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EFFECT OF HCG OR HCG+FSH TREATMENTS IN THALASSEMIC PATIENTS WITH HYPOGONADOTROPIC HYPOGONADISM (HH). R. Balducci, M. Toscano, G. Finocchi, M. V. Adamo, G. Mucicchi, A. Mangiantini, B. Boscherini. Clinica Pediatrica II Università, V. Clinica Medica I Università ROMA-ITALY

HH is common (40%) in beta thalassaemic patients, therefore has to be adequately treated (avoiding hepatotoxic drugs) to ensure a well adjusted sexual life. Considering that in HH patients we achieved

good results in pubertal development, using hCG treatment (1500 IU every 6 days), 8 HH thalassaemic patients (14 5/12-17 yrs; all with bone age more than 13 8/12) were treated with the same regimen. In 4 out of these 8, purified FSH (75 IU every 3 days) was added to hCG to evaluate the FSH effect on testosterone (T) response. (1° group hCG alone, 2° group hCG+FSH; Profasi HP and Metrodin Serono). In order to evaluate the kinetic of T response, plasma levels of T were determined basally and 1, 2, 4 and 6 days after hCG injection. This dynamic study and a clinical examination were performed at the beginning, at 4th month and 12 month of therapy. Results obtained in the 1° group confirmed our previous data obtained in HH, reaching stage G2-G3 after 12 month of therapy. In the 2° group, however testis size and T response were significantly higher with respect to the 1° group. At 4th month in the 1° and 2° group testis size and the area under T curve were (M±SE) 3.18±0.5ml and 13447±1325 vs 4.6±0.25ml (p<0.02) and 17427±962.5 (p<0.016) respectively. At 12th month in the 1° and 2° group testis size and T area were increased to 6.1±0.25ml, 23541±1361 and 8.1±0.6ml (p<0.01) and 34515±2664 (p<0.01) respectively. Our results demonstrate that hCG or hCG+FSH treatments are able to induce a good sexual development; moreover FSH addition seems to improve not only testis size but also T development.

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SERUM IMMUNOREACTIVE ERYTHROPOIETIN (sIEP) IN CHILDREN WITH CYANOTIC AND ACYANOTIC HEART DISEASE
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Erythropoietin (Ep) is the main regulator of red blood cell production. It is a commonly held notion that Ep levels are increased in conditions with decreased oxygen supply. The following study shows that this does not always hold true.

We measured sIEP in 79 children with cyanotic and acyanotic heart disease, aged 1 day to 10 years. During the first days of life sIEP varied widely, and the levels seemed mainly to reflect intrauterine conditions and events taking place around birth.

Cyanotic infants aged 2 weeks to 4 months had significantly higher sIEP values than their older counterparts. After 4 months of age the levels were the same in cyanotic and acyanotic children, and similar to normal adults.

sIEP did not correlate significantly with haemoglobin, haematocrit, arterial PO₂ or saturation.

The sIEP levels in cyanotic children display the same pattern as observed in man and animals exposed to prolonged hypobaric hypoxia, in which after an initial rise in Ep concentrations the levels fall to normal while increased erythropoiesis is sustained.

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USE OF A PANEL OF TRADITIONAL AND NEW MARKERS OF GERM CELL TUMORS

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Eighty patients with germ cell tumors were diagnosed at the Children's Hospital, University of Helsinki, between 1968 and 1986. The primary tumors were malignant in 22 and benign in 58 patients. 35/58 of the benign tumors were sacrococcygeal teratomas of newborns. Four of these 35 patients subsequently developed a malignant yolk sac tumor. Serial samples from patients with a primary malignancy were assayed for AFP, placental proteins hCG and SP₁, and two new tumor markers CA 12-5 and CA 19-9. AFP was elevated at diagnosis in 8/11, placental protein in 4/8, CA 12-5 and CA 19-9 in 1/8 patients. Ten patients operated for a sacrococcygeal teratoma in the newborn period were followed monthly by a panel of tumor markers. Three of these patients developed a malignant yolk sac tumor at age 8-19 months. AFP values became elevated in all 3 cases and SP₁ in one. One recurrence was also associated with a slightly elevated CA 12-5. Patients with germ cell tumors should be carefully followed up by a panel of tumor markers in order to gain information how to detect the potential cases of subsequent malignancy.

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Factors predictive for chronic course of idiopathic thrombocytopenic purpura (ITP) in children

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Childhood ITP is generally a self-limiting syndrome. However, 5-15% of patients develop a chronic thrombocytopenia. We tried to find out factors predictive for chronic course in a series of 91 consecutive children who got their primary diagnosis of ITP at the Helsinki University Children's Hospital 1975-84. Age at onset varied between 0.4 and 16.0 (median 4.1) yrs; 42 were females. 51 patients had had an acute viral illness 2-4 weeks prior to diagnosis. 20 patients received corticosteroid treatment to their early bleedings. Platelet count reached 100x10⁹/l within 3 months in 89% of the patients. Eight children were thrombocytopenic still at 6 months (and also at 12 months) after diagnosis. Only one of these 8 patients with chronic ITP had had an antecedent viral infection. They were significantly older (median 10.0 yrs), and all but one were females. Their acute symptoms had been insidious. Use of steroids did not have any influence on outcome. These results suggest a different etiology and pathogenesis in patients with acute and chronic ITP of childhood.

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A FOLLOW-UP STUDY OF 24 CHILDREN BORN TO INTRAVENOUS DRUG USERS INFECTED WITH HIV.

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Between October 1985 and February 1987, screening for HIV antibodies was carried out in 39 pregnant heroin addicts. Twenty four women (15 prostitutes) were found to be antibody positive on both enzyme-linked immunosorbent assay and Western blot procedures. Detection of HIV antigens in serum was negative in all cases and there was no clinical evidence of AIDS or AIDS-related complex in any of the mothers at the time of delivery. Twenty four infants were born to these women. Antibodies against HIV were detected at birth, by both techniques in all children. Identification of anti-HIV antibodies by Western blot electrotransfer showed identical patterns when serum samples from each mother and child were compared. Two additional patients, aged 7 and 14 months, whose mothers were also drug abusers were referred to our hospital with clinical and laboratory features of AIDS-related complex. Their antibody response against viral proteins was different from the response of their seropositive mothers. Mean period of follow-up was 8 months (range 2 to 17). Of the ten infants above the age of 6 months, 6 became seronegative (at a mean age of 8 months) and 4 were lost of the follow-up study. The remaining 14 children (below the age of 6 months) were still positive but clinically healthy. Antibodies against polypeptide p24 were found to be the most persistent. Conclusions: High incidence of HIV infection among pregnant addicts in our city. Passively transferred antibodies disappear at a mean age of 8 months.

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Berlin prospective Study of children born to HIV (human immunodeficiency virus)-positive mothers

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The presence of HIV-IgG-antibodies (AB) in newborns does not provide conclusive evidence of an actual infection with the virus. This is determinable only by

long-term observation concerned in accordance with clinical, immunological and virological criteria.

Consequently, since July 85, all NB of HIV-AB-positive mothers are being kept under close control and 38 newborns have so far been included in the study.

During an observation period of now over 20 months, none of the children have as yet contracted AIDS. A slight neurological abnormality was apparent in 3 children. 1 child who had become seronegative at 6 months of age suffered a bacterial meningitis at 11 months of age. Now the HI-virus was identified in CSF despite further seronegativity.

As expected, 15 children whose virus culture was negative, became seronegative at between 3 to 7 months, however also 7 children with a positive virus culture became seronegative. These results clearly show that AB-Screening is not a sufficient method of course control but that long-term observation is essential for accurate classification of HIV-AB in newborns.