg/m², or 1.5 mg/kg (maximum of 50 mg), for other 6 weeks.

4.3.2 Prevention and treatment of relapses of NS in children

Recommendation 4.3.2.1: For children with frequently relapsing and steroid-dependent nephrotic syndrome who are currently taking alternate-day glucocorticoids or are off glucocorticoids, we recommend that daily glucocorticoids 0.5 mg/kg/d be given during episodes of upper respiratory tract and other infections for 5–7 days to reduce the risk of relapse (1C).

Practice Point 4.3.2.1: The initial approach to relapse should include oral prednisone/prednisolone as a single daily dose of 60 mg/m²/d or 2 mg/kg/d (maximum 60 mg/d) until the child remits completely for ≥3 days.

Practice Point 4.3.2.2: After achieving complete remission, reduce oral prednisone/prednisolone to 40 mg/m² or 1.5 mg/kg (maximum 50 mg) on alternate days for ≥4 weeks.

Practice Point 4.3.2.3: For children with frequently relapsing nephrotic syndrome or steroid-dependent nephrotic syndrome without glucocorticoid toxicity, the same glucocorticoid regimen may be employed in subsequent relapses.

Practice Point 4.3.2.4: For children with frequently relapsing nephrotic syndrome without serious glucocorticoid-related adverse effects, low-dose alternate-day oral prednisone/prednisolone (optimally ≤0.5 mg/kg/d) can be prescribed to prevent relapse.

Recommendation 4.3.2.2: For children with frequently relapsing nephrotic syndrome who develop serious glucocorticoid-related adverse effects and for all children with steroid-dependent nephrotic syndrome, we recommend that glucocorticoid-sparing agents be prescribed, rather than no treatment or continuation with glucocorticoid treatment alone (1B).

Practice Point 4.3.2.5: Patients should ideally be in remission with glucocorticoids prior to the initiation of glucocorticoid-sparing agents such as oral cyclophosphamide, levamisole, mycophenolate mofetil (MMF), rituximab, or calcineurin inhibitors (CNIs). Coadministration of glucocorticoids is recommended for ≥2 weeks following initiation of glucocorticoid-sparing treatment.

Practice Point 4.3.2.6: Choosing the most appropriate glucocorticoid-sparing agent from among oral cyclophosphamide, levamisole, MMF, rituximab, and CNI is a decision that requires careful consideration of specific patient-related issues such as resources, adherence, adverse effects, and patient preferences. Oral cyclophosphamide and levamisole may be preferable glucocorticoid-sparing therapies in frequently relapsing nephrotic syndrome. MMF, rituximab, CNIs, and to a lesser extent, oral cyclophosphamide may be preferable to glucocorticoid-sparing therapies in children with steroid-dependent nephrotic syndrome (Figure 41¹⁷⁸).