53 ACTION OF SOMATOSTATIN AND CB 154 IN THE DIENCEPHALIC SYNDROME. P.Stubbe and G.Lenard (Intr. by H. Helge). Dept. of Pediatrics, University of Göttingen, Göttingen, W. Germany.

Increased levels of plasma growth hormone (GH) represent a characteristic feature of diencephalic syndrome (DS) in childhood and may in part be responsible for extreme emaciation and loss of subcutaneous tissue. Somatostatin and CB 154 were shown to lower GH in acromegaly. Both substances were applied to a 3 1/2 yr old girl with DS who was unsuccessfully irradiated for a spongioblastoma of the 3rd ventricle at 1 1/2 yr of age. Somatostatin following arginine infusion or during sleep lowered GH levels immediately with a T 1/2 of 17 and 14 minutes respectively. This gives evidence of GH hypersecretion instead of impaired clearance in DS. The action of CB 154 is more protracted and doses of 4 to 10 mg daily are effective in adults. The patient received 3 mg CB 154 daily and preliminary results indicated a reduction of GH basal levels to about 30 per cent of pretreatment values. CB 154 may have favourable therapeutic effects in DS by preventing metabolic changes due to GH hypersecretion.

54 FAMILIAL MIXED GONADAL DYSGENESIS. Ch.Sultan and R. Jean (Intr. by J.Bertrand), Clin. des Maladies des Enfants, Montpellier, France.

Mixed gonadal dysgenesis is characterised by female ductal differenciation in spite of presence of testicular tissue. Two cases of familial mixed gonadal dysgenesis are reported. The first patient had a female phenotype and presented at 13 years undeveloped breast, primary amenorrhea and enlarged clitoris. She had normal female external genitalia and a narrow vaginal introitus distinct from urethra. Normal vagina, uterus and tubes were opacified by external genitography. At laparotomy uterus appeared small but Falloppe tubes were normal. Streak gonad was found in the place of the right ovary and the site of the left gonad was occupied by an ovoid tumor which histologic examination occurred a gonadoblastoma with abnormal testicular tissue. The caryotype in blood culture was XY. The brother presented at birth hypospadias with bilateral cryptorchidism. He was 8 years when referred to the Paediatric Center because of ambiguous external genitalia. He had a male phenotype with labial fusion and fusiform urogenital sinus opening at base of phallus. Gynecography disclosed normal vagina and outset of uterus. Exploratory laparotomy revealed a right gonad: dysgenetic testis was found at biopsy. A next surgical laparotomy is programmed on opposite gonad. Caryotype was XY in blood culture. These reports may be classified in mixed gonadal dysgenesis group. Original findings = Same asymetric testicular dysgenesis in two siblings, but with different phenotype. Its origin can be found in a structural abnormality of Y chromosome.

55 "KETOTIC HYPOGLYCEMIA" IN PITUITARY DWARFS. H.U.Tietze (intr. by R.P.Zurbrügg), Cnopf'sche Kinderklinik, Nürnberg, Germany.

Hypoglycemic attacks and delayed recovery of blood glucose (G) during insulin tolerance test (ITT) are seen in both, in "ketotic hypoglycemia" (KH) and in pit. dwarfism. Recently the occurence of impaired cortisol regulation in "KH" has been reported. Thus a comparism of G regulation in both diseases seemed to be indicated. A ketogenic diet was given to 5 children suffering from pit. dwarfism. A critical drop of G within 24 hours was seen in 4 of the 5 dwarfs, while in a contrast group of 4 healthy children none exhibited a similar hypoglycemia under the same regime. When compared with a group of 4 children with proved "KH" the pit. dwarfs showed not only the same hypoglycemic reaction on ketogenic diet, but also the same delayed G recovery during ITT. The rise of plasma cortisol and of urinary epinephrine excretion during the same test was diminished in both groups. GH secretion during ITT was lowered in 2 of 4 children with "KH" and absent in the pit. dwarfs. The findings may indicate a similar disturbance of blood glucose regulation in pit.dwarfism and in "KH", probably located in the hypothalamus.

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56 PLASMA 17 HYDROXYPROGESTERONE AND PROGESTERONE IN NORMAL NEWBORNS AND INFANTS. J.E. Toublanc, N.T. Tea, M. Roger, and N. Joab (Intr. by P. Canlorbe), Hôpital Saint Vincent de Paul, Paris and Fondation de Recherche en Hormonologie, Fresnes, France.

Plasma levels of 17 Hydroxyprogesterone (17 OHP) and Progesterone (P) were measured by specific radioimmunoassays in 76 newborns and infants (36 boys, 40 girls), for 17 OHP and 42 newborns and infants (16 boys and 26 girls) for P. Cord blood values of 17 OHP and P were also evaluated in, respectively, 25 and 20 cases.
17 OHP mean level (ng/ml ± SEM) in cord blood was 45.91±3.45, in the newborns during the first 12 hrs. 7.55±0.59 (n=14), from 12 to 24 hrs. 3.60±0.72 (n=8). After the 24th hr., up to 1 month, 17 OHP remains in plateau at 1.31±0.10 (n=33). After 1 month there is a decrease of 17 OHP which is, at 6 months, in the prepubertal range (0.24±0.14, n=7).
P mean level (ng/ml±SEM) in cord blood was 413.20±39.76, in the newborns during the first 12 hrs. 32.23±5.23 (n=10), from 12 to 24 hrs. 11.56±1.41 (n=11), by the 6th day of life P was in the prepubertal range : 0.29±0.06 (n=13).
The data presented, namely 1-sharp decrease of P, 2-first month plateau of 17 OHP, 3-absence of any sex difference, suggest, for 17 OHP, an adrenal contribution.

57 METABOLIC ACTIVE PEPTIDES ISOLATED FROM THE URINE FROM PATIENTS WITH CONGENITAL GENERALIZED LIPODYSTROPHY, "METABOLIC" OBESITY, AND ANOREXIA NEVROSA. O. Trygstad and I. Foss. Section of Endocrinology, Institute of Pediatric Research, Rikshospitalet, Oslo, Norway.

Urine from the listed groups of patients and from normal controls were fractionated by protein precipitation and Sephadex gel filtration. The chromatograms were different for the three groups of patients, and they differed from that of the normal controls. Fractions with mol.wt. in the range of 1000 were observed to be metabolic active in mice. Daily injections to groups of animals were given over three weeks. The fraction from patients with generalized lipodystrophy produced persistent lipodystrophy in infantile animals, and lipoatrophic diabetes in adult animals. The corresponding fraction from the metabolic obese patients induced obesity, and the mean body weight of the mice increased from 36g to 70g. The obesity persisted for four months. The fraction from the patients with anorexia nervosa had an anorexogenic effect that persisted for four weeks. The body weight decreased 15% for albino and 30% for obese yellow mice. The amino acid compositions of the urinary fractions was similar to that of thalamic agents. The active factors are supposed to be hypothalamic.

58 TRH INDUCED TSH RELEASE IN COELIAC DISEASE. M. Vanderschueren-Lodeweyckx, E. Eggermont, C.Cornette, P. Malvaux, C. Beckers and R. Eeckels, Dept. of Paediatrics, Katholieke Universiteit Leuven and Centre de Médecine Nucléaire, Université Catholique de Louvain, Belgium.

TRH (thyrotrophin releasing hormone) induced TSH (thyroid stimulating hormone) release has been studied in 11 patients with active gluten-sensitive enteropathy and in 21 normal children. Blood samples were taken before and 20, 40 and 60 min after intravenous administration of 200 µg of synthetic TRH. The data reported are means ± 1 S.E.M. In normal children basal plasma TSH of 5.9±1.5 µU/ml increases to 32.6±3.5 µU/ml at 20 min and decreases to 21.3±3.0 µU/ml at 60 min. In 9 out of the 11 coeliac patients an exaggerated response is observed. Basal plasma TSH level of 6.3±1.3 µU/ml is not significantly different from the value observed in normal children. But plasma levels recorded 20 min (52.1±5.8 µU/ml) and 60 min (51.7±7.5 µU/ml) after the administration of TRH are significantly higher than in normal children (at 20 min $p = 0.01$-0.001; at 60 min $p < 0.001$).
These data give additional evidence of a dysfunction in the hypothalamo-hypophyseal system in coeliac disease (Helv.paediat. Acta 28: 349, 1973). It is not clear whether it is due to malnutrition, to some direct action of gluten-peptides or to other factors.