47 REACTIONS OF PARENTS OF CHILDREN WITH MALIGNANCY.

A.Kreuger, K.Gyllenskjöld, G.Pehrsson, S.Sjölin. Dept of Pediatrics, University Hospital, Uppsala, and Pedagogical Institution, University of Stockholm, Sweden.

The parents of eight children who had died of malignant diseases were interviewed about their experiences concerning the medical and social care of the children. There was a great need for repeated and thorough information about the disease and the therapy to all in the family. The risk of isolation of the family could be diminished by active measures. There was a great need for continuity with as few people as possible involved in the care. It proved important to recognize at an early stage the economical and social problems of the family and help the family to solve them. Through this investigation we hope to achieve a better understanding of the difficulties of the family problems and how to manage them.

48 SERUM LIPOPROTEINS IN CHILDREN WITH ANEMIA.

M.Seip, S.Skrede. Dept. of Pediatrics, National Hospital, Univ. of Oslo, Norway.

An association between uncomplicated anemia and low serum lipid levels, particularly low cholesterol, has been found in several studies. In a previous study of 17 children with various forms of uncomplicated anemia a positive correlation between hemoglobin and serum cholesterol levels was found. The present study aims at a further characterization of the serum lipoprotein pattern in anemic children. Nine children with different types of anemia and 9 healthy control children were studied. The anemic children had rather high plasma triglycerides, but low total cholesterol and phospholipids. The levels of HDL cholesterol were particularly low, with a mean of only 70 % of the controls. A positive correlation was found for hematocrit with HDL cholesterol, but not with LDL cholesterol. A striking, inverse correlation between HDL cholesterol and VLDL triglycerides was present. Anemia thus represents another pathophysiological situation where the metabolism of these lipoproteins shows interdependent changes. The findings are not compatible with the previous hypothesis that the hypolipidemia accompanying uncomplicated anemia is due solely to the diluting effect of increase in plasma volume in anemias.

49 FOETAL HEXOKINASE ISOENZYMES AND HAEMOGLOBIN F IN CHILDREN WITH HAEMATOLOGIC DISEASES.

M. Gahr, W. Schröter.
Universitätskinderklinik Göttingen, West-Germany.

Foetal erythropoiesis is characterized by many properties of the erythrocytes the best known of which is the presence of haemoglobin F in the erythrocytes. A distinct pattern of hexokinase isoenzymes is a further typical sign of foetal erythrocytes.

We have investigated the hexokinase isoenzymes of 4 children (age: 2, 8, 9, and 11 years) with acute myeloid and juvenile chronic myeloid leukaemia, erythroleukaemia and aplastic anaemia whose erythrocytes exhibited in - creased haemoglobin F-concentrations (8, 65, 53, and 10 %). Their hexokinase isoenzymes were partially purified by treatment with DEAE-Sephadex and separated by analytical isoelectric focussing in polyacrylamide gels.

The foetal type of hexokinase isoenzymes was observed in the erythrocytes of all patients. This suggets that the synthesis of haemoglobin F and of the hexokinase isoenzymes are under common control as well in malignancies as in non-malignant disease.

50 HEMOGLOBIN LEPORE IN SICILY: HEMATOLOGICAL AND BIOSYNTHETIC STUDIES.

G. Russo, G. Schiliro, S. Musumeci, G. Pizzarelli, A. Fischer, M.A. Romeo. Dept. of Pediatrics, University of Catania, Italy.

The results of hematological and biosynthetic studies in 15 subjects carriers of Hb Lepore and in 5 double heterozygotes for Hb Lepore and thalassemia are presented.

In the carriers the hematological and biosynthetic data are compared with carriers of thal., while the five double heterozygous patients were compared with ^{o} and $^{+}$ thal. major subjects. In the carriers of Hb Lepore no synthesis of chains was observed in the peripheral blood cells, on the contrary we found a peak in the bone marrow. Double heterozygous subject with circulating nucleated red cells showed in the peripheral blood chains synthesis.

51 FETAL HEMOGLOBIN IN SICKLE-CELL ANEMIA.

S. Özsoylu, Section of Hematology, Department of Pediatrics, Hacettepe University, Faculty of Medicine, Ankara, Turkey.

Fetal hemoglobin values with one exception were found elevated (> 2) in 89 patients with sickle-cell anemia and the mean level of Hb F was of 11.45 %. In 47 of these patients Hb F was more than 10 % by Singer's method. Hemoglobin and/or hematocrit values were not correlated with the Hb F concentrations of these patients and no difference was found between the Hb or Hct values of the patients with less than or more than 10 % of Hb F. Retics counts and normoblastemia were not found correlated with Hb F concentrations of the patients.

More than one patient with sickle-cell anemia was present in at least 9 families. Although the Hb F values of the siblings differed significantly from each other, their anemia did not seem to be affected with their Hb F concentrations either. Since the genetic differences were less between the siblings, there is no evidence that Hb F level in patients with sickle-cell anemia would influence the hematological findings of these patients.

52 EFFECTS OF PHOSPHORUS ADMINISTRATION ON IRON BALANCE IN OMOZYGOUS β-THALASSEMIA.

S.Pantelakis, A.Karaklis, C.Vretos, P.Lapatsanis, S.Doxiadis. Institute of Child Health Athens, Greece.

Previous work has shown that thalassemic children have increased phosphaturia and that phosphorus administration restores phosphorus balance and results in improvement of their bone lesions. In addition, the absorption of iron is known to be decreased by the presence of oxalate, phosphate and phytate in the food. In order to study the effect of phosphorus administration on iron balance in thalassemia, twenty iron and phosphorus balance studies were performed in 14 thalassemic children, 8 boys and 6 girls, aged 13 months to 16 years. Ten studies were performed while the patients were on oral phosphate administration (1.5-2.0g / day) and ten while they were not. Phosphorus intake was found to have a negative correlation with net iron absorption ($r = 0.462$, $p < 0.05$) and iron balance ($r = 0.455$, $p < 0.05$). In addition a linear correlation was found between fecal phosphorus and fecal iron ($r = 0.442$, $p < 0.05$) and between net phosphorus absorption and urinary iron excretion ($r = 0.487$, $p < 0.05$). In conclusion it seems that in thalassemia, phosphorus administration, in addition to the beneficial effects on the bone lesions has an effect on iron balance. Oral phosphorus not only prevents iron absorption but it increases to a certain degree iron elimination in the stools. The effect of net phosphorus absorption on iron elimination in the urine although statistically significant does not play an important role in the iron balance.