499 SOMATOSENSORY EVOKED POTENTIALS IN CHILDREN TREATED FOR CONGENITAL HYPOTHYROIDISM. M. Vanasse, E. Laureau, J. Letarte, J. Glorieux, M. Desjardins, Université de Montréal, Centre de Recherches Pédiatrique, Hôpital Sainte-Justine, Department of Pediatrics, Montréal, Canada.

We have recently reported that despite early treatment, children with congenital hypothyroidism have lower scores in hearing-speech performance scales and practical reasonning at 18 and 36 months of age, using the Griffith developmental test. In order to clarify these observations, we studied somatosensory evoked potentials (SEP) in these children. We undertook both a retrospective and prospective study in hypothyroid children detected by the Quebec Screening Program and we report here results of SEP, obtained in 20 treated congenitally hypothyroid children age 5-12 as compared to 19 age and sex matched controls.

Two types of abnormalities were observed:

1° Increase (≥ 2 S.D.) in interpeak latencies were seen in 7 children affecting N13-N19 interpeak in 6 patients and N19-P22 in 2 children (one child showed an increased in both).

2° The morphology of the SEP was different in hypothyroid children with the P22 wave being much smaller and biphasic in shape.

These findings suggest that cerebral function is affected, at least electrophysiolocally, in this group of patients even if they are adequately treated. However the clinical significance of these abnormalities is not clear at the present time and is under study.

500 CALCEMIC AND CALCIOTROPIC HORMONE RESPONSES TO GLUCOSE INGESTION IN NEONATES: LOWERED SERUM CA, MG, P AND ELEVATED SERUM PARATHYROID HORMONE (PTH). PS Venkataraman, KE Blick, HK Fry, G Dasarathy (spon by OM Rennert). Pediatrics, Univ. Okla., Okla. City.

In neonates we studied the theses that glucose ingestion would: lower serum Ca, Mg, P and blood iCa; serum calcitonin (CT) would rise; and serum PTH would respond to serum Ca. In 10 normal term infants at age of 52±1 hr, serum Ca, Mg, P, iCa, PTH and CT were studied at 0, ½, 1 and 2 hr post ingestion of 1.8±0.1 g/kg of glucose over 20 min.

	0	+½ hr	+1 hr	+2 hr
Serum Ca, mg/dl	8.6± 0.4	7.8± 0.3*	7.9± 0.4*	8.0±0.3
Serum Mg, mg/dl	1.79±0.07	1.66±0.06†	1.62±0.06†	1.62±0.06†
Serum P, mg/dl	7.3± 0.3	6.9± 0.3*	6.9± 0.3*	6.8± 0.3*
Blood iCa, mg/dl	4.57±0.12	4.74±0.07	4.62±0.08	4.53±0.12
Serum glucose, mg/dl	52±4	97±10‡	118±11‡	102±12‡
Serum PTH, pmol/l	147±39	185±38*	233±68*	134±18
Serum CT, pg/ml	198±37	198±31	197±45	172±27

mean±s.e. * $p < 0.05$. † $p < 0.025$. ‡ $p < 0.005$.

In response to glucose ingestion serum glucose rose and serum P, Ca and Mg declined significantly. At +½ hr amount of glucose ingested correlated with serum glucose, r=0.65, $p < 0.05$, and with decline in serum Ca, r=0.66, $p < 0.05$. Serum PTH (RIA mid-mol, N 30-85) rose significantly and blood iCa and pH were unaltered. Serum CT was elevated $p < 0.005$ vs. adult values (RIA N< 102) and did not change. Thus, in neonates glucose ingestion: 1) lowers serum Ca, Mg and P; 2) serum CT is unaltered; and 3) rise in serum PTH and lowered serum P maintains blood iCa. We suggest that in neonates oral glucose lowers serum Ca and P with compensatory rise in serum PTH concentrations.

● **501** SOMATOMEDIN (Sm-C) in FETAL GROWTH. RA Vileisis and AJ D'Ercole, Dept. of Peds., Duke Univ. and Univ. of N. Carolina, Durham and Chapel Hill, N. C.

In the postnatal rat, suboptimal nutrition leads to a decrease in Sm-C which may account for the resultant decrease in growth. To study the effect of fetal malnutrition on serum and tissue Sm-C concentrations, intrauterine growth retardation (IUGR) was induced by uterine artery ligation on day 17 of gestation in 15 pregnant rats. Fetuses of the non-ligated horns served as appropriately grown (AGA) controls. After surgery the animals were allowed ad libitum feeding. On day 21 fetal blood and organs were obtained and analyzed for Sm-C and insulin by radioimmunoassay and glucose by glucose oxidase. Data are presented in relation to uterine position with 1 denoting the fetus nearest the ovarian artery. At each position, IUGR fetuses had decreased weight, serum and liver Sm-C, glucose, and insulin concentrations. (x±sem, *p<.05)

	weight		glucose		liver Sm-C		serum Sm-C	
	AGA	IUGR	AGA	IUGR	AGA	IUGR	AGA	IUGR
1:	4.4±.2	3.2±.2*	66±3	41±3*	.29±.03	.20±.02*	1.53±.05	1.08±.10*
2:	4.4±.2	3.0±.2*	64±7	30±5*	.30±.03	.18±.02*	1.53±.08	0.95±.10*
3:	4.4±.2	2.8±.2*	71±9	34±5*	.32±.03	.15±.02*	1.49±.11	0.89±.16*
4:	3.5±.5	2.6±.1*	67±8	37±8*	.22±.02	.12±.03*	1.34±.07	0.79±.26*

Fetal weight was significantly related to glucose (r=.730), liver Sm-C (r=.682) and serum Sm-C (r=.452). Stepwise linear regression demonstrated that these three factors in combination correlated highly with fetal weight (r=.836). A weaker correlation existed for insulin (r=.378) and lung Sm-C (r=.209). These data indicate that, in the fetus, Sm-C is influenced by nutrition and may have a mediating role in the control of growth.

502 THE DETERMINATION OF 17 HYDROXYPROGESTERONE AND 17 HYDROXYPREGNENOLONE IN PATIENTS WITH ELEVATED ANDROGENS. D. Wiener, T. Moshang, A. Bongiovanni, J. Cara, B. Marx and J. Paradise. The Children's Hospital of Phila., The University of Pennsylvania, Philadelphia, PA.

Inborn errors of steroidogenesis present either with the classic clinical findings of congenital adrenal hyperplasia (CAH) or, as more recently described, in women with the clinical findings associated with the polycystic ovarian syndrome (PCOS). The present study evaluated serum 17 hydroxyprogesterone (17 OH-Prog) and 17 hydroxypregnenolone (17 OH-Preg) levels in documented cases of 21 hydroxylase deficiency (untreated and treated patients), a male patient with 3 beta hydroxy steroid dehydrogenase deficiency (3 BHSD) and 14 adolescent females with hirsutism, oligomenorrhea and elevated androgens clinically diagnosed as having PCOS. The PCOS group was also studied with ACTH stimulation. In children with 21 hydroxylase deficiency, the 17 OH-Prog/17 OH-Preg ratio was always > 1. The 17 OH-Prog/17 OH-Preg ratio was < 1 in the patient with 3BHSD deficiency. In the females with PCOS, the baseline ratios of 17 OH-Prog/17 OH-Preg were < 1 in 10 of the 14 patients. In the 4 patients with a ratio > 1, 1 had a 17 hydroxyprogesterone response to ACTH similar to the patients with partial 21 hydroxylase deficiency. However she did not respond to glucocorticoid therapy. Of the 10 PCOS patients with a 17 OH-Preg ratio < 1, 2 had a 17 hydroxy pregnenolone response similar to patients with partial 3 BHSD deficiency. We did not find as high an incidence of late onset CAH in our patients presenting with apparent PCOS as reported in the adult literature. 17 OH-Prog/17 OH-Preg ratios did not correlate with response to glucocorticoids.

503 SUSTAINED CATCH-UP GROWTH IN HYPOPITUITARY CHILDREN TREATED WITH INTERMITTENT GROWTH HORMONE (GH) AND FLUOXYMESTRONE (F). Joyce E. Wise and Leon Librik (Spon. by George R. Honig), University of Illinois College of Medicine, Peoria, Illinois.

Children with GH deficiency exhibit rapid "catch-up" growth during the first year of therapy, but growth often slows later. In an effort to prolong "catch-up" growth, 9 children with hypopituitarism, initial chronological age (CA) 12.2 yr., initial bone age (BA) 8 yr., were treated for a mean of 3 years with four different regimens of intermittent GH (dose - 2 U/day X 28 consecutive days every 3 months X 2) and/or F (dose - 2.5 mg/m²/day). Each regimen lasted 6 months, and the order of regimens was chosen at random. The four regimens were: GH alone, GH + F on the same days that GH was given (GH + FS), GH + daily F (GH + FL), and F alone. The growth rates ($\bar{X}$±SE) and mean change in BA/change in height age (HA) for each regimen are listed below:

Treatment Regimen	Growth Rate cm/yr.	ΔBA/ΔHA
GH alone	7.2 ± .69	1.3
GH + FS	8.2 ± .51	.66
GH + FL	9.1 ± .54	1.06
F alone	8.3 ± .88	1.6

Mean growth rates and ΔBA/ΔHA were similar in 5 children who have stopped therapy (initial CA 13.3 yr., initial BA 9.5 yr., mean treatment length 2.6 yr., growth rate 8.2 cm/yr., ΔBA/ΔHA 1.16) and 4 children who continue treatment (initial CA 8 yr., initial BA 6.1 yr., mean treatment length 3.5 yr., growth rate 8.4 cm/yr., ΔBA/ΔHA 1.14). The combination of intermittent GH and F has maintained rapid growth for at least 3 years without excessive BA maturation.

504 EARLY PUBERTY IN KLINEFELTER SYNDROME DUE TO A CHORIONIC GONADOTROPIN (hCG)-PRODUCING TUMOR. Janaki Yadlapalli, Mariano Castro-Magana, Sujatha Kosuri, Moris Angulo, Boris Espinoza and Platon Jack Collipp, Nassau County Medical Center, SUNY, Stony Brook Health Sciences Center, Department of Pediatrics, East Meadow, NY 11554

A 15 year old white male presented with a 3-week history of productive cough, hemoptysis and weight loss. His past history revealed the presence of pubic hair at age 10 and pubertal growth spurt at 12 years of age. Physical examination revealed pubic hair stage V, axillary and facial hair stage III. Penile length was 12 cm and testicular volume 4 ml. The small size of the testes led to determination of karotype, which was 47,XXY. Chest X-ray revealed opacification of the right hemithorax. CT scan of the chest showed a large heterogeneous density with solid, fluid and calcified areas. Mediastinal shift to the left was noted. Thoracotomy was performed, and biopsied tissue revealed an immature Teratoma with histologic characteristics of malignancy. Serum Alpha-Fetoprotein (16.600 mcg/L), hCG (2,810 U/L) and LH (125 mIU/ul) were found to be greatly elevated. Serum FSH (8 mIU/ml) and testosterone (623 ng%) were normal. Probably the early puberty seen in this patient resulted from the secretion of hCG, from the thoracic teratoma.

This patient as well as the others reported in the literature demonstrate that the association of a gonadotropin-secreting teratoma with Klinefelter syndrome must be more than accidental.