824 THE VARYING CLINICAL SPECTRUM OF PRESENTATION OF BURKITT'S LYMPHOMA IN CHILDREN. K. R. Kini, S. Raman, R. P. Warrier, E. Abdulkadir, M. Poel, A. Shringeri. Spon. by L. Weiss. Henry Ford Hospital, Department of Pediatrics, Detroit, Michigan.

Burkitt's lymphoma is a rare neoplasm in children and has varied clinical presentations and course. Six Caucasion children between the ages of 5 and 18 years were studied at Henry Ford Hospital. Five were male and one was female. Five out of six of the patients were initially diagnosed as having gastroenteritis. Their initial presentation was abdominal pain, vomiting and distention. Two of the children had evidence of acute intestinal obstruction at the time of diagnosis. Leukemic transformation (4/6), meningeal involvement (3/6), testicular involvement (1/6), bilateral ovarian involvement with ascites and hypomenorrhea (1/6), monarticular arthritis with bone involvement (1/6), massive pleural effusion (1/6), submandibular lymphadenopathy (1/6) were other clinical signs present at the time of diagnosis. Two out of six achieved complete remission on chemotherapy and are long-term survivors at 70 months and 66 months. Serum lactic dehydrogenase (LDH) levels closely correlated with recurrence and extent of tumor mass. Both patients who had complete remissions had only localized disease and had LDH levels below 400 IU/cc. Metabolic complications encountered consisted of uric acid nephropathy (1), hypocalcemia and hyperphosphatemia (2) with uncontrolled seizures. Morphological, cytochemical and ultrastructural studies were performed to confirm the pathological diagnosis.

825 HEMATOLOGICAL STUDIES IN HEMOGLOBIN SO ARAB AND CO ARAB DISEASES: CLINICAL AND LABORATORY EVALUATION. K. R. Kini, R. P. Warrier, K. Maeda, J. Rebuck, D. L. Rucknagel. Spon. by L. Weiss. Henry Ford Hospital and University of Michigan, Department of Pediatrics, Detroit, Michigan.

Ten year old, black female originally diagnosed as having SC disease had multiple thrombotic crises resembling SS disease. Because of her unusually severe clinical course, Hb electrophoresis on citrate agar and finger printing was done and SO Arab disease was diagnosed. The clinical presentation of Hb SO Arab in this patient resembled that of Hb SD disease. Her hemoglobin varied from 9.9 to 10.5 gm/dl and reticulocyte count from 1.2 to 2.5%. The peripheral blood smear showed moderate anisopoikilocytosis, moderate polychromasia and occasional sickle forms. The RBC survival study revealed a half life of 19 days (normal 22-28 days). Hb CO Arab was found in two siblings who are asymptomatic. Numerous target cells and mild anisocytosis was seen on the peripheral blood smear. RBC survival study in one of the siblings showed a half life of 16 days with sequestration in the spleen. Oxygen dissociation curve in the patient with SO Arab showed marked decrease in oxygen affinity of P_{50} 66.5 mm/Hg resembling the pattern seen in sickle cell anemia. In the siblings normal or slightly increased P_{50} of 35.4 mm/Hg was seen. This is the first report of decreased RBC survival with RBC sequestration in the spleen in CO Arab disease. We wish to emphasize the importance of citrate agar electrophoresis and finger printing for the diagnosis of SO Arab disease.

826 ACYCLOVIR: THERAPY OF V-Z INFECTIONS IN CHILDREN WITH MALIGNANCIES. Thomas R. Kinney, Laura T. Gutman, Catherine M. Wilfert, Ronald Keeney, Paulo deMiranda, Christine Rudd, John M. Falletta. From Dept. Peds. Duke Univ. Med. Center and Wellcome Res. Labs, Durham, N. C.

7 immunocompromised children (4-16 yrs) whose primary diagnoses were ALL (2), Burkitt's lymphoma and Hodgkin's (2 each), Ewing's tumor (1) had primary varicella with pneumonia (2) or zoster (5). Virus isolation was attempted from all pts. and was successful in 3. Six pts. were on multiagent chemotherapy for malignancy; the 7th pt. last received chemotherapy 3 mo before zoster. Lymphopenia (<1200/mm^3; mean 654/mm^3) occurred in 5 of 7 children and neutropenia of 368/mm^3 in 1 child. Acyclovir (ACV) therapy was administered in a dosage of 250 mg/m^2/day (1 pt.), 750 mg/m^2/day (2 pts.) and 1500 mg/m^2/day (4 pts.) as 3 doses/day for 5 days.

	T½ hr.	Mean Peak	Mean Nadir
250 mg/m^2/d	2.5 hr.	2.2 µgm/ml	.07 µgm/ml
750 mg/m^2/d	2.4 hr.	5.7 µgm/ml	.65 µgm/ml
1500 mg/m^2/d	2.4 hr.	20.8 µgm/ml	2.30 µgm/ml

All patients recovered from their infections. Vesicular lesions crusted in 1-4 days after initiation of ACV. Viral cultures were negative within 1 day of initiating therapy. There was no noted toxicity associated with ACV. There were no relapses of V-Z although cancer chemotherapy was given concomitantly in 1 pt. and resumed in 4-15 days in 6 pts. Follow-up was 4 mo (1) to 1 yr (1) and no sequelae of infection or recurrences have been noted. One Burkitt's pt. died, 1 has active Hodgkin's disease, and 5 are in remission with respect to their malignant disease.

827 CONGENITAL SIDEROBLASTIC ANEMIA (CSA): A MICROENVIRONMENT DEFECT. Joseph Kochen, Nader Ibrahim, John Lutton and Richard Levere. Cornell Univ. Med. Col., North Shore Univ. Hosp., Dept. Ped, Manhasset, NY and New York Med. Col. Dept. Med., Valhalla, NY

Erythroid colony (EC) formation and heme biosynthesis were studied in marrow cultures of a 2 year old girl with severe CSA and neutropenia. Bone marrow showed micronormoblastic erythroid hyperplasia, ringed sideroblasts and decreased granulopoeisis. The patient had not responded to hematinics and prednisone. EC formation was determined in plasma clot (pc) and methyl cellulose (mc) culture systems after 7 days incubation by benzidine staining and expressed as CFU_E/$8x10^5$ marrow cells. In pc cultures, EC(140) were fewer than normal adult controls (1100). However, cultures in mc resulted in exuberant EC growth (1400). Good EC formation (900) also occurred without added erythropoeitin (ep), in contrast to no growth in controls. Addition of hemin stimulated EC formation (2000) and reversed pyridoxal PO_4 inhibition. δ-aminolevulinic acid (ALA) synthetase activity of marrow itself was substantially decreased and heme oxygenase was increased. However, in mc culture ALA synthetase, ALA dehydratase, C-^{14}ALA incorporation into heme and H-^{3} leucine incorporation into protein were normal and increased by addition of hemin. This patient's excellent capacity to synthesize hemoglobin and form EC in mc culture, even in the absence of added ep, suggests that CSA may result from an unfavorable marrow microenvironment.

828 CEREBROSPINAL FLUID (CSF) PROCOAGULANT IN ACUTE LYMPHOBLASTIC LEUKEMIA. D.M. Komp, L.P. Clyne, E.Sullivan, Yale Univ., Depts. of Ped. and Lab. Med., New Haven, CT

CSF of children with leukemia treated with cranial radiation and intrathecal methotrexate (IT MTX) can shorten the recalcification time of normal plasma. Graeber postulated that this procoagulant activity of CSF reflects release of phospholipid from the demyelinating effects of central nervous system (CNS) prophylaxis. We have confirmed their findings and report further studies of CSF from 28 samples during CNS prophylaxis, 15 samples during treatment with IT MTX alone given for prophylaxis and 14 samples during IT MTX for active CNS leukemia. Procoagulant activity was equally demonstrable in the three patient populations. CSF from these patients shortened the recalcification time of normal plasma and plasmas deficient in factors VIII, IX, X or XII but not those deficient in II, V or X. Procoagulant activity was independent of chamber nucleated cell count but mononuclear cells could be demonstrated in all CSF samples by cytocentrifugation. Removal of cellular material from CSF by millipore filtration or a special modification of cytocentrifugation diminished the expression of procoagulant activity. Ultracentrifugation or millipore filtration of substrate plasma to more effectively remove platelets rendered the plasma insensitive to the CSF procoagulant. Similarly, complete glass or celite activation of platelet-containing plasma produced a plasma no further responsive to CSF. These observations suggest that the CSF procoagulant is a tissue factor (lipoprotein complex) associated with mononuclear cells in the CSF following IT MTX and reflect arachnoiditis rather than demyelination.

● **829** DISACCHARIDASE LEVELS IN THE INTESTINE OF IRON DEFICIENT INFANTS AND RESPONSE TO IRON TREATMENT. Philip Lanzkowsky, Gungor Karayalcin, Frederick Miller, Bernard Lane. Sch. of Med., Health Sciences Ctr., SUNY at Stony Brook and Long Island Jewish-Hillside Med. Ctr., Dept. of Pediatrics and Pathology, New Hyde Park, N.Y.

During the course of an investigation into gut function in severe iron deficient anemia, we observed impairment of lactose tolerance and a deficiency in small intestinal disaccharidase activity. Of 10 subjects with severe iron deficiency anemia (mean hemoglobin level 6.4 gm/dl, range 5.2-7.1 gm/dl) 6 had evidence of abnormal lactose tolerance tests (mean maximum blood glucose rise before iron therapy after lactose load was 19.0 mg/dl, range 5-28 mg/dl compared to 58.3 mg/dl, range 38-86 mg/dl post iron therapy). Of the 6 who had abnormal lactose tolerance tests 5 had intestinal biopsies. All of these showed reduced disaccharidase activities (lactase, sucrase, maltase) both quantitatively and qualitatively and reduction in cytochrome oxidase and succinic dehydrogenase. Light and electron microscopy were normal in all these patients. Following oral iron therapy, the abnormal lactose tolerance tests returned to normal in all the patients and the one patient in whom an intestinal biopsy was carried out following iron therapy there was a quantitative increase in the disaccharidase activity.

Although abnormalities of gut function are well known to occur in iron deficiency anemia, impairment of small intestinal disaccharidase activity has not previously been described in human iron deficiency. This impairment of lactose tolerance is transient and responds to oral iron therapy.