139 NEPHROTIC SYNDROME IN CHILDREN

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Experience with 103 children suffering from nephrotic syndrome with a follow up even for the period of 20 years is presented. Congenital nephrotic syndrome, differing from the finnish type was found twice. 69 children suffered from the minimal changes disease. The affection mostly began in the preschool age.No correlation as to the recovery or number of relapses in connection with age at onset or with the frequency of adjoining allergic disease could be found. The prognosis was excellent, none of the patients died from renal insufficiency in spite of the fact that they were observed even for 20 years. The most dangerous complication was hypovolemic shock and thrombosis of great vessels. The response to prednisone was mostly very good, cyclophosphamide was used in some patients, but its effect on gonads should not be forgotten. In 32 children with other types of nephrotic syndrome 8 died in chronic renal failure and renal insufficiency developed in 3 others. No therapy in this group gave satisfactory results with exception of some cases of lupus nephritis. Minimal changes disease differs distinctly in clinical course and prognosis from other forms of the primary nephrotic syndrome.

140 FOCAL GLOMERULAR SCLEROSIS IN CHILDHOOD: LONG TERM FOLLOW-UP.

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Of renal biopsies from 300 children with idiopathic nephrotic syndrome (INS), 32 showed histological lesions of focal glomerular sclerosis. From this latter group, 25 had segmental and focal hyalinosis (FSH) with or without mesangial hypercellularity, and 7 focal glomerular obsolescence (FGO). The overall outcome after a mean observation of 7 years (0.5-18) was as follows: 11 patients (34%) were in complete remission, 12 (38%) had persistent proteinuria and/or recurrent nephrotic syndrome, 6 (19%) were in chronic renal failure and 3 patients (9%) died from non renal disease. Patients with FGO had a lower incidence of hematuria, hypertension and tubular defects, a better response to steroid and cyclophosphamide therapy and a better overall outcome. These results are similar to those reported by Habib and Gubler (Ped. Nephrology, Ed. Rubin and Barratt, p. 499-515) and indicate a more favorable outcome than that reported by Cameron et al. (Clin. Nephrol. 6:213, 1978). A progressive decrease in GFR was observed in some patients with persisting proteinuria or recurring nephrosis indicating that ultimate outcome of FGS in children cannot be established from pediatric age population studies.

141 MEMBRANOPROLIFERATIVE GLOMERULONEPHRITIS (MPGN): A DOUBLE BLIND TRIAL OF ALTERNATE DAY PREDNISONE (ADP). International Study of Kidney Disease in Children.

A double blind trial of ADP is being conducted in children with MPGN and the nephrotic syndrome. Thirty-four children have been randomly allocated to either ADP (60 mg/m^2/q 48 hrs in 18) or placebo (16). All have had at least 2 yr followup with renal biopsy. There were no significant differences initially between the groups. The types of MPGN were not separately distributed (20 Type I, 9 Type II and 5 Type III). Overall GFR decreased by ≥30% in 6; increased ≥30% in 8 and was unchanged in 20. Initial histopathologic findings of mesangial proliferation (MP), crescents and adhesions (CA), glomerular sclerosis (GS), tubular atrophy and interstitial fibrosis (TAIF), or capillary wall abnormalities (CWA) did not predict the ΔGFR. At the 2 yr biopsy, GS and TAIF were associated with decreased GFR, but not MP, CA, or CWA. MP decreased more in those on ADP. The proportion of patients with decreased GFR at 2 yrs was less with ADP (1/18) than with placebo (5/16) but no differences were noted at 3 yrs (3/14 vs 5/11), 4 yrs (5/12 vs 7/10), or 5 yrs (5/11 vs 8/11). Preliminary results indicate that ADP may retard progression early but the effect is lost by 3-5 yrs. The trial is continuing because of the small numbers of patients followed past 2 yrs.

142 THE STUDY ON RELAPSE OF IDIOPATHIC NEPHROTIC SYNDROME IN CHILDREN

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One of the main subjects of idiopathic nephrotic syndrome in children today is prediction and prevention of relapse. In this study, we predicted relapses of idiopathic nephrotic syndrome immunologically. Detection of inhibitory factors for Con-A induced lymphoblastogenesis in the nephrotic plasma was carried out, and the factors were expressed as inhibition index.

In remission, the inhibitory factors seemed to disappear, while in relapse and 2 to 4 weeks prior to relapse as well as at the onset, the indices were abnormally high. Molecular weight identification of the inhibitory factors was carried out on the patients' sera at the onset and in relapse by Sephadex G-200 gel chromatography. The results revealed three peaks with strong inhibitory activity. One is probably glycoprotein at more than $6X10^5$ molecular weight, and the others are multifactors with $3 - 7X10^4$ in molecular weight and peptide at $6X10^3$ molecular weight.

We believe the inhibitory factors exist in nephrotic sera at the onset as well as in relapse. Prediction of the relapse is possible 2 to 4 weeks prior to actual relapse. Preventive therapies can be applied successfully.

143 LONG-TERM STUDY OF STEROID SENSITIVE NEPHROSIS (SSN).

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312 children with SSN were followed for 2 to 24 years (y). Age at onset was under 1 in 8 who never relapsed over 2 to 14y, and between 1 and 8y in 80 % of cases. 77 children were unselected : of them, 32 (42 %) never relapsed, 8 recovered after 1 or 3 relapses, 36 were steroid (S) dependent (D). Among the 312 children representing a highly selected series, 4 children died from complications and 8 developed renal failure (RF) : five were SD for high doses, remained untreated and nephrotics for at least 15y : 3 reached terminal RF (TRF), 19, 20 and 21y after onset. Three children with initially SSN became resistant and were referred for TRF 2, 5 and 6y after onset. The 300 other patients had normal renal function after 5 to 10y in 118, 10 to 15y in 51, 15 to 24y in 23. 62 patients never relapsed, 24 recovered after 1 to 3 relapses, and 214 were SD. Of the latter, 104 received immunosuppressants (IS) resulting in prolonged remission in 74. Interrupted or alternate day S therapy was used for respectively 1 to 7y in 38 children, and 1 to 6y in 35. The latter allowed a good control of the disease with little toxicity and normal growth. In SD, therapy could be discontinued without IS in 26 children after 2 to 5y, in 15 after 5 to 10y, between 10 and 20y in 13, and remained necessary after 10 to 18y in 17 children, all of them with normal renal function. The high incidence of a single attack in unselected series, the unpredictable duration of disease in SD patients, the possibility of poor outcome in untreated patients and the good prognosis in the others may be stressed out.

144 *Serum Concentration of Prednisolone in Children with Nephrotic Syndrome:*

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[Purpose] Recently prednisolone has been used clinically for the therapy in children with nephrotic syndrome. But little information was available about the study of the absorption and excretion of prednisolone in the nephrotic syndrome. So we invented new easy radioimmunoassay(RIA) for prednisolone, and measured the serum concentration of prednisolone in children with nephrotic syndrome. [Methods] Three rabbits were injected with prednisolone 21-BSA hapten and Freund's adjuvant. The rabbit serum(anti-prednisolone antibody) and goat anti-rabbit gamma globulin was used for RIA by double antibody technique. By heating unknown serum in 70°C for 30 minutes, the native prednisolone binding protein was denatured, and it was possible to analyze prednisolone in unextracted and nondiluted serum. Twenty children with idiopathic nephrotic syndrome were administered orally prednisolone several times. The quantity of prednisolone was equal in the each patient. The serum concentration of prednisolone was measured serially. [Results and Discussion] This RIA was capable of detecting 50 picograms of prednisolone in 50 μl of undiluted serum. The peak concentration of prednisolone in patients had a tendency to increase gradually with the decrease of the urine protein. The tendency had little correlation with the change of the total protein in serum.