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Autosomal Dominant Inheritance of the Idiopathic Hypercalcemic Syndrome

A 16-month-old girl with idiopathic hypercalcemia /failure to thrive, characteristic face, supravalvular aortic stenosis/ has been under observation for 12 months. Her serum calcium level was between 14 and 19 mg per 100 ml, which could not be decreased permanently by any therapeutic efforts including administration of Calcitonin, Prednisolone, Furosemide, sodium sulphate, phosphates and a diet of low calcium intake. Both her father and a 17-year old brother were mentally retarded, and had calcium deposits in their corneae. Their serum calcium values were 11.9 and 13.0 mg per 100 ml, respectively, with increased urinary excretion of calcium. Two other members of the family on the paternal side were also suspicious of the syndrome, while the mother proved to be normal. The family histories are suggestive of autosomal dominant inheritance of the disease.

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Further studies on the pathogenesis of hypocalcemia in primary hypomagnesemia. Evidence for a defect in the relase process of parathyroïd hormone(PTH). L. DAVID* MC. CHAPPUIS*et R.FRANCOIS; Lyon France.

Recent studies by Anast et al (Science 117-606 1972) and by Suh et al (J.Clin.Invest. 52-153 1973) have shown that severe magnesium deficiency induces impaired parathyroïd function and that this impairment is responsible at least in part for the secondary hypocalemia However it is not known if the synthesis or the release of PTH or both, are affected in this syndrome. The present study was undertaken to investigate the type of impairment of the parathyroïd function during hypomagnesemia.

The clinical course of the 5 years old boy in whom the present investigations were carried out have been reported previously (Pouillaude et al. Arch.Franc.Ped.28-1021-1971). During the hypocalcemic and hypomagnesemic state we confirmed that the bone system and the kidneys were responsive to exogenous bovine PTH as demonstrated by the normalisation of the calcemia and the increase of urinary hydroxyproline and c AMP. Low to non detectable levels of serum immunoreac tive PTH gived also clear indication of impaired parathyroïd function. By contrast IV injection of SO4 Mg induced an instantaneous and sharp increase of serum IPTH as shown by levels above the normal range 10 mn after the injection. This was followed by a regular decrease of IPTH level reaching the normal range 2 hours later. We believe that this finding favors the view that magnesium depletion induces impaired PTH release in man.

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Effect of parathyroid extract (PTE) on electrolytes, phosphorus and protein content of parotid saliva.

The excretion pattern of calcium (Ca), magnesium (Mg), potassium (K), sodium (Na), phosphorus (P) and protein of the parotid saliva was studied before and during PTE administration in 6 normal subjects aged 24 to 32 years. The Stensen's duct was cannulated and the flow rate of saliva measured before and after stimulation by lemon juice. After a control period, 50 U of PTE were given i.v. followed by an infusion of 2 U/min. for 60 to 90 min. In the absence of any change in serum Ca and P, PTE induced a significant increase of salivary Ca ($p < 0.001$), P ($p < 0.005$) and protein ($p < 0.001$) concentrations. These changes were independent of the flow rate. Na excretion was also increased ($p < 0.005$) but only at high flow rates. PTE had no significant effect on Mg and K concentrations. These results provide evidence that PTE acts on the parotid gland by affecting Ca and P transport and protein secretion.

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EVIDENCE FOR TWO PARALLEL SODIUM PATHWAYS ACROSS ILEUM.

Intestine activity is characterized by an efficient absorption of nutrients from lumen to blood; it is also characterized by large transmural fluxes of water and electrolytes and low electrical resistance. The effect of diffusional forces on net Na transport was therefore investigated. Rabbit ileum was mounted as a flat sheet between two chambers. Temperature, pressure and electrolyte solution (Ringer +10 mM glucose) were identical on both sides of the epithelium. Transmural Na fluxes were studied as a function of applied electrical potential, ψ_{ms}, (from +9 to -9mV). The net Na flux, J^{Na}_{net}, was found to be a linear function of ψ_{ms} ($J^{Na}_{net} = -0.20\ \psi_{ms} + 1.89$; $r = 0.99$). In absence of electrochemical potential difference J^{Na}_{net} had the value of 2.07 ± 0.79 µeq/hr cm^2). Each transmural unidirectional Na flux comprised a PD-dependent, J_d, and a PD-independent flux, J_c. In absence of electrochemical potential the ratio of J_d from mucosa to serosa/J_d from serosa to mucosa was 1.04, and the difference between J_c from mucosa to serosa minus J_c from serosa to mucosa was 2.07 µeq/hr cm^2. These results clearly indicate the presence of two parallel pathways for Na across rabbit ileum. It is proposed that the PD-dependent pathway represents a paracellular pathway and the PD-independent pathway represents a cellular pathway.