71 H.KRAWCZYŃSKA,K.WÓJCIK*,E.MAŁUNOWICZ*,M.CHMURA* Endocrine Unit and the I.Department of Pediatrics,Children's Hospital in Dziekanów, Department of Laboratory Diagnostics,Medical Centre of Postgraduate Education, Warsaw, Poland.

Thyrotropin-Induced Hyperthyroidism. A four years follow up study in a 10 year old girl with goitre.

At the age of 6 the girl presented a small goitre without clinical signs of hyperthyroidism but T_4 and T_3 and also TSH were elevated. She was treated with exogenous thyroid hormones for one year. We observed total suppression of TSH but T_4 and T_3 levels became even more elevated and signs of hyperthyroidism appeared. During next 3 years without treatment with thyroid hormones T_4, T_3 and basal TSH were still elevated and the response of TSH to TRH stimulation increased to the hypothyroid range. Acceleration of bone age and Growth velocity confirmed hyperthyroidism.The size of goitre did'nt change.The HGH after insuline induced hypoglycemia,cortisol,PRL after TRH, LH and FSH after LHRH stimulation were all in normal range. Skull Rtg was normal.

A selective pituitary resistance to thyroid hormones or microadenoma of thyrotrophs is supposed.

72 M.J. MARTINEZ*, J.L. CHAUSSAIN, J. LEMERLE*, M. ROGER, P. CANLORBE, and J.C. JOB. Hôpital Saint-Vincent de Paul, Paris, France.

Klinefelter syndrome, tumor and sexual precocity.

Klinefelter syndrome was discovered in 3 boys with precocious puberty and small testes (20x10 mm) in whom adrenal function was normal, thus excluding the diagnosis of adrenal hyperplasia or secreting tumor. In 2 of these patients, aged 7 and 12 years, Leydig-cell secretion (plasma testosterone 9 and 5.2 ng/ml) was secondary to an HCG-secreting mediastinal teratoma, with high plasma levels of HCG (75 and 355 mIU/ml). After surgery, plasma HCG was undetectable, but plasma testosterone (2.9 and 0.9 ng/ml) remained elevated and plasma gonadotropins rose (LH 5 and 4.7, FSH 2.2 and 10 mIU/ml). In the 3rd patient, aged 1 year, plasma testosterone (2.5 ng/ml), basal (LH 4.9, FSH 7.1 mIU/ml) and post-LHRH (LH 20, FSH 14.5 mIU/ml) gonadotropins levels were increased. HCG was undetectable in plasma, cranial computerized tomography demonstrating an hamartoma of the 3rd ventricle. Karyotype was XXY in these 3 patients. These cases suggest 1/ that chromatine and/or karyotype must be studied in cases of male sexual precocity with small testes and normal adrenals; 2/ that there may be a relationship between the supernumerary X chromosome and the occurrence of tumors, particularly polyembryomas.

73 H.MOELLER*,G.OETTLING* and D.GUPTA, Department of Diagnostic Endocrinology, University Children's Hospital, D-74 Tubingen, FRG

Development of DHT-receptor (DHT-R) estimation through (3H)-DHT exchange.

The convenient dihydrotestosterone-(DHT)-R estimation with (3H)-methyltrienolone (R1881)in human tissue is unspecific as it is also bound by the progestin receptors. We have, therefore, developed a highly specific (3H)-DHT exchange assay for cytoplasmic and nuclear DHT-R. In parallel to the suppression of DHT metabolism during incubation by NADase, suppression of low capacity binding was carried out in the presence of an overdose (100-fold) of R1881. In this way, the residual metabolism of (3H)-DHT remained equal in both assays. Free (3H)-DHT and sex hormone binding globulin (SHBG) were separated by agar gel electrophoresis (R. Wagner, 1972). Under these conditions, the binding characteristics of DHT and R1881 were not different in relation to K_S value (5.08×10^{-9} mol/l), association and dissociation velocities, and number of estimated binding sites (free 0.5 fmol/mgDNA; total 1.0 fmol/mg DNA). The data indicate that the higher affinity of R1881 to the DHT-R may not be real as DHT concentration diminishes in the usual assay systems.

74 B.PEITERSEN[X], H.HERTZ[X], B.BROCK JACOBSEN and S.KRABBE[X]. University Clinic of Paediatrics, Children's Hospital, Fuglebakken and Department of Paediatrics, Rigshospitalet, University of Copenhagen, Denmark.

Myelofibrosis and T_3-thyreotoxicosis in a girl with McCune-Albright Syndrome.

The pathogenesis of McCune-Albright Syndrome involving bone, skin, and endocrine organs remains unknown. The syndrome might be associated with dysfunctions of the hematopoietic system. A girl with severe polyostotic fibrous dysplasia and vaginal bleedings from 3 months of age is presented. The concentration of serum T_3 was increased from the age of 13 months but se-T_4 was normal. A T_3-thyreotoxicosis was diagnosed at the age of 3½ years (no TSH response to TRH). Aged 3½ years she developed severe anemia, thrombocytopenia and splenomegalia. Bone marrow biopsy showed myelofibrosis. Following splenectomi the hemoglobin concentration increased and the platelet count was normalized. The patient was treated with medroxyprogesterone-acetat (MPA) during the first 3 years of life and with propylthiouracil (PTU) from the age of 3½ years. The myelofibrosis developed before unset of PTU, excluding any causal relationship. Nor the myelofibrosis seems to be associated with MPA since no such side effect of MPA is known, and the only patient previously reported with McCune-Albright syndrome and myelofibrosis was not treated this way.

75 L.D.Peneva and R.Slavova
Endocrine Division,Research Institute of Pediatrics,Sofia,Bulgaria.

Effects of human growth hormone treatment for 1-8 years in pituitary dwarfism.

The effects of long term/1-8 years/treatment with HGH(Sotropin H 3x5E/week,Rumanian HGH 3x4mg/week and Grorm 3x4mg/week)in 17 children with growth hormone deficiency were followed up.In 10 patients there was birth injury,in particular assisted breech delivery, in 5 other-family cases.Two patients(brother and sister)had isolated GH-deficiency and other 15 had multiple GH-deficiency.The latter group were treated with additional thyroid hormones.In the first year of treatment it was "catch-up growth" till 10cm/year and after that growth rate lowered to 4-5cm/year.Total height gain was from 42cm/8years till 14cm/2 years.In the first year height age increased more than bone age in reverse to the second year.Growth index(Soyka)raised with treatment over 1.Interruption of HGH-treatment resulted in decrease of growth rate and changing of the medicine improved it.After the age of 17 HCG-treatment in two-months courses had a very good effect.Thyroid medication in appropriate substitution dose is advisable.Best effects were obtained in small children with retarded bone age and in continual treatment.

76 K.E.PETERSEN, B.BROCK JACOBSEN, T.IVERSEN[X] and T.BILLE[X]. Children's Hospital Fuglebakken, Dep. of Pediatrics G, Rigshospitalet, Copenhagen, Dep. of Pediatrics, Århus and M.O.H., Narssak, Denmark.

X-linked inheritance of congenital cortisol and aldosterone insufficiency (adrenal hypoplasia).

Congenital adrenal hypoplasia is a rare cause of adrenocortical insufficiency in infancy. Two different modes of inheritance, sex-linked and autosomal, have been suggested. Three male infants with severe clinical signs developed within the first week of life are described. Biochemical data indicated both glucocorticoid and mineralocorticoid insufficiency. Clinical remission followed substitution therapy. The infants descend from the same Greenlandic family. Informations concerning 82 family members in five generations were obtained. Fourteen male infants had a clinical picture suggesting adrenocortical insufficiency, and died in early infancy. No female was affected. The pedigree of the family demonstrated no father-to-son transmission. The pedigree indicated an X-linked transmission of the adrenal hypoplasia gene locus; the locus seems to originate from the great-great-grand mother.