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Growth Hormone in Silver Syndrome

Eight children with typical features of Silver syndrome are described(6 girls and 2 boys,2 girls were twins).Stature was from -11% to -19% retarded.Only one of twin girls was at 3rd centile.Delay in skeletal maturation was found in 7 patients.Growth hormone(g.h.)levels were estimated after insulin hypoglycaemia(o,1 to o,15 units/kg)in all patients.Fasting g.h. values were from 1,o to 9,6 ng/ml,mean 3,4 ng/ml.G.h. reached maximum of 24,o-3o,o-5o,o ng/ml at 30 min in 3 patients.In other 5 cases there was less marked increase of g.h.:7,o;7,9;8,1;9,5; 1o,o ng/ml,with peak values after 3o or 45 min. Mean peak serum g.h. concentration were 18,3 ±16,7 ng/ml(range 7-5o).In a control group(n=6) mean fasting g.h. values were 5,3 ng/ml(1,2-11,7) and a mean peak 25,8±8,7 ng/ml(range 15,1-4o,5). Analysis of results demonstrated smaller g.h. response to insulin hypoglycaemia and decreased fasting g.h. values in patients with Silver syndrome, compared with control group.

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HbA_{1C} determined by colorimetric method in the management of juvenile diabetic children.

HbA_{1C} was determined by a modification of a colorimetric method using thiobarbituric acid. The mean percent of HbA_{1C} in a control non-diabetic population (n=26) was 6.44±0.2 (mean±SEM). The mean HbA_{1C} percent of the diabetic patients group (n=40) was 9.31±0.38 (mean±SEM) and was significantly different from the normal group ($P < 0.001$). There was no correlation between fasting blood (37 patients) and urinary (36 patients) sugar determination and HbA_{1C} conc. obtained the same day (r=0.10 and r=-0.12, respectively). However, serial HbA_{1C} determination over a 4-month period in 15 patients revealed a good correlation with the individual status of diabetic control, as evaluated by clinical and laboratory criteria. HbA_{1C} colorimetric determination seems to be a simple, reproducible method which provides the clinician with an objective,useful indicator of carbohydrate control

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Long-term treatment with high dose gonadotrophin releasing hormone to induce puberty.

Gonadotrophin releasing hormone (LHRH, Hoechst) 500 µg has been administered subcutaneously twice daily to four patients (two male, two females) with hypogonadotrophic hypogonadism due to LHRH deficiency for a minimum of one year. The initial results were extremely encouraging in terms of the biochemical responses and physical changes but in all four patients gonadotrophin responsiveness waned, levels of sex steroids were not maintained and pubertal development failed to progress.

The diminishing responsiveness of the pituitary to continual stimulation with LHRH appears severely to limit the use of LHRH as a therapeutic agent, although it is possible that smaller doses given more intermittently may be effective. Theoretical considerations and experimental data obtained in dogs suggest that this may be the case.

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Determination of serum T_4 and TSH in blood dot for newborn screening

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In order to assess the efficacy of newborn screening for congenital hypothyroidism serum T_4 and TSH have been analysed by taking a dot of blood in filter paper from 71 newborns from Athens and 68 from the country. In selected cases the results of the dot paper have been compared with those obtained by conventional methods. Of the newborns from Athens, 9 out of 71 (12,6 %) had a serum T_4 below 7.0 µg/dl, 25 (35,2 %) above 12 µg/dl, but none a serum TSH above 50 mU/l. Of the newborns from the country, 11 out of 68 (16,1 %) had a serum T_4 below 7.0 µg/dl and 21 (30,9 %) above 12.0 µg, but again none a serum TSH above 50 mU/l. The cases with abnormal results are being sought for a more complete evaluation. The results until now suggest that serum determinations of T_4 but not of TSH overestimate the frequency of congenital hypothyroidism in areas with iodine deficiency.

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Glycosylated Hemoglobin (GHb) as an index of diabetes control in diabetic children and adolescents.

Increased levels of GHb (HbA1a-c) serve as an indicator of the degree of hyperglycemia over the life span of the erythrocyte. Blood GHb was measured in 29 normal (N) and 50 diabetic children and adolescents (D) in the postprandial state. GHb was determined by cation exchange chromatography with "Isolab" columns. In the N subjects mean GHb was 7.7±1.1% of total HbA (Range: 6.0-9.5%). In the D patients the range was 7.7-20.1%. A significant negative linear correlation ($r=0.68$ $p < .001$) was found between GHb and urine glucose excretion measured 3 times daily for 3 months. Significant differences in GHb were also found when the patients were grouped according to the frequency distribution of the daily urine glucose excretion; GR 1: 0-½% - GHb=9.9±1.8%; GR 2: 1-2% - GHb=12.2±0.9%; GR 3: 2-4% - GHb=14.0±1.9%; GR 4: ≥ 5% - GHb=16.7±1.8% ($p < 0.01$ - 0.001). A significant curvilinear relationship was found between GHb and postprandial blood glucose levels ($y=a+b \ln(x)$ $r=0.6$ $p < .001$) suggesting that GHb reflects a saturable system of glycosylation. Concentration of GHb serves as a useful index for diabetes control.

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Pituitary adenoma: hipersecretion of GH pre and post-surgical operation.

Male 13,2/12 years old.Height: 187,0 cm (P 97 Tanner). Abnormal growth since seven years of age. Emergency admision by intracraneal hypertension. After neuroradiological (neumo and arterial) and gammagraphic studies, a diagnosis of supraselar tumor was done.

Hormonal studies showed hyperproduction of GH (basal: 220-max: 277,5 ngr/ml; insulin test). After oral administration of glucose there was not supression of serum GH levels (basal: 220-minimal: 150 ngr/ml).

After surgical intervention the conventional and ultraestructural studies of the tumor demonstrated the presence of a GH-hypersecretor adenoma. Seven months latter, the GH levels were high and other pituitary hormones were low.