151 IDIOPATHIC NEPHROTIC SYNDROME: ADRENOCORTICAL SUPPRESSION (ACS) AFTER PREDNISONE MEDICATION.

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In children with nephrotic syndrome ACS after prednisone medication was detected in half of the new patients, when measured with the 2-hour ACTH stimulation test (Pediat.Res.:12:272:1978). Severe ACS is potentially dangerous in stress (infections, surgery). A spontaneous recovery of the adrenocortical function takes place if the patient stays in remission for a few months.

In a study of 23 children with their first attack of nephrotic syndrome a deficient response in the ACTH test was found in 11 instances. Ten of these children relapsed within a year. However, a normal response in the ACTH test did not guarantee a prolonged remission. In a subsequent double-blind cross over study of 26 relapses in 13 children partial cortisol substitution and placebo were compared. At three months 8 children in the cortisol and 1 in the placebo group were in remission, at six months the difference was less marked.

We suggest that the ACTH test should be performed after prednisone treatment; those with deficient response should be placed on partial cortisol substitution at least during stress for three months or until adrenocortical function is normalized. Treatment with prednisone should be adjusted and tapered in a way causing minimal ACS.

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PATHOLOGY

152 GLOMERULAR MORPHOMETRY IN THE NEPHROTIC SYNDROME.

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Glomerular morphometry was performed in needle biopsy specimens from 53 children with the nephrotic syndrome with minimal change (MC), focal global glomerulosclerosis (FGS), and segmental glomerulosclerosis (SGS). Glomerular cell counts and diameter measurements were performed on methacrylate-embedded, 1μ thick and PASM-stained sections by direct microscopy. Duplicate analyses showed a high degree of reproducibility. There were significantly fewer epithelial cells and more mesangial cells in SGS than in MC and FGS. The number of epithelial cells decreased with age in MC and FGS, but remained constantly low at all ages in SGS. There were no significant differences in differential cellularity between MC, with or without focal glomerular obsolescence or focal tubular atrophy, and FGS. Glomerular diameter increased with age in MC and FGS, but not in SGS. These findings support the view that SGS is an entity rather than a variant of MC, and it is suggested that the lower epithelial cellularity may have aetiological significance.

153 HIGH RESOLUTION LIGHT MICROSCOPY IN RENAL PATHOLOGY

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The purpose of this paper is to report the reliability, reproducibility, and usefulness of plastic embedding and metachromatic stains in diagnostic renal pathology. Six hundred consecutive renal biopsies were fixed in noncoagulant fixatives, embedded in epoxy resin, cut with glass knives, and stained with metachromatic dyes (Toluidine Blue or Paragon). Cell boundaries, intracellular organelles, basement membranes, the deposition of different substances, and other pathologic changes were readily recognized in these sections.

It is concluded that this method is an excellent procedure for the light microscopic examination of renal biopsies. Its high resolution of detail supersedes paraffin embedding results. The method negates the need for special stains, and thus permits better utilization of more expensive techniques (i.e., electron microscopy).

154 RENAL SEGMENTAL "HYPOPLASIA" (ASK-UPMARK): PATHOLOGIC ASPECTS.

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Segmental "hypoplasia" of the kidney is a form of severe renal atrophy commonly associated with vesicoureteric reflux (VUR). The scarred segments contain atrophic tubules and hyperplastic blood vessels; glomeruli are often absent or sparse. These lesions have been identified in 21 patients (12F, 9M), 6-27 yr: 13 (62%) hypertensive (HT) and 8 normotensive (NT); 13 with prior urinary tract infection; 18/19 (95%) with radiographic VUR; 8 (38%) with impaired renal function (GFR <40 ml/min/1.73 m^2); none with severe proteinuria. Renal atrophy commonly progressed after surgical repair of VUR, even in absence of infection. Lesions were bilateral in 5 HT patients and 2 others had unilateral agenesis; bilateral in only 1 NT patient. HT responded to nephrectomy in all patients. Histologic examination of the scars showed no differences between HT and NT patients in numbers of sclerotic glomeruli or in severity of interstitial fibrosis, arteriolar sclerosis, and inflammatory cell infiltration. Examination of cortical tissue adjacent to scars showed HT patients to have more severe tubular atrophy and vascular change, possibly as the result of HT. Thioflavin-T staining showed the sclerotic glomeruli and hypertrophic arterioles to contain more JGA granules in the scars of HT than of NT patients. Segmental "hypoplasia" appears to be an acquired abnormality, although its association with renal dysplasia in two specimens indicates an occasional origin during intrauterine life.

155 THE THICKNESS OF GLOMERULAR BASEMENT MEMBRANE IN FAMILIAL HEMATURIA

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We measured the thickness of glomerular basement membrane in 7 patients in familial hematuria and compared with that in control group, 2 cases of orthostatic albuminuria, 1 case of urolithiasis and 1 case of hematuria with stomatitis. Needle biopsies were performed on the 7 patients with familial hematuria. 2 patients had a third renal biopsy. The measurements of glomerular basement membrane were taken from photographs and at intervals of 1 from cytoplasmic membrane of the endothelial cells to that of epithelial foot process. Comparing with control group, the thickness of glomerular basement membrane in familial hematuria is remarkable thin. (Table 1 and Table 2)

Table 1

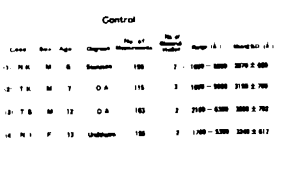
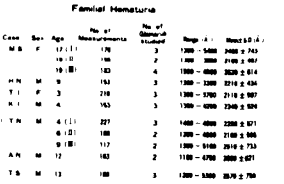


Table 2



156 EXPERIMENTAL REFLUX NEPHROPATHY: ASSESSMENT OF THE PATHOLOGICAL CHANGES IN EARLY SCAR FORMATION.

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In an experimental model of reflux nephropathy in the pig, segmental pyelonephritic scarring occurs rapidly in the presence of infected vesicoureteric reflux with intrarenal reflux in the course of 1 - 4 weeks and the process may be modified by early treatment with antimicrobial drugs. These lesions were assessed histologically in terms of their extent and the type of tissue response. Even after this brief period of infection parenchymal damage was often extensive. Early changes were of active chronic inflammation with tubular destruction but little fibrosis. Later features were a variable degree of fibrous scarring with lymphoid follicle formation. The histological appearances suggest that direct damage by microorganisms initiates the damage, but other mechanisms such as ischaemia and leakage of urinary constituents may also play a part.

Animals treated with antimicrobial drugs after 1 week of infection showed much less extensive damage. Scarring was focal, but both tubular and glomerular changes were found. In controls with scars older than two weeks, an often considerable degree of irreversible damage had occurred which would not be affected by treatment at this stage.