5 CLINICAL AND ELECTROPHYSIOLOGICAL FINDINGS IN ADOLESCENT PATIENTS WITH PHENYLKETONURIA (PKU)

Ludolph A. C., Ullrich K., Nedjat S., Masur H., Bick U.
Dept. of Neurology and Pediatrics, University of Münster, FRG

18 early treated adolescent patients with PKU (mean age 18.2 y) were tested clinically and with electrophysiological techniques inclusing sensory and motor conduction velocities such as evoked potentials (SSEP, BAEP, VEP). Three patients showed slight impairment of proprioception, clinical examination was otherwise normal. The level of education in our patients did not reach that of the average population. The central motor and sensory latencies to the upper but to the lower limbs were significantly prolonged as compaired to controls ($p < 0.001$). BAEP revealed a significant increase of interpeak latencies I-V and III-V ($p < 0.005$ and < 0.025). 30 % of the patients had subclinical visual deficits and in 50 % the sensory conduction velocity of the right sural nerve was slightly decreased. Changes in central latencies and BAEP but not VEP correlated to the degree of metabolic control.
- The different vulnerability of pathways in the central nervous system might be related to the timing and sequence of maturation in the human nervous system. Morphological findings of de/dysmyelination (MRI) did not correlate to the functional deficits, suggesting that these are caused by morphological as well as pharmakological deficits like dopamine deficiency.

6 THE LOCAL AND SYSTEMIC ANTIBODY RESPONSE IN INFANTS AFTER ORAL ADMINISTRATION OF INACTIVATED ENTEROPATHOGENIC E.COLI SEROTYPES 0111 AND 055

Raja Lodinová-Žádníková, Bohuslav Korych
Institute for Care of Mother and Child, Prague, ČSSR

In 10 infants 1/up to one month of age 2/from 2-7 months of age, the dynamics and formation of different antibody isotypes after orally administered inactivated enteropathogenic E.coli strains 0111 and 055 was followed after the first and booster dose, by indirect immunofluorescence method.

Infants up to one month of age produced antibodies of IgM and IgA isotypes in stool after the first and booster dose of antigens. The serum IgG antibody increased after 48 hours following the first and second dose of antigens and decreased during 3 days. The infants aged 2-7 months, however, expressed predominantly the IgA isotype response in stool after the first and booster dose of antigens. The serum IgG level did not change.

The results help to explain the good therapeutic effect of the oral inactivated vaccine we have been using for treatment of gastrointestinal infections.

7 INFLUENCE OF pO_2 ON CEREBRAL BLOOD FLOW VELOCITIES IN PRETERMS < 32 GESTATIONAL WEEKS (GA)

Rabe H, Jorch G
Childrens' University Hospital, Münster, FRG

Animal studies show that beside pCO_2, pO_2 influences brain circulation. Since fluctuations of pO_2 occur frequently in ventilated preterms, we investigated whether changes within the therapeutic range of 40-90 mmHg affect blood flow velocity of internal carotid artery or not. Within continuous Doppler studies in preterms < 32 GA over 1 h we looked for accidental fluctuations of pO_2 > 10 mmHg occuring within 10 min. We questioned whether there was a concomitant change of Doppler parameters. Continuous pulsed Doppler measurements were performed with a miniature transducer fixed onto the infants fontanelle in 10 preterms (mean GA 28 w, mean birthweight 1321 g). A connected microcomputer evaluated the Doppler curves. A total of 15 pO_2 changes > 10 mmHg occured. In 12 of 15 events there were no concomitant changes of mean maximum velocity > 8 % (coefficient of variation of method) which was significant by sign-test, $p < 0.05$. There were no significant changes in the enddiastolic or peak systolic flow velocities outside the normal range. In conclusion there were no significant changes of cerebral Doppler parameters with accidental pO_2 changes.

8 NUCLEAR MAGNETIC RESONANCE AND IRON OVERLOAD IN THALASSEMIA MAIOR AFTER BONE MARROW TRANSPLANTATION

F. Schettini, D. De Mattia, V. Sabato, N. Santoro, G.C. Del Vecchio, G. Martinelli, P. Di Bartolomeo°, G. Torlontano°
1st Pediatric Clinic, University of Bari, Italy.
° Division of Hematology, University of Chieti-Pescara, Italy.

Nuclear magnetic resonance (NMR) imaging is a new and noninvasive technique to detect tissue iron. In fact the signal intensity on spin echo images is as low as tissue iron load is high. We used this technique to assess tissue iron load in 10 B-thalassaemic maior patients (mean age 11 ± 4.8 years) after bone marrow transplantation (BMT); the follow-up ranged between 10 and 54 months (mean 27.3 months) from transplant. In all patients a significant correlation has been observed between the summation of signals given out from all the organs examined (liver, spleen and pancreas) with iron overload versus serum ferritin levels ($r = 0.64$; $p < 0.05$). Signals obtained from each organ were not correlated with serum ferritin levels, but the only exception was the signal obtained from liver ($r = 0.69$; $p < 0.05$).

We conclude that NMR is of great diagnostic validity for the evaluation of iron load of each organ in patients with B-thalassemia subjected to BMT, while serum ferritin levels are related only with total body iron overload. At last these findings lead us to speculate about the need in these subjects to continue a chelation therapy or to undergo periodic phlebotomy.

9 A. Moustouannis, N.G. Beratis. Department of Pediatrics, University of Patras, Patras, Greece.KILLING OF ECHINOCOCCUS GRANULOSUS SCOLICES BY SENSITIZED LYMPHOCYTES.

The immune response of lymphocytes from subjects with hydatid disease (active or operated) was studied by cultivating the mononuclear cells from such patients in the presence of human hydatid fluid and measuring the [^{3}H]thymidine incorporation into the cells. Maximum transformation was observed on the 6th to 7th day of culture at a hydatid cyst protein concentration of 15 µg/ml of culture medium. The stimulation index (SI) of cells from 10 hydatidosis patients was 4.3±1.6 (mean±SD), with a range from 2.1 to 7, whereas in the control cultures it was 1.1±0.3, with a range from 0.5 to 1.5 ($p<0.0005$). Cocultivation of lymphocytes from hydatidosis patients and from normal subjects with E. granulosus scolices resulted in a greater killing of the scolices by the sensitized than the nonsensitized lymphocytes. The diffence was significant after culturing for 1 to 7 days ($p<0.005$). Similarly, the viability of the sensitized lymphocytes cocultivated with scolices was significantly lower than that of nonsensitized lymphocytes cultured in parallel under the same conditions ($p<0.005$). The findings show that sensitized lymphocytes from subjects with active or cured hydatid disease due to E. granulosus can kill scolices in vitro demonstrating the importance of cellular immunity in the host defense against the parasite.

10 CEREBROVASCULAR RESPONSE TO HYPOCAPNIA IN HYPOTENSIVE NEWBORN PIGLETS

Andrew Whitelaw, Bente Karlsen, Kirsti Haaland, Petter Andreas Steen, Marianne Thoresen. Depts of Paediatrics & Anaesthesia, Ullevål Hospital, Oslo, Norway.

It has been suggested that hypocapnia (which normally produces cerebral vasoconstriction) may be a cause of periventricular leukomalacia. This study tested the hypothesis that hypocapnia superimposed upon hypotension produces a further reduction in cerebral blood flow velocity (CBFV).

In 12 newborn piglets, CBFV was measured continuously through an artificial fontanelle. Hypotension was induced by removing 30 ml/kg of blood over 30 minutes. Increasing the ventilator rate from 20 to100/m min reduced the average pCO2 from above 5.0 to below 2.0 kPa. When mean arterial pressure (MAP) was held steady at 45 mm Hg or above, hypocapnia produced a signficant drop in CBFV but , in all the piglets with a steady MAP below 38 mm Hg, hypocapnia failed to change CBFV. Jugular vein or sagital sinus blood showed no increase in hypoxanthine during hypocapnia. Hyperventilation (without haemorrhage) produced a signficant drop in MAP, preventable by infusing colloid.

Hypocapnia itself does not further reduce CBFV in the hypotensive piglet. However, the pressure effect of hyperventilation may significantly impair the cerebral circulation.