International Journal of Contemporary Pediatrics
Pandya NK et al. Int J Contemp Pediatr. 2018 Jul;5(4):1172-1175
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: http://dx.doi.org/10.18203/2349-3291.ijcp20182057

Original Research Article

Clinical profile of patients with steroid sensitive nephrotic syndrome at tertiary care centre in Gujarat, India

Nimisha K. Pandya[1]*, Kedar G. Mehta[2]

[1]Department of Pediatrics, [2]Department of Preventive and Social Medicine, GMERS Medical College, Gotri, Vadodara, Gujarat, India

Received: 08 March 2018
Accepted: 27 April 2018

***Correspondence:**
Dr. Nimisha K. Pandya,
E-mail: nimisha.pandya@hotmail.com

ABSTRACT

Background: Nephrotic syndrome is a significant cause of chronic renal disease in children. The objective of the study was to analyze demographic profile, response to steroids and associated complications, in children with Nephrotic syndrome.
Methods: A retrospective study of all patients referred to Renal diseases clinic at GMERS Medical College Gotri Vadodara was done. Period of study was from year 2014 to June 2017. Variables assessed were sex distribution, age at presentation for first attack, occurrence of complications, steroid responsiveness and use of steroid sparing agents. Study was done from special clinic cards used for documentation of visits of patients in Nephrotic disease special clinic.
Results: 59 patients were studied. Mean age at presentation of was 4.08 years. Sex distribution ratio was 1.18:1. 88% of cases were infrequent relapsers. Steroid dependence was observed in 8.4% of cases. Complications were noted in 38% children. UTI was the commonest complication 13.5%. Associated renal conditions were present in 5% of children.
Conclusions: In the present study clinical profile of children with Nephrotic syndrome was concordant with typical nephrotic syndrome in children. Pattern of nephrotic syndrome and response to treatment did not differ significantly from other studies.

Keywords: Complications, Nephrotic syndrome, Steroid sensitive

INTRODUCTION

Nephrotic syndrome is an important chronic disease in children. Incidence is reported to be 2-3/100000 children in western countries while as its incidence is slightly higher (2-7/100000) in children with South Asian origin and its prevalence is 12-16/100000 children.[1] About 80% children with idiopathic Nephrotic syndrome show remission of proteinuria following treatment with corticosteroids and are classified as 'steroid sensitive'. Most patients have multiple relapses, placing them at risk for steroid toxicity, systemic infections, and other complications.[2] Nephrotic syndrome is defined as heavy proteinuria, hypoalbuminemia (serum albumin <2.5 g/dl), hyperlipidemia (serum cholesterol >200 mg/dl), and edema. Nephrotic range protienuria is present if early morning urine protein is 3+/4+ on dipstick or spot protein/creatinine ratio >2 mg/mg, or urine albumin excretion >40 mg/m^2 per day.[2]

The standard medication for treatment is prednisolone or prednisone. The medication is administered after meals to

reduce its gastrointestinal side effects. The treatment of first attack is prednisolone 2 mg/kg per day (maximum 60 mg in single or divided doses) for 6 weeks, followed by 1.5 mg/kg (maximum 40 mg) as a single morning dose on alternate days for the next 6 weeks. Remission is defined as urine albumin nil or trace for 3 consecutive early morning specimens.

Relapse is defined as urine albumin 3+ or 4+ 4 for 3 consecutive early morning specimens. Viral and bacterial infections may occasionally precipitate relapses in patients previously in remission.

Frequent relapsers are defined as children having two or more relapses in initial six months or more than three relapses in any twelve months. Frequent relapsers respond promptly to prednisolone and are managed using the regimen for each relapse. Such children are at a low risk for developing steroid toxicity.[2]

Steroid dependence is defined as two consecutive relapses when on alternate day steroids or within 14 days of tapering. It is usually not necessary to perform a renal biopsy in these cases.

Following treatment of a relapse, prednisolone is gradually tapered to maintain the patient in remission on alternate day dose of 0.5-0.7 mg/kg, which is administered for 9-18 months. A close monitoring of growth and blood pressure, and evaluation for features of steroid toxicity is essential.

If the prednisolone threshold, to maintain remission, is higher than 0.5 mg/corticosteroid toxicity are seen, additional use of steroid sparing agents like Levamisole and Cyclophosphamide is suggested.

Children with Nephrotic syndrome are prone to many complications. Complications of NS are divided into two categories: disease-associated and drug-related complications. Disease-associated complications include infections peritonitis, sepsis and cellulitis, acute renal failure, anaemia, hypocalcemia and bone disease.[3] The incidence of infections in NS are still a major problem in developing countries.

The most frequently encountered infections include: upper respiratory tract infections, urinary tract infections, peritonitis, pneumonia, acute gastroenteritis and tuberculosis.[4] Sepsis remains one of the main causes of death in children with Nephrotic Syndrome. Infections due to *S. pneumonie*, *β-hemolytic streptococci*, *Hemophilus influenza -B* and Gram-negative bacteria are frequently found.[3-5] Reduced serum concentration of immunoglobulin, impaired ability to make specific antibodies, decreased levels of alternative complement pathway and immunosuppressive treatment are linked to increased risk of infection in children with Nephrotic Syndrome.[6] All children with Nephrotic syndrome should receive immunization against pneumococcal infections as children may develop serious pneumococcal infections such as peritonitis and sepsis. Immunization Committee of the Indian Academy of Pediatrics recommends the heptavalent conjugate pneumococcal vaccine for children below 2 years of age followed later, by a dose of the 23-valent polysaccharide vaccine. Children older than 5 years require only a single dose of the polysaccharide vaccine.[7]

Corticosteroids are known to cause potentially serious side effects such as cushingoid features, obesity, growth retardation, hypertension, osteoporosis and cataracts.[8]

METHODS

A retrospective study of all patients referred to Renal diseases clinic at GMERS Medical College and General hospital, Gotri, Vadodara was done. Our medical college runs special clinic for patients with Nephrotic Syndrome every Thursday 3-5 pm. Monthly follow up of all patients is done.

All patients referred to special clinic are investigated for protienuria. Complete clinical examination is done to rule out complications. Bi annually all children undergo ophthalmic examination to screen for development of cataract. Period of review was from year 2014 to June 2017. Review was done from special clinic cards used for documentation of visits of patients in Renal disease special clinic. Variables assessed were sex distribution, age at presentation for first attack, occurrence of complications, steroid responsiveness and use of steroid sparing agents. Biopsy was performed on 2 patients with steroid dependence. Histopathology was suggestive of minimal change disease.

RESULTS

59 children presented with Steroid sensitive Nephrotic syndrome between the ages of 1-12 years with mean age at presentation for first attack as 4.08 years. There were 32 males (54.2) and 27 females (45.8) with male to female ratio of 1.18:1. 3 children above 9 years of age were diagnosed with steroid sensitive Nephrotic syndrome.

Table 1: Sex distribution and presentation of steroid sensitive nephrotic syndrome cases.

Total patients n = 59	n	(%)
Male	32	54.2
Female	27	45.8
Steroid sensitive Nephrotic patients n = 59		
Infrequent relapses	52	88.1
Frequent relapses	2	3.3
Steroid dependence	5	8.4

Majority of patients were infrequent relapsers 88.1%. Steroid dependence was observed in 8.4% patients. 1

patient on follow up was classified as frequent relapser and 2 patients developed steroid dependence on follow up. This is in concordance with prevalence of steroid sensitive Nephrotic syndrome in literature and other studies.

Table 2: Age of onset of first attack of Nephrotic syndrome.

Age at first attack (in years)	Number
1	4
2	10
3	13
4	9
5	10
6	4
7-8	5
9-12	3

All patients were evaluated for complications. Serious complications were observed in 40% of patients. Infections were seen in 27% patients of cases, commonest infection being urinary tract infections in 8 patients (13.5%). 3 (5%) patients were diagnosed with Pneumonia and 2 patients developed spontaneous bacterial peritonitis (3.3%).

42% of patients with urinary tract infection were culture positive, most common organism isolated being *E. coli*, followed by *Enterococcus*. Patients with culture positive UTI were treated on indoor basis. All patients with Pneumonia and SBP were treated as indoor patients.

Table 3: Complications in Nephrotic syndrome.

Complications n = 59	n	(%)
Urinary tract infections	8	13.5
Hypertension	4	6.7
Pneumonia	3	5
Tuberculosis	2	3.3
Spontaneous bacterial peritonitis	2	3.3
Septic shock	1	1.6
Cataract	3	5

Table 4: Associated renal conditions.

Associated conditions n = 59		
Renal dysgenesis	2	3.3
Renal tubular acidosis	1	1.6

Associated renal conditions noted were renal dysgenesis and RTA. Of 2 patients diagnosed with renal dysgenesis, 1 patient was diagnosed with unilateral kidney and other horse shoe kidney. 1 patient was diagnosed with Renal tubular acidosis type 1 with medullary nephrocalcinosis. Both patients presented as Infrequent Relapser Steroid Responsive Nephrotic Syndrome.

DISCUSSION

59 children diagnosed with Steroid sensitive Nephrotic syndrome were reviewed. The mean age at presentation for first attack as 4.08 years with male to female ratio of 1.18:1. These observations are similar to data available from other centers.[9] The mean age of presentation was similar to other studies. A study in Auckland observed mean age at diagnosis as 5.4 years.[10] A single center study done in Iran reported mean age of presentation as 4.87 years.[11] According to observational studies, the prevalence of Nephrotic Syndrome in children has a 2 to1 male to female ratio. Other studies report an incidence of 1.45-1.9/1.

Majority of patients were infrequent relapsers 88.1%. Steroid dependence was observed in 8.4% patients. biopsy was suggestive of minimal change disease in patuients with steroid dependence. This is in concordance with prevalence of steroid sensitive Nephrotic syndrome in literature and other studies.[9]

Serious complications were observed in 40% of patients.[3-5,8] Infections were seen in 27 % patients of cases, commonest infection being UTI, 13.5% followed by pneumonia, tuberculosis and peritonitis.[3-5] Three patients had features of steroid toxicity namely posterior capsular cataract 5%.[8]

Hypertension was observed in 6.7 % of children with steroid sensitive nephrotic syndrome. Minimal change nephrotic syndrome disease is usually not associated with Hypertension, but Hypertension was noted in 12% patients with minimal change disease in other studies too.[9] These children were started on antihypertensive. Response was noted in all patients and drugs were tapered within 3-4 months.

Associated renal conditions noted were renal dysgenesis and renal tubular acidosis. Genetic conditions linked with renal malformations may lead to functional changes in glomerular membrane resulting in manifestations of Nephrotic Syndrome.

CONCLUSION

Hence, we can conclude that there is no significant difference in the clinical and demographic profile of patients with Steroid Sensitive Nephrotic Syndrome. Major concern is the occurrence of complications, disease as well as drug related. Detailed examination on all visits, vaccination, patient education and routine screening for corticosteroid toxicity is required to prevent these complications in this chronic disease.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: The study was approved by the Institutional Ethics Committee

REFERENCES

1. Eddy AA, Symons JM. Nephrotic Syndrome in childhood. Lancet. 2003;362(9384):629-39.
2. Ali U, Bagga A, Banerjee S, Kanitkar M, Phadke KD, Senguttuvan P, et al. Revised guidelines for management of steroid-sensitive nephrotic syndrome. Indian Pediatr. 2008;45:203-14.
3. Gulati S, Kher V, Gulati K, Arora P, Gujral R. Tuberculosis in childhood nephrotic syndrome in India. Pediatr Nephrol. 1997;11:695-8.
4. Gorensek MJ, Lebel MH, Nelson JD. Peritonitis in children with nephrotic syndrome. Pediatr. 1988;81:849-56.
5. Alwadhi RK, Mathew JL, Rath B. Clinical profile of children with nephrotic syndrome not on glucorticoid therapy but presenting with infection. J Paediatr Child Health. 2004;40:28-32.
6. Anderson DC, York TL, Rose G, Smith CW. Assessment of serum factor B, serum opsonins, granulocyte chemotaxis, and infection in nephrotic syndrome of children. J Infect Dis. 1979;140:1-11.
7. Shah RC, Shah NK, Kukreja S. Pneumococcal vaccines. In: Shah RC, Shah NK, Kukreja S, editors. IAP Guide Book on Immunization. Mumbai: Wyeth; 2007:35-6.
8. Grenda R, Webb NJ. Steroid minimization in pediatric renal transplantation: early withdrawal or avoidance? Pediatr Transplant. 2010;14:961-7.
9. Sahana KS. Clinical Profile of nephrotic syndrome in children. J Evol Med Dent Sci. 2014;3(4):863-70.
10. Simpson AK, Wong W, Morris MC. Pediatric nephrotic syndrome in Auckland, New Zealand. J Paediatr Child Health. 1998;34(4):360-2.
11. Safaei A, Maleknajed S. Spectrum of childhood nephrotic syndrome in Iran: a single centre study. Indian J Nephrol. 2009:19(3):87-90.

Cite this article as: Pandya NK, Mehta KG. Clinical profile of patients with steroid sensitive nephrotic syndrome at tertiary care centre in Gujarat, India. Int J Contemp Pediatr 2018;5:1172-5.