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Calcitonin (pCT) in the first hours of life.

High values of immunoreactive CT (iCT) found in newborns are yet inexplicated. The results of the iCT assays are impaired by the immunoheterogeneity of the hormone causing a discordance of the results obtained with different antisera. A purification by hydrophobic interaction chromatography may yield a high accuracy of the determation of the hormone. iCT, pCT and T3, T4, TSH were assayed in the cord and at 3.12.24.48 hours from delivery in 15 newborns, hospitalized for nonendocrine diseases. 1 ml blood/point was drawn only if venipuncture was otherwise clinically indicated. iCT values (cord 135 ± 33.6 pg/ml) significantly increased ( $p < 0.01$) at 12h (402 ± 53) and persisted high at 24h (611 ± 66) and at 48h (371 ± 43). pCT significantly increased ($p < 0.01$) at 3h (from 17.3 ± 4.0 to 28.5 ± 6.0) with a peak at 12h (100 ± 28.2) and decreased at 24h (72 ± 28.2) and at 48h (33 ± 2.8). In conclusion: a peak of CT in the neonate seems to be related to the stress typical for delivery.

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*Delayed puberty in pseudohypoparathyroidism type I (PHP-I).*

*PHP-I is a genetic disease characterized by target organ resistance to the action of parathyroid hormone (PTH) and sometimes of other peptide hormones. Recent evidence suggests that the adenylate cyclase defect in PHP-I resides in the guanine nucleotide binding regulating protein (N-protein), which mediates the response of the adenylate cyclase to peptide hormones.*

*The present study describes a 15 yr old girl (karyotype XX) with hypocalcemia, elevated serum iPTH, decreased urinary cyclic AMP response and Albright's osteodystrophy who presented with delayed puberty. Despite accelerated bone age (17 yr at the chronological age of 13 yr) the girl did not go beyond a Tanner stage III. Serum estradiol was low (20-35 pg/ml) despite elevated basal and LHRH-stimulated LH and FSH secretion suggesting partial gonadotropin resistance of the ovary. Target organ resistance to the action of TSH, ACTH and ADH as well as prolactin deficiency could be excluded. Measurement of N-protein activity (kindly done by H. Radeke, Hannover, FRG) in membrane extracts of erythrocytes, using cyc⁻ S49 mouse lymphoma cells gave normal results (105.9 %, normal 100 ± 40 %). It is concluded that the disease in this girl is caused by a molecular defect different from other patients with PHP-I affecting accumulation of cyclic AMP in the target organs of PTH and the gonadotropins.*

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Parathyroid hormone, calcitonin and 1,25 $(OH)_2D_3$ levels in betathalassemia major.

Pathogenesis of osteoporosis in beta-thalassemia is still nuclear. In 13 thalassemic children: (7,aged 3 to 5 yrs, group 1) and (6aged 10 to 13 yrs, group 2), who had never had vitamin D therapy, we determined serum Ca, P, PTH, CT, 25 $OHD_3$ and 1,25 $(OH)_2D_3$ levels both in winter and in summer, in comparison with 2 groups of 14 controls of the same age. The results were similar in thalassemic and in controls. In thalassemic patients of group 2, serum PTH (0.61±0.43 ng/ml), 25 $OHD_3$ (23,9±11.2. ng/ml), 1.25 $(OH)_2D_3$(14.9±6.7 pg/ml) levels were lower than in controls (PTH: 1.52±0.47ng/ml;25 $OHD_3$: 32.7±12.4 ng/ml 1,25 $(OH)_2D_3$: 36.1±13.7 pg/ml), whereas serum CT levels (305±79pg/ml were higher than in controls (107±35pg/ml). No significant differences exist between winter and summer; only 25 $OHD_3$levels were higher in summer than in winter in thalassemic patients and in controls. Conclusions: advancing age induces in thalassemic patients a decrease in PTH secretion and consequently a deficit in synthesis of 1.25 $(OH)_2D_3$.This may explain some aspects of osteoporosis, which CT hypersecretion may tend to counter to.

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Urinary cyclic AMP in spot urine of healthy children.

Reference values for the excretion of cyclic adenosine 3´, 5´monophosphate (cAMP) in children are few. Data are mostly based upon a 24 h urine collection We measured the urinary cAMP by a competitive protein-binding assay. Reference values for the urinary cAMP in spot urine obtained in the morning from 142 healthy children aged 2 - 2oo months are reported. A significant positive correlation between urinary cAMP and creatinine excretion appeared (r o.68, p<o.ol), and for which reason the cAMP excretion was creatinine corrected (cAMP/crea.).

The mean value (and 95% significance limits) for (cAMP/crea.) were 747 µmol/mol (254-22o6 µmol/mol).

A logarithmic transformation of the (cAMP/crea.) ratio showed an even distribution. Log (cAMP/crea.) values were related to age or body surface with decreasing values for increasing age or body surface area (r-o.55 and -o.57 respectively, p<o.ool).

It is concluded that spot urine for measurement of urinary cAMP excretion appears simple and preferable as compared to a 24 h urine collection in children.

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Hormonal and DNA studies in XX males.

8 male patients with 46XX caryotype were studied at age 1 mon. to 15 yrs. 3 had normal genitalia, 5 had hypospadias, 4 with cryptorchism. Biopsy performed in 5 showed normal testis before age 8 and testicular dysgenesis at age 8-12 yrs, without any ovarian structure. Follow-up for 5-18 yrs was obtained in 6: puberty was normal up to stages 3-5, with small testes in all, gynecomastia in 4, final height 163 cm, normal mental development in 5 and slight impairment in Hormonal studies: 1) before puberty, (5 patients) plasma FSH and LH (basal and peak after LH-RH) and testosterone (T) were normal; 2) during puberty (n=5) FSH and LH were increased with normal T; 3) 2 adult patients had very high gonadotropins with decreased T, showing that XX testes go to pubertal involution.

Chromosomal studies of the 8 patients failed to show any Y material. However DNA, studied in 3 patients by means of DNA probe, showed specific sequences of chromosome Y. One more patient studied by immunological method had HY antigen. These data demonstrate translocation of Y material on another chromosome.

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Biologic and immunologic measurements of serum erythropoietin (EPO) in children.

Within recent years different test systems have been developed for EPO determination basing on either its biologic(b)activity or its immunologic(i)criteria. To eludicate the relationship of these two test principles an in-vitro bioassay, measuring 59-Fe incorporation in hematologically active liver cells of 15 days old fetal mice, was compared with a hemagglutination inhibition test.Only b-EPO,significantly(r=-0.96) correlated with hemoglobin(Hb)concentration in normal and anemic children without renal failure(n=30). In spite of a greater variation of i-EPO, median values in normal children did not significantly differ from each other in both test systems. In renal failure EPO activity was decreased with mean i-EPO higher than b-EPO,only the latter,however,significantly correlating with Hb and its half saturation pressure in contrast to i-EPO. Accordingly no relationship between b-EPO and i-EPO could be detected(r=0.08). Data suggest that b-EPO,as measured by the fetal mouse liver cell test, determines biologically valid EPO concentration, whereas i-EPO,as measured by the hemagglutination inhibition test, additionally detects inactive EPO fragments especially accumulating in renal failure.