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PRIMARY CULTURE OF TESTICULAR CELLS FROM A LARGE CELL CALCIFYING SERTOLI TUMOR OF A PREPUBERTAL BOY WITH GYNECOMASTIA. E. Berensztein, A. Belgorosky, MTG. de Dávila, MA. Rivarola. Lab. de Inves. y Servic. de Anat. Patol. Hosp. Garraham. Bs.As., Argentina.

We have studied in vitro the steroidogenic activity of a bilateral large cell calcifying Sertoli tumor and of its surrounding testicular tissue,in a 9-year old boy with gynecomastia, absence of androgenic clinical signs and bilateral testicular hard nodules. Basal and post LH-RH levels of serum LH and FSH were low.Cells from 2 fractions were separated: tumoral (Tu), (hard and calcified) and extratumoral (ExTu),(soft,composed of prepubertal testicular tissue with scarce micronodules of tumoral cells. A control (C) group of prepubertal testicular cells obtained at necropsy were also cultured. Cells in Tu, Extu and C were stimulated with insulin (I) 5000 ng/ml, hCG 10 ng/ml and hFSH 10 ng/ml.On day 6 of culture,estradiol (E2)and testosterone(T) were determined by RIA and aromatase activity (ARO: E2 under 0.5uM T as substrate)was also estimated.Basal T was(X±SD);C,0.91±0.20, ExTu 13.6 ±8.83 and Tu, 0.52±0.26 ng/millón cel.24 hrs and it was stimulated by I and hCG in C(23.5±8.68 and 4.30±0.79 resp)and in ExTu (58.3±26.0 and 29.1±14.2) but not in Tu (0.37±0.28 and 0.11±0.10). E2 was non detectable in every instance. Basal ARO was obsent in C, ExTu and Tu but it was stimulated by I and hFSH in ExTu (120±43.8 and 83.6±22.5 pgE2/10 6 cell 24 hrs) and in Tu (250±14.0 and 479±147). It is concluded the tumor a) Can stimulate steroidogenesis in the surrounding tissue by a paracrine factor, b) it did have ARO and c) it did not have steroidogenic activity by itself. The estrogenic activity in the patient could be explained by the interaction of tumoral and testicular cells combining themselves to secrete estrogens autonomously.

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GROWTH IN TURNER SYNDROME. (TS) C. García, A. Martínez, A. Keselman, JJ. Heinrich y C. Bergadá. CEDIE Div. de Endocrinología. Hosp. de Niños R. Gutierrez. Buenos Aires. Argentina.

The growth pattern of a population of 254 patients with TS, seen between 1960 and 1992, was analyzed and their data compared with those of normal girls and of other TS series, in order to have local reference parameters. A combination of cross-sectorial and longitudinal data consisting of 2308 height determinations was studied. Mean birth weight of 156 term infants was (X±SD) 2812 ± 487 grs, significantly reduced when compared to normal controls 3340 ± 490 grs ($p < 0.001$). The mean birth lenght of 14 patients was 46.7 ± 2.67 cm below the mean of the control group: 50.0 ± 1.80 cm ($p < 0.001$). Our data shows moderate growth retardation already apparent at birth; lenght -1.77 SDS. This difference increase in the first year of life to -2.91 SDS, stabilizes up to 10 years -2.48 SDS, and then increases again during pubertal age up to reach a final height of 138.7 ± 5.46 cm at -3.62 SDS of the mean normal adult stature, different from that of other TS populations. Positive correlation between final and target height was found ($r=0.42$ $p < 0.004$) in a group of 44 patients.

Conclusion: The growth pattern of patiens with Turner Syndrome studied is similar to that of other reported TS populations. The observed differences may be explained by height differences between normal female populations.

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PRENATAL TREATMENT IN CONGENITAL ADRENAL HYPERPLASIA (CAH): FIVE CASES EXPERIENCE IN TWO YEARS. G. Guerra*; ATM. Guerra**; W. Pinto**; APM. Faria**; R. Barini***; SHVL. Marini*; MTM. Baptista ****. Departments of Paediatrics*, Medical Genetics**, Gynecology and Obstetrics*** and Medical Clinic - F C M UNICAMP.

Since there is a 25% probability for a couple with a CAH affected child to have another affected child, the CAH prenatal diagnosis has been searched in the last 10 years. The main objective is to avoid the virilization of the external genitalia and, so, to reduce the social and psychological implications repercussions, and to reduce surgical corrections. We are following 30 families with CAH in our Services. Two years ago we initiated the prenatal treatment of the pregnant women with CAH children. We followed 4 pregnancies (1 of them were twins). The protocol used was: pregnancy diagnosis with BHCG; Dexamethasone (0.75 mg twice) starting at the 8th week and amniocentesis to perform the karyotype close on the 16th week. The treatment was stopped when the fetus was a male, and it was maintained the whole pregnancy when the fetus was a female. Among the 4 male newborns, 2 were normal (monozygotic twins) and 2 were affected (both with virilizing form); the female newborn had a severe saltlosing form, a cleft palate and no genital ambiguity. Our next aim will be the exact CAH prenatal diagnosis.

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EFFECT OF ACTH ON PLASMA MET ENKEPHALIN (ME) AND FREE AND CONJUGATED CATECHOLAMINES (CA). MC. Negueruela, ML. Figuerola, L. Contreras, M. Barontini. Hospital de Niños "Ricardo Guetierrez", CEDIE, Buenos Aires, Argentina.

It was reported that ME and CA are costored and co-released from chromaffin vesicles in the adrenal medulla. The aim of this study is to investigate the relationship between ACTH and ME and CA (free and conjugated). We studied 8 healthy children (aged 6-13 yr) by performing a ACTH i.v. test (0.25 mg). Basal blood samples at 30' and 60' after ACTH injection were obtained. In 4 cases blood samples were obtained also at 5',10' and 15' after the i.v. injection. Plasma ME and cortisol (F) were measured by RIA. Free and conjugated plasma CA were determined by a radioenzimatic method. ACTH rose F ($p < 0.01$ basal vs 30' and 60') without significant changes in plasma adrenaline and noradrenaline. However, a significant decrease in conjugated dopamine (DA) was observed (0': 1522 ± 314; 30': 1115 ± 203; 60': 1129 ± 278 pg/ml; $p < 0.01$ basal vs 30' and 60'). A decrement in conjugated DA was also seen 10' after ACTH stimulus 0': 1154 ± 267 vs 10': 992 ± 100 pg/ml; $p < 0.01$). Plasma ME rose significantly from basal values (0': 0.29±0.06 vs 30': 0.44±0.04 pmol/ml; $p < 0.05$). Our data show that ACTH acutely decreases total plasma DA levels by reducing its conjugated fraction and increases plasma ME leveles. These results suggest that the adrenal may not be the only source of ME secretion.

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PSEDOPRECOCIOUS PUBERTY IN A BOY WITH PEUTZ-JEGHERS SYNDROME (PJS) AND FEMINIZING SEXCORD TUMOR (SCT). EFFECTS OF TREATMENT WITH KETOCONAZOLE (KTZ). L. De Lacerda, SK. Kohara, MCS. Boguszewski, MC. Schmitt-Lobe, and R. Sandrini. Department of Pediatrics, Universidad Federal of Parana, Curitiba - Brazil.

The association of PJS and SCT is rare. Gynecomastia and advanced bone age in the affected individuals depends of estradiol secretion. KTZ inhibit p450-cythochrome dependent enzyme 17,20 - 1 lyase and hence testosterone synthesis. We report a case of a 6,5 y old white boy with gynecomastia. Height 90th and weight 25th centiles; melanotic spots in nose and lips; breasts Tanner stage III at right and II at left; no pubic hair; penile length was at 50th centile: testes 4 ml with no palpable masses. BA was 9,5 y. Brain CT and ultrassonograpy (US) and CT of the abdomen were normal; US of the gonadas showed multiple areas of hyperechogenicity. GI R^{x} series with polips in stomach and duodenum. Alpha-fetoprotein and carcinoembrionic antigen were negative.17KS: 11,4 umol,and 17 OHCS: 21,8 umol; undetectable b-HCG;normal prolactin; two LH-RH tests without response of both FSH and LH; random E2 serum concentrations were normal in one occasion (42.2 pmol/L) and elevated in another (205.6 pmol/L); basal and post-HCG serum testosterone were 0.4 and 17.3 nmol/L respectively, Bilateral testicular biopsy revealed a SCT.The father,father's brother and sister,and a 9 old sib have PJS.Started on KTZ until reaching 600 mg/day (for 19 months).Liver function not affected. Gynecomastia waned completely at left and has been stage II (soft) at right. Testes are 6ml; BA (11 y) advanced 1.5 years in the last 2.1 years, though the treatment started 6 months after the first BA evaluation.PAH was 170 cm before therapy, and is currently 1.75 cm.,KTZ appears to be an promising therapeutical approach

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GnRH AGONIST THERAPY IN TRUE PRECOCIUS PUBERTY SECONDARY TO HYPOTHALAMIC HAMARTOMA (IHH). S. Domenice; AC. Latrônico; BB. Mendonca; IJP. Arnhold; W. Bloise. Division of Endocrinology and Metabolism - HCFMUSP - Sao Paulo - SP. Brasil.

Hypothalamic hamartomas, are benigns tumors of central nervous system, related to development of the true sexual precocious puberty, have been more frecuently diagnosed due to improvement of the radiologic technology (computed tomography and MRI scanning).

We studied two children with precocious sexual development (Case 1: boy, onset the development of public hair and growth of the penis at 6 months of age; Case 2: girl with breast development and menarche at 6 months.

In both patients hypothalamic hamartomas were demonstrated by MRI (Case 2) and CT and MRI (case 1).

At the first evaluation we observed enlargenment of the penis (8,0 cm) and a testicular diameter of 2,5 cm in case 1; and b reast stage Tanner II in case 2. Both had a pubertal response to the GnRH stimulation test (100 mg IV), LH 51,4 and 28,3 mUI/ml, respectively and normal levels of DHEAS for age.

The children were treated with GnRH analog depot (D-Trp6-GnRH) in dosage of 1.5 mg IM every month for 36 and 18 months respectively. Gonadotropin secretion was inhibited with use of GnRH analog. Physical signs of puberty did not progress futher, without menstrual periods and with height velocity remained between normal range. Laboratory data showed normalization of testosterone level in case 1 (144,5 to 15 ng/dl).

We conclude that GnRH analog therapy is efficient in patients with true precocious puberty secondary to hyphotalamic hamartoma.