153 S. Garne, W. Petersen, C. Heilmann, O. Andersen, K.E. Petersen, B. Marner, C. Binder and J. Nerup. University of Copenhagen, dep. of Pediatrics G, dep. of Gynecology and Obstetrics, Rigshospitalet and Steno Memorial Hospital, Copenhagen, DENMARK.

BETA-CELL FUNCTION IN CYSTIC FIBROSIS.

Little is known about beta-cell function in patients with advanced cystic fibrosis (CF). To study this 22 CF patients (12 males, 10 females, age 15.1 years (10-17 years)) who never had glucosuria were compared with 21 age and sex matched healthy children (controls). All controls had normal oral glucose tolerance tests (OGTTs), but 10 (45 per cent) of the CFs had abnormal OGTTs, 8 impaired glucose tolerance and 2 diabetic. The area under the glucose curves in CFs with normal OGTTs was larger than that of controls ($p<.05$). Insulin and C-peptide responses were reduced and delayed in all CFs. Abnormal HbAlc values (>6.4 per cent) were found in 7 (32 per cent) of the CFs, but in no controls. Insulin receptor-binding to erythrocytes was similar in CFs and controls. Islet cell antibodies were negative in all CFs and controls.

Conclusion: Abnormal beta-cell function is frequently present in CF. With increasing longevity of CF patients, this may represent an important problem.

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EVOLUTION OF PLASMA PURIFIED CALCITONIN LEVELS IN CHILDREN.

CT radioimmunoassays are hampered by the heterogeneity of the hormone that often causes results not consistent with clinical findings. For this reason a purification method based on reversed phase chromatography was developed and purified CT (pCT) measured in parallel with non purified CT (nCT) in the plasma of 145 normal children and adults in occasion of routine samples for minimal pathologies. Results ± SD in pg/ml were:

Age	5 days	1-10 month	1-3 year	3-8 year	Adults
N	10	25	30	40	40
nCT	110±30	56.6±18.6	63.4±18.8	82.8±34.7	56.6±18
pCT	25.3±9	49.8±5.6	46.4±20.2	44.5±25.1	15.1±40.6

In addition in 11 athyreotic children aged 1 to 6 years nCT were 64.4±21 and pCT 52.4±21.6 pg/ml. The data show 1) newborns have significantly lower pCT compared to children ($p<0.01$), 2) nCT/pCT ratio is higher in children than in adults (0.59 vs 0.26, $p<0.01$),3) athyreotic patients have normal CT levels.

In conclusion higher levels of pure, and probably more active CT are present at time of active bone mineralisation independantly of the presence of the thyroid gland.

154 STEROID CELL AND ADRENAL ANTIBODIES IN AUTOIMMUNE POLYENDOCRINOPATHY - CANDIDOSIS - ECTODERMAL DYSTROPHY (APECED)

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We studied steroid cell (ovary, testis, placenta) (SCA) and adrenal (AA) antibodies in 311 serum samples collected from the Finnish whole-population-based series of 47 patients with APECED during 1.3-13 (mean 7.3) years. 15 patients had no antibodies. 3 patients had AA alone, 28 had AA+SCA and 1 patient had SCA alone. AA and/or SCA were observed in 30/34 patients with adrenocortical failure (A) and in 4/13 without A ($p=0.0002$), in 13/13 females with ovarian failure (O) and in 7/13 females without O ($P= 0.01$). A was observed to develop in 12/25 patients. AA or SCA appeared in 11/12 of them in contrast to 4/13 of patients maintaining normal adrenal function ($p=0.003$). O was observed to develop in 10/23 patients. In 10/10 of them AA and SCA appeared in contrast to 7/13 females maintaining normal ovarian function ($p=0.02$). SCA were detected in 29/47 patients, in contrast to 6/7 patients of Elder (JCEM 1981:52:1137) who all had a multicomponent disease. 5/16 of our patients with no or 1 endocrinopathy, 8/13 of those with 2 and 16/18 of those with 3 or more endocrinopathies had SCA. Thus SCA were commoner in patients with multicomponent endocrinopathy ($p=0.003$).

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VITAMIN D METABOLISM IN MALE PUBERTY

The increased growth and mineralization together with higher serum levels of $1.25(OH)_2D_3$ in puberty compared to adult levels may indicate a regulatory role of sexhormones in vitamin D metabolism. Twenty pubertal boys, aged 11.0-12.6 years, were included in a 2-year study. Local bone mineral content (BMC), serum levels of testosterone (T), $25OHD_3$, $1.25(OH)_2D_3$, $24.25(OH)_2D_3$, and $25.26(OH)_2D_3$ have been determined at 3 months intervals. A curve fitting analysis was performed to define the time (t_m) at which T and BMC displayed the maximal increase. t_m for T occurred about 5 months before t_m for BMC. The seasonal variation in the vitamin D metabolites was taken into account by calculating the ratios to $25OHD_3$. No significant changes (Student's t test for paired data) in neither $1.25(OH)_2D_3$, $24.25(OH)_2D_3$ nor $25.26(OH)_2D_3$ could be found within a period of 1 year before to 1 year after t_m for T and BMC.

The data demonstrate that the changes in testosterone secretion during puberty seem to have no significant short-term influence on vitamin D metabolism.

155 A REDUCTION IN CRANIAL SCLEROSIS DURING CALCITRIOL THERAPY IN A PATIENT WITH CRANIOMETAPHYSEAL DYSPLASIA L. Lyndon Key, Jr. Roland Baron, and Constantine S. Anast Harvard Medical School, Department of Pediatrics, Boston, MA and Yale University Medical School, Department of Internal Medicine, New Haven, Conn. USA

A 1 month old male presented with a left facial nerve palsy, hepatosplenomegaly, and sclerosis of long bones and the base of the skull. The original diagnosis of osteopetrosis was revised to craniometaphyseal dysplaisa (CMD) based on mandibular sclerosis and normal density of the vetebrae. by 2 months of age, hepatosplenomegaly had resolved, but the base of the skull had become more sclerotic and there was no improvement of facial nerve function. Parameters from an iliac crest biopsy included: trabecular bone volume, 15.1% (25+3%); osteoclast surface area, 2.6% (1.1+0.3%); and osteoclast number/mm trabecular bone surface, 0.37 (0.12+0.02). All these parameters differ significanly from childhood norms (+SD). Osteoblastic parameters were normal. The patient was begun on high dose (1.5 mcg/kg/d) calcitriol (Hoffmann-La-Roche) and a low calcium (15 mg/kg/d) diet (Ross Laboratories). After 7 months of therapy, the facial nerve palsy had improved. Raiographs of the base of the skull demonstrated decreased density. A CT demonstrated increased size of optic foramina and internal auditory canals. A reapeat iliac crest biopsy showed decreased osteoclast number and increased trabecular bone volume.

In CMD, sclerosis of the cranium has increased throughout early childhood in all reported cases. In our patient, high dose calcitriol reversed the cranial sclerosis without stimulating excessive bone loss from other, less sclerotic bones.

158 RENAL EFFECT OF PHYSIOLOGICAL AND PHARMACOLOGICAL DOSES OF HUMAN PTH, 1-34, IN MEN

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Recent study of Puschett et al. (Mineral Electrolyte Metab. 6:190, 1981) showed that infusion of bovine PTH at physiological dose (0.3-0.5 u/kg) in thyroparathyroidectomized dogs produced phosphaturia and a reduction in urinary Ca excretion, but no increase in urinary excretion or renal tissue content of cAMP, suggesting that some effects of PTH may be mediated by mechanisms other than cAMP. In order to assess whether similar phenomena can be observed in human, we infused human PTH, 1-34, at 0.3 u/kg and 3 u/kg in 5 normal adults and measured urinary cAMP (UcAMP), %TRP, Ca, Mg, Na, K, and plasma cAMP (PcAMP) in the next 3 hours. As shown in the table, PTH at 0.3 u/kg induced phosphaturia of a similar magnitude and a comparable urinary excretion of electrolytes to that of 3 u/kg but PcAMP and UcAMP were markedly different. After 0.3 u/kg, there was no rise in PcAMP, and a small rise in UcAMP indicating a difference in handling and production of cAMP after physiological and pharmacological dose of PTH.

PTH (u/kg)	ΔPcAMP (pmol/ml)	ΔUcAMP (nmol/min/100mlCcr)	ΔTRP (%)
0.3	1.5±3.1	1.6±0.3	-9.1±1.1
3.0	52.6±5.0*	117.0±30.9*	-8.9±1.8

* $P<0.05$

Δ=Post - Pre PTH