

EXPRESSION OF INTERLEUKIN-2 RECEPTOR ON LEUKEMIC CELLS IN VITRO.

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In order to induce biological effects which are conditioned by IL-2, it is required to combine it with a specific membrane receptor. It may be assumed with great probability that the IL-2R and its modulation can be included in uncontrolled growth of leukemic cells. That is why it has been decided to examine expression of IL-2R on leukemic cells obtained from the blood of 70 patients, aged 1-18 years with uncured acute lymphoblastic leukemia. The control group consisted of 20 healthy persons aged 1-18 years. The following factors have been estimated: receptor modulation and proliferation kinetics of unstimulated cells, as well as of stimulated by following mitogens: phytohemagglutinin, concanavalin A and pokeweed mitogen. The main identifying test was the test of indirect immunofluorescence. On newly isolated leukemic cells the IL-2R was present only in a small percent. Already after 24h. of incubation with mitogens its expression increases to a greater degree on leukemic cells than on lymphocytes of the control group. The possibility of inducing a IL-2R on leukemic cells implies their sensibility to IL-2 and the fact that they can proliferate under its influence.

GROWTH (G) AND SEXUAL MATURATION (SM) PATTERNS IN THALASSEMIA MAJOR CHILDREN (Th.).

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G and SM are frequently delayed in Th., but data are disomogeneous because of different transfusional and iron-chelation protocols. Height (H), weight (W), Tanner stages (T), penis length, testis volume (TV), serum ferritin (SF) and bone age (BA) were evaluated in 83 Th., aged 3-22y, transfused to maintain pretransfusional Hb levels >9 and >11 g/dl before and after 1980, respectively, treated with DFO, im until 1978, sc later on. Th. were divided by sex and age: A(3-8y), B(8-12y), C(>12 y). MALES(43): H was <3 centile(c) in 60%A, 50%B, 72%C; >25c in 0%A, 29%B, 6%C. H delay(HD) and age (CA) (r=0.69), BA and CA(r=0.95) and H age(HA) (r=0.93) were correlated*. W was <3c in 10%A, 22%B, 55%C; >25c in 40%A, 44% B, 11%C. HA and WA(r=0.69), HD and WD(r=0.92) were correlated*. In C, 63% had a penis length <5 cm, 51% a TV <5 ml and 45% T I pubic hair (PH). FEMALES(40): H was <3c in 9%A, 9%B, 67%C; >25c in 54%A, 45%B, 16%C. HD and CA(r=0.85), BA and CA(r=0.93) and HA(r=0.96) were correlated*. W was <3c in 0%A, 18%B, 44%C; >25c in 70%A, 64%B, 39%C. HA and WA(r=0.94), HD and WD(r=0.90) were correlated*. In C, 58% had no menarche, 11% T I breast and 21% T I PH. In both sexes auxological parameters were not correlated with iron stores(=SF). Our data show a precocious and progressive impairment of growth and bone maturation, especially in males, worst after 12 years in both sexes because of the lack of pubertal growth spurt.*p<0.001.

CORD PLASMA ERYTHROPOIETIN IN RELATION TO FETAL GROWTH RETARDATION

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From the first trimester the fetus can produce erythropoietin (EP). The only known stimulus for EP is hypoxia. Cord EP is elevated in pregnancies complicated by preeclampsia. Our aim was to assess if cord plasma EP is increased in fetal growth retardation (IUGR, <-2SD) of unknown cause (no maternal or fetal etiologic risk factor), or if it is a specific feature associated with preeclampsia.

We measured cord plasma EP by RIA in 74 infants born after preeclampsia of pregnancy, (geometric mean; range 98;18-7900 mU/l, p<0.001 compared with controls), in 23 singleton infants with IUGR (56;12-805, NS), in 13 pairs of twins (33;11-185, NS), and in 57 (20 preterm and 37 term) control infants (40;13-486). EP did not correlate with gestational age in control infants, nor in any other study group. EP correlated weakly with the SD score of the birth weight (r=-0.284, p=0.014) in the preeclampsia group, but not in IUGR. The EP levels were similar in first and second born twin, no difference was seen in discordant twins. EP correlated with umbilical arterial pH in preeclampsia group (r=-0.31, p=0.007), but not in IUGR or twin group.

We conclude that cord EP level is unrelated to gestational age, its correlation in preeclampsia with the degree of growth retardation suggests a relationship between this and hypoxia. However, EP is unrelated to the severity of IUGR of unknown cause, suggesting that most of these cases are unrelated to lack of oxygen.

RESULTS OF LONG TERM TREATMENT IN CASE OF DIHYDROBIOPTERIN SYNTHETASE DEFICIENCY

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Patient R.M. - a boy treated for phenylketonuria since the age of 3 weeks with very good dietary control. At age of 5 months admitted to the clinic because of psychomotor retardation. Muscular hypotonia and convulsions were observed. Diagnostic examinations for atypical PKU; BH loading test /with evident decrease in 4th and 8th hour/, analysis of bipterins in the urine /high neopterin, low biopterin/ pointed for the defect in dihydrobiopterin synthesis. Treatment with Dopa + Carbidopa and 5HT was introduced and at age of 1 year supplementation with BH, allowed to withdraw low phe diet and to decrease the doses of drugs /Dopa between 7-9mg/kg bw., 5HT 3-5 and now 8,9mg/kg bw., BH, 20-30mg/24h/. During 6 years observation period, almost full normalisation in psychomotor development was observed. DQ from the lowest 49 increased and since the age 3 1/2 IQ is in the range of 90. Physical development is at 10 percentile. Blood phe levels -around 3 mg%. Renal and liver function tests normal. The only complication is backward bone age.

WEIGHT REDUCTION AND EXERCISE CAPACITY IN OBESE ADOLESCENTS K. Zwiauer, K. Widhalm, M. Götz, J. Zarkovic Dept. Pediatrics, Univ. Vienna, A-1090 Austria

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Physical activity is more strenuous for obese children and adolescents compared to normalweight healthy peers. The purpose of the present investigation was to study the cardiorespiratory response to exercise and exercise capacity of obese female adolescents before and after a three week weight reduction. We studied highest oxygen uptake (VO2 max), resting heart rate (RHR), systolic blood pressure (SBP) and time to reach a pulse rate of 170/min (HF 170) during bicycle ergometer test in 8 grossly obese adolescents (x±SD 12.8±0.8 years, 63±17% overweight), treated for 3 weeks with a 2.4MJ/d mixed diet were studied before (B) and at the end (E) of the weight reduction period.

n=8	+ p<0.05, * p<0.01 (mean±SD)				
	weight	VO2 max	SBP	RHR	HF170
	(kg)	(ml/min/kg)	(mmHG)	(/min)	(min)
B	74.0±9.5*	24.0±4.1	128±17+	101±18+	5.7±0.6*
E	67.8±8.8	25.3±3.2	112±16	86±14	6.4±0.5

Resting heart rate and systolic blood pressure decreased significantly during the weight reduction regimen. Moreover, the HF170-time increased significantly indicating an improvement of the exercise capacity after weight reduction. At the end of the diet period the VO2 max was raised slightly but not significantly. Our data indicate that cardiorespiratory performance and physical capacity during moderate and strenuous exercise improved during weight reduction treatment in obese female adolescents.

CARDIAC FUNCTION DURING A LOW CALORIE DIET (2000 KJ) IN OBESE ADOLESCENTS K. Zwiauer, K. Widhalm, H. Schmidinger Dept. Pediatrics, Univ. Vienna, A-1090 Austria and * Dept. Cardiology, Univ. Vienna, A-1090 Austria

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Malignant cardiac arrhythmias are reported during prolonged drastic weight reduction with very low calorie diets (VLCD). Moreover, we could monitor potentially cardiac arrhythmias in a previously healthy obese adolescent girl treated with a mixed low calorie diet (2400 KJ) for only two weeks.

Therefore we studied cardiac function in 36 grossly obese adolescents (aged 12.1±0.6 years, overweight 54±4%, mean±SE) who were treated with a 2000 KJ diet for weight reduction by 24 hour Holter monitoring before and during the treatment at one week intervals (Del Mar Avionics, Model 500, USA). Weight loss during the low calorie treatment was 5.7±0.3 kg.

Results:	(mean±SD)				
	HR(avg)	HR(min)	HR(max)	VES(n)	SVES(n)
week 1	83±2	53±2	185±6	6	16
week 2	83±2	51±2	177±5	7	17
week 3	76±2	49±2	161±5	10	21
week 4	75±2 *	45±1 *	156±7 +	3	22

* p<0.008 p<0.03

Analysis of variance showed a significant decrease of average heart rate (HRavg), minimal heart rate (HRmin) and maximal heart rate (HRmax) during the weight reduction. However, the number of patients who revealed ventricular (VES) and supraventricular (SVES) extrasystoles did not change significantly. Neither before nor during the period of weight loss dysarrhythmias were seen.