

IAP - IJPP CME 2018

NEPHROTIC SYNDROME - MANAGEMENT GUIDELINES

***Sangeetha G**

Abstract: *Steroid sensitive nephrotic syndrome is the most common form of nephrotic syndrome in children. Earlier, dysregulation of T cells was considered as the cause for proteinuria. Molecular mechanisms like podocyte injury, presence of circulating lymphocytotoxin, vascular permeability factor, impaired lymphocyte response with cross talk between T and B cells, etc., have given new insights in the understanding of nephrotic syndrome. Hypothesis about the mechanism of edema is also changing, with more focus on tubular epithelial sodium channels. Glucocorticoid is the cornerstone of treatment as the majority of children achieve complete remission after prednisolone treatment.*

Keywords: *Nephrotic syndrome, Steroid sparing drugs, Diuretics.*

Points to Remember

- *Idiopathic nephrotic syndrome is the most common among childhood nephrotic syndrome.*
- *Nephrotic syndrome should be treated adequately with corticosteroids both in terms of dosage and duration.*
- *In case of relapse, adequate treatment of infection may result in spontaneous remission.*
- *Low dose steroid is always coadministered with steroid sparing drugs in the initial period of treatment of FRNS and SDNS.*
- *All steroid sparing drugs have their own benefits and adverse effects and needs serial monitoring.*
- *Rituximab which selectively targets CD20-positive B cells is useful in difficult SDNS and FRNS. It may have variable results for SRNS.*
- *Nephrotic edema should be treated cautiously with serial monitoring of electrolytes.*
- *Parents of nephrotic syndrome children should be counselled regarding the need for vaccination during remission, particularly pneumococcal and varicella vaccination.*
- *Children with risk of suppression of hypothalamic-pituitary-adrenal axis should get stress dose steroids during the period of stress.*

References

1. Bagga A, Ali U, Banerjee S, Kanitkar M, Phadke KD, Senguttuvan P, et al. Indian Pediatric Nephrology Group, Indian Academy of Paediatrics. Management of Steroid Sensitive Nephrotic Syndrome: Revised guidelines. Indian Pediatr 2008; 45: 203-214.
2. Gulati A, Bagga A, Gulati S, Mehta KP, Vijayakumar M. Indian Society of Pediatric Nephrology, Indian Academy of Pediatrics. Management of Steroid Resistant Nephrotic Syndrome. Indian Pediatr 2009; 46(1):35-47.

* Assistant Professor,
Department of Pediatric Medicine and
Division of Pediatric Nephrology,
Sri Ramachandra Institute of Higher Education
and Research, Chennai.
email: sangeethaperungo@gmail.com

3. Niaudet P, Boyer O. Idiopathic nephrotic syndrome in children: clinical aspects. In: Avner ED, Harmon WE, Niaudet P, Yoshikawa N, Emma F, Goldstein SL (Eds). *Pediatric Nephrology*, 7th Edn, Verlag Berlin Heidelberg: Springer; 2016; pp839-882.
4. McCaffrey J, Lennon R, Webb NJ. The non-immunosuppressive management of childhood nephrotic syndrome. *Pediatr Nephrol* 2016; 31(9):1383-1402.
5. Andolino TP, Reid-Adam J. Nephrotic Syndrome. *Pediatr Rev* 2015; 36(3): 117-125.
6. Gbadegesin R, Smoyer WE. Nephrotic syndrome. In: Geary DF, Schaefer F (Eds). *Comprehensive Pediatric Nephrology*, 1st Edn, Philadelphia: Mosby Elsevier; 2008; pp204-218.
7. Siddall EC, Radhakrishnan J. The pathophysiology of edema formation in the nephrotic syndrome. *Kidney Int* 2012; 82(6):635-642.
8. Saravanan G, Amish Udani, Vijayakumar M. Steroid sensitive nephrotic syndrome. In: Vijayakumar M, Nammalwar BR (Eds). *Principles and Practice of Pediatric Nephrology*, 2nd Edn, New Delhi: Jaypee Brothers Medical Publishers; 2013; pp324-343.
9. Steroid-sensitive nephrotic syndrome in children. *Kidney Int Suppl* (2011) 2012; 2(2): 163-171.
10. Bagga A, Sinha A. Pathogenesis and therapy of nephrotic syndrome. *Indian J Pract Pediatr* 2017; 19(2):140-155.
11. Sinha A and Bagga A. Rituximab therapy in nephrotic syndrome: implications for patients' management. *Nat Rev Nephrol* 2013; 9(3):154-169. doi:10.1038 nrneph.2012.289.