Nephrotic syndrome: first presentation

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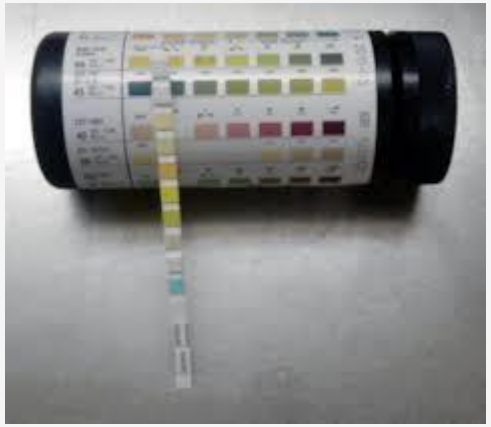
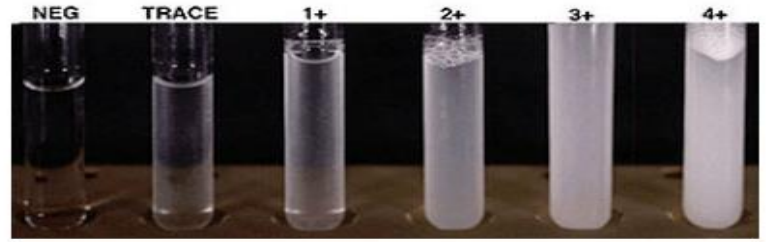
Nephrotic syndrome..

- Proteinuria
- Edema
- Hypoalbuminemia (serum albumin <2.5 g/dl)
- Hypercholesterolemia (serum cholesterol >200 mg/dl)

Nephrotic range proteinuria??



- Urine protein > 3g/day
- 24 hour urine protein:
  - >40mg/m2/hour
  - 50mg/ kg/ day



- Spot urine protein creatinine ratio >2mg/mg
- Urine dipstick 3+ (300mg/dl), 4+

Etiology

Nephrotic Syndrome

Idiopathic

Infections:
Post streptococcal, hepatitis B, HIV, malaria, schiztosomiasis

Vasculitis:
HSP, SLE

Hematological:
Leukemia, lymphoma

Drugs:
lithium, penicillamine

History

- Cardinal symptoms:
  - swelling and its progression, exclude cardiac and hepatic causes
  - Decreased urine, hematuria
  - UTI

- History of etiology
  - Preceding viral illness
  - Sore throat, skin infections
  - Rash, joint swelling
  - Medications
  - Weight loss
  - Allergies

History

- History of complication
  - Abdominal pain
  - Persistent vomiting, dizziness
  - infections
- History of treatment taken
- Family history
- Immunisation

Examination

- Vitals: pulse rate, peripheral pulses, BP (orthostatic BP), capillary refill
- Anthropometry
- Extent of edema
- Lymphadenopathy
- Skin: rash, cellulitis
- Joints
- Respiratory: pleural effusion, pneumonia
- Abdo: ascites, signs of peritonitis
- CVS: murmur

Investigations

- CBC
- Urine routine
- Urine protein/creatinine ratio
  or
- 24-hour urine protein
- Serum albumin
- Serum cholesterol
- Blood urea/serum creatinine
- Xray chest, Mantoux test

If indicated

- Serum electrolytes Na, K
- USG KUB
- Complement C3 levels
- ASO titre
- ANA
- Hep B, Hep C, HIV- high risk

Investigations

- **Diagnosis and severity of nephrotic syndrome**:
  - Urine protein/creatinine ratio or 24-hour urine protein;
  - Serum albumin
  - Serum cholesterol
- **Acute kidney injury**
  - Serum creatinine/ BUN
  - Na/K
- **USG KUB**
  - Structural anomaly
- **Intravascular volume status:**
  - Hematocrit
  - Fractional excretion of sodium
- **Infection:**
  - Leucocytosis
  - Urine routine
  - Chest Xray, MT
- **Secondary causes (if indicated):**
  - Complement C3 levels
  - ASO titre
  - ANA
  - Hep B, Hep C, HIV- high risk

Early renal biopsy

(i) age at onset <1yr

(ii) gross hematuria, persistent microscopic hematuria or low serum C3

(iii) renal failure, not attributable to hypovolemia

(iv) suspected secondary causes

(v) sustained severe hypertension.

When to hospitalize?

- Severe edema compromising ambulation or respiratory compromise
- Unstable vital signs
- Urine output less than 0.5ml/kg/hr
- Severe hemoconcentration
- Severe infections requiring iv antibiotics and monitoring
- Suspicion or evidence of thromboembolism

What to monitor?

- Pulse
- Respiratory rate
- Blood pressure
- Daily Weight
- Abdominal girth
- Input/ output
- Spot Urine albumin

Management

Supportive
- Fluids
- Diet
- Edema control

Medications
- Corticosteroids

Management of complications

Patient and parent education

Supportive

- Fluid restriction: only if child is oliguric

- Dietary salt restriction **(NO ADDED SALT):**

<5 years- 1-1.5g/day sodium, school aged- 2g/ day
(1 teaspoon salt = 2.3g sodium)

- Adequate protein intake **(not high protein diet)**

- Not more than 30% calories should be derived from fat and saturated fats avoided.

- Can attend school and have full activity

Medical treatment of the first episode

- **Why is it important?**
- Appropriate treatment of the first episode plays an important role in long term outcome

- **Which?**
- Corticosteroids: Prednisone- mainstay of treatment
- dexamethasone, betamethasone, triamcinolone or hydrocortisone: **not recommended**

Prednisone induction regime

- **Duration**
- 12 weeks (6+6)

- **Dose**
- Prednisone 60mg/m^2 (2mg/kg), max 60mg, daily for 6 weeks followed by 40mg/m^2 (1.5mg/kg) on alternate days for 6 weeks
- Single daily or divided doses
- After meals

Side effects of Corticosteroids

- Short term
  - Nausea, vomiting
  - Behavioral changes
  - Cushingoid facies
  - Increased appetite, weight gain
  - Increased hair
  - Gastritis
  - suppression of the hypothalamic pituitary axis (>2 weeks high dose prednisone)
- Moderate to Long term
  - Hypertension
  - Glucose intolerance
  - Cataract
  - Muscle weakness
  - Bone health
  - Skin changes

Response to corticosteroids

- 80-85% will respond to corticosteroid therapy
- Proteinuria resolves in 50% by 2 weeks and in 90% by 4 weeks
- 60% will have relapses

Patient and Parent Education

- Adequate information: disease, associated complications, expected course
- Parental motivation and involvement
- Urine examination for protein at home using a dipstick.
- Maintain a diary
- Ensure normal activity and school attendance
- Infections, immunisations

Take home messages

- Prompt diagnosis
- Send a urinanalysis when in doubt
- Complete examination and look for secondary causes
- Prednisone: correct dosing and duration
- Parent education

To be covered in future topics

- Pathology
- Pathogenesis
- Infrequent/ frequent relapses/ steroid dependent
- Complications and management
- Immunisation in children with nephrotic syndrome

Referral to Pediatric Nephrologist

- Onset below 1-year of age; family history of nephrotic syndrome

- Nephrotic syndrome with hypertension, gross/persistent microscopic hematuria, impaired renal function, or extrarenal features (e.g., arthritis, serositis, rash)

- Complications: refractory edema, thrombosis, severe infections, steroid toxicity

- Resistance to steroid therapy

- Frequently relapsing or steroid dependent nephrotic syndrome