MMed and DCH Lectures

Weekly by Zoom

Prof Trevor Duke

MMed and DCH Lectures

Common kidney diseases in children

June 1, 2020

Prof Trevor Duke

Common kidney diseases in children

1. Nephrotic syndrome
2. Post-streptococcal glomerulonephritis
3. Chronic kidney disease

Nephrotic syndrome

- Heavy proteinuria, hypoalbuminaemia and oedema
- Most common is idiopathic (90%)
- SLE, Henoch-Schonlein purpura
- Key acute complications are hypovolemia, infection and thrombosis
- 80-90% steroid responsive, 80% will have one or more relapses

Oedema – differential Dx

Hypoalbuminaemia

- Inadequate intake - kwashiorkor
- Increased losses
  - Kidney: nephrotic syndrome
  - GI tract – protein losing enteropathy, worm infestation
- Inadequate production
  - Liver failure

Fluid overload

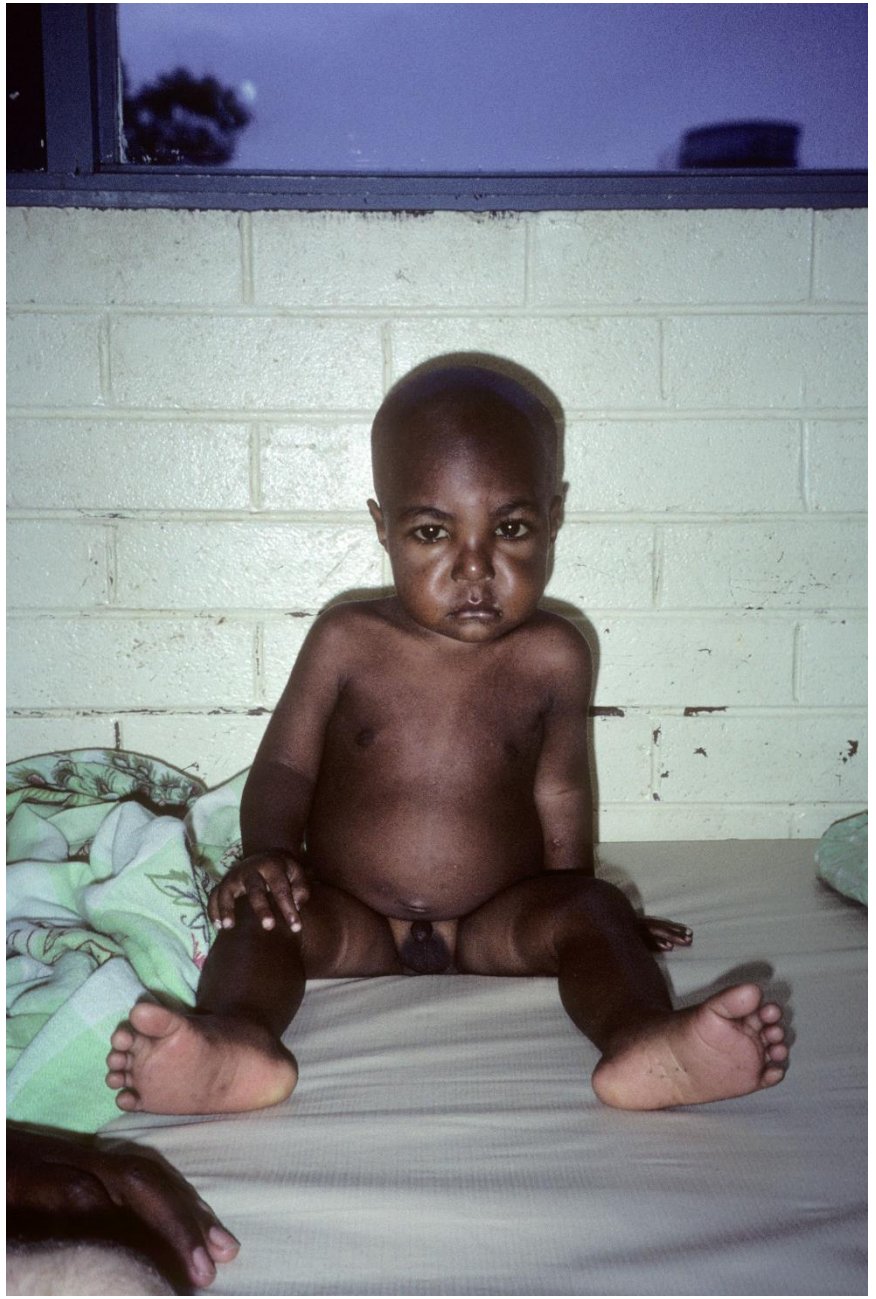
  - Anaemia
  - Congestive heart failure
  - Iatrogenic

Lymphatic obstruction

  - Tuberculosis, other

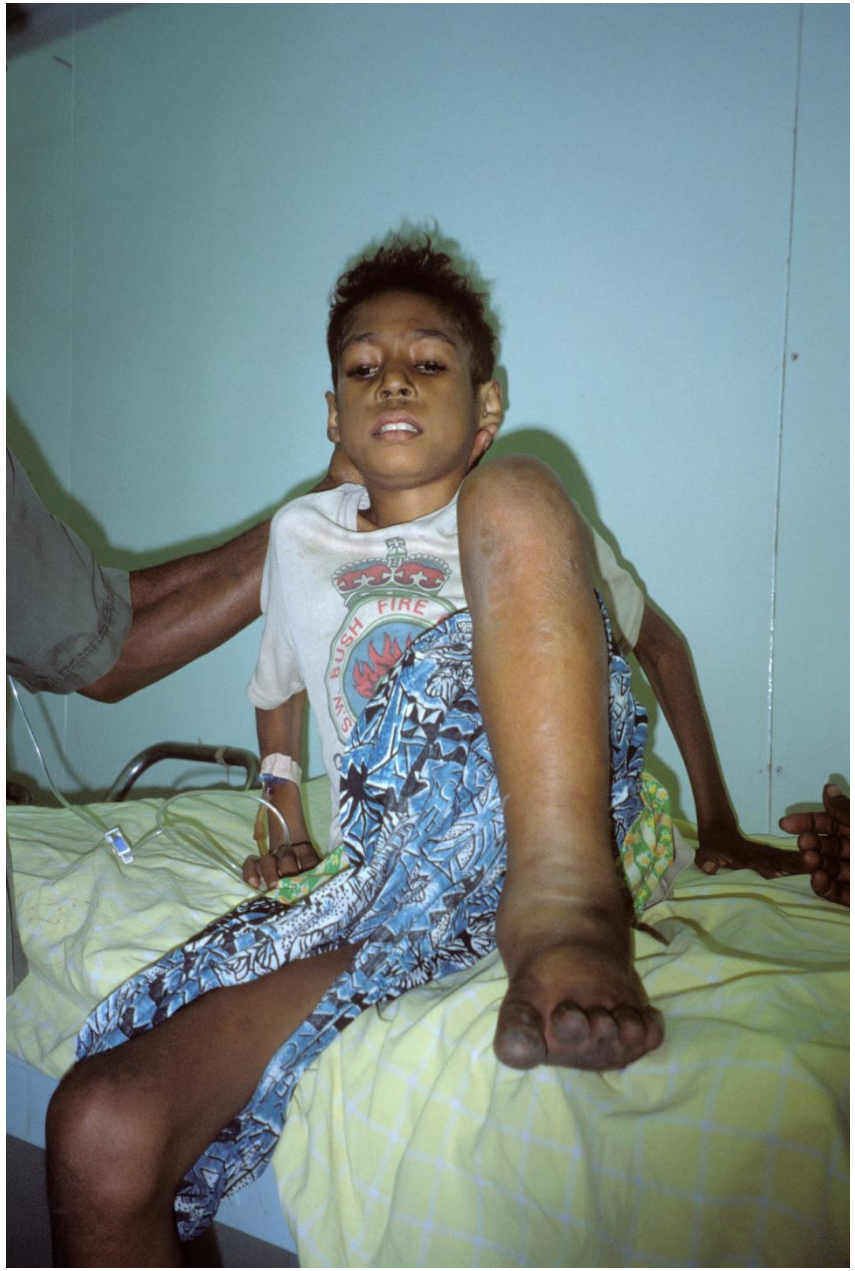
Generalised capillary leak

  - Severe sepsis

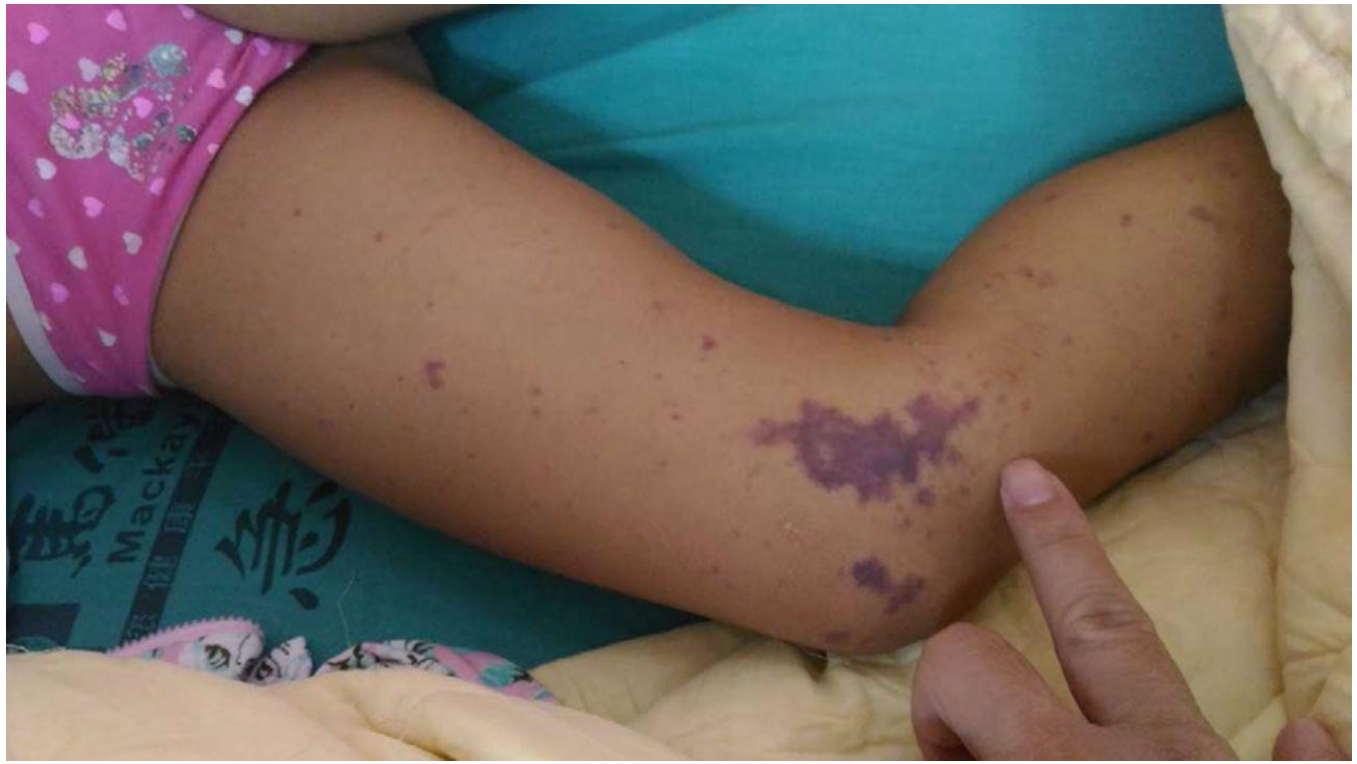


Assessment

- Sick or well
- Generalised or focal
- Ascites, pleural effusions, pericardial effusion
- Pallor, jaundice
- Other signs of PEM
- Signs of TB
- Signs of lymphatic obstruction
- Rash – SLE, HSP


BUSH FIRE

- Well looking
- Abdominal pain, vomiting, bloody diarrhoea
- Arthralgia
- Foot and ankle oedema
- WCC normal
- Haematuria



Nephrotic: assessment severity and complications

- Intravascular volume depletion
  - Cold hands or feet, capillary refill time >3 seconds
  - Tachycardia, low volume pulses
  - Oliguria, hypotension, narrow pulse pressure (e.g. 60/80).
- Severe oedema
  - Gross scrotal / vulval oedema
  - Skin breakdown / cellulitis
  - Pleural effusion, ascites
- Infection
  - Cellulitis
  - Spontaneous bacterial peritonitis – abdominal pain, tenderness
- Thrombosis

Investigations

- Heavy proteinuria 3-4+
- Check urine for blood – if large suggests nephritic syndrome
- Creatinine:
  - may be normal
  - mild elevation of serum creatinine if volume depletion.
  - If creatinine very high, consider *nephritic* syndrome
- LFT including albumin (<25 g/L)
- FBC: check for anaemia, neutrophilia or neutropenia (infection).

Treatment

- Manage oedema
  - No added salt diet
  - Daily weights, daily urine dipstick
  - Strict fluid balance with close attention to volume status
- Do not give frusemide, as can make intravascular volume depletion worse
  - If shock, give albumin if you can +/- frusemide
- Penicillin V (phenoxymethylpenicillin) to prevent pneumococcal peritonitis until oedema subsides

Treatment

- **Prednisolone**: to induce remission, then a slow wean to reduce risk of relapse
  - 60 mg/m^2/day (max 60 mg) for 4 weeks
  - then 40 mg/m^2/day (max 40 mg) on alternate days for 4 weeks
  - then 20 mg/m^2/day on alternate days for 10 days
  - then 10 mg/m^2/day on alternate days for 10 days
  - then 5 mg/m^2/day on alternate days for 10 days
  - then cease

Body surface area (m^2)

$$\frac{\text{Weight (kg) x height (cm)}}{3600}$$

Discharge planning – teaching the family

- Teach the family to test urine protein each morning to identify a relapse (3-4+ protein for 3 consecutive days), at which point the family should re-present
- It is better to detect relapse *before* edema develops, much more effective to reinstitute steroids (lower dose shorter course)
- Tell parents: child will likely respond to therapy, they will likely have relapses (80% chance)

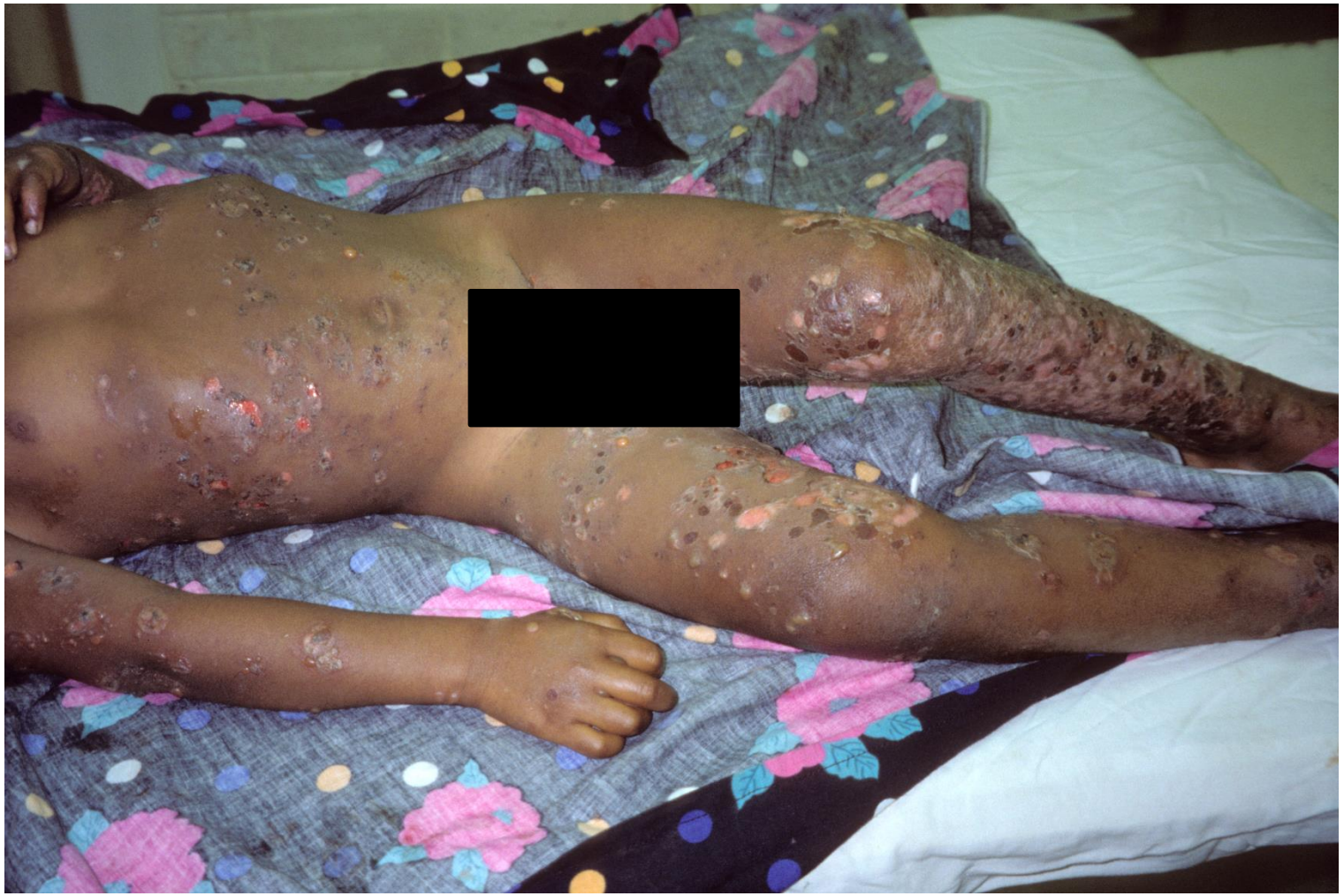
Red flags – when to think again

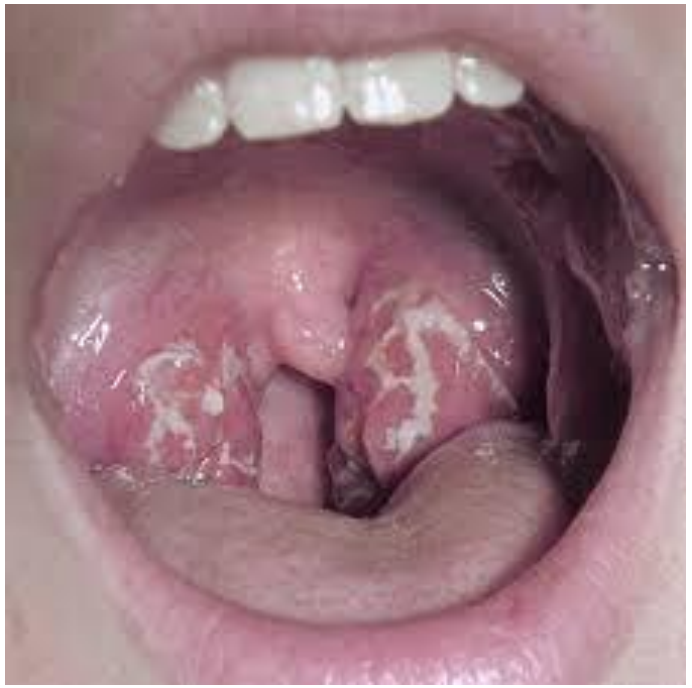
- Features suggesting diagnosis other than INS
- Severe, difficult to control oedema
- Elevated creatinine despite correction of hypovolemia
- Not in remission after 4 weeks of steroid therapy
- Relapses (while taking steroids or within two weeks of cessation)
- Steroid toxicity prompting consideration of alternative agent

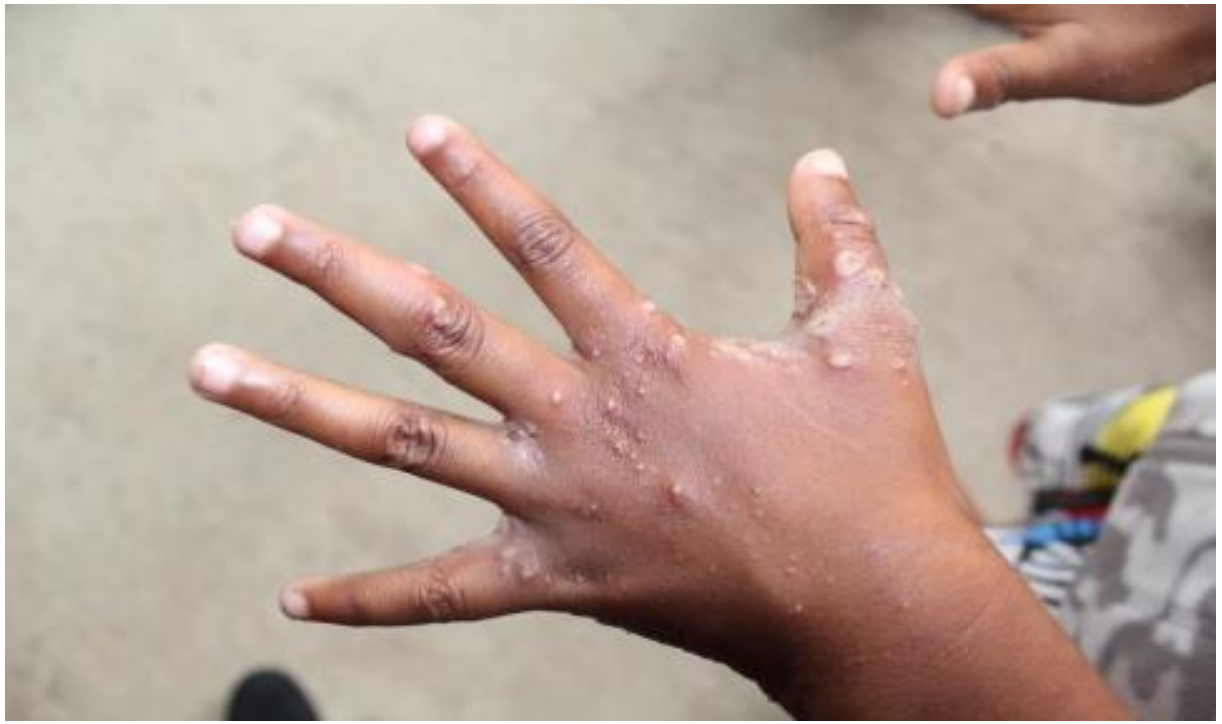
Post-streptococcal glomerulonephritis (PSGN)

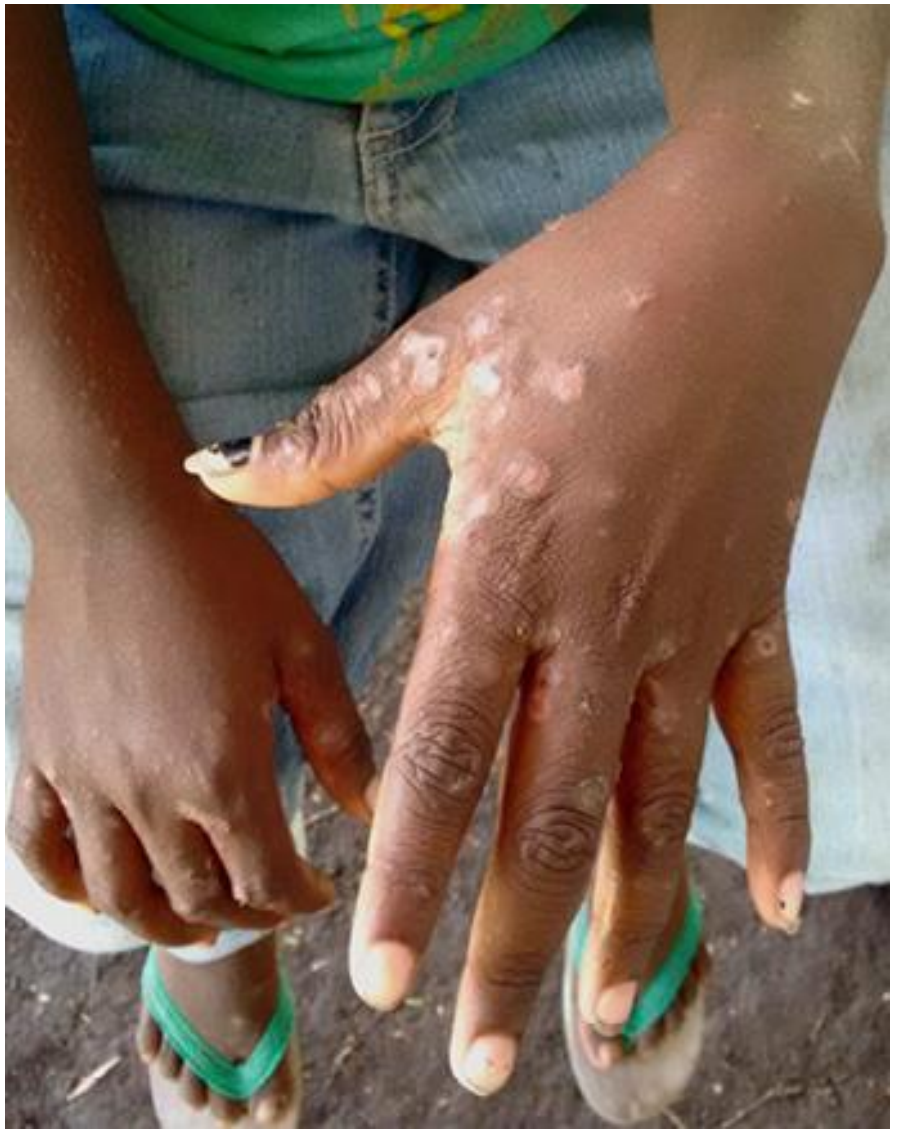
- Children 5-12 years
- Post throat infection (1-3 weeks) or skin infection (3-6 weeks) from Group A streptococcus
- Immune complex deposition in glomerulus, complement activation
  - Haematuria
  - Proteinuria (can reach the nephrotic range)
  - Oedema
  - Hypertension – salt and water retention, sometimes encephalopathy
  - Acute kidney injury

Group A streptococcus skin and throat infections

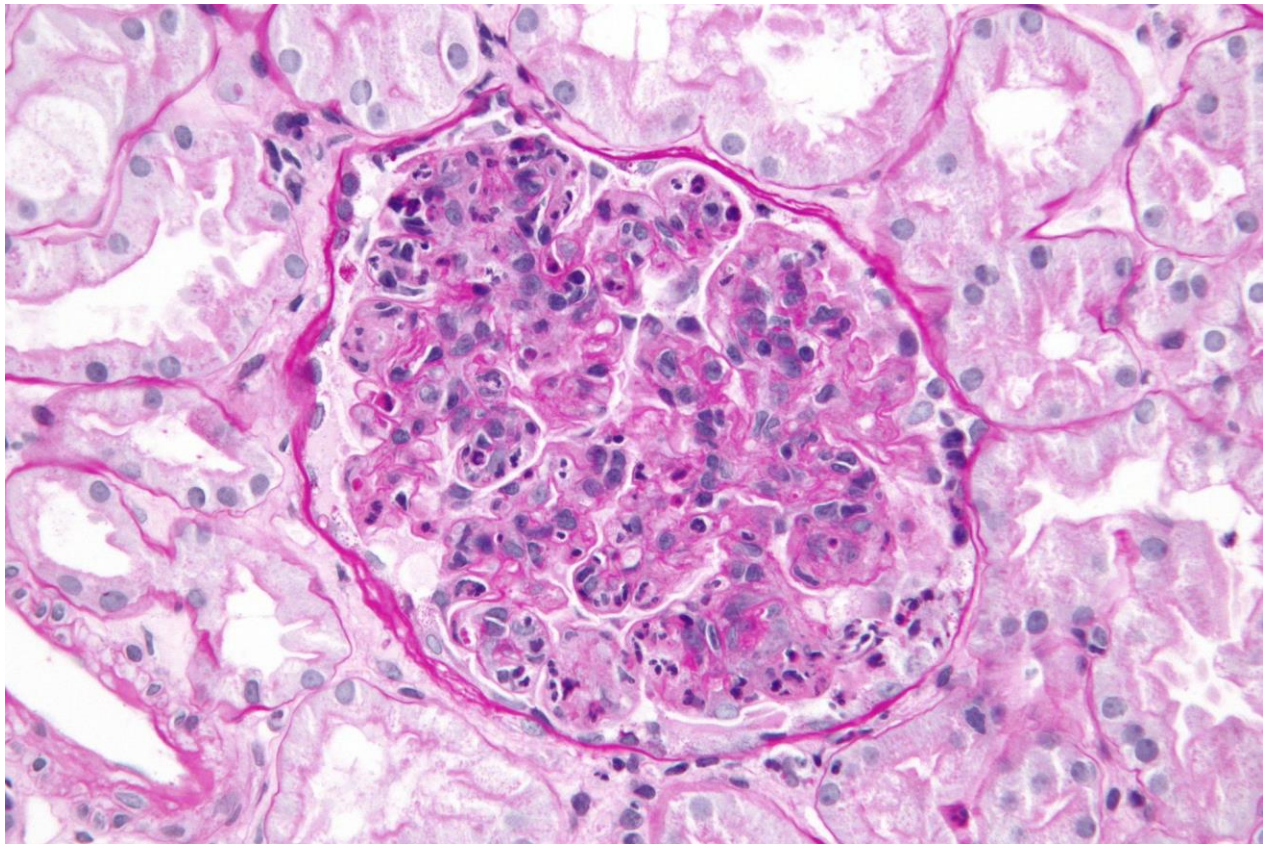






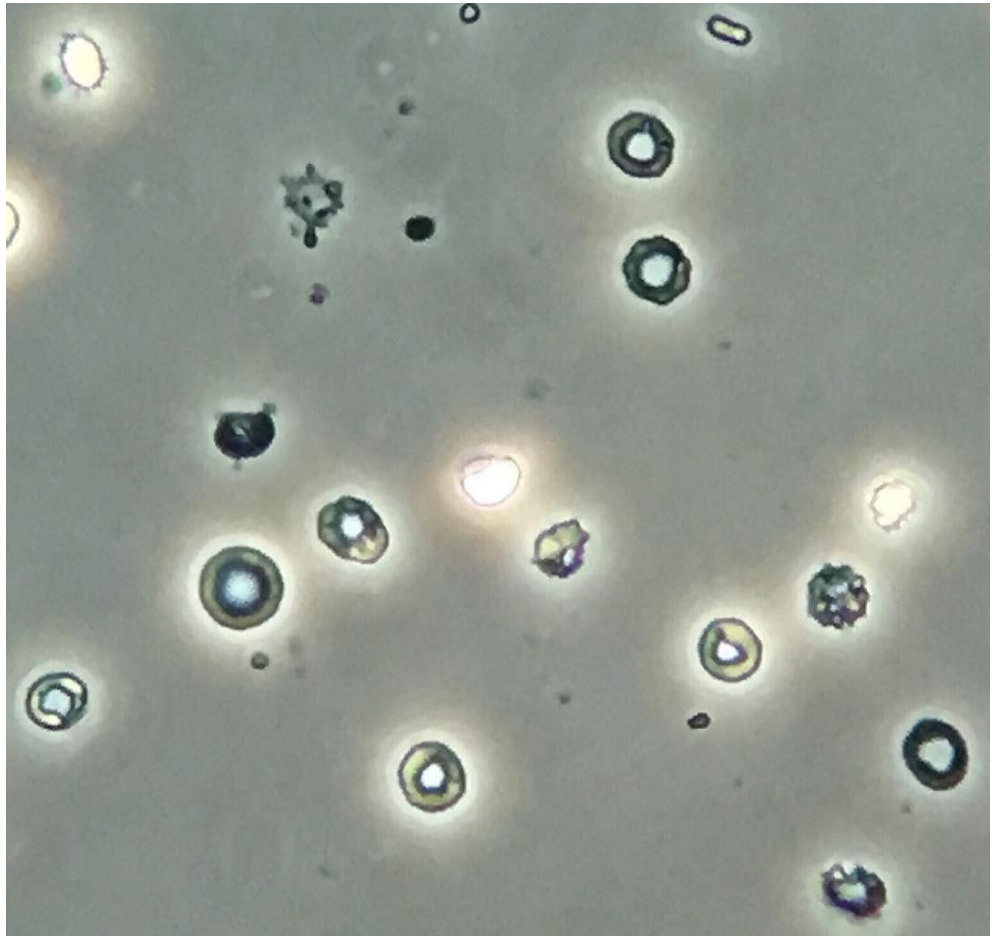


- Deposition of streptococcal antigens within the glomerulus: Streptococcal pyrogenic exotoxin B (and others)
- Complement activation → C3↓↓
- Neutrophil infiltrate ++++
- “Acute diffuse proliferative GN”



Investigations

- Urine for red cells, casts, glomerular red cells
- Creatinine ↑
- WCC ↑
- Antibodies to Streptococcal antigens
  - Anti-streptolysin (ASO)
  - Anti-DNase B antibodies



Differential diagnosis

- IgA nephropathy (HSP)
- Hepatitis B nephritis
- Endocarditis nephritis
- Lupus nephritis
- Drug induced nephritis
  - Antibiotic-associated nephritis (eosinophilic): cephalosporins, ciprofloxacin, ethambutol, isoniazid, macrolides, penicillins, rifampicin, sulfonamides, tetracycline
  - Other: NSAIDS

Treatment

- Penicillin to treat the Streptococcal infection if still present
- Treat hypertension
  - Frusemide 0.5-1mg/kg QID
  - Nifedipine
  - Captopril (watch for hyperkalaemia)
- Most start to resolve within 2 weeks, normal creatrinine by 4 weeks
- Dialysis only for progressive renal failure, acidosis, hyperkalaemia, intractable hypertension

Complications and prognosis

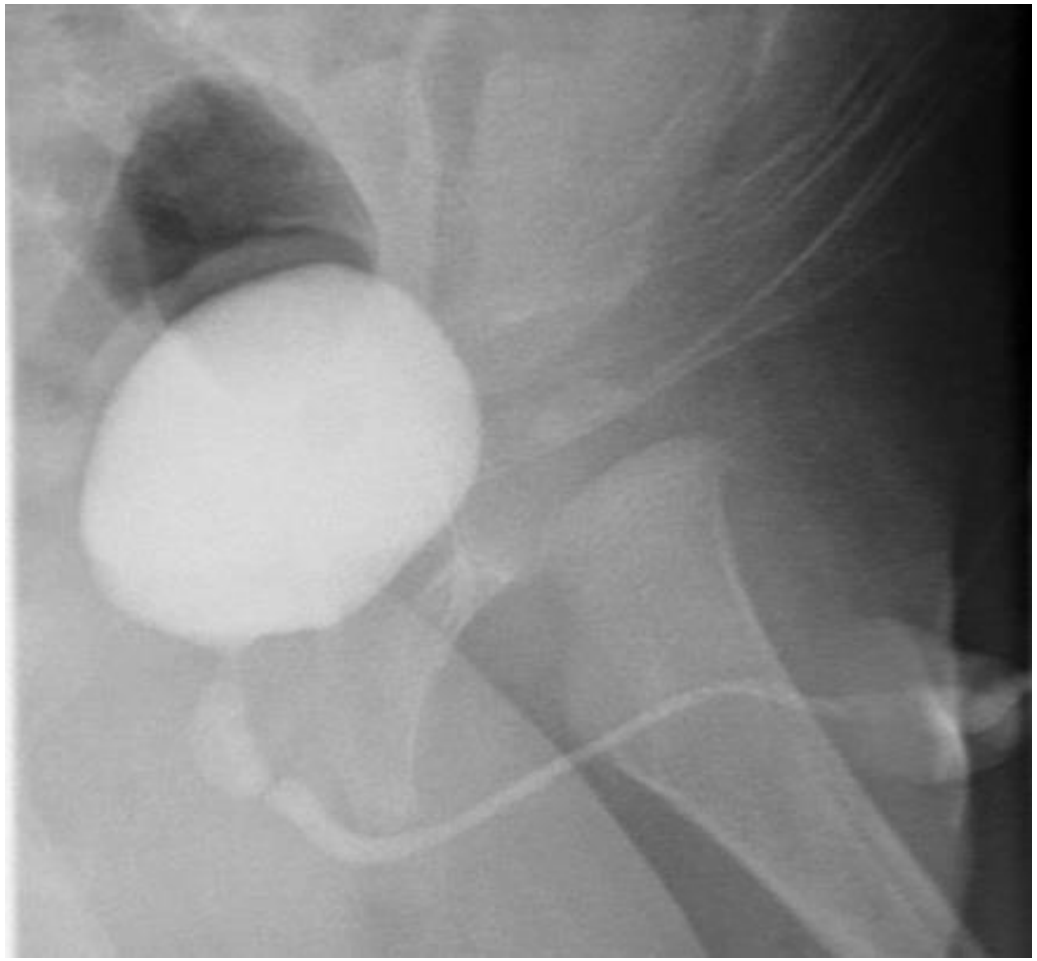
- Almost all resolve
- Some relapse if exposed to GAS antigens again (like ARF)
- Hypertension
- Albuminuria
- Adult renal impairment - glomerulosclerosis

Chronic kidney disease

Causes of chronic kidney disease in children	Infants	>12 years
Congenital abnormalities of kidney or urinary tract (CAKUT) • Obstructive (PUJ obstruction, posterior urethral valves) • Reflux • Hypoplasia	50%	↓↓
Steroid resistant nephrotic syndrome	10	↑↑
Chronic glomerulonephritis	8	↑↑
“Renal cilioathies”: e.g. cystic kidney disease (AD PKD)	5	
HUS		
Nephrolithiasis		
Infections		

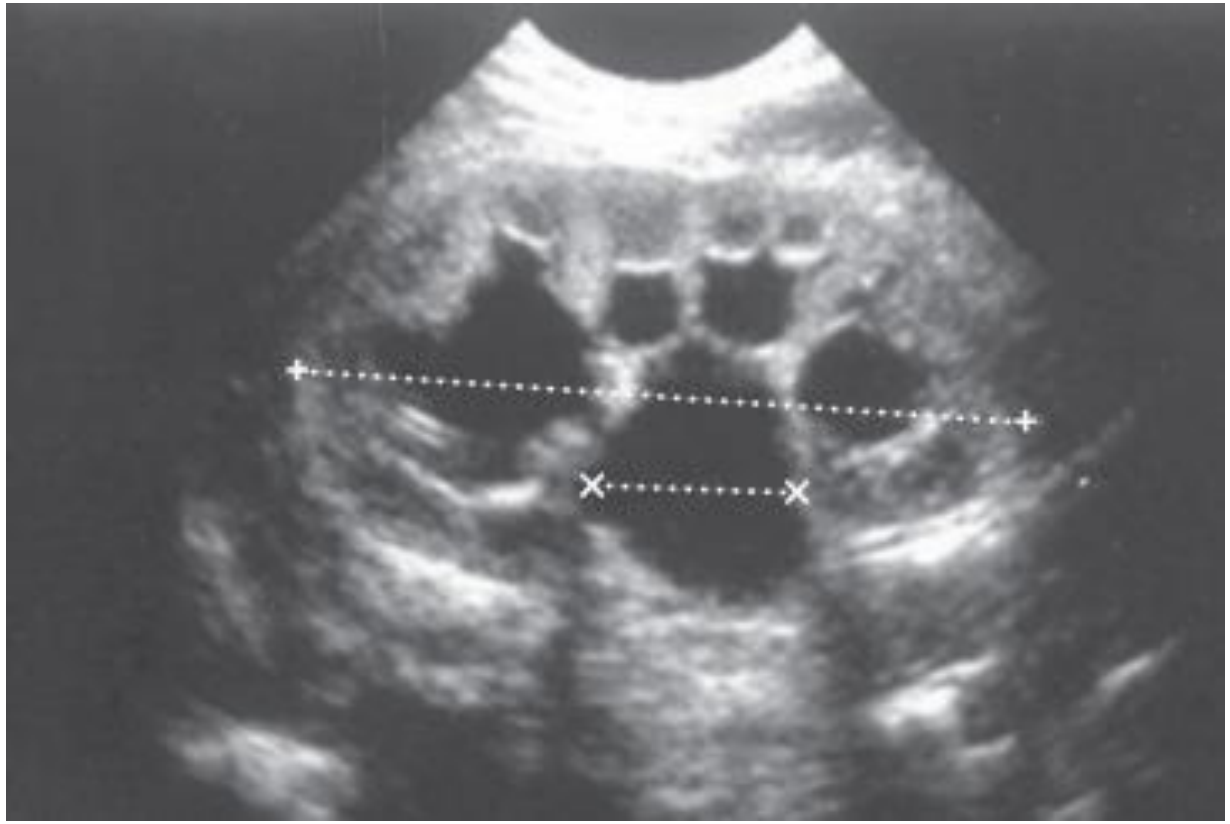
Posterior urethral valves

- Congenital obstructive posterior urethral membrane
- Posterior urethral valves:
  - boys, minimal stream, marked bladder distension, hydronepohrosis and hydroureter)
- Micturating Cysto-Urethrogram
- Surgically curable, urgent



Congenital obstructive renal diseases

- Pelvi-ueretric junction (PUJ) obstruction
  - Can be bilateral or unilateral
  - Many resolve spontaneously
  - Infection because of stasis
  - How to know if a kidney is working?



Chronic renal disease: complications

- Growth impairment
  - Especially common in congenital renal anomalies (as rapid growth occurs in the first 2 years), stunting.
  - Inadequate nutrition, low growth hormone-IgF-1, bone failure, acidosis, anaemia
- Bone and mineral disorders
  - Kidneys do not clear phosphate → hyperparathyroidism, ↓ calcium, bone resorption
  - Damaged kidneys cannot convert D3 into it's active form calcitriol→ hypocalcaemia
  - Treatments
    - Calcium supplements
    - *Active* forms of vitamin D supplements (calcitriol)
    - Phosphate binders (calcium carbonate)

Chronic renal disease: complications

- Anaemia
  - ↓ Erythropoietin
  - Iron deficiency
- Hypertension
  - Needs to be controlled to limit left ventricular hypertrophy and progression of renal disease (especially proteinuria)
  - Endothelial dysfunction, arterial thickening, calcification, LVH
- Prevention of cardiovascular complications
  - Treat hyperphosphataemia, hyperparathyroidism, anaemia, hypertension

Care of children with chronic kidney disease

- Correct any correctable problems (surgical, medical)
- Renal impairment has effects on many organs. Look after the whole child: growth, bones, anaemia, cardiovascular system, prevention of infection, avoid drug toxicity (e.g. steroids, nephrotoxic drugs).
- A written care plan and a trusted primary doctor
- Teach the family and the child
- Attention to schooling, mental health, family stress.