23 GEL-ELECTROPHORETIC STUDIES ON THE PROTEOGLYCANS OF THE NORMAL AND ABNORMAL GROWTH CARTILAGE.

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The gel-electrophoretic pattern of the proteoglycans of the tibial epiphyseal plate was studied in 7 foetuses, 5 premature newborns, 4 term newborns, 4 infants and 4 children. The proteoglycans obtained from small cartilage biopsies in cases with bone growth disturbances (achondroplasia ; pseudo-achondroplasia, 3 cases ; Kniest's disease, 2 cases ; diastrophic dwarfism, 2 cases ; parastrematic dwarfism ; pycnodysostosis ; mucolipidosis type III ; Blount's disease and multiple exostoses) were compared with the proteoglycans of the normal cartilage.

The tissues were extracted with 4 M Guanidinium chloride After dialysis against 8 M urea at pH 7, the proteoglycans were obtained by ion-chromatography in urea on D.E.A.E. cellulose and submitted to gel-electrophoresis on polyacrylamide-agarose gels.

Gel-electrophoresis of proteoglycans showed a different pattern in foetuses from that found in children. In the groups studied, the change occurs in the first months of extrauterine life. In the pathological subjects studied, abnormal patterns were found in pseudo-achondroplasia and in Kniest's disease.

24 SERTOLI CELL FUNCTION DURING INITIATION AND MAINTENANCE OF SPERMATOGENESIS

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Testicular tubular function has traditionally been judged by morphological criteria. Very recently, we have demonstrated a biochemical marker of Sertoli cell function; the testicular androgen binding protein (ABP). ABP is synthesized in Sertoli cells as a response to FSH stimulation. It disappears completely following hypophysectomy, but reappears after FSH administration. No other pituitary hormones are active in stimulating ABP synthesis. ABP is transported with the testicular fluid into the caput epididymis, where it is partly taken up by the lining epithelial cells. We have proposed that ABP serves as an important store of androgenic hormones (mainly testosterone and dihydrotestosterone) that are necessary for initiation and maintenance of spermatogenesis.
ABP was found to be present at low concentrations in testes of prepubertal rats, but increased rapidly during sexual maturation. Experimental cryptorchidism dramatically decreased the testicular and epididymal content of ABP demonstrating an impaired Sertoli cell function under these experimental conditions. The finding indicates that Sertoli cell damage may be an important factor in the overall impairment of spermatogenesis found in this condition.

25 PLASMA 5α-ANDROSTANE 3α, 17β-DIOL IN BOYS DURING PUBERTAL DEVELOPMENT.

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Recent in vitro studies have shown that 5α-androstan-3α,17ß-diol(DIOL) is a major conversion product in man from labelled precursors. This report describes a specific and highly sensitive RIA for the estimation of plasma DIOL in children during their adolescent growth. Plasma DIOL values appear to be quite low(0.5ng/100ml) at puberty stage 1. A marked increase was noted from stage 2(1.8ng/100ml) to stage 3(3.5ng/100ml), but without showing any significant difference between stage 3 to 4(3.8ng/100ml). Stage 5 boys(7.3ng/100ml) demonstrate a steep rise in the plasma level, which increased further in adulthood(11.5ng/100ml). The pattern of the mean trend in the increament of DIOL paralleled more with that of DHT than with T. When the ratio of DIOL/T is considered, there is a sharp rise from stage 1 to stage 2 followed by a decreament to stage 3. In bilateral cryptorchidism the levels of DIOL were significantly lower when related to values seen in comparable age-groups.
Although the distribution and metabolism of DIOL has been found to be different from that of T and DHT, it is possible that like DHT, this substance, as another steroid messenger, has certain actions on the male reproductive organs during sexual maturation.

26 17 HYDROXYLASE DEFICIENCY IN AN ADOLESCENT GIRL.

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The 17 hydroxylase deficiency remains a rare entity which has never been diagnosed before adulthood. We present a new case observed in a young teenage girl. This 13, 1/2 years old girl presented with hypertension (150 mm Hg), absence of puberty, delayed boneage (= 9 years), growth retardation (statural age : 11 years) hypokalaemic alkalosis (K : 2,7 mEq/l). There was no remarkable family history. Infancy and childhood had been uneventfull except for frequent tonsillitis and bronchitis.

Laboratory data included : low urinary 17 ketosteroids (2,68 mg/24 h and 17 hydroxycorticoids increasing very little following ACTH (respectively 3,66 and 1,61 mg/24 h). Plasma gonadotrophins were markedly increased (FSH 53 an LH 116 mui/ml) as well as plasma ACTH levels (207 pg/ml). Low plasma aldosterone levels (15 pg/ml) increased sharpedly 12 h after Synacthen 1 mg IM (1500 pg/ml) while plasma renin activity remained low (1,1 ng/l/mn). Determinations of urinary steroids derivatives were as follow :

	basal	ACTH		basal	ACTH
THF	ND	ND	THB	774	636µg/24h
THE	125	348µg/24h	Pregnandiol	3700	6700 "
THDOC	128	184 "	Pregnandiol	240	860 "

To our knowledge this is the first case of 17 hydroxylase deficiency diagnosed before adultwhood.

27 CHARACTERISATION OF IDIOPATHIC ADDISON'S DISEASE BY ANTIBODY STUDIES.

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The existence of three genetically distinct types of idiopathic Addison's disease has been proven by family analysis (Spinner et al, 1968). Antiadrenal antibodies are demonstrable in the majority of patients with the different types of disease using complement fixation and immunofluorescence (IF) techniques. - Using IF and gel filtration (GF) techniques we have demonstrated distinction of the endocrinopathy - moniliasis syndrome (EMS) by presence of precipitating antiadrenal antibodies. These GF-detectable antibodies are not the same as those demonstrable by IF, but the occurrence is mostly parallel. - Of 21 patients, 3-23 years of age, with EMS, antiadrenal antibodies were demonstrable by IF in 16, and also by GF in 11 of these. 10 of the 16 had Addison's disease, of these 9 were positive by IF and 6 by GF. In 5 of them the antibodies were detected before a distinct clinical manifestation of this disease. One, who had the disease already for ten years prior to the study, was negative by both IF and GF. 11 had so far only other components of the syndrome, 7 of them were positive by IF and 5 by GF. - Of 15 patients, 8-48 years of age, with Addison's disease alone, antiadrenal antibodies were demonstrable by IF in 14, but in none by GF.
- Of 5 patients with diabetes mellitus, thyroiditis, or both (Schmidt's syndrome), 22-60 years of age, antiadrenal antibodies were demonstrable by IF in 4, but in none by GF.

28 A COMPARATIVE STUDY ON THE INSULIN DOSAGE IN THE TREATMENT OF DIABETIC KETOACIDOSIS IN CHILDREN

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Two insulin regimens (A and B) were compared in the treatment of diabetic ketoacidosis in children: Regimen A, the initial dose of insulin being 2-3 U/kg ($\frac{1}{2}$ i.v., $\frac{1}{2}$ s.c.), followed by 0.5U/kg s.c. at intervals of 1-4 hours, and regimen B, the initial dose being 0.5 U/kg i.m., followed by 0.25 U/kg i.m. at intervals of $\frac{1}{2}$ -2 hours. Fluid and electrolyte therapy were similar in both regimens. - Eight diabetics admitted in severe ketoacidosis have so far been studied during regimen B. Recovery from severe ketoacidosis (Kussmaul breathing +→-, rise of the pH above 7.2) took place as rapidly with both regimens, within approximately four hours. The mean dose of regular insulin (Actrapid®) to achieve this was in regimen A 2.4±0.2 and in regimen B 1.3±0.1 U/kg ($P < 0.001$). Also complete recovery from acidosis took place with less insulin in regimen B than in A. During regimen A a too rapid change in plasma osmolality and hypokalemia occurred in some cases despite an early start of the administration of glucose and potassium containing solutions. - Our results suggest that the initial treatment of diabetic ketoacidosis in children with smaller doses of insulin than previously used is as effective as with larger doses. In addition, the risks of hypoglycemia, hypokalemia and cerebral oedema are smaller when insulin is administered in relatively small doses.