E-ISSN 2250-0944
ISSN 2250-1150
doi: 10.31838/ijprt/15.02.27

Research Article

Evaluation of Idiopathic Late Onset Nephrotic Syndrome in Children

Dr. P. Ravindranath Reddy[1*], Dr. Grace Aparanji[2], Dr. D. Nagarjuna[3]

[1*]Associate Professor, Department of Paediatrics, Kurnool Medical College, Kurnool.
[2]Assistant Professor, Department of Paediatrics, Kurnool Medical College, Kurnool.
[3]Assistant Professor, Department of Paediatrics, Kurnool Medical College, Kurnool.

***Corresponding Author:** Dr. P. Ravindranath Reddy
Associate Professor, Department of Paediatrics, Kurnool Medical College, Kurnool.
Received: 10.04.25, Revised: 14.05.25, Accepted: 18.06.25

Abstract

Introduction: Idiopathic nephrotic syndrome is defined by the combination of a nephrotic syndrome (proteinuria, hypoalbuminemia, hyperlipidemia, and edema) and non-specific histological abnormalities of the kidney. The prevalence of NS is estimated at 2 to 7 per 100,000 children around the world. Children between 2 to 7 years of age get affected by this condition more often, particularly boys.

Materials and Methods: A Retrospective study of hospital records. Case sheets of children admitted in department of pediatrics, Government General Hospital, Kurnool with nephrotic children above the age of 10 years From January 2020 to December 2024. Children aged more than 10 years at onset fulfilled in the diagnostic criteria for nephrotic syndrome were included in the study. Secondary causes of nephrotic syndrome. (eg:SLE, Hepatitis B/C, Diabetic mellitus) were excluded from the study. Clinical data including age at onset, gender, presenting sympotoms and laboratory findings will be recorded histopathlogical finding will be categorized. Treatment response to steroids will be evaluated.

Results: The study included 108 children who were followed for a minimum duration of one year. The mean (±sd) age of the population was 13.3 (±1.4) years. The gender distribution of the population was males accounting for 66.7% and females at 33.3%. Hypertension was observed in 8(14.8%). Haematuria in microscopy was found in 28 (51.9%) study participants. Anti-Nuclear Antibodies investigation was done for 50 children and only 3(5.6%) were found to be positive. After the six weeks of steroidal therapy complete remission was found in 58 (53.7%) participants. No remission and partial remission were observed in 26 (24.1%) and 24 (22.2%) members of the study population respectively. Based on the response to steroidal therapy the participants were diagnostically classified as SRNS in 52 (48.1%) children followed SSNS in 50 (46.29%) children.

Conclusion: In our study most of the patients were diagnosed with SRNS and SSNS, and most common histological findings were MCNS, MES HC, FSGS and MESPGN. Late onset NS has higher frequency of atypical features, steroid resistance, and histopathology showing lesions other than MCD. Early biopsy may be useful guide to management.

Keywords: Idiopathic Nephrotic Syndrome, Proteinuria, Hypoalbuminemia, Hyperlipidemia.

INTRODUCTION

Idiopathic nephrotic syndrome is defined by the combination of a nephrotic syndrome (proteinuria, hypoalbuminemia, hyperlipidemia, and edema) and non-specific histological abnormalities of the kidney. The prevalence of NS is estimated at 2 to 7 per 100,000 children around the world. Children between 2 to 7 years of age get affected by this condition more often, particularly boys.[1]

Nephrotic syndrome can be caused by amyloidosis, minimal change disease (MCD), focal segmental glomerulosclerosis, diabetesrelated nephropathy, lupus, and membranous nephropathy in adults. The most noticeable symptom is swelling-especially around the eyes and face. Additional signs and symptoms include frothy urine, weight gain, fatigue, loss of appetite and infection due to loss of antibodies through urine.[2]

Nephrotic syndrome is characterized by proteinuria, hypoalbuminemia, Edema, Hyperlipidemia. Most cases in children occur before age 6, late on set nephrotic syndrome refers to cases that appear after age 10.[3] Late on set idiopathic nephrotic syndrome has three distinct histological varients – minimal change Nephrotic syndrome (MCNS), focal segmental glomerulosclerosis (FSGS), membranous nephropathy (MN).[4]

Steroid sensitivity is most often associated with minimal change histology and a more favorable long term prognosis, whereas steroid resistance is frequently associated with focal segmental glomerulosclerosis and carries high risk of end stage renal disease[2, 3]. The initial response to steroid therapy has been established as one of the important prognostic marker for children with NS.[4, 5] Complications of Nephrotic syndrome are-hematuria, hypertension, acute and chronic kidney disease, infection.

Advanced age at presentation was associated with increased incidence of hypertension hematuria, steroid resistance and progress to end stage renal disease.[7]

Aim of our study is to evaluate the presence of atypical features like hypertension, hematuria, and also to note the response to steroids, as these are increased in children with nephrotic syndrome when presented after age 10. Presence of these features leads to renal biopsy and management alterations.

OBJECTIVES

1. To study the clinical profile in late onset nephrotic syndrome in children.
2. To observe response to treatment in late onset nephrotic syndrome in children.

MATERIALS AND METHODS

Study Design: Retrospective study of hospital records.

Study Population: case sheets of children admitted in department of pediatrics, Government General Hospital, Kurnool with nephrotic children above the age of 10 years.

Study Period: January 2020 to December 2024.

Sample Size: All the cases admitted in department of pediatrics, Government General Hospital, Kurnool with nephrotic children above the age of 10 years.

Inclusion Criteria: Children aged more than 10 years at onset fulfilled in the diagnostic criteria for nephrotic syndrome.

Exclusion Criteria: Secondary causes of nephrotic syndrome. (eg:SLE, Hepatitis B/C, Diabetic mellitus)

Data Collection Procedure: Clinical data including age at onset, gender, presenting sympotoms, and laboratory findings will be recorded histopathlogical finding will be categorized. Treatment response to steroids will be evaluated.

Statistical Analysis: Data will be entered in Microsoft excel and analysis will be done using SPSS Version 23

Descriptive data is presented in percentages while inferential data with appropriate tests of significance.

RESULTS

The study included 108 children who were followed for a minimum duration of one year. The mean (±SD) age of the population was 13.3 (±1.4) years. The gender distribution of the population was males accounting for 66.7% and females at 33.3%. Hypertension was observed in 8(14.8%). Haematuria in microscopy was found in 28 (51.9%) study participants. Anti-Nuclear Antibodies investigation was done for 50 children and only 3(5.6%) were found to be positive. After the six weeks of steroidal therapy complete remission was found in 58 (53.7%) participants. No remission and partial remission were observed in 26 (24.1%) and 24 (22.2%) members of the study population respectively. Based on the response to steroidal therapy the participants were diagnostically classified as SRNS in 52 (48.1%) children followed SSNS in 50 (46.29%) children.

Table 1. Descriptive Analysis of Background Parameters in the Study Population (N=108).

Parameters	N (%)
Age in years (Mean ± SD)	13.3 ±1.4
Gender	
Male	72 (66.7%
Female	36 (33.3%)

Hypertension	
Yes	16 (14.8%
No	92 (85.2%
Hematuria in Microscopy	
Yes	56 (51.9%)
No	52 (48.1%)
Antinuclear Antibodies	
Positive	6 (5.6%)
Negative	94 (87.0%
Not done	8 (7.4%)

Table 2. Remission Status and Diagnosis Observed in the Study Population (N=108)

Remission status (N=108)	
Response	**N (%)**
Complete Remission	58 (53.7%)
No Remission	26 (24.1%)
Partial Remission	24 (22.2%)
Diagnosis	
LUPUS NEPHRITIS	2 (1.9%)
SDNS	4 (3.7%)
SRNS	52 (48.1%)
SSNS	50 (46.29%)
SDNS: Steroid Dependent Nephrotic Syndrome, SRNS: Steroid Resistant Nephrotic Syndrome, SSNS: Steroid Sensitive Nephrotic Syndrome	

Table 3. Renal Function Parameters Pre and Post Steroidal Therapy

Parameters	**Mean ± SD**	**Minimum**	**Maximum**
Pre -Albumin (g/dl)	2.1 ±0.9	1	4.6
Post_Albumin (g/dl)	3.7 ±1.0	1.2	5.1
Pre_Creatine (mg/dl)	0.8 ±0.8	0.25	6.18
Post_Creatine (mg/dl)	1.0 ±1.4	0.2	10.7
CHOLESTROL (mg/dl)	335.5 ±186.9	123	1100
Hemoglobin (g/dl)	12.1 ±1.5	8	14.7
Urine_Protein	8.0 ±8.3	0.03	54
Urine_Creatine	3.3 ±8.8	0.015	58

Table 4. Frequency Distribution of Biopsy Findings in the Study Population

Biopsy findings	
Parameters	**N (%)**
MESPGN: Mesangial Proliferative Glomerulonephritis	13 (24.1%)
FSGS	24 (22.2%)
MN	8 (7.4%)
DPGN	16 (15.8%)

MES HC	36 (33.3%)
MCNS	42 (38.9%)
FSPGN	2 (1.9%)
PROLIF GN IGG	2 (1.9%)
MPGN IGG,C3	2 (1.9%)

Table 5. Comparison of Response and Biopsy Background Parameters

Parameter	Response				P-Value
	Complete remission	No remission	Partial Remission	Total	
MESPGN	16 (61.5%)	2 (7.7%)	8 (30.8%)	26	0.263
FSGS	14 (58.3%)	8 (33.3%)	2 (8.3%)	24	0.377
MN	2 (25%)	6 (75%)	0 (0%)	8	0.043
DPGN	2 (25%)	8 (50%)	6 (37.5%)	16	0.037
MES HC	28 (77.8%)	4 (11.1%)	4 (11.1%)	36	0.043
MCNS	34 (85%)	4 (100%)	4(100%)	4	0.006
FSPGN	0 (0%)	0 (0%)	2 (100%)	2	0.168
PROLIF_GN_IGG	2 (100%)	0 (0%)	0 (0%)	2	0.645
MPGN_IGG_C3	0 (0%)	0 (0%)	2 (100%)	2	0.168
FSPGN_IF_FH	0 (0%)	0 (0%)	2 (100%)	2	0.168

MESPGN: Mesangial Proliferative Glomerulonephritis, FSGS: Focal Segmental Glomerulonephritis, MN: Membranous Nephropathy,
DPGN: Diffuse Proliferative Glomerulonephritis, MES HC: Mesangial Hypercellularity, MCNS: Minimal Change Nephrotic Syndrome.

Table 6. Comparison of Response and Diagnosis Background Parameters

Parameter	Response				P-Value
	Complete remission	No remission	Partial Remission	Total	
SRNS	12 (23.1%)	26 (50%)	14 (26.9%)	52	0.001
SSNS	42 (84%)	0 (0%)	8 (16%)	50	0.001
SDNS	4 (100%)	0 (0%)	0 (0%)	4	0.409
Lupus Nephritis	0 (0%)	0 (0%)	2 (100%)	2	0.618

SRNS: Steroid Resistant Nephrotic Syndrome, SSNS: Steroid Sensitive Nephrotic Syndrome, SDNS: Steroid Dependent Nephrotic Syndrome.

DISCUSSION

In this retrospective study we have assessed the clinical profile and outcome of Idiopathic late onset Nephrotic syndrome in children. All the children included were followed for a period of one year. For this study we have taken the 108 children, of age group 10-16 years of age. Most of the patients were diagnosed with SRNS and SSNS, few were diagnosed with SDNS and lupus nephritis. All the patients have undergone biopsy after the treatment, in which most of the patients were diagnosed with MCNS, MES HC, DPGN, FSGS and MESPGN.[6]

During the study we have observed high level of cholesterol and low level of urine creatinine in patients. During the study we have seen the improvement in albumin level after the treatment of 6 weeks. Most of the patients in our study were

not having hypertension and were having negative antinuclear antibodies.[7] As per the International Study of Kidney Disease in Children (ISKDC), most of the studies are going for outcomes which are classified on the basis of patient's response to corticosteroids or other therapy and by NS relapse. Few or no relapses are linked to the least severe SSNS course. Individuals with SSNS who experience relapses frequently or who become dependent on steroids are more complex. Few patients with initial steroid response develop SRNS (late non-responders) and others present with SRNS.[8]

At last, the most severe NS instances are resistant to corticosteroids as well as other treatments. Childhood NS can result in longterm renal problems, such as adult NS relapse, hypertension, chronic kidney disease (CKD), and end-stage renal disease (ESRD).[9] Hala Wannous et al, have reported the main indication of kidney biopsy was steroidresistant nephrotic syndrome (SRNS) and the main histopathological patterns were minimal change disease (MCD) and focal segmental glomerulosclerosis (FSGS). FSGS was the most common histopathological pattern in idiopathic SRNS and had the worst prognosis. Calcineurin inhibitors could be an effective therapy to induce complete remission in SRNS. In our study main indication of biopsy was SRNS and SSNS and the main histopathological patterns were MES HC, MCNS, MESPGN and FSGS.[10]

CONCLUSION

In our study most of the patients were diagnosed with SRNS and SSNS, and most common histological findings were MCNS, MES HC, FSGS and MESPGN. Late onset NS has higher frequency of atypical features, steroid resistance, and histopathology showing lesions other than MCD. Early biopsy may be useful guide to management.

REFERENCES

1. Nephrotic-syndrome-Children.pdf. [Cited 2024 Sep.3]. Available from: https://med.stanford.edu/content/dam/sm/pednephrology/ documents/secure/Nephrotic-syndromeChildren.pdf
2. Del Rio M , Kaskal F(2008) evaluation and management of steroid – unresponsive nephrotic syndrome. Curr Opin Pediatr 20: 151-156.
3. Mc B ryde K, Kershaw-D , SmoierW(2021) pediatric steroid resistance nephrotic syndrome. Curr Probl Pediatr Adolesc Health Care 31: 275-307. 4. Letavernier B, Letavernier E, Leroy S, at all. Prediction of high degree steroid dependency in pediatric idiophathic nephrotic syndrome. Pediatr Nephrol. 2008; 23::2221-6. DoI:10.1007/s00467008-0914-y.
5. Yap HK , Han EJ, Heng – CK, gong WK. risk factors for steroid dependency in children with idiophatic nephrotic syndrome. Pediatr Nephrol. 2001 ; 16:1049-52. doi:10.1007/s004670100024.
6. Nephrotic syndrome-Nephrotic syndrome - Symptoms &causes. Mayo Clinic [Internet]. [Cited 2024 Sep.2]. Available from: https://www.mayoclinic.org/diseasesconditions/nephroticsyndrome/symptoms-causes/syc-20375608
7. Chang – JW , TSAI HL , Wang HH , Yang LY . clinico pathological features and prognosis of Chinese children with idoiopathic nephrotic children between different age groups. Eur J Pediatr . 2009; 168: 1189 – 1194. Doi:10.1007/s00431-008-0902-z.
8. Niaudet P, Boyer O. Idiopathic Nephrotic Syndrome in Children: Clinical Aspects. In: Avner E, Harmon W, Niaudet P, Yoshikawa N, eds. Pediatric Nephrology: Sixth Completely Revised, Updated and Enlarged Edition. Springer; 2009:667-702. 9. Esezobor C, Ademola AD, Adetunji AE, Anigilaje EA, Batte A, Jiya-Bello FN, et al. Management of idiopathic childhood nephrotic syndrome in sub-Saharan Africa: Ibadan consensus statement. Kidney International. 2021;99(1):59-67. 10. Tapia C, Bashir K. Nephrotic Syndrome. In: StatPearls. StatPearls Publishing; 2024. [Cited 2024 Sep.2]. Available from: http://www.ncbi.nlm.nih.gov/books/NBK470444/.