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Radioimmunoassay of urinary LHRH.

LHRH immunoreactivity in the urine has been reported with suggestions that the excreted product is intact LHRH or a 2-10 or 3-10 fragment. To better elucidate the unclear nature of urinary LHRH, we have developed radioimmunoassay systems utilizing 8 antisera. Using 30 analogs of LHRH, we tested each antiserum for binding characteristics. Three, the highly specific Niswender R-42 and the two $Kelch_{13}$ antisera appeared to require the entire molecular configuration for activity; two required only 3-4 C-terminal amino acids; and three bound non-specifically to different portions of the sequence. Utilizing the sensitive $Kelch_{13-10}$ antiserum, basal immunoreactive urinary LHRH in a normal man was 8 pg per ml of unconcentrated urine. After an I.V. injection of 50 µg LHRH, the concentration of urinary immunoreactive LHRH was 430 pg/ml in the first hour. An excretion rate of 101 ng/hr was found in the first hour, falling to 6.5 ng/hr by the 4th hour, with a total of 150 ng excreted in 4 hours. It is concluded that LHRH immunoreactivity is detectable in the urine in the basal state and after a bolus injection.

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PLASMA PROLACTIN (PRL) BEFORE AND AFTER TRH IN 23 CHILDREN WITH CRANIOPHARYNGIOMA.

PRL has been determined before and after TRH IV (200 ug/m^2) in 23 children (11 ♀/12 ♂) with craniopharyngioma using reagents provided by the NIAMDD, Bethesda USA. As controls served 10 endocrinologically healthy children (4♀/6♂), aged 4-16 yrs, who had TRH tests for other reasons. 13 patients, aged 1-13 yrs, have been studied before surgery, 10 of them and 10 additional patients, aged 1-20 yrs, after surgery.

RESULTS:	PRL ng/ml	n	0'	20'	60'
controls	($\bar{x}\pm1$ SD)	10	4.9 ± 2.0	18.8 ± 4.7	10.6 ± 3.1
patients preop.(n=13)	normal	6	6.2 ± 1.9	14.2 ± 4.3	11.4 ± 3.9
	no rise	3	6.1 ± 2.0	7.3 ± 2.1	6.2 ± 1.2
	elevated	4	17.9 ± 3.7	29.9 ± 2.7	24.7 ± 7.9
patients postop.(n=20)	normal	6	6.9 ± 3.3	15.9 ± 8.3	11.8 ± 5.0
	no rise	2	4.3 ; 3.0	5.8 ; 4.8	4.7 ; 4.5
	elevated	6	38.9 ±31.5	47.5 ± 27.4	44.3 ± 28.3
	deficient	6	0.7 ± 0.3	1.1 ± 0.6	1.0 ± 0.6

Assuming that PRL secretion is controlled by PIF (PRL inhibiting factor), elevated PRL levels as seen before and after surgery would indicate hypothalamic disorder or disruption of the hypothalamo-pituitary axis. PRL deficiency, observed only after surgery, would reflect pituitary damage.

68 M. ROGER*, J.-C. CHAUSSAIN, M.-C. FEINSTEIN-SOLDAT*, J-E. TOUBLANC* and P. CANLORBE. Fondation de Recherche en Hormonologie et Hôpital Saint-Vincent-de-Paul, Paris, France. Prolactin response to TRH in children with hypopituitarism.

Prolactin (PR) and TSH responses to IV TRH (0.2 mg/m2) were studied in 49 hGH deficient children (33 boys, 16 girls): 29 tumoral hypopituitarisms (TH) including 20 craniopharyngioma (5 before and 15 after surgery) and 20 idiopathic hypopituitarisms (IH). Results were expressed as ng/ml of NIH hPR F3. In IH, the mean PR basal value was 19.8 (range from non detectable ND to 60); 5 values were above the 95 th. percentile of normal range; the mean peak value was 41.6 (range 5 - 114, 6 values were under the 5 th. percentile). In CP, the mean PR basal value was 6.9 (range ND - 16) and the mean peak value was 15.8 (range ND - 33, 12 values under the 5 th. percentile). In other TH, the mean basal value was 21.9 (range 3 - 55) and the mean peak value was 42 (range 9 - 90, 3 values under the 5 th. and 3 values above the 95 th. percentiles). In both groups no correlation was found between PR responses and TSH responses or thyroxine (T4) levels. However, in IH normal or high PR responses were always associated with normal or high TSH responses, while low T4 levels were encountered in 7 subjects of this subgroup. In CP, low PR responses with low TSH responses and low T4 levels were observed only after surgical removal. These data demonstrated that PR response to TRH is normal in about 50 % of hGH deficient children, while low responses are frequently observed in CP before and after surgery. Elevated basal and peak levels in some IH or TH lead to speculate a prolactostatin (PIF) deficiency.

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Prevalence of b^o and b^+ thalassemia genes in Greek patients with thalassemia.

In an attempt to estimate the prevalence of b^o and b^+ thalassemia genes in Greece chromatographic analysis of hemoglobins was performed in 30 patients with thalassemia prior to any transfusion. In 12(40%) no Hb A was detected, suggesting the presence of b^o gene in the homozygous state (b^o / b^othal.) In the remaining 18, Hb A showed a bimodal distribution with values ranging from 4-35%. The detection of Hb A suggests the presence of b^+ gene, while the bimodal distribution could be explained by the assumption that the b^+gene in single dose and in combination with b^o gene (b^o/b^+thal) results in the production of small amount of Hb A ranging from 4-12%, (first curve), while in double dose (b^+/b^+thal.), in the production of higher amounts, of Hb A ranging from 18-35% (second curve). The b^o/b^+thal., was observed in 11(37%) and the b^+/b^+thal., in 7 (23%).

It is concluded that both b^o and b^+genes are common in Greece and chromatographic analysis helps to determine the genotype.

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HLA antigens, serum and salivary amylase activity and serum amylase isoenzymes in homozygotes and heterozygous-gene-carriers of cystic fibrosis.

12 homozygote patients and 32 heterozygous-gene-carriers from families with cystic fibrosis /CF/ were HLA-typed for 29 HLA-antigens. The HLA-B18 proved to be more frequent in the groups of homo- and heterozygotes 5o and 31 per cent respectively compared with 14 per cent in a control group of 13o healthy blood donors*. Serum and salivary amylase activity were investigated with the using of Phadebas-amylase method. The average of total amylase activity in serum was 269 U/l±113,7, in the CF homozygotes' group, in the heterozygotes' group was similar as in the healthy persons /21o.7 U/l±86/. The CF homozygotes' salivary amylase activity was more increased*/148.7 U/l±65.1/ than in the heterozygotes' group /118.2±67.o/ and in the control ones /97.7±34.9/ The salivary amylase activity was moderately elevated in the cases of CF heterozygotes without the change of the serum total amylase activity. The amylase isoenzymes were identificated with flat-bed polyacrylamide gel-electrophoresis. Significant differences have been found in the distribution of amylase isoenzymes too.

* Change statist significant.

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The diagnosis of mucolipidosis II in a pregnancy at risk by studies on amniotic materials.

Mucolipidosis II (ML II, I-cell disease) is a fatal inborn error of metabolism with altered levels of certain lysosomal hydrolases. In a pregnancy at risk, in which ML II was suspected, we were able to monitor the activity of several lysosomal hydrolases in cultured amniotic fluid cells and in amniotic fluid. In amniotic fluid an increase in hydrolase activity was observed, whereas cultured amniotic cells showed a decrease, together with an accumulation of ^{35}S-labeled glycosaminoglycans (^{35}S GAGs). The pregnancy was therefore interrupted in the 22nd gestational week. The diagnosis was clearly confirmed by analysis of material from the aborted fetus. In fetal serum the activities of several lysosomal hydrolases were elevated approximately 10 to 70-fold, whereas the enzyme levels in fetal fibroblasts were considerably reduced. There was also an excessive accumulation of ^{35}S GAGs in fetal fibroblasts. Our results, and those of others (Aula et al, J.Pediat. 87, 1975; Huijing et al, Clin.Chim.Acta 44, 1973; Matsuda et al, Humangenetik 30 1975), indicate that increased levels of these enzymes in amniotic fluid may suffice as a prenatal diagnosis of ML II. However, due to considerable variations between individual samples, the diagnosis should also be confirmed by analyses of these enzymes and by ^{35}S incorporation studies in cultured amniotic cells.