Childhood Nephrotic Syndrome

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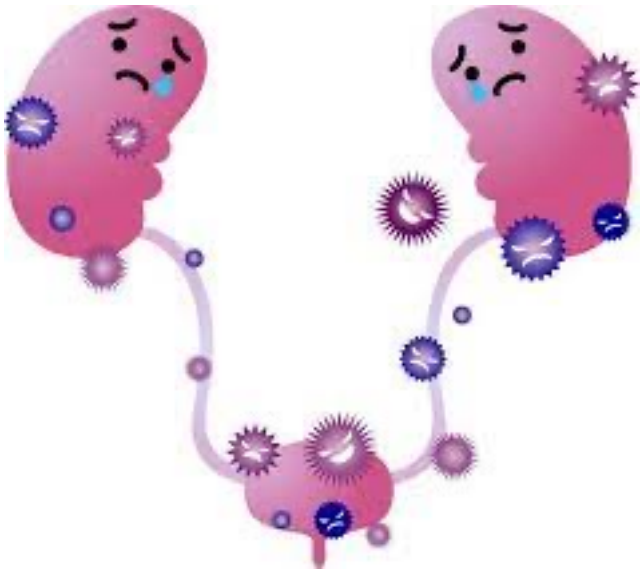
Overview

- Diagnosis
- Definitions
- Disease course
- Long term management and monitoring



Background and Definition

- 2-4 cases / 100,000 children in the UK
- Commonest glomerular disease of childhood
- More common in South Asian populations
- Most children have idiopathic form

- **Triad of**
- Proteinuria 3+ or >300mg/mmol
- Oedema
- Hypoalbuminaemia (<25g/l)

First Presentation

- Basic invesitgations

UE, FBC, LFT, UaUc or UpUc, VZV Ab status

- Atypical features – early discussion with renal centre

- <1yr or >12 yrs
- Macroscopic haematuria
- Significant renal impairment
- Hypertension – not pred related
- Slower onset / borderline proteinuria

- Further tests to consider

Complement, ESR, ANA, dsDNA, ASOT

Initial Management

- Management Goals
- Achieve remission
- Prevent / reduce acute risks
- Avoid / reduce risks of long term steroids
- Monitor for complications

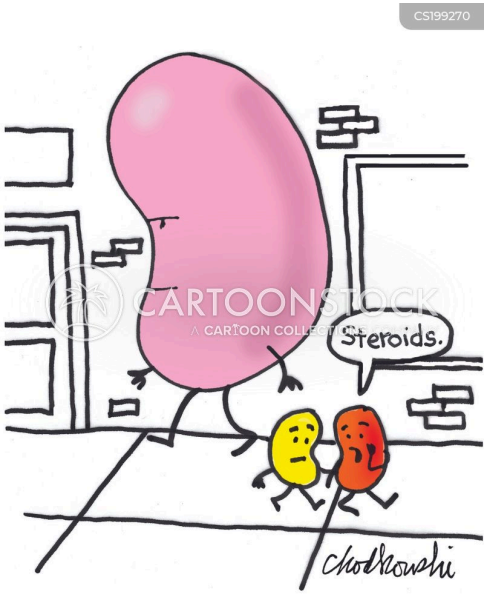
Steroid Treatment

4 weeks 60mg/m2/d (max 60mg OD)

Then if in remission

4 weeks 40mg/m2/ alternate days

Then stop



Also Consider

- **IV methylpred –** Give if not tolerating oral steroid
- **PenV** – can be given as prophylactic cover for bacterial peritonitis
- **PPI** – for gastritis cover
- **Warn parents** – appetite, sleep and behaviour disturbances are common at these steroid doses

What happens next?

- Remission – neg or trace protein 3 consecutive days
- Steroid sensitive – patient enters remission within 28 days of steroid
- Steroid resistant – does not achieve remission after 28 days (including pulsed methylpred)
- Steroid dependent – enters remission with high dose steroid but relapses when pred is weaned or within 14 days of stopping
- Relapse – 3 consecutive days of 3+ proteinuria
- Frequently relapsing – 2 relapses in 6/12 or 3 in 1 yr

Disease Course

- ~10% steroid resistant
- Will need tertiary referral. Usually Tacrolimus and ACE +/- renal biopsy. Worse long term prognosis

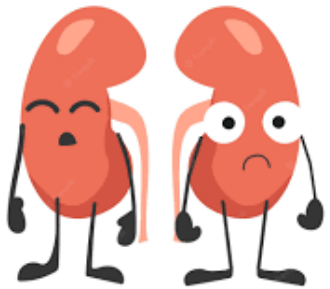
- ~20% will never have another relapse
- Can be followed up for 1-2 yrs with safety netting

- ~70% will have relapses – about 50% of them will have frequently relapsing / steroid dependent nephrotic syndrome

Relapse

- Can often be managed as outpatient if not severe

- Relapse protocol
  - 30mg /m2 (max 30) until remission
  - 1 week 20mg /m2/alternate days
  - 1 week 10mg / m2 / alternate days
  - 1 week 5 mg /m2/alternate days then stop



- If significant oedema or not responding at 7 days or if previously needed higher doses then start at 60mg/ m2

- Ensure daily dipping of urine to monitor response

Frequent Relapse and steroid Dependence

- Steroid burden is high in this group and more likely to have steroid side effects
- Steroid sparing agents should be used in this group to reduce risks
- Usually In conjunction with a tertiary centre
- Levamisole (best where relapse are mild and very steroid responsive)
- Tacrolimus
- MMF
- Rituximab – only approved by NICE where other therapies have failed

Immunisations

- Should follow normal schedule as much as possible

- Live vaccines should not be given if
- On immunosuppression
- Within 6 months of Ritxuimab
- High dose steroids and for 3 months after

Caution with MMR booster and intranasal flu

- Response to vaccines may be inadequate if immunosupressed

Infection

- Increased risk of infection with initial presentation and relapse – especially peritonitis
- VZV status should be checked at diagnosis
- If significant chicken pox exposure then should have VZV repeated even if previously immune and any non-immune child should have VZIG given
- Confirmed chicken pox while on pred or immunosuppression (or within 3/12 of stopping) should be treated with Aciclovir – low threshold for IV if unwell.

Psychosocial Support

- Frequent relapse / steroid dependence places significant burden on children and families
- High anxiety / uncertainty
- Steroid side effects are hard on children, young people and parents
- Explanation, reassurance, support and communication with school and nursey are really important

Summary

- Nephrotic syndrome diagnosis and definitions
- Initial management
- Atypical features and when to involve tertiary centre
- Disease course
- Management of complex cases
- Immunisaiton and infection in nephrotic syndrome
- Support