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ACQUIRED STOMATOCYTOSIS IN INFANTS RECEIVING PARENTERAL FAT EMULSION. J.D Ross and P.E. Mendoza. Dept. of Peds., W.Va. Univ. Med. School, Charleston Div. (Spon. by Barbara Jones).

A premature infant on total parenteral nutrition (TPN) with Intralipid$_R$ (IL) developed hemolytic anemia with striking stomatocytosis, which cleared when IL was discontinued; 10-month hematologic follow-up has been normal. Another infant on IL also developed hemolytic anemia and stomatocytosis, but expired from complications of prematurity. Ten additional high risk infants on TPN with standard amounts of IL were studied. All developed stomatocytosis. In 9 cases 25% to 50% of RBC's were affected. Stomatocytosis, absent before IL administration, appeared by day 4-7 of IL, peaked at day 5-18, and disappeared 7-10 days after discontinuing IL. One of these infants developed severe hemolytic anemia. Routine transfusions obscured signs of hemolysis in the others. Three infants receiving TPN without IL showed no stomatocytosis; 2 of these developed stomatocytosis when switched to IL.

Stomatocytosis is associated with severe abnormalities of RBC membrane composition and function and, occasionally, hemolysis. Also, fetal RBC membranes are different from those of adults. Our findings suggest: (1) IL or its metabolic byproducts interact with newborn RBC's to induce a structural or functional membrane change and may increase susceptibility to hemolysis: (2) Infants on IL should be monitored for signs of hemolysis; (3) An experimental model for the study of membrane physiology is suggested.

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THE USE OF TISSUE LEUKEMIA BURDEN (TLB) IN THE STAGING OF CHILDHOOD ACUTE LYMPHOCYTIC LEUKEMIA (ALL) Shende, Ashok and Lanzkowsky, Philip, SUNY at Stony Brook, Health Sciences Center and Long Island Jewish-Hillside Medical Center, Department of Pediatrics, New Hyde Park, NY 11040

Ninety eight patients, aged 5 months to 15 years with ALL diagnosed between April 1971 and November 1977 were staged on the basis of TLB using a modified staging system of non-Hodgkin's lymphoma (NHL). Twenty-two of these patients had stage 0 disease (marrow involvement only), 17 stage 1 (marrow and single nodal or extranodal site involvement), 11 stage 2 (marrow and two or more nodal or extranodal sites involvement on the same side of the diaphragm), 45 stage 3 (marrow and tumor bulk on both sides of the diaphragm or intrathoracic or extensive abdominal disease) and 3 stage 4 (marrow and central nervous system disease). The median initial white blood cell count (W.B.C.) was 10,250/cub.mm. (range 400 to 900,000). Of the 98 patients 39 were evaluable for 3 years disease free survival (DFS).

	White Cell Count		Disease Free Survival	
Stage	< median	> median	< 3 years	> 3 years
0,1,2	31	19	6	12
3,4	18	30	15	6
p value	< 0.02 by x^2 analysis		< 0.02 by x^2 analysis	

The data reveals that initial white blood cell count and the 3 year DFS are associated with stage. This staging system permits the inclusion of ALL patients as subclasses of stage IV (NHL) and thus allows more accurate interpretation of results of clinical trials of NHL.

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OUTCOME OF EMERGENCY UNCROSSMATCHED BLOOD TRANSFUSION IN CHILDREN. Michael M. Siegel, Jorge A. Ortega, Jackie M. Dillinger, Sabu Dzinovic (Sponsored by Harry T. Wright), University of Southern California, Childrens Hospital of Los Angeles, Department of Pediatrics, Los Angeles.

Transfusion of uncrossmatched blood is only done in an emergency situation because of the risk of transfusion reaction. We reviewed the records of 65 patients who received uncrossmatched blood between 1969 and 1976. Emphasis was placed on the presence of major transfusion reactions. Forty six of these patients received group specific blood. Of the 19 patients that received non type specific blood 15 were transfused with O- blood, two with O+ and 2 others with B-. In one patient receiving non type specific blood the crossmatch was found to be incompatible after the blood had been given. Eighteen patients had been previously transfused and 30 received subsequent transfusions. No documented transfusion reaction occurred in any of the 65 patients. Pre-existing shock and bleeding diathesis made this evaluation difficult in some cases. Eight patients died within 24 hours of receiving their uncrossmatched blood, none with evidence of transfusion reaction. In life threatening situations when time will not permit compatability testing, uncrossmatched O- or type specific blood should be used because the risk of a major transfusion reaction is minimal.

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DIRECT INTERFERENCE CONTRAST MICROSCOPY (DICM) AS A MEASUREMENT OF SPLENIC FUNCTION. Richard H. Sills and Frank A. Oski. Dept. of Peds., SUNY, Syr., N.Y.

The presence of red blood cell (RBC) membrane surface "pits" as revealed by DICM has been demonstrated to commote splenic hypofunction. DICM was employed to study splenic function in a variety of pediatric disorders and to compare its sensitivity with that of Howell-Jolly body (HJB) counts. Normal adults exhibited pitting on 0.70± 0.82% of their RBCs as compared to 0.90±0.71% for infants and children 6-108 mos. of age. In 14 pts. without spleens the mean pit count was 38.7±12.6%. In 6 children with sickle cell anemia (SS) ages 7 mos. to 16 yrs., the pit count averaged 35.8%. A 7 mo. old with SS and 22.5% pitting was hypertransfused over 2 wks. and his % pitting fell to normal within 1 wk. and remained there for 3 mos. This documents the presence and reversibility of splenic hypofunction in young SS pts. A pt. with S-C disease and one with S-thalassemia also exhibited splenic hypofunction (48.4 & 54.6% pits respectively). 5 newborns with sepsis and 5 children with sepsis &/or meningitis were evaluated as to the possible role of hyposplenism in septicemia. No abnormality in RBC pitting was noted. DICM was compared to the presence of HJB from peripheral smears. No pt. had HJB without an elevated pit count. Surface pits always preceded the appearance of HJB in splenectomized pts. In 32 samples with elevated pit counts, 7 had no HJB. These data indicate that the use of DICM for detection of surface "pits" is a rapid and sensitive technique for the detection of splenic hypofunction and is superior to the use of HJB counting.

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HERIDITARY ELLIPTOCYTOSIS, RED CELL FRAGMENTATION, AND DECREASED GLUTATHIONE (GSH). Clark M. Smith II, Peter F. Coccia, William Krivit, Elaine Berger, James G. White, John W. Eaton. Univ. of Minnesota, Depts. of Pediatrics and Medicine, Minneapolis, 55455.

Marked poikilocytosis accompanied by membrane fragmentation and microspherocyte formation have been described in occasional cases of hereditary elliptocytosis. We have studied a healthy 10 year old black male with a compensated hemolytic anemia, elliptocytosis and severe red cell fragmentation. Several parameters known to affect RBC membrane stability were found to be normal: pyruvate kinase activity, RBC ATP content, unstable hemoglobin evaluation, direct and indirect Coomb's tests, membrane thermal sensitivity, RBC calcium and calcium permeability, RBC cholesterol and phospholipid content, and the distribution of membrane phosphatides including phosphatidyl ethanolamine. The only abnormality found was a RBC GSH content approximately 50% of normal (below that seen in most G-6-PD deficiencies). Furthermore, in centrifugally-separated cells, GSH content decreased progressively with increasing cell density. Unlike G-6-PD deficiency the low GSH content was not accompanied by abnormal oxidant sensitivity; GSH was not diminished by incubation with ascorbate-aminotriazole and no methemoglobin or sulfhemoglobin was formed. Enzymes involved in GSH metabolism (GSH reductase, G-6-PD, GSH peroxidase) were slightly elevated. Incubation of RBC with H^3-glutamate, with subsequent molecular weight separation of the GSH intermediates, suggests an intact GSH synthetic pathway but diminished H^3-glutamate content. The low RBC GSH may be related to the RBC morphologic features and hemolysis.

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RECURRENT PNEUMOCOCCAL MENINGITIS IN INFANTS UNDER 15 MONTHS WITH SICKLE CELL DISEASE. Sreedhar P. Rao., Audrey K. Brown., Dept. of Pediatrics., SUNY-Downstate Medical Center, Brooklyn, NY. (Spon. by C.D. Cook)

Children with sickle cell disease have a markedly increased susceptibility to pneumococcal septicemia and meningitis, attributed primarily to the loss of splenic function. Review of 24 cases of pneumococcal meningitis in sickle cell patients revealed a high frequency of recurrence chiefly among those infants whose initial episode of pneumococcal meningitis occurred before the age of 15 months. Twelve of 24 patients (50%) were under the age of 15 months; eight (33%) were below the age of 12 months (5,6, 6½,8,9,11,12, and 12 months) at the time of initial episode of meningitis. Five (20%) of 24 patients developed recurrent pneumococcal meningitis; four patients had one recurrence and one had three recurrences. All recurrences were seen in infants who were below the age of 15 months at the time of initial episode of meningitis. The interval between the initial episode and the recurrence ranged between one and 24 months. Evaluation of blood smear at the time of initial diagnosis of meningitis in 3/4 patients with recurrence revealed the presence of Howell-Jolly bodies (5,9, and 15 months) indicating splenic hypofunction. In this study pneumococcal meningitis in sickle cell patients occurred earlier than is generally reported and the recurrence rate was high when the initial episode was at less than 15 months, suggesting a defect in host defense in addition to the absence of splenic function in these very young patients. This suggests the need to evaluate antibody forming capacity in such infants.