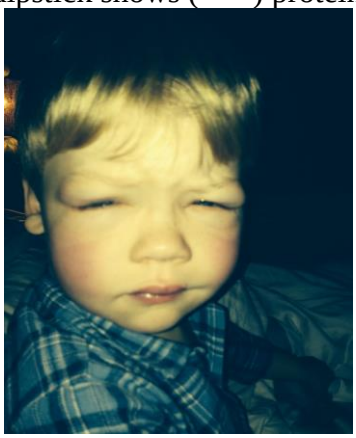
- **Case 1**: a 3 years old boy presents with puffy eyes (not responding to anti-histamine), abdominal swelling and urine dipstick shows (+++) protein.



- **What is your diagnosis?**
  - ✓ Nehprotic syndrome. The criteria which are needed to diagnose nephrotic syndrome are:
    - ❖ Heavy proteinuria (+++) on dipstick or protein:creatinine ratio > 200 mg/mmol.
    - ❖ Hypoalbuminemia < 25 g/L
    - ❖ Edema (especially peri-orbital).
    - ❖ Hypercholesterolemia.
- **What is the pathophysiology?**
  - ✓ The basic physiologic defect is a loss of the normal charge and size selective glomerular barrier to the filtration of plasma proteins.
  - ✓ Excessive urinary protein losses lead to the hypoalbuminemia of nephrotic syndrome.
  - ✓ Hypercholesterolemia is a consequence of hypoalbuminemia.
- **Epidemiology:**
  - ✓ Male:female ratio is 2:1
  - ✓ Commonly between ages of 2-5 years.
  - ✓ More common in southeast Asia and among Arabs.
- **Causes:**
  - ✓ Primary nephrotic syndrome: minimal change disease (90% of all childhood cases), Focal Segmental Glomerulosclerosis (FSGS), Membranoproliferative Glomerulonephritis (MPGN) and membranous nephropathy.
  - ✓ Secondary nephrotic syndrome: SLE, infections, obesity, drug exposure, Henoch-Schonlein purpura and malignancy (which is rare in children).
- **Investigations which you must order are:**
  - ✓ CBC.
  - ✓ Renal function test (urea and creatinine).
  - ✓ Liver Function Test (for albumin).
  - ✓ Serum electrolytes.
  - ✓ Urinalysis and protein:creatinine ratio (done in the morning).
  - ✓ Complement C3 and C4.
  - ✓ ANA and anti-dsDNA
  - ✓ HBV, HCV

- **Treatment:**
  - ✓ Corticosteroids (e.g. prednisolone). Adverse effects of steroids include: osteoporosis and growth retardation, immunosupression, diabetes mellitus, hypertension, cushingoid appearance, abdominal striae, acne and hirsutism.
    - ❖ *When do you expect to see a response?*
      - ➢ Within 2 weeks there will be negative proteinuria.
      - ➢ If there is no response after 1 month of treatment, 3 daily pulses of IV methylprednisolone will be given.
      - ➢ If still there is no response, a biopsy is indicated.
    - ❖ *Steroid-dependent nephrotic syndrome*: means that patients relapse whenever steroids are stopped.
  - ✓ Salt restriction.
  - ✓ Gentle fluid restriction.
  - ✓ Diuretics (if severe edema is present).
- **Complications of nephrotic syndrome:**
  - ✓ Hypovolemia (due to shifting of fluids).
  - ✓ Immunodeficiency.
  - ✓ Pro-thrombotic state (due to loss of anti-thrombin III in the urine).

- **Acute glomerulonephritis:**
  - **It is characterized by**: hematuria, proteinuria, hypertension and renal impairment (↑ urea and creatinine).
  - **Etiology**: deposition of immune-complexes in glomeruli.
  - **Clinical classification:**
    - ✓ Acute and sudden.
    - ✓ Chronic.
    - ✓ Recurrent.
    - ✓ Rapidly progressive.
  - **Histopathology classification:**
    - ✓ Post-streptococcal glomerulonephritis (representing 80% of cases in childhood):
      - ❖ Commonly preceded by pharyngitis or skin infection with group A β-hemolytic streptococci 1-2 weeks before.
      - ❖ Clinical manifestations: cola-colored urine, oliguria, proteinuria, hypertension and edema.
      - ❖ Investigations: ↑ASO titer, ↑ blood pressure, urinalysis showing RBC casts, throat swab is obtained for culture, ↓C3 (which will return to normal value after 6-8 weeks; if it remains decreased → your differential diagnosis is SLE or MPGN).
      - ❖ Treatment: diuretics, salt and water restriction and antibiotics (controversial).
    - ✓ Alport syndrome.
    - ✓ IgA nephropathy (more common in adolescents).
  - **Indications for biopsy for any glomerulonephritis:**
    - ✓ Rapidly Progressive Glomerulonephritis (RPGN).
    - ✓ Abnormal creatinine at 6 weeks.
    - ✓ ↓C3 for more than 3 months.
    - ✓ Proteinuria for more than 6 months.