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Renal Amyloidosis in Children.

The authors are relating 4 cases of renal amyloidosis in children between 2-14 years of age, with nephrotic syndrome. Clinically, the oedema were variably developed. The most characteristic laboratory data indicated: proteinuria 18-4,5 g%, hypoproteinemia, inconstant and moderate hypercholesterolemia, alpha-2 fraction increased,up to 33%. In one case,was associated mild hematuria and hypertension. The diagnosis of amyloidosis was relied on renal biopsy(3 cases) and gingival biopsy (1 case). The diagnosis of primary amyloidosis was based on clinical criteria: absence of chronic suppurative infections(tuberculosis, Bronchiectasis,osteomyelitis) accompanying amyloid nephrosis. In one child, the amyloidosis was associated with a pulmonary tumor (bronchogenic dysembryoma). The site and extent of depots of amyloid in renal tissue was variable:initially, in glomerular and tubular basement membrane, subsequently arterio-arteriolar, finally affecting all the glomeruli. The evolution of all cases was progressively rapid(8-14 months) to renal failure.The corticoids were inefficient and the chlorambucil deteriorated the clinical and biological evolution of the illness.

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Congenital Hepatic Fibrosis (CHF) and Nephronophtisis.

A 12 year-old boy was referred for pallor and general weakness for at least six months. Previously the child had been hospitalized at the age of five, for mental retardation and hepato (7cm)spleno(6cm)megaly.At that time, no explanation could be found for the first symptom, whereas all liver function tests gave normal results.A surgical biopsy was thought to be compatible with the diagnosis of CHF. At physical examination,height was 14cm below the 3rd percentile.The boy's vision was very poor. Blood pressure was 130/70mm. Fundoscopy revealed tapeto-retinal dysplasia.Blood values on admission:hematocrit 16%,hemoglobin 6g/100ml, Calcium 6,7,Phosphorus 7,1,Urea 234,Creatinine 9,2mg/100ml.The diuresis was about 2000ml/24hrs. Urine sediment was negative.Proteinuria between 500 and 1600mg/24 hrs.Creatinine clearance : 7ml/min/1.73m^2.Except for slight increases of serum GOT and GPT values,there was no anomalies in liver function.A liver puncture biopsy was inconclusive.A kidney biopsy showed diffuse tubular atrophy and interstitial fibrosis. Autopsy findings confirmed the hepato (1500g) spleno(380g)megaly and both diagnoses of CHF and nephronophtisis.This observation is very similar to the cases reported by Biochis et al. (Quart.J.Med.42,221,1973) and seems to confirm the existence of this new syndrome.

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Pathomorphological study of focal and segmental sclerosing glomerulopathy (-nephritis).

From 44 patients with f.s.s.g. 75% suffered from a nephrotic syndrome, 25% from proteinuria, and 48% from erythrocyturia. The characteristic glomerular lesion is a progressive increase in mesangial matrix (sclerosis) which in early stages concerns only few glomeruli and within these only a portion of the capillary tuft. Histologically the remaining glomeruli show minimal changes, only. Electron microscopy, however, reveals these glomeruli as pathologically altered, too. PAS-positive hyaline deposits are identified with the light microscope in about 80% of all patients. Electron dense deposits, identical with immune deposits are regularly found within sclerosing areas, but rarely occur in minimally changed glomeruli, too. Immunofluorescence microscopy reveals a corresponding segmental deposition of IgM and C3. The findings suggest that the underlying mechanism is a diffuse one, but associated with focal accentuation which might depend on or be promoted by deposited immune aggregates.

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The Renal Lesion in Congenital Chloride Diarrhea (CCD).

Juxtaglomerular hyperplasia, hypertensive arteriolar changes and nephrocalcinosis have been reported in CCD. To elucidate the pathophysiology of these changes we have studied the renal function and histology, and measured the activity of renin, angiotensin II and aldosterone in 19 patients. The older children were earlier treated with a KCl supplementation, the dose being adjusted to maintain normal serum electrolyte levels. They had high hormone levels and developed extensive kidney changes. Therefore, we instituted substitution with both NaCl and KCl adjusting the dose to maintain normal blood pH and chloriduria in addition to normal serum electrolyte concentrations. In the patients treated thus from birth renal changes were absent, expect for some glomerulosclerosis, and the hormone levels were normal. Glomerular destruction may be due to acute juxtaglomerular swelling, because even the slightest infection tends to disturb the balance and causes transitory hypovolemia and high renin levels. The latest biopsies of the older patients show clear improvement during the present treatment.