Nephrotic Syndrome

This condition is COMPLETELY different in pathophysiology and clinical manifestation – apart from oedema – to acute nephritis. Managing one as the other may lead to death.

Clinical features
Characterised by oedema and the triad:
1) Proteinuria (4+ on dipstix)
2) Hypoalbuminaemia
3) Hypercholesterolaemia

Important things to look for on clinical examination
- Ascites
- Pleural effusions
- Evidence of infection (remember peritonitis)
- Evidence of other disease process (which may indicate the underlying aetiology e.g. SLE)
- Hypertension

Admit all cases of suspected nephrotic syndrome

Investigations
You WILL miss the diagnosis of nephrotic syndrome if you base the diagnosis on 4+ proteinuria on dipstix. In cases where HYPOALBUMINAEMIA is extreme, there is no more albumin to leak out. ALL children presenting with oedema should have their albumin levels checked

1) Make the diagnosis of nephrotic syndrome
   - Do a urine protein:creatinine ratio on the admission urine sample or the next morning, and repeat at least once
     - > 200mg protein per 1mmol creatinine is diagnostic (make sure you get the units right: protein in milligrams and creatinine in millimoles)
   - Total protein and albumin
   - Cholesterol
   - Complement

2) Determine renal function
   - U&E and creatinine

3) Find a cause
   - Infections: Hepatitis B, VDRL, ASOT, AntiDNAse B, HIV
   - Other: C3, C4, ANA

4) Rule out a urinary tract infection
   - Urine M,C&S (expect hyaline and granular casts)

5) Rule out tuberculosis (in case steroids are indicated)
   - Household contact, chest X-ray and mantoux

6) Consult a renal service (IALCH and Grey's) for further management plans
   - Do this soon after presentation for appropriate care planning
Immediate management in the ward while initiating above steps

- Daily urine dipstick for protein
- Salt restriction (no added salt to food) while oedema is present
- Prophylactic penicillin VK 250mg 12 hourly

**Diuretics and fluid restriction are NOT part of the management and in a child who is already intravascularly depleted. This may have dire consequences.**

- Indications for 20% ALBUMIN 15mg/kg over 4 hours and FUROSEMIDE 1mg/kg added (use only in consultation with a paediatrician):
  - Severe peri-orbital oedema leading to impaired vision
  - Hypoperfusion i.e. cold peripheries, weak pulses, hypotension
  - Severe ascites compromising diaphragmatic function
  - Severe uncomfortable scrotal oedema
- Steroids can be started if:
  - The child is >1 year old and < 10 years
  - No hypertension
  - No renal failure
  - No macroscopic haematuria
  - No evidence of other disease (Hep B/SLE/Syphilis/TB/HIV)
  - Complement levels are normal
- Steroid regimen for initial episode:
  - PREDNISONE 2mg/kg 24H PO for 6 weeks then, 2mg/kg PO on alternate days for 4 weeks, then taper over 2 weeks
- Prevent thrombotic complications:
  - Give ASPIRIN 75mg – 150 mg 24H PO until in remission

Arrange a Greys Hospital Renal Clinic consultation urgently. The child will be referred to a to Nephrologist in Durban if:

- The child did not fit the criteria for starting steroids (as above)
- Steroid resistance (ongoing proteinuria after 6 weeks of prednisone)
- Steroid dependency (recurrence of proteinuria when tapering)
- Frequent relapses
- Unacceptable steroid side effects
- Hypocomplementaemia
- Secondary forms of nephritic syndrome other than congenital syphilis
- Impaired renal function
- Persistent hypertension
- Congenital or infantile nephritic syndrome
- First presentation > 10 years of age

**If a renal biopsy becomes necessary, this should only be done by an individual/in a centre where renal biopsies are performed regularly. DON’T dabble in renal biopsies.**