

Epidemiological Study of Nephrotic Syndrome in Egyptian Children

(A Single Center Study)

Thesis

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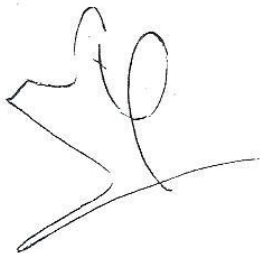
عنوان الرسالة:

دراسة خصائص المتلازمة النفروزية في الاطفال المصريين (دراسة من مركز واحد)

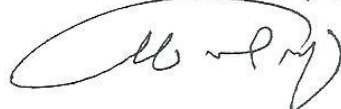
الملخص :

الهدف من هذه الدراسة كان لتحليل الخصائص والصفات الاكلينيكية والمعملية وسمات التكوين المرضي
النسجي للمتلازمة النفروزية في الاطفال المصريين الذين حضروا الي عيادة أمراض الكلي بمستشفى
الاطفال جامعة القاهرة خلال الفترة من يناير ٢٠١١ وحتى يناير ٢٠١٣.

١٥٠ ملف لحالات المتلازمة النفروزية تم اختيارهم عشوائيا وتم فحصهم استيعاديا. ناقشت الدراسة
النتائج المستخلصة من حيث السن و الجنس والاعراض الاكلينيكية و الاستجابة للعلاج بالكورتيزون
ومضاعفات المرض.



وترى اللجنة قبول البحث



Abstract

The aim of this study was to analyze the epidemiological, clinical, laboratory and histopathological characteristics of idiopathic nephrotic syndrome in Egyptian children attending the pediatric nephrology outpatient clinic of CUCH during the period from January 2011 to January 2013 and to determine their response to various therapeutic modalities and current outcome.

150 files of nephrotic syndrome patients were randomly selected and retrospectively reviewed; they included 92 males and 58 females with a male : female ratio of 1.6:1, the age of patients ranged between 1.3-15 years with a mean of 5.13 ± 2.92 years, the age at onset ranged between 0.7-12 years with a mean of 4.45 ± 2.49 years.

As regards to the clinical findings at presentation; 150 patients (100%) had puffiness of eye lids and lower limb edema, 101 patients (67.3%) had ascites, 64 patients (69.6% of the males) had scrotal edema, 3 patients (2%) had pleural effusion, 1 patient (0.7%) had pericardial effusion, 30 patients (20%) had hypertension, 2 patients (1.3%) had gross hematuria, 1 patient (0.7%) had mild hepatomegaly and 1 patient (0.7%) had mild splenomegaly. Outcome of NS patients according to steroid response was SRNS in 34 cases (22.7%) and SSNS in 116 Cases (77.3%) (62 cases of them were IR while 54 cases were FR). Renal biopsies were done in 39 cases where the most frequent pathological diagnosis was FSGS in 15 cases (38.4%). The commonest complication among our studied group was infection in 81 patients (54%). Among the patients who had infections, the most common pattern encountered was UTI which occurred in 56 patients (37.3%).

The treatment of NS in this study, all patients (150) (100%) took steroids, 37 patients (24.7%) took CPO (orally taken in 35 patients and intravenously taken in 2 patients), 31 patients (20.7%) took CSA, 12 patients (8%) took MMF, 10 patients (6.7%) took azathioprine and 40 patients (26.7%) took levamisole.

Key Words: Epidemiology – Nephrotic syndrome - Egyptian children.



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List of Abbreviations

ACEIs	: angiotensin converting enzyme inhibitors.
ACTN4	: α actinin 4.
AFP	: alpha fetoprotein.
ANA	: antinuclear antibody.
ARBs	: angiotensin II receptor blockers.
ARF	: acute renal failure.
AZA	: azathioprine
BMI	: body mass index.
BUN	: blood urea nitrogen.
C3b	: complement 3 b.
CD2AP	: CD2 adaptor protein.
CKD	: chronic kidney disease.
CNS	: congenital nephrotic syndrome.
CPO	: cyclophosphamide.
CR1	: complement receptor 1.
CSA	: cyclosporine A
DAG	: diacylglycerol.
DMP	: diffuse mesangial proliferative.
DMS	: diffuse mesangial sclerosis.
MCNS	: minimal change nephrotic syndrome
DN	: diabetic nephropathy.
ER	: endoplasmic reticulum.

List of Abbreviations

ERSD	: end stage renal disease.
ESRF	: end stage renal failure.
FPs	: foot processes.
FR	: frequent relapser
FRNS	: frequent relapser nephrotic syndrome.
FSGS	: focal segmental glomerulosclerosis.
FSGS NOS	: focal segmental glomerulosclerosis not otherwise specified.
GBM	: glomerular basement membrane.
GCW	: glomerular capillary wall.
GFB	: glomerular filtration barrier.
GFR	: glomerular filtration rate.
GLEPP-1	: glomerular epithelial protein-1.
HBV	: hepatitis b virus.
HDL	: high density lipoprotein.
HLA	: human leucocyte antigen.
HMG-COA	: hydroxy-methyl-glutaryl coA.
IGAN	: immunoglobulin A nephropathy.
INF2	: member of the formin family of actin-regulating proteins.
INS	: idiopathic nephrotic syndrome.
IP3	: inositol 1,4,5-triphosphate.
IR	: infrequent relapser

List of Abbreviations

ISKDC	: international study of kidney disease in children.
LDL	: low density lipoprotein.
LM	: laminin.
LN	: lupus nephritis.
LP	: lipoprotein.
MCD	: minimal change disease.
Mes-PGN	: mesangio-proliferative glomerulonephritis.
MGN	: membranous glomerulonephritis.
MMF	: Mycophenolate mofetil.
MN	: membranous nephropathy.
MPGN	: membranoproliferative glomerulonephritis.
NPHS1	: gene encoding nephrin.
NPHS2	: gene encoding podocin.
NPS	: nail-patella syndrome.
NS	: nephrotic syndrome.
NSAIDs	: nonsteroidal anti-inflammatory drugs.
ORG	: obesity-related glomerulopathy.
PCNA	: proliferative cell nuclear antigen.
PF	: permeability factor.
PGN	: proliferative glomerulonephritis.
PI3K	: phosphatidylinositol 3-kinase.
PKC	: protein kinase c.
PLCϵ1 (NPHS3)	: phospholipase c-epsilon-1.

List of Abbreviations

RAAs	: renin angiotensin aldosterone system.
RCTs	: randomized controlled trials.
ROS	: reactive oxygen species.
SD	: slit-diaphragm.
SDNS	: steroid-dependent nephrotic syndrome.
SGP	: sialoglycoprotein.
SLE	: systemic lupus erythematosus.
SNS	: sympathetic nervous system.
SRNS	: steroid-resistant nephrotic syndrome.
SSNS	: steroid-sensitive nephrotic syndrome.
TPA	: tissue plasminogen activation
TRPC6	: transient receptor potential cation channel 6.
URTI	: upper respiratory tract infection
UTI	: urinary tract infection
VEGF	: vascular endothelial growth factor.
WASP	: Wiscott-Aldrich syndrome protein.
WT-I	: Wilms tumor-1.
ZO-I	: zonula occludens-1.

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Introduction and Aim of the Work



Introduction and Aim of Work

Childhood NS is one of the most common pediatric kidney diseases (**Kerlin et al., 2014**).

The incidence and the histologic pattern of NS are affected by geographic location and ethnic origin (**Gbadegesing and Smoyer, 2008**).

The most common form of NS in childhood is primary idiopathic NS (INS). Minimal change nephrosis is the main pediatric form of INS and is usually a benign condition responsive to standard steroids treatment (**Alsalloum et al., 2012**).

Recent studies have reported an increasing incidence of focal segmental glomerulosclerosis (FSGS) during childhood where the prognosis is bad, with the majority of cases evolving to terminal renal insufficiency within several years (**Reusz et al., 2006**).

The Underlying histopathology usually affects the course of the disease and the response to treatment (**Kari et al., 2009**).

Aim of the work:

To study the pattern of idiopathic nephrotic syndrome in Egyptian children attending the pediatric nephrology outpatient clinic of Cairo university children's hospital (Abo-Elrish) during the period from January 2011 to January 2013, to document the epidemiologic as well as certain clinic-pathological characteristics of the disease in Egyptian children and to determine the long term effects of various therapeutic modalities used during the course of treatment.