29 PLASMA ADRENALINE RESPONSE IN KETOTIC HYPOGLYCEMIA. INVESTIGATIONS IN PATIENTS WITH OR WITHOUT ADRENAL CALCIFICATIONS.

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In some patients with ketotic hypoglycemia a reduced urinary output of catecholamines has been reported. Emplying a double isotope derivative technique (Clin. Sci.1973,45:163) plasma adrenaline and noradrenaline were determined in six children with ketotic hypoglycemia. One of the patients had extensive bilateral adrenal calcifications. Adrenal cortical insufficiency was excluded.
The plasma adrenaline values (mean value,ng/ml) before - during hypoglycemia were: (A) in patients without adrenal calcifications 0.05-0.10, (B) in one patient with adrenal calcification 0.01-0.04, (C) in healthy children (n=3) 0.22-1.33, (D) in healthy adults (n=5) 0.09-0.69. The plasma noradrenaline changed insignificantly in patients and controls.
Estimated from blood glucose level during hypoglycemia the plasma adrenaline was only four per cent of the expected values.
These preliminary results seem to indicate that in patients with ketotic hypoglycemia the immediate plasma adrenaline response during hypoglycemia is greatly reduced in patients with as well as without adrenal calcifications.

30 SCREENING FOR CONGENITAL HYPOTHYROIDISM.

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The early diagnosis of congenital hypothyroidism relies essentially on the biological evidence of the defect. Preliminary data of a systematic screening of neonates are presented. The values of serum thyroxine (T4) and TSH were determined in the cord blood of 271 infants. In 152 of these children serum TSH was again measured on a finger puncture sample at 5 days of age. Serum T4 was measured by the competitive binding method of Murphy and Patty. Serum TSH was determined by double antibody radioimmunoassay with the TSHk of CEA-IRE-SORIN. Results are as follows :

	Birth (cord blood)	5th day
Thyroxine µg/100 ml	11.9 ± 2.6a (249)b 6.7 - 24.6c	-
TSH µU/ml	7.6 + 5.1 (271) 1.5 - 31.5	3.5 ± 1.9 (152) 1.5 - 12.1

a : mean ± SD b : number of cases c : range

At birth the variability is important for T4 and TSH. On the 5th day the scattering of TSH values is much smaller and the mean is lower than at birth (< 0.001). In agreement with previously reported data we found serum TSH levels of 160 and 500 U/ml in 2 hypothyroid infants younger than 1 month. The striking difference between these high TSH values and the control values observed on the 5th day allow to support the view that this screening procedure is reliable for detecting congenital hypothyr.

31 NEONATAL DETECTION OF HYPOTHYROIDISM BY RADIOIMMUNOASSAY OF THE THYROXIN IN THE ELUATE OF DRIED BLOOD.

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The assay of Thyroxin (T_4) has been made by radio-immunoassay in the eluate of a few drops of blood deposited on a piece of blotting paper. Six disks (40 µl of blood) are eluated in 750 µl of barbital buffer for 24 hours at 4°.
The comparative assay of T_4 in eluate and in fourty microliter of blood allows to find a close connection with a regressive coefficient of 0,80. The effectivness of elution is 48 %.
The results from 6 disks (40 µl of blood) in 150 normal newborns are 600 pg ± 12.
In 5 confirmed hypothyroidians infants the average is 160 ± 60 pg. In 2 cases of athyreosis the average is 80 pg.
This method was used for detection of hypothyroidism in one thousand of newborns by using samplings obtained from the phenylcetonuric detection center. We detected by this way a case of hypothyroidism which was confirmed by traditional tests.
This method seems to totally justifying a neonatal detection of hypothyroidism on a big scale.

32 GOITER DUE TO CONGENITAL DYSHORMOGENESIS : FOLLOW UP STUDY IN 28 CHILDREN.

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We have studied 17 boys and 11 girls (7 pairs of siblings) with goiter over a period of 2-15 yrs. Methods : In all patients, ^{131}I-uptake, PBI^{131}, CR, SCNK discharge, PBI, BEI or T_4, column chromatograms of radioiodinated aminoacids in serum have been performed ; in some, chromatograms have been done also in thyroid tissue and urine.

Based on laboratory data, 5 groups have been formed. Although synthesis of thyroid hormones is blocked in all enzyme defects, the course of the disease is variable but characteristic for each group : 1) in 7 of 8 children with complete peroxydase defect, brain damage and idiocy developed despite thyroid therapy beginning shortly after birth. 2) 4 patients with Pendred syndrome were euthyroid and had small goiters ; of 2 sisters with the same clinical picture, one showed the typical I^{131} discharge whereas the other one had a normal SCNK test. 3) 9 patients with coupling defect had goiters around the age of 2 yrs and continuously increased in size inspite of thyroid therapy. 4) 3 children with dehalogenase defect were born with goiters ; hypothyroidism developed slowly. In 2 of them goiters had disappeared and laboratory and clinical findings became normal without therapy 2 yrs after compulsory iodination of salt in Argentina. 5) In 4 children, the enzyme defect was not established. Goiter and hypothyroidism appeared around 11 months. Thyroid therapy was very successful.

33 EFFICIENCY AND MODE OF ACTION OF CONJUGATED OESTROGENS IN THE TREATMENT OF TALL GIRLS.

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In 1972, we have reported on the successfull therapy of tall girls by continous administration of high doses of conjugated oestrogens, combined with an oral progestin for 5 days per month. This is a report on 23 such treatments, - in 14 cases with 7.5 mg Presomen (R) daily, in 9 cases with 4.5 mg Presomen In 9 cases the therapy has meanwhile been terminated. At the start these girls showed bone ages of 11 to 14 cm ($\bar{x}$ = 8,91cm) This growth rate was reduced to 1.9 cm on the average for the first year. The growth reduction was 7.22 cm in the mean. In the second group, which we treated with 4,5 mg, we saw further growth between 2,5 and 4,2 cm within the first six months under therapy. Hence we recommended treatment with a daily dose of 7.5 mg Presomen. Serious side effects were not encountered under this dosage. The almost abrupt growth-stop under Presomen in spite of open epiphyseal lines caused us to investigate plasma somatomedin levels and growth hormone. After 4 to 6 months of treatment the somatomedin levels, calculated as ratios of SM before and under therapy, were decreased to ratios between 0.65-0.8 while GH secretion increased under therapy with Presomen.

34 EXCESSIVELY TALL STATURE IN GIRLS. FURTHER EXPERIENCE WITH SEX HORMONE TREATMENT IN HIGH DOSES.

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We have previously reported our experience in treating children with sex hormones. A difficulty of that preliminary study was the inaccuracy of bone age (BA) estimation (Greulich and Pyle) and of adult height prediction (Bayley and Pinneau). BA was reassessed in an extended series of 40 girls using the Tanner-Whitehouse II RUS technique and the new regression equations of Tanner for prediction of adult height. All girls were in puberty at start of treatment (BA >12). They were divided into 3 BA-groups (12.1-13, 13.1-14, 14.1-15) and received ethinylestradiol 0.3 mg daily and norethisterone for 5-7 days every 4th week. The mean reduction of adult height was 4.6 ± 2.4 cm, slightly more (5.8) in the youngest and somewhat less (4.5) in the oldest BA-group. BA-velocity during the whole treatment period was 1.5 ± 0.5yrs per yr in the youngest and 1.1 ± 0.4 yrs/yr in the oldest BA-group. The individual variation, however, was large in all groups. Postthera-peutic amenorrhea of 0.2-0.5 yrs occurred in only 7 patients.

This improved method of analysis confirms the effectiveness of sex hormone treatment in tall girls.