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TRANSIENT NEONATAL HYPERTHYROTROPINEMIA. Sgarbi J.A., Novaes M., Biancalana M.M., Dualibi L., Werner R.S., Romaldini J.H. Dept of Endocrinology and Neonatology, HSPE-IAMSPE, CP 8570, Sao Paulo, Brasil.

Transient neonatal hyperthyrotropinemia (TNH) is a rare condition that must be included in the differential diagnosis of congenital hypothyroidism (CH) to avoid inappropiate thyroid hormone replacement. Our screening program for congenital hypothyroidism evaluated 4,075 newborns with simultaneous determinations of TSH and T4 in the umbilical cord blood after immediate clamping. Newborns presenting TSH concentrarions higher than 40 uU/ml (n= 38; 0.7%) were recalled. In this group, 9 out of them (0.2%) had TSH values above 60 uU/ml. All 38 newborns presented normal T4 levels and their TSH values were observed to fall within the normal range (1-7 uU/ml) between the 6th and the 30th week after delivery. TNH was associated to fetal or maternal stress. Fetal stress causes were anoxia (39%), jaundice (37%), birth injury (24%), prematurity (16%), respiratory distress syndrome (10%), hemolytic disease (5%) and congenital anomalies (5%). Maternal causes were hypertensive disease (10%), premature separation of placenta (5%), eclampsia (5%), premature amniorrhexis (2.6%) and diabetes mellitus (2.6%). We concluded that 1) the frequency of TNH is higher in our population than the one observed in other studies. 2) the cutoff point for isolated TSH determination should be higher than the one usually utilized and 3) simultaneous TSH and T4 determinations should be employed in the screening programs for congenital hypothyroidism.

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ABSENCE OF SIALIC ACID IN MATURE THYROGLOBULIN (Tg) CAUSING CONGENITAL GOITER AND HYPOTHYROIDISM. Doi SQ, Grollman EF, Shifrin S, Weiss P and Medeiros-Neto GA. National Institutes of health, Behtesda and Thyroid Laboratory, Div Endocrinology, Hospital des Clínicas FMUSP

Tg Glyosylation is considered necessary to be secreted into the follicular lumen. The present study was carried out on Tg isolated from a patient with congenital goiter and hypothyroidism. The patient's parents were first cousins and had 5 siblings who were goitrous an hypothyroid. Previous studies have indicated the Tg was normal on ultracentrifugation and reacted normally with anti-Tg. Thyroid peroxidase activity in the gland was markedly increased, due to the intense TSH stimulation. Tg from the patient was isolated from the thyroid homogenate and had an elution pattern on Sepharyl S-200 that was normal, although poorly iodinated. This Tg acted like normal Tg on SDS-PAGE under non-reducing conditions, but behaved abnormally after 2-mercapto-etanol reduction. One of the most outstanding features of the Tg was the virtual absence of SIALIC ACID (N=24.3, Abnormal Tg: 1.0 nmol/ug) although normal levels of manose, galactose and glucosamine were found. The T3+T4 released from Tg after Pronase hydrolysis was less than 20% of normal values. In conclusion, the absence of sialic acid from the Tg molecule caused extreme structural changes that prevents normal synthesis of T3+T4 causing congenital goiter and hypothyroidism.

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24 HOUR GROWTH HORMONE SECRETION IN GIRLS WITH SEXUAL PRECOCITY. Mendonça B.B., Villares S.M., Domenice S., Arnhold I.J.P., Dahia P. L.M., Golabeck J., Liberman B., Bloise W. and Nicolau W. Division of Endocrinology - Hospital das Clínicas - University of São Paulo Medical School, São Paulo, Brasil.

Twenty four hour growth hormone (GH) secretion has been determined to classify patients with growth disorders, a mean (x) 3 ng/dl considered GH neurosecretory deficiency. We studied GH secretion in 6 girls with sexual precocity submitted to 24 hour evaluation of LH and FSH secretion. 5 patients had breast development (Tanner stage II-III), advanced height and/or bone age (cases 1-5) and one girl had premature pubarche (case 6-Breasts stage I, pubic hair stage II) as well as advanced height and bone ages. Blood was withdrawn through an I.V. catheter every 20 min. for 24 hours (total volume <7ml/Kg) during normal daily activities and meals and nocturnal sleep. GH for each case was determined in duplicate in the same RIA and the results analysed by the algoryth-detect method using a IBM-PC computer to verify the number and area under the pulses. The diurnal period was considered 8-20 h and nocturnal 20.20 - 7.40 h. Somatomedin -C levels were elevated for chronological age in case 3 and in cases 1, 2 and 4.

CASE	C.A. yrs.	sds height	B.A. yrs.	GH (ng/ml)					
				$\bar{x}$ 24 h	MAX. PULSE	NIGHT PERIOD		24 h. PERIOD	
						n pulses	area	n pulses	area
1	4.83	+2.5	6.83	1.12±0.97	4.7	3	33	8	61.2
2	5.58	-0.52	6.5	2.18±3.29	5.4	5	106	8	137.9
3	6.5	+2.8	7.83	3.7 ±5.5	26.6	6	200	10	240
4	7.58	-0.14	7.83	4.83±7.85	44.5	5	226	9	324
5	7.33	+0.21	10.5	3.1 ±4.4	18.4	4	158	7	202
6	5.83	+0.43	6.83	4.25±6.24	27.5	3	245	6	286

Great variation in GH secretion was observed among the patients, but with predomigance of nocturnal secretion. There was no correlation of height SDS with either X GH secretion or diurnal or nocturnal-GH area. In 2 patients, despite normal (case 2) or elevated (case 1) height, X 24 h GH secretion was <3 ng/ml. We concluded-that 24 h GH secretion in girls with sexual precocity is heterogeneous, and X can be lower than 3 ng/ml challenging this values as a limit for normal in the evaluation of growth disorders.

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24 HOUR LH AND FSH SECRETION AND RESPONSE TO GNRH IN GIRLS WITH SEXUAL PRECOCITY. Domenice, S.; Mendonça, B.B.; Villares, S.M.; Arnhold, I.J.P.; Frade, E.M.; Midori, M.; Mazi, C.R.; Nicolau, W. and Bloise W. Gonadal and Intersex Unit, Hospital das Clínicas, University of São Paulo, São Paulo, Brazil.

The laboratory diagnosis of true puberty (Tpp) relies on LH response to GnRh administration (ΔLH>15mIU/ml in our laboratory) and nocturnal LH secretion. Nevertheless, some patients with TPP may have prepubertal ΔLH on the GnRH test. We compared the 24 h. LH and FSH secretion with the GnRH test in 4 girls with TPP (cases 1-4), 1 girl with previous ovarian cyst (case 5) and 1 girl with premature pubarche (case 6). Blood was drawn every 20 min (vol <7/ml/Kg/24hs) through an I.V. catheter; meals, daily activities and nocturnal sleep were maintained. LH and FSH determinations were done in duplicate, in the same assay, and pulses were analysed by the algorithm-DETECT method in an IBM-PC compatible computer. Diurnal period was 8-20 hs and nocturnal 20:20- 7:40 hs. Basal estradiol levels were below sensitivity of the assay (<10 pg/ml) throughout the 24 hs.

CASE	C.A. (years)	B.A. (years)	PUBERTAL STAGE		AFTER GnRH 100 ug		LH(mIU/ml)		NOCTURNAL LH/ DIUERNAL LH	
			BREAST	PUBIC HAIR	ΔLH mIU/ml	ΔFSH mIU/ml	MEAN 24	MAXIMUM PULSE	NUMBER OF PULSE	AREA UNDER PULSE MIU/ml
1	5.58	6.5	II	I	27	41	2.49±0.29	3.2	3/1	10/3
2	6.5	7.83	III	I	36	15	2.89±0.29	3.9	9/7	74/79
3	7.33	10.5	III	II	11.5	6.4	3.28±2.63	13	6/2	120/19
4	7.58	7.83	III	II	12	29	3.12±1.52	8.4	5/2	119/15
5	4.83	6.83	III	I	4.1	17	3.1 ±0.2	3.8	3/1	35/9
6	5.83	6.83	I	II	4.3	22	3.1 ±0.34	4.0	7/4	57/51

A predominance of nocturnal LF secretion (area and number of pulses) was observed in 3 patients with TPP. The 2 patients with TPP and ΔLH<15 mIU/ml also had predominant nocturnal LH secretion. Cases 1 and 2 with TPP had ΔLH>15 mIU/ml and case 1 with a pubertal ΔLH had the lowest area under LH pulses, even lower than the patient with premature pubarche. We concluded that analysis of 24h. LH and FSH secretion did not offer a definitive method for diagnosis of TPP due to the heterogeneity of gonadotropin secretion.

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XX TRUE HERMAPHRODITISM ASSOCIATED TO MULTIPLE MALFORMATION:ABSENCE OF Y-SPECIFIC DNA SEQUENCES AND NORMAL H-Y ANTIGEN EXPRESSION. Arnhold IJP 1; Mendonça BB 1; Bloise W 1, Frade EM 1; Russo FO 2; Medeiros MA 1; Ferraz-Costa TE 2; Moreira-Filho CA 2. 1 Intersex Unit, Division of Endocrinology, Hospital das Clínicas and 2. Department of Immunology, Biomedical Sciences Institute, University of São Paulo, Brazil.

True hermaphroditism occurs when ovarian and testicular tissues are present in the same patient and usually is not associated to other malformation. We studied a patient with ambiguous genitalia from day 6 of life, phallus measured 3 cm, urethral and vaginal orifices were separate, slight posterior labial fusion and left(L) gonad palpable in the inguinal region, X chromatin on buccal smear was positive 28% and karyotype in peripheral lymphocytes 46,XX. hCG stimulation resulted in great testosterone increase (<10 to 832 ng/dl). Ultrasonography revealed uterus and tubes but no intrabdominal gonads. The patient was assigned a female sex and at 16 month exploratory laparotomy revealed uterus, tubes, an ovary on the right(R) and an ovotestis on the L, which was removed. The patient had multiple malformation: cranial assymetry, telecantum greater on the L, low nasal bridge, fat and cutaneous hypertrophy in lower limbs and gluteus with keratosis, specially on the dorsum of R foot. On the feet: normal halux on L and with clinodactilia on the R, 2nd. finger with bilateral macrodactilia and R Clinidactilia and 4th. finger with bilateral campodactilia and L macrodactilia. Growth was normal (Ht. 73.6 cm at 1y. 1mo.). The parents were young and not consanguineous and denied affected relatives. The mother was medicated with piperidolate hydrochloride in the first trimester and ampicilin and cephalotin for urinary infection on the 4th. month of gestation. The patient was born full term with 3.400 g and 46 cm length and had hypoglycemia.
The patient had no Y-specific DNA sequences, as detected by ZFY probe in Southern hybridizations (the probe corresponds to a Y chromosome region encompassing the socalled testis determining factor gene, or TDF). Expression of H-Y antigen assayed with monoclonal anti-H-Y antibody and an ELISA was normal.
Although hermaphroditism and malformations could be unrelated, one can speculate that alteration of a single autosomal or X-linked locus could have caused the patients's condition.

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PREMATURE TELARCHE(PT): STUDY OF ITS FRECUENCY AND ETIOLOGICAL FACTORS. PRELIMINARY REPORT. Youlton R., Valladares L.,García H., Cattani A., Jara A., Tijmes J. and Venegas P. University of Chile, Catholic University of Chile, Metropolitan Halth. Services of Santiago and Clinica Las Condes, Santiago, Chile.

In the last 10 years there has been an apparent increase in the number of cases of PT. 460 normal healthy girls of different socio-economic (SE) groups were specifically examined to detect the presence of PT. 65 of them had palpable breast tissue ≥ 1cm(14.1%); this incidence was not different in higher (15.7%) than in lower SE groups (12.7%) 53 girls with PT have entered in a study protocol; none of them have additional signs of precocious development. In 49% of the patients PT presents the persistance of neonatal thelarche. There is no history of known contact with estrogenic coumpounds. A detailed nutritional investigation did not show significant differences between affected and control girls of similar age and SE group. Heights and weights of patients are normally distributed around the mean for age. Bone age was significantly advanced in only two cases and retarded in two. Ovarian size was X 0.81 ml + 0.48 in PT and 0.47 ml + 0.22 in controls. Cysts > 0.7 cm havenot been observed. Basal serum Prolactin, LH and FSH were 20.2 ng/ml + 11.7, 0.48 and 2.7 + 1.4 respectively in PT and 20.6, 0.35 + 0.42 and 1.86 + 1.1 respectively in controls. After LHRH patients with PT showed a peak of LH= 5.05 + 2.78 and of FSH= 29.17 + 17.6 mIU/ml. Plasma Estradiol was 29.2 + 12.1 in PT and 30.6 + 12.0 in controls, but total estrogenic activity in plasma by RRA was 201.1 and 78.8 pg of E2 equivalents/ml. respectively. Presence of Stilbestrol (DES) and Zearalenone (Z) was excluded by TLC. This method and a sensitive RRA were used to detect presence of DES, Z and total estrogenic activity in meat; up to know, 30 samples of beef have been negative. Conclusions: The hormonal prolife of the patients is not different from that observed in normal girls of similar age, with the exception of increased estrogenic activity in plasma measured by RRA, a highly sensitive assay; 2 We have nor found estrogenic contamination in the samples of chicken and beef assayed; a larger number of samples as well as other foods need to be studied; 3 We cannot rule out the possibility that PT is a variant form of normal development whose prevalence has not been previously established.