0032-3998/92/3202-0249$03.00/0
PEDIATRIC RESEARCH

Vol. 32, No. 2, 1992
Printed in U.S.A.

SOCIEDAD LATINOAMERICANA DE ENDOCRINOLOGIA PEDIATRICA

Selected Abstracts from the 5th Annual Meeting October 27–30, 1991—Viña del Mar, Chile

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1

ENDOCRINE AND ABSORPTIVE FUNCTION AFTER 95% PANCREATECTOMY. 6-YEAR FOLLOW UP. 2 CLINICAL CASES. Asenjo S.;Gleisner A.;Wilhelm V.;Venegas G. (1); Rojas S.(2).

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Persistent hyperinsulism in the newborn (NB) entails a risk of recurrent hypoglycemia and secondary neurological damage. When performing pancreatectomy, a 95% extirpation is advised to prevent recurrence of hyperinsulinism. Two female 6 year old patients are presented, submitted to 95% pancreatectomy in the NB period for hyperinsulinism verified by determinations of insulin and plasma glucose and diagnosis of nesidioblastosis with components of endocrine dysplasia (Case A) and endocrine dysplasia with nesidioblastosis component (Case B). The following evaluations were done: Anthropometric (weight, size:Table NCHS-WHO), neurologic and endocrine function; glycemia (GLYC), insulinemia (INS), relation insulinemia uU/ml/glycemia (mm/L) (RelIG) VN=6, peptide C (pepC) in fasting (0') and post-prandial (90'), test of tolerance to oral glucose 1.75/Kg (OGT) and digestive absorption study:carotenemia, d-xylosemia, and fat absorption balance (Van de Kamer test). Case A, stature 116 cm, weight 21 Kg p 50 (NCHS). Case B, stature 118 cm, p 75, weight 24 Kg p 90 (NCHS). Cases A and B had normal neurologic test.

ENDOCRINE FUNCTION	GLYC mm/l	INS uU/ml	REL I/G VN=6	PEP C ng/ml	O.G.T. 0 - 30 - 60 - 90 - 120
CASE A:0'	5.0	3.0	0.6	0.6	76-160 - 68 -120 - 95
90'	6.4	17.1	2.7	1.6	
CASE B:0'	5.9	10.8	1.8	1.2	114-200 -207 -200 -126
90'	6.2	39.7	6.4	2.4	

ABSORTIVE FUNC.	CAROTENE mcg%	D-XYLOSE (VN=30%)	ABS FATS
CASE A:	93	35%	93%
CASE B:	130	61%	94.5%

In summary, in 6-year follow up, 95% pancreatectomy did not affect growth, endocrine function or the classical parameters of digestive absorption. In case B, a slight increase REL I/G was observed wihout clinical expression.

2

REPRODUCIBILITY OF GROWTH HORMONE (GH) SPONTANEOUS SECRETION AND RESPONSE TO CLONIDINE (CLO) IN BOYS WITH CONSTITUTIONAL SHORT STATURE
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There is still controversy on the diagnostic advantage of measuring spontaneous GH levels over pharmacological tests in children with short stature. There are few data available to confirm the reproducibility of the responses to these tests. The aim of this study was to evaluate the variability of the GH response to pharmacological test (CLO) and during sleep in the same child. Eight prepubertal boys with constitutional growth delay were studied, mean chronological age was 10.4±2.50 years mean bone age 7.86±2.77 years and their height standard deviation score were between -2.20 to -3.41. For the GH release studies during sleep, blood samples were drawn every 20 minutes from 7:00 PM until 7:00 AM. At 7:00 AM, all patients received an oral dose of 100 ug/m^2 clonidine and blood samples were drawn at 0,30,60,90 and 120 minutes. Both tests were performed twice, separated by a week, in every child. Serum GH concentrations were measured by RIA and the 12 hours profiles were analyzed by the CLUSTER program. The highest spontaneous peaks occurred during sleep in all children. The occurrence of the peaks was similar for each child in sleep test performed twice.There was no statistically significant difference between repeated sleep tests in either mean GH levels (first test, 4.57±1.78 ng/ml; second test, 4.37±1.73 ng/ml) or mean amplitude of the peaks (first test, 13.1±3.86 ng/ml; second test 13.9±1.78 ng/ml). GH response to clonidine test was equal or greater than 10 ng/ml in at least one occasion in seven out of eight patients.

We conclude that the GH spontaneous secretion showed good reproducibility while the clonidine test was markedly more variable.

3

SALT-WASTING CONGENITAL ADRENAL HYPERPLASIA (SWCAH) GROWTH DURING THE FIRST YEAR OF LIFE RELATED TO TREATMENT.
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SWCAH constitutes a metabolic emergency of difficult diagnosis and management during the newborn period. We have analyzed restrospectively the data of 97 (M=33,F=64) patients with this disease treated at our Hospital in order to characterize their clinical form of presentation, laboratory, treatment and follow-up of growth during the first year of life. Their chronological age (CA) on admission was F=13.2±11.7 days vs M=18.2±11.7 (p=0.05). Hiponatremia was significantly lower in males than females 118±10.7 vs 125±7.0 mEq/l, p=0.01. Serum potassium was similar in both sex M=7.97±1.2 vs F=7.9±1.5 mEq/l. Clinical signs on admission were ambiguous genitalia in 100% of girls, dehydration 52%, failure to thrive 48%, vomits 45% and diarrhea 19%. During the first year ol life a progressive impairment of height was observed (CA 0.15 years SDS=0.65±1.3, CA 0.51 years SDS=-1.47±1.2, CA 1.05 SDS=-1.73±1.1, p = 0.001). The time to normalize plasma sodium was significantly longer in M=58.4±39 days than F=28.9±21.9 days, p=0.036. There was a significant negative correlation between recovery of serum sodium and hiponatremia at onset (r=-0.42 p= 0.01), and with SDS of height at 12 months(r=-0.28, p= 0.05). Most of the patients were initially treated with parenteral corticosteroids. In approximately 75% of patients the treatment of maintenance was with oral hidrocortisone (25 mg/m^2/day) fluohidrocortisone 0.1 mg/day and sodium chloride 2 gr/day. In 25%, higher doses of hidrocortisone at varying times were used. The patients who received initially hidrocortisone at 25 mg/m^2/day had a better SDS of height at 12 months than those who were treated with higher dosis of hidrocortisone (-1.44±1.0 vs -2.53±1.2,p=0.002) In summary the significant growth retardation observed during the first year of life in patients with SWCAH could have been due to the delay of patients to normalize the serum sodium and/or to an excess of corticosteroids administered during the first months of life. With these results we should emphasize the importance of an adequate therapy from the moment of diagnosis to improve their height.

4

AUTOIMMUNE THYROID DISEASE (AID) IN CHILDREN AND ADOLESCENTS WITH INSULIN-DEPENDENT DIABETES MELLITUS (IDDM). Gruñeiro L.; Chiesa A.; Trifone L.; Bergadá I.; Raizman H.; Bergadá C.

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A greater incidence of AID has been described in children with IDDM nevertheless the incidence is variable in different series. We studied the AID in 88 children and adolescents (56 girls, 36 boys) range between 4.6 and 20 years (X±DS: 12.7±3.26 years). Serum T4, T3, TSH and/or TSH response to TRH, and antimicrosomal thyroid antibodies (AM) were measured at diagnosis or during follow up. Goiter was noted in 46%, AM in 39.7%, thyroid disfunction in 22.7%, hypothyroidism in 20.5% and hyperthyroidism in 2.3%. Hypothyroidism occurred in 16/18 patients following the onset of IDDM whereas hyperthyroidism preceded the appearence of IDDM. A positive family history of autoimmune disease (IDDM type I or thyroid disease) was present in 23.9% of all group and in 66.6% of hypothyroids. Autcantibody titters identified subjects with thyroid dysfunction with a sensitivity of 80%, specificity of 61.8%, a positive predictive value of 38% and a negative predictive value of 91.3%. We recommend that all children and adolescents must be screened after diagnosis of IDDM determinating TSH to identified thyroid dysfunction and testing AM to characterize the risk of future dysfunction and the need ot future testing in their follow up.