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Communications


Details of the number of patients treated, age, survival, primary renal disease, cause of death, and rehabilitation were given, supplemented by the results of a special inquiry on paediatric patients into growth, bone age, pubertal status, and osteodystrophy. The report has been published in full in *Proceedings of the EDTA* (1976), Volume 13, Pitman Medical, London.


Plasma and erythrocyte amino acids were studied before and after haemodialysis in 7 children aged 4 to 17 years. Increased concentrations of plasma glutamate, methyl histidine, glycine, and citrulline were found before dialysis but tended to normalize after dialysis. Amino acid clearances on dialysis varied from 40–70 ml/min. Erythrocyte amino acid concentrations were raised particularly in the essential group. These results suggest malnutrition in children on chronic dialysis.

Plasma amino acids in children on haemodialysis. R. Counahan, M. M. El-Bishti, B. D. Cox, and C. Chantler. Guy's Hospital, London SE1 9RT.

Sixteen children on chronic haemodialysis were studied. Significant decreases in valine, leucine, isoleucine, lysine, serine, histidine, and tyrosine, and increases in glycine, proline, and citrulline concentrations were found. Children with poor glucose tolerance had high plasma alanine levels which became normal after intravenous glucose. It is suggested that the changes may reflect increased utilization of amino acids because of decreased energy intake or defective metabolism of energy sources.

Plasma lipid abnormalities and energy metabolism in children on regular haemodialysis. M. M. El-Bishti, R. Counahan, and C. Chantler. Guy's Hospital, London SE1 9RT.

Plasma triglycerides, cholesterol, insulin, and growth hormone concentrations were significantly raised in 16 children on haemodialysis; glycerol concentrations were decreased. Basal hyperglycaemia and glucose intolerance was shown. Plasma nonesterified fatty acid concentration fell after intravenous glucose. There was a significant correlation between fatty acid levels and growth hormone. No correlation existed between food intake expressed as percent of normal intake and plasma lipids. It is suggested that the prime derangement in these uraemic children is of normal energy metabolism.


Nine of 80 children on haemodialysis developed pericarditis; in 3 it was recurrent. Heart failure occurred in 8 children but other symptoms were inconstant. 7 children presented with a friction rub but only 2 had typical ECG and x-ray changes. Routine echocardiography is recommended for all uraemic children because it allows early diagnosis. Since its introduction 3 children have been successfully treated by conservative management with frequent dialysis and with indomethacin in one.

1-α-hydroxycholecalciferol therapy in children with renal osteodystrophy. R. J. Postlethwaite and I. B. Houston. Royal Manchester Children's Hospital, Pendlebury, Manchester.

Ten children with renal osteodystrophy, aged 5–15 years, were treated for up to 9 months with 1-α-hydroxycholecalciferol (1-α-HC). In the 5 children studied, intestinal calcium absorption increased. 6 children showed definite signs of healing. The other 4 have only recently started treatment. 1-α-HC in a dose of 0.04–0.08 µg/kg per day was effective and safe in these preliminary studies.

Ten children on chronic haemodialysis, all HBsAg negative, were treated with hepatitis B immunoglobulin (HBIG titre 1 : 20 000, dose 0·2 ml/kg i.m.) every 4 weeks. No side effects were observed. HBsAg hepatitis, HBs antigenaemia, and active HBs antibody synthesis did not occur. Resorption of injection was complete in 1·5 days; the half life was 20 to 26 days except in 3 malnourished children where the half life was 7·8 days. This procedure may be useful in preventing the spread of HBsAg in dialysis units.

Aseptic necrosis after renal transplantation. C. Uittenbogaart, A. Isaacson, P. Stanley, A. Pennisi, M. Malekzadeh, and R. N. Fine. University of Southern California School of Medicine and D & T Program, Childrens Hospital of Los Angeles, California.

Eight of 87 patients developed clinical and/or radiological manifestations of aseptic necrosis after renal transplant. The affected areas were femoral head, femoral condyles, talus, and navicular bones. Pain preceded radiological changes by up to 5 months, though 2 children were asymptomatic. All the affected children had osseous disease before transplant but there was no correlation with dosage of prednisone, hyperparathyroidism, serum Ca or P levels. Rest and reduction in steroids were ineffective and femoral or humeral head replacement was required in 5 children.


The urine of 25 patients and family members with familial haematuria was examined by immunoelectrophoresis for excreted break-down products of glomerular basement membrane (GBM). 6 patients were biopsied and splitting of the lamina densa of the GBM, typical of Alport's syndrome, was found. In 11 cases (including the biopsied patients) GBM products were found in the urine; all had the clinical features of the syndrome.

Studies on urine immunoglobulins in children with pyelonephritis. M. Ignatova, D. Stefani, V. Issachenko, and N. Korovina. Institute of Paediatrics and Child Surgery, Moscow, USSR.

Urinary IgA, IgM, IgG, and IgG-splitting (Fab-Fc-fragments and free L-chains) were assessed in children with pyelonephritis and compared with controls. IgG exceeded IgG-fragment excretion in severe pyelonephritis, IgM was only detected in progressive pyelonephritis with metabolic or obstructive uropathy, and high levels of secretary IgA were found in active pyelonephritis.


Skin biopsies were examined by immunofluorescence in 79 children and 183 adults with various nephropathies diagnosed by renal biopsy. Granular IgA and late acting complement factors were detected in superficial cutaneous capillaries of 45 patients, most of whom had similar mesangial deposits with a clinical diagnosis of anaphylactoid purpura, benign haematuria, and focal nephritis.

New congenital type of primary proximal renal tubular acidosis with extremely low bicarbonate threshold. A. Winsnes and E. Monn. Rikshospitalet, Oslo, Norway.

Intellectual and growth retardation, congenital nystagmus, recurrent coma, and corneal and lens opacities were features of 2 brothers with severe hyperchloraemic metabolic acidosis. Rickets and nephrocalcinosis were absent. Distal tubular acidification appeared normal but bicarbonate infusion showed a threshold of 12 mEq/l. No other proximal tubular defects were present. Erythrocyte osmotic fragility was reduced. Necropsy on one child was essentially normal apart from swollen vacuolated tubular cells.


Two children exhibited growth retardation, polyuria, hypercalcuria, nephrocalcinosis, and metabolic acidosis with high urinary pH. At low plasma bicarbonate levels (7 and 12 mmol/l), the urinary excretion was higher than observed in classical distal renal tubular acidosis. Bicarbonate titration, sodium excretion on a low sodium diet, and hypotonic saline perfusion suggested impaired proximal sodium reabsorption. Acidosis and hypercalcuria improved after hydrochlorothiazide administration.

Renal magnesium wasting with incomplete distal tubular acidosis in sils. F. Manz, K. Scharer, and I. Lombeck. University Children's Hospitals, Heidelberg and Düsseldorf, FRG.
Polyuria, hyposthenuria, nephrocalcinosis, hypercalciuria, low citrate excretion, and low glomerular filtration rate, with interstitial fibrosis and glomerular sclerosis were features of 2 female sibs followed over 8 years. Acid loading suggested incomplete distal renal tubular acidosis. Intestinal calcium and magnesium absorption was increased. Hypomagnesaemia was present with urinary magnesium wasting. It is suggested that the tubular acidosis is the result of a defect in tubular transport of magnesium.


Two children with Fanconi's syndrome, due to glycogen storage disease and idiopathic with bilateral cataracts, were studied. Tubular dilatation, slight interstitial fibrosis, thickening of tubular basement membrane with negative immunofluorescence were found at renal biopsy. Electron microscopy showed a high incidence of giant mitochondria which might be directly associated with the tubular disorder.


Muscle electrolyte content and fluid compartments of 6 children with Bartter's syndrome were compared to 16 controls. Samples were obtained by needle biopsy and analysed by neutron activation. The data showed muscle potassium depletion in untreated children, increased muscle sodium secondary to increased cellular transfer, and low intra-cellular potassium despite normokalaemia and normal total muscle potassium.

**Gradient-type nephrogenic diabetes insipidus.** D. Bretsch, A. Blumberg, and O. Oetliker. University Children's Hospital, Bern, Switzerland.

Four patients with nephronophthisis, oligomeganehphric hypoplasia, nephrocalcinosis, and sarcoidosis had a quantitative defect in urine concentrating ability without response to exogenous vasopressin. Water loading showed a defect in diluting capacity and exogenous vasopressin decreased the diuresis and produced slight urine concentration. These findings indicate a failure to build up an adequate medullary concentration gradient as the cause of the hyposthenuria. This should be termed gradient-type nephrogenic diabetic insipidus.


Three of 21 children, of whom 12 were corticosteroid-dependent, were treated with an average of 2500 mg prednisone for glomerulonephritis and developed bilateral posterior subcapsular cataracts. Vision was not impaired. It is suggested that the observed correlation between length of treatment, total prednisone dose, and the incidence of cataracts may be an indication for cyclophosphamide in children with corticosteroid-dependent nephrotic syndrome.

**Antenatal diagnosis and fetal renal pathology in congenital nephrotic syndrome.** J. Rapola, M. Seppälä, P. Aula, O. Karjalainen, and N-P. Huttunen. Children's Hospital and Department of Obstetrics and Gynaecology, University Central Hospital, 00290 Helsinki 29, Finland.

Amniotic alphafetoprotein (AFP) was measured between the 15th and 20th weeks of pregnancy in 8 women who had previously had a child with Finnish-type congenital nephrotic syndrome (CFN). 7 women with normal levels had healthy babies but one, with increased levels at 15 weeks (150 mg/l) and 19 weeks (190 mg/l), had a termination at 19 weeks. Fetus and placenta were normal but electron microscopy of glomeruli showed absence of normal epithelial cell foot processes with increased mesangial material. It was concluded that measurement of alphafetoprotein is a reliable method to detect CFN.


Thirty-seven children aged 2-26 years with hypertension, from 17 centres, had renal vein renin determinations. Surgery cured 19 of 24 children with improvement in 3; 12 were treated medically. 17 children had a renal vein renin ratio of more than 1.5. 13, all but one of whom had unilateral disease, were cured by surgery, though uninephrectomy failed to cure one. Of 20 with a ratio of less than 1.5, 9 were cured (all had unilateral lesions). One with bilateral renal artery stenosis was not cured, and 10 were treated medically. Routine renal vein renins cannot be recommended for diagnosis and treatment of children with renal hypertension.
Prognosis of Henoch-Schönlein nephritis. M. H. Winterborn, R. Couhanan, H. Swetschin, J. S. Cameron, C. Chantler, D. Turner, and R. H. R. White. Children's Hospital, Birmingham; Guy's Hospital, London.

Eighty-six children with Henoch-Schönlein nephritis (HSN) previously reported in 1972 were reassessed 5 years later. Overall mortality (including renal deaths) had risen from 3.5 to 8%; 4.5% of children with residual proteinuria or haematuria had developed chronic renal failure. Some children had developed hypertension. Contrary to the previous report, children with residual urine abnormalities 2 years after onset of HSN may develop chronic renal failure.

Cryoglobulinaemia in the Henoch-Schönlein syndrome. M. Garcia-Fuentes, C. Chantler, and D. Gwyn Williams. Department of Paediatrics and Renal Medicine, Guy's Hospital, London.

The presence of cryoglobulins was used to assess circulating immune complexes in children with Henoch-Schönlein disease (HSP). Significant titres of cryoglobulins were found in 50% of children investigated within one month of onset of HSP. The cryoglobulins contained combinations of IgG, IgM, IgA, and C3. There was no correlation with renal involvement. Cryoglobulins disappeared as the HSP resolved, but children with persistent renal disease had persistent cryoglobulins.


Three girls aged 3–7 years developed renal failure, 2 acutely. Systemic symptoms were noted in all. In the acute cases serum complement was initially low. Lupus tests were negative. One acute child showed panarteritis of medium sized arteries, the other had endo/extracapillary proliferative nephritis, with 77% crescents, with IgA, C3, and fibrin deposition. The chronic case had focal interstitial nephritis with microaneurisms on arteriography. One child died, the child with crescentic nephritis recovered renal function after 3 months, and the chronic case developed chronic renal failure.


Fourteen children aged 5–15 years had glomerular nephritis with necrotizing angiitis or multiple arterial aneurysms. 13 cases had extra renal symptoms which could mimic Henoch-Schönlein purpura. Renal failure and hypertension were common. Crescentic nephritis with necrotizing lesions was the commonest biopsy finding, but lesions resembling membranoproliferative disease were also seen. Subendothelial Ig and C3, and humps with and without C3 were noted. 8 of 11 deaths were not related to renal failure which though severe could recover.


Four children aged 8–19 years with low α-galactosidase activity were diagnosed as Fabry's disease and presented with pain, angiokeratoma, or cornea verticillata. Renal function was normal. Renal biopsy showed lipid storage with cytoplasmic vacuolization, focal tubular atrophy, and interstitial fibrosis. Dense, osmiophilic lamellated bodies were seen in foam cells on electron microscopy. In 2 children pain improved with carbamazepine or diphenylhydantoin.


Two cases of acute renal failure occurred during recurrence of severe herpes simplex (HSV) type I neuroinfections. New Zealand albino rabbits were therefore infected with HSV type I, and viral antigen was detected in the kidney in 8 of 10 animals, IgG in glomerular basement membrane of 9 of 19 animals, and viruria in 12 of 29 animals. Electron microscopy showed immune complex in the glomerulus.


Experimental evidence suggests that MGN results from deposition of immune complexes on the epithelial side of the basement membrane. 28 of 81 children with MGN had extra-renal disorders: acute hepatitis with circulating HBAg, SLE, sickle cell disease or trait, congenital syphilis, streptococcal sensitivity, thrombocytopenic purpura, proximal tubular dysfunction, myelomonocytic leukaemia, or polyarthritis. Prognosis histology including immunofluorescence and complement studies were similar to other children with MGN. A number of antigens have been shown in MGN deposits and the detection of the responsible antigen may be important for treatment and prognosis.

Medium-term outcome of acute poststrepococcal glomerulonephritis. D. Gill, H. Richardson, and
C. Chantler. Children's Hospital, Dublin. A. Comley, R. H. R. White, and E. Glasgow. Children's Hospital, Birmingham.

Thirty-eight children, mean age 7.7 years, who developed acute poststreptococcal glomerulonephritis (ASGN) were followed for 5–13 years. 2 died in the acute phase, but 33 of the survivors have been reassessed; 2 others are in good health and 1 could not be traced. 3 had microscopical haematuria and 6 had mild proteinuria, one had membranoproliferative glomerulonephritis; glomerular filtration rate was normal in all. No child was hypertensive. The medium-term outcome of ASGN in childhood is good.


Endotoxin has the capacity to induce acute intrinsic renal failure. In 18 children with HUS endotoxin was not found using limulus lysate assay, though it was present in 10 children with Gram-negative septicaemia. Primary endothelial cell damage may be important in HUS but in 4 children with acute HUS circulating endothelial cells could not be found. This may relate to the time of sampling for in rats with generalized Schwartzman reactions endothelial cells were found one hour after injection of liquidou but not after 24 hours.


Aetiological factors were analysed in 57 infants aged less than 2 months with renal thrombosis. In 60–75% an association with concentrated milk formula was noted. Hypertonic dehydration, hyperchloraemia, hyperkalaemia, and acidosis were common. It was concluded that hyperosmolar dehydration and dehydration were the principle causes of renal vein thrombosis in newborns.


Cuprous thiocyanate (CuSCN) was used as a continuous marker to assess intestinal calcium absorption in 15 children aged 9 months to 11 years on 500–900 mg oral calcium intake daily. Plateau of copper excretion started between the 1st and 5th days with almost complete faecal recovery. Cumulative and day-to-day calcium absorption were similar and in good agreement with carmine marker technique. It seems to be possible to limit faecal collection to 1 or 2 days after 4 days of constant diet and CuSCN assimilation.

Assessment of renal function without urine collection. J. P. Guignard, A. Torrado, and E. Gautier. Department of Paediatrics, Centre Hôpitalier Universitaire Vaudois, Lausanne, Switzerland.

Standard inulin and PAH clearance technique was compared to a constant infusion technique (where the rate of infusion equals clearance after constant plasma concentration is reached) in 27 neonates and 25 children. In children the correlation for inulin was 0.74%; in neonates the correlation was 0.76, but the infusion technique overestimated glomerular filtration rate by 108%. PAH clearance by infusion was higher in both groups. The infusion technique is useful where urine collection is difficult and in neonates where it overestimates glomerular filtration rate by a constant factor.


A 7-year-old boy was obese, permanently sweating, had thickened skin, and pyrexia and pseudoparalysis of the limbs. Pain threshold and thirst were diminished. Growth, intelligence, bone age, and pneumoencephalography were normal. Hypernatraemia (169 mEq/l) was present on a normal sodium intake. Plasma ADH levels did not change appropriately with changes in serum osmolality, though they did in relation to hypovolaemia. The essential hypernatraemia is due to the resultant disturbance in thirst control.

Sodium homoeostasis in the newborn. A. Spitzer. Albert Einstein College of Medicine, Bronx, New York.

Enhanced sodium (Na) reabsorption in the distal nephron is present in newborns. Micropuncture experiments in guinea pigs aged 1–56 days showed that the rise in glomerular filtration rate between 15 and 22 days was not accompanied by increased proximal tubular Na reabsorption, though this did increase in older animals. Experiments in puppies aged 2–24 days indicated that plasma renin activity is high but falls in response to intravenous saline. It is suggested that Na deposition in bone from growth stimulates renin release and the resultant increased distal tubular Na reabsorption is important for Na homoeostasis until the proximal tubular reabsorption and intrinsic distal reabsorption catches up with the rise in glomerular filtration rate after birth.

Renal haemodynamics in the perinatal period. A study of lambs. A. Aperia, O. Broberger, P. Herin, and I. Joelsson. Karolinska Institutet, St Goran’s
Hospital, Sabbatsberg's Hospital, Stockholm, Sweden.
Renal blood flow (using microspheres) and filtration capacity (using ferrocyanide injection) were studied in fetal lambs on placental circulation, in the newborn after cord clamping, and during the first 9 days of life. Blood flow did not increase much at birth but did during the first week; cord clamping caused increased outer cortical perfusion and this further increased during the first week, accompanied by increased nephron perfusion. Almost all juxtamedullary nephrons were filtering at birth and 98% of superficial and 100% of juxtamedullary nephrons were filtering at 3 days postnataally.

Correlation between ultrastructure and function during development of renal proximal tubule. L. Larson. Department of Anatomy, Karolinska Institutet, St Gorans Hospital, Stockholm, Sweden.
The ultrastructure, particularly the lateral and basal cell membranes and mitochondria, of rat proximal tubular cells were studied by electron microscopy and correlated with tubular function assessed by fluid reabsorption in isolated nephrons at different ages postnataally. The increases in the area of the membranes and the mitochondria paralleled the increased fluid reabsorption, suggesting that the increase in membrane is a determinant of net fluid transport and the increased energy requirement is reflected by the mitochondrial increase.

Reduction of glomerular filtration rate in rats to 20% of normal was achieved by subtotal nephrectomy and irradiation of remaining renal tissue. Delayed transformation of cartilage into primary spongiosa, with chondro-osteoid, and delayed secondary spongiosa was noted. Osteoid increased but 25-hydroxy-vitamin D3 levels increased. Modest hyperparathyroidism (osteoclast counts; urinary cAMP) was present. Longitudinal growth was normal but weight gain was poor. Reduced calorie and protein intake was the major determinant of growth failure in these rats.

Short case reports

Maximal daily urea synthesis in acute glomerulonephritis. C. Giordano, N. G. De Santo, S. Rinaldi, M. Pluvio, and S. Steri. 1st Medical Faculty, Policlinico, Pizza Miraglia, Napoli, Italy.


Transient distal renal tubular acidosis: Lightwood's syndrome? E. P. Leumann and K. Baerlocher. University of Zurich and Children's Hospital of St. Gallen, Switzerland.


Charcot-Marie-Tooth disease and chronic nephropathy. Th. Lennert, F. Hanefeld, and J. Bernstein. Kinderklinik, Frei e Universität Berlin, Germany, and William Beaumont Hospital, Royal Oak, Michigan, USA.