NEPHROTIC SYNDROME – AT A GLANCE

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INTRODUCTION
Nephrotic Syndrome is the clinical entity characterised by tetrad of signs & symptom namely heavy proteinuria (>40mg/m²/hr), hypoalbuminemia (serum albumin < 2.5g/dl), hyperlipidemia (serum cholesterol >200mg/dl) and oedema. It has been seen that child below 6yr are mostly affected. Histopathologically in children nephrotic syndrome is usually idiopathic with minimal change disease (MCD) as the most common followed by MCD variant (Mesangial Proliferative variant) and F.S.G.S (focal segmental glomerulosclerosis). About 80% of nephrotic syndrome do respond to steroid. 70% shows relapse and good percentage become frequent relapses (≥2 relapse in 6 months or ≥ 4in 12 months) or steroid dependent (≥2relapse within 2 weeks of stopping steroid or while tapering steroid dose. Relapse is diagnosed by urinary dipstick of 3+ or more of proteinuria for 3 or more days remission despite 4 weeks of full dose steroid & has been shown to predict worse prognosis.

PATHOGENESIS
As we know that podocyte are terminally differentiated epithelial cells which form part of glomerular capillary wall. The podocyte are attached to the basement membrane with foot processes and their architecture is maintained by an actin cytoskeleton. The defect in podocyte is considered as a primary cause responsible for nephrotic syndrome. Now, proteinuria in nephrotic syndrome is due to T. cell dysfunction that results in the production of glomerular capillary wall & cause proteinuria. Interleukin 13 and CD 80 cause injury to podocyte leading to proteinuria.

CAUSES
A. Idiopathic Nephrotic Syndrome
• Minimal change nephrotic syndrome 85%
• Mesangial Proliferative glomerulonephritis– 5%
• Focal segmental glomerulosclerosis
B. Secondary Nephrotic Syndrome
• Chronic glomerulonephritis-MPGN, Membrane Glomerulonephritis
• Hereditary Nephropathies:- Polycystic dis
• Collagen disorders:- SLE,HSP
• Tumors:- Leukemia, Lymphoma, Renal tumor
• Toxic:- Insect bite, heavy metal poisoning
• Metabolic:- Diabetic, Amyloidosis
• Congenital nephrotic syndrome

CLINICAL FEATURE
Edema is the cardinal feature of nephrotic syndrome which is typically more prominent in periorbital region and during morning hours in early stages. Gradually, it progress to a stages of generalised anasarca with marked scrotal and abdominal wall oedema. Pleural effusion may present in severe cases.
• Acute abdominal pain and diarrhea is common due to mesentric and intestinal wall oedema
• Oliguria is common in MCNS (Minimal Change nephrotic Syndrome) due to intravascular volume depletion.

EVALUATION
• Blood pressure Measurement
• Look for associated defection eg fever rash, joint pain, hepatomegaly
• Nephrotic range proteinuria is defined as urinary protein 3+ or 4+ by dip stick or spot urine protein to creatinine ratio ≥ 2 or urine protein excretion of > 40 mg /m2 / hr .

RECOMMENDED INITIAL INVESTIGATION
• R F T
• Sr. Cholesterol
• Serum complement C3
TERMINOLOGY IN NEPHROTIC SYNDROME

- Remission: Urine Albumin trace or nil or Proteinuria < 4 mg/m2/hr for 3 consecutive days.
- Relapse: Urine Albumin 3+ or 4+ or proteinuria > 4mg/m2/hr for 3 Consecutive days.
- Frequent Relapse: Two or more relapse in 6 month or initial episode or >3 relapse in 12 month.
- Steroid dependence: 2 Consecutive relapse when in alternate day steroid or within 14 days of its disease continuation.
- Steroid resistance: No remission despite therapy 4 weeks of daily prednisone in close of 2mg/kg/day.

TREATMENT

(1) Initial Episode of Nephrotic Syndrome

- Prednisone at dose 2 mg/kg/day (max dose 60 mg) in single dose or divided dose daily for 6 weeks followed by 1.5mg/kg (max 40mg) as a single dose on alternate days for next 6 weeks.
- As per Cochrane renal group, the duration of initial prednisone therapy should be for 12 weeks.

(2) Treatment of Relapse

- Treatment of infection result in spontaneous remission and they don’t require steroid therapy.
- 2 In sustained relapse prednisone initiated with 2mg/kg/day till urine, is negative or nil for protein 3 consecutive days subsequently 1.5 mg/kg in alternate day for 4 weeks is given. Total duration of treatment = 6 weeks
- If frequent relapse or steroid dependent we can try low dose alternate day steroid or can give levomisole (2-2.5 mg/kg alternate day for 18-24 months with prednisone 1.5 mg/kg/alternate day for 2 – 4 weeks them reduced gradually by 0.25 mg/kg/every 4-6 weeks or maintenance dose 0.25mg/kg/alternate day for 6 – 12 months.
- In case of steroid resistance calcineurim inhibitors are drug of choice.
- Renal biopsy - (Patient with Atypical feature) - Age of Patient <14r
- Persistent [low serum complement] Gross hematuria or Persistent Hypertension those with steroid resistance.

REFERENCES