

Abstracts (Supplement)

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1 ASPHYXIATING THORACIC DYSTROPHY
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Respiratory and others physiological functions were followed in four infants presenting clinical and radiological findings of Jeune's syndrome. Three infants died. The principal respiratory disturbances were tachypnoea, hypoxemia / $paO_2=40-60$ mmHg/ compensated respiratory acidosis / $paCO_2=42-60$ mmHg/ compensated respiratory acidosis / $paCO_2=42-60$ mmHg/. It was present also a decrease of V_T and an increase of V_E an increase of physiological dead space /VD phisiol./ and of venous admixture. The signs of hypoxia were evident on the EEG. Electromyographical examinations revealed in several sites including intercostal muscles widespread signs of neurogenic muscular sufference. The surviving infant shows at the age of 11 months: respiratory rate=66/min, $VT=0,81$ ml/cm, $V_E=599$ ml/kg, VD phisiol.=35,2%, QVA/QT=25,4%. It is suggested that should be taken into account, these findings when considering surgical therapy.

2 EFFECTS OF A 24 HOUR FAST IN OBESE CHILDREN
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Effects of a 24 hour fast were studied in 22 obese children aged 7 to 14 years and in 8 controls. Mean blood glucose /BG/ during fast dropped more in controls /0.90 to 0.50g/l/ than in obese /0.92 to 0.63g/l/. Plasma cortisol changes were similar in the 2 groups. FFA increased /p 0.01/ in the 2 groups, but the 24 hour mean level was higher in controls /4.0mEq/l/ than in obese /2.06mEq/l/. At the end of the fast, a ketonuria was present in all children. Serum alanine dropped similary in obese /28 to 24uM% and in controls /30 to 22uM%. All obese exhibited at the end of the fast a significant rise /p 0.001/ of branched chain aminoacids, not observed in controls. Responses to g glucagon /0.03mg/kg I.M./ were studied before and after fast. At time 0, BG response was higher and more prolonged in obese in spite of hyperinsulinism. At time 24 hour, the response to glucagon was strongly decreased in controls /0.50 to 0.74g/l for BG, 8 to 35 uU/ml for insulin/ while in obese BG raised from 0.63 to 1.06g/l and insulin from 25 to 88uU/ml. Thus 24 hour fast evidence in obese children 1/a sustained hyperinsulinism 2/concomitant criteria of hypoinsulinism suggesting a peripheral resistance to insulin 3/a limited lipolysis.

3 CHROMOSOME INSTABILITY DURING IMMUNOSUPPRESSIVE TREATMENT
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The mutagenic effect of cytostatic therapy of children suffering from non-tumorous diseases was studied. The chromosomes of cultured peripheral blood of children suffering from autoimmune diseases and membranoproliferative glomerulonephritis were analysed. 26 children treated with cyclophosphamide /3-5 mg/kg/day/ were examined. On the 4th-6th week of the treatment the number of chromosomal breaks increased to 18,6%, in contrast to the previously found 1,5%. The frequency of aberrations remained on a higher level during the therapy, afterwards it decreased, and reached again the control level 6 month later. At a smaller dose /1-2 mg/kg/day/ the frequency of chromosomal breaks was 5,5% /2 children examined only/. Chlorambucyl and 6-mercaptopurine also induced chromosomal aberrations in 12% and 13,5% respectively. The in vitro chromosome stability was examined in 5 patients during cytostatic therapy. The number of mutations induced in vitro by Lycurim was about the double that of healthy untreated children. Presumably the higher number of chromosomal mutations and the decreased chromosome stability induced both by the immunosuppressive treatment contribute to the higher frequency of malignancies in this group of patients.

4 BRONCHOSPASM PRODUCED WITH LOADING IN CHILDREN SUFFERING FROM ASTHMA BRONCHIALE
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Asthmatic patients with different duration of the disease and in different severity stage have been examined - on the basis of the general conventions - with ergospirometric method. The examinations have been carried out with medium loading during 6 minutes. It was found that the bronchospasm developed after physical loading was in no relation with the duration and the severity of the disease or the degree or duration of the loading either. After loading in 20 % of the patients bronchospasm has been observed. The authors' observations seem to be very useful in the prescription of the care of the patients suffering from asthma bronchiale.