Current Practice

The treatment of steroid sensitive nephrotic syndrome

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(Key words: steroid sensitive nephrotic syndrome, SSNS, treatment)

Due to an oversight, a part of the above article was omitted from the last issue of the journal. We are now rectifying this omission.

Frequently relapsing and steroid dependent MCNS

These children are difficult to treat and develop steroid toxicity. The first step in management is to identify frequent relapers and steroid dependence accurately, using the definitions given. This requires the paediatrician to spend time evaluating the course of the disease and the treatment given throughout. The definitions are applicable only if standard relapse treatment has been used in the preceding relapses.

Levamisole

The relative efficacy of levamisole is not known. In the University Paediatric unit, at the Lady Ridgeway Hospital for Children, we have been using this drug since 1993. A 10 year review of 110 children with SSNS followed up by us showed that levamisole was able to induce remissions lasting 2 years or longer and there was no statistically significant difference in efficacy when compared with cyclophosphamide. However, it appears as though levamisole is more effective in the milder forms of the disease. We have not encountered any serious side effects with this drug to date.

Cyclosporin A

This drug is nephrotoxic and therefore can be used only if facilities are available to monitor drug levels very regularly. The Medical Research Institute has recently commenced assessing blood levels of cyclosporin, but this is done only on 2 specified days of the week. The very high cost of the drug and the problems of assessing blood levels regularly makes it difficult for us, in Sri Lanka, to use this drug at present.

WHAT WE NOW KNOW

- Children with frequent relapses usually remain steroid responsive.
- There are no randomized controlled trials comparing repeated courses of standard relapse therapy with long term low dose alternate day prednisolone therapy for frequent relapses.
- 8 week courses of cyclophosphamide or chlorambucil and prolonged courses of cyclosporin or levamisole substantially reduce the incidence of relapses.
- Cyclosporin and levamisole effectively induce remission, but are unable to sustain this effect once treatment is withdrawn.
- There is insufficient data at present to show that alkylating agents are more effective than cyclosporin or levamisole in reducing the incidence of relapses.

Treatment protocol for children with SSNS

This protocol is a modified version of the guidelines set out by the British Association for Paediatric Nephrology and Research Unit.

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Treatment Protocol

1. Initial episode
   - Treatment as discussed above.

2. 1st and 2nd relapse
   - Standard relapse therapy

3. Frequent relapses
   - Maintenance Prednisolone
     0.1-0.5mg/kg/EOD for 6 months
     Slow withdrawal over another 6 months

4. Relapse on Prednisolone >0.5 mg/kg/EOD
   - Levamisole
     2.5 mg/kg/EOD for 12-18 months.

5. Relapse while on Levamisole or Prednisolone >1.0mg/kg/EOD
   - Cyclophosphamide
     2.5 mg/kg/day for 8 weeks.

6. Post Cyclophosphamide Relapse
   - Treat as in 2 & 3 above.

7. Relapse on Prednisolone > 0.5 mg/kg/EOD
   - Cyclosporin 5 mg/kg/day for one year or?
     Chlorambucil
References


