



Nephrotic Syndrome

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Outline

- Introduction
- Nephrotic syndrome classifications
- Signs and symptoms
- Diagnoses
- Management
- Complications
- Monitoring
- Case presentation

Introduction

- Nephrotic syndrome (NS) is caused by renal diseases that increase the permeability of the glomerular filtration barrier without compromising GFR.
- Occurs at all ages but is most prevalent in children between the ages 1.5-6 years
- It affects more boys than girls, 2:1 ratio
- Most studies put the incidence at 2-7 per 100,000 population.

Primary nephrotic syndrome

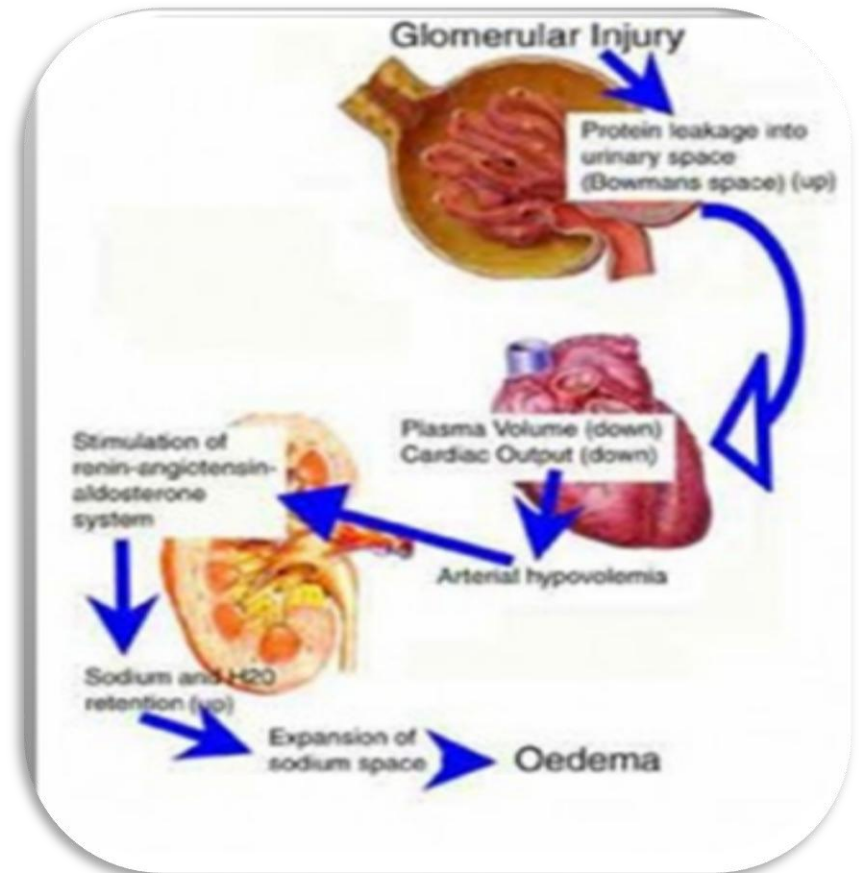
- Minimal Change Nephrotic Syndrome (MCNS)
- Focal Segmental Glomerulosclerosis (FSGS)
- Membranoproliferative Glomerulonephritis (MGPN)
- Congenital nephrotic syndrome (Cong NS)

Secondary nephrotic syndrome

- SLE
- Infection
- HIV
- Hepatitis B and C
- Malaria
- Syphilis
- Drug exposure
- NSAID
- Malignancy (Rare in children)
- Diabetes is not a cause of NS in children due to long latency

Signs and symptoms

- Proteinuria
- Hypoalbuminemia
- Hypercholesterolemia
- Edema



Edema

- Edema is the primary presenting feature. History can also include weight gain, poor urine output, dizziness, or discomfort as a result of the edema (including abdominal pain).

Degree of edema ranges from:

- Mild
- Moderate with peripheral pitting edema of the limbs.
- Severe with gross limb edema, ascites and pleural effusions.

Diagnosis

- Urinalysis
- Urine protein quantification
- Serum albumin
- Lipid panel
- CBC
- Electrolytes
- C3, C4
- ANA

In selected patients

- Genetic studies
- Kidney ultrasonography
- Chest radiography
- Kidney biopsy

Management

Steroids:

- To induce remission, followed by a slow wean to reduce risk of relapse.
- Most patients who respond to steroid therapy will do so within the first four weeks of therapy.

Management

- **Steroid-sensitive NS:** Almost all patients with steroid-sensitive NS have an excellent outcome with few patients developing end-stage renal disease or chronic kidney disease.
- **Steroid-resistant NS:** Patients who fail an initial course of steroid therapy should undergo renal biopsy to determine the underlying diagnosis to guide further therapeutic choices.

Relapse

A relapse is defined as proteinuria 3+ or 4+ for 3 consecutive days, and should prompt re-introduction of full dose prednisolone

- **Frequent relapses:** 2 or more in first 6 months or 4 or more in any 12 month period.
- **Infrequent relapses:** 1 relapse in 6 months or 1–3 relapses in 12 months.

Management

Nonsteroidal therapy:

- In children with significant side effects of steroid therapy, other steroid-sparing agents that prolong remissions and reduce the dose of steroids should be considered.
- These drugs include alkylating agents, levamisole, cyclosporine, mycophenolatemofetil, and rituximab.

Management

Diuretics:

- Loop diuretics, such as furosemide may improve edema.
- Metolazone may be beneficial in combination with furosemide for resistant edema.

Management

Antihypertensive agents:

- Angiotensin-converting enzyme (ACE) inhibitors and angiotensin II receptor blockers (ARBs) can reduce hypertension and may also contribute to reducing proteinuria.
- Calcium channel blockers and beta blockers may also be used as first-line agents for hypertension.

Management

- Statins may be used for managing hyperlipidemia.
- Pneumococcal vaccine and Flu Vaccine.
- Calcium and vitamin D

Complications

- Infection including peritonitis
- Thromboembolism
- Renal insufficiency
- Hypovolemia
- Impaired growth

Monitoring

- Ongoing monitoring for proteinuria is required to detect relapses early, and initiate therapy to prevent significant fluid accumulation (edema) and minimize the complications.
- Patients and their parents are taught to routinely measure body weight.



CASE PRESENTATION

General data

- BK is a 4 month Saudi male. His weight is 2.7 kg and his height is 41 cm.
- He has no known allergies.
- His parents are first degree cousins with no renal diseases history.

Chief complaint

- Generalized edema

History of present illness

- He is a known case of congenital nephrotic syndrome.
- After birth, he was in NICU at another hospital for about one month on albumin then discharged home.
- At age of 2 months, he went back to ICU and then to the ward but family discharged him from that hospital and come here for further investigations.

Past medication history

- Thyroxine
- Vitamine D
- Ferrous sulfate
- Folic acid

Physical examination

- General: Conscious, alert, pale with puffy face
- Chest: Normal with good equal air entry
- CVS: S1+S2
- Abdomen: Distended with positive shifting dullness
- Others: Edema

Vital sign

- Temp: 36.4
- Blood pressure: 90\47
- Heart rate: 125
- Respiratory rate: 60
- O2 saturation: 100% on room air

Lab results

WBC	8.81	cl	100
Hbg	9	PO4	1.31
Plts	549	HCo3	22.8
Cr	9	Mg	0.80
Urea	2.9	Albumin	15
Na	133	Protein	28
K	4.7	Glucose	1.6

Problem list

- Hypoglycemia
- Hyponatremia
- Anemia
- Hypothyroidism
- Congenital nephrotic syndrome

Plan

- 10% glucose 20 ml over 30 min.
- Lasix 5 mg IV OD
- 20% albumin 5 g IV OD
- NCI 10 ml PO BID
- Iron syrup 20 mg PO OD
- Folic acid 1 mg PO OD
- Thyroxine 25 mcg PO OD
- Vitamine D 4 drops PO OD
- Multivitamine syrup 0.5 ml PO OD
- Order thyroid function test and urine dipstick

Day 2-5

Subjective	Objective	Assessment	Plan
<p>Edema with increased weight</p>	<p>Wt. 2.82 Temp. 36.9 BP 106\59 HR 125 RR 52 O2 saturation 99% Hbg 7.5 Hco3 30.8 Mg 0.60 Cholesterol 3.8 Triglyceride 4.5 LDL 2.3 HDL 0.6 TSH 24.4 T4 5.6</p> <p>Urine depstick RBC -ve Protein +3 I\O 380\220</p>	<p>The patient's edema is not improving. He also has proteinuria</p>	<ul style="list-style-type: none"> Start enalapril 0.5mg PO OD Mg sulfate 200mg PO BID Increase the frequency of lasix to BID Order abdomen ultrasound Continue on the other meds

Day 6-10

Subjective	Objective	Assessment	Plan
<ul style="list-style-type: none"> • Vomiting • Oxygen desaturation • Edema 	<p>Wt. 3.23 Temp. 37 BP 92\59 HR 130 RR 40 O2 saturation 83% on RM, 100% on NC Hbg 6.50 Hco3 34 Albumin 8 K 3 I\O 265\430 Ultrasound shows both kidneys are echogenic and moderate ascites is seen</p>	<p>The patient is oxygen dependent and he is not improving. His abdomen still distended</p>	<ul style="list-style-type: none"> • PRBC • Start KCl 2 ml PO BID • Omeprazole 4mg PO OD • Domperidone 0.8mg PO OD • Continue on the other meds

Day 11-17

Subjective	Objective	Assessment	Plan
Edema	Wt. 3.8 Temp. 36.5 BP 92\59 HR 130 RR 40 O2 sat. 98% on NC Hbg 9 Hco3 31.5 Albumin 9 K 3.6 I\O 300\245 Ultrasound shows stable ascites without any significant changes	The patient condition is not getting better. He has minimal response to lasix	<ul style="list-style-type: none">• Start metolazone 0.4 mg PO BID• Continue on the other meds• Order iron study• Order thyroid function test

Day 18-25

Subjective	Objective	Assessment	Plan
<p>Slightly improving edema and abdomen distention</p>	<p>Temp. 36.9 BP 116\51 HR 138 RR 40 O2 sat. 98% on NC RBC 2.77 Hbg 8 MCV 87 Iron 5.2 Unsaturated iron binding capacity 11 Ferritin 1282 TSH 18.780 T4 5.2 I\O 340\360</p>	<p>The abdomen distention and edema clinically slightly improved but still need further management. He is not responding to oral iron.</p>	<ul style="list-style-type: none"> • IV iron • Increase thyroxin dose • Increase metolazone dose • Consider adding spironolactone if there is hypokalemia

Medication list

- Lasix 5 mg IV BID
- 20% albumin 5 g IV BID
- Metolazone 0.5 mg PO BID
- NCI 10 ml PO BID
- Iron IV 25 mg
- Folic acid 1 mg PO OD
- Thyroxine 37 mcg PO OD
- Vitamine D 4 drops PO OD
- Multivitamine syrup 0.5 ml PO OD
- Mg sulfate 200 mg PO BID
- KCl 2 ml PO BID
- Omeprazole 4mg PO OD
- Domperidone 0.8 mg PO OD

THANK YOU!

