906 POSTNATAL VARIATION IN VISCOSITY AND HEMATOCRIT OF POLYCYTHEMIC AND NORMAL INFANTS: IMPLICATIONS FOR DIAGNOSIS. RS Ramamurthy, (Spon. by C Grose) Univ

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Prompted by the need for standardization of diagnostic parameters for neonatal polycythemia-hyperviscosity syndrome 89 infants were studied with parental consent. Cord and peripheral venous (PV) blood sampling was done at 2, 6, 12 and 18 hrs of age. 16 infants with PV hct $>64\%$ were considered polycythemic. Viscosity was determined at shear rate 11.5, 23 & 46 sec^{-1} with a Wells Brookfield viscometer and hct was measured in duplicate.

In normal infants the mean $\pm 2\text{SD}$ hct rises from cord value of $49 \pm 8\%$ to 2 hr (54 ± 10), 6 hr (53 ± 10) and 12 hr (52 ± 10) ($p < .01$) and drops at 18 hr (50 ± 10). The viscosity remained constant and the best correlation with hct was at 2 and 18 hrs ($r = .6$). In the polycythemics the hct rises from a cord value of 57 ± 4 to 2 hr (66 ± 4), 6 hr (64 ± 4 , $p < .01$) and drops at 12 hr (60 ± 6) and 18 hr (60 ± 8). The PV viscosity rises from cord value of (9 ± 8) cps to 2 hr (12 ± 4) but drops at 6 hr (11 ± 4) and 12 hr (10 ± 4), rising again at 18 hr (13 ± 6 , $p < .01$). Only 6 of 16 polycythemics had PV hct $>64\%$ beyond 18 hrs. All were asymptomatic.

Cord blood hct $>50\%$ is a good screening procedure to select infants for PV blood sampling. In polycythemic infants the hct and viscosity peak by 2 to 6 hrs, hence PV sampling should be done by 6 hrs. In asymptomatic infants PV sampling should be repeated after 18 hrs and viscosity should be determined before therapeutic decisions are made.

907 TREATMENT OF PELVIC SARCOMA IN CHILDREN: EXPERIENCE AT ONE INSTITUTION. Beverly Raney, M.D., Andrew

Carey, B.A., Louise Schnauffer, M.D., Philip Littman, M.D., and Jane Chatten, M.D. The Children's Cancer Research Center (CCRC) of the Children's Hospital of Philadelphia and Department of Pediatrics, University of Pennsylvania, Philadelphia, PA 19104.

Seven children aged 2-16 years with sarcomas arising in the retroperitoneum or pelvis (not in bladder or prostate) were treated at the CCRC from 1974-1983. 6 presented with abdominal distention and a palpable mass; the other had back pain and weight loss. 5 had localized tumors, and 2 had lung metastases. Median primary tumor diameter was 15 cm. All pts underwent exploratory laparotomy and biopsy; none had complete excision. All pts received local radiotherapy (med, 4000 rad) and intravenous vincristine, actinomycin D, and cyclophosphamide; 5 also received Adriamycin. Four pts responded completely and 2 of them survive free of recurrence at 1.5 and 7.5 yrs. The other 5 pts died of tumor 2-62 months (med, 18 mos) from diagnosis after local (3 pts) or distant (2 pts) recurrence. By contrast, all 5 pts with bladder-prostate sarcoma treated at the CCRC from 1974-1983 are free of recurrence at 1.3-8.7 yrs (med, 4.3 yrs). These 5 pts presented with acute urinary obstruction and had smaller tumors, without metastases. We conclude that retroperitoneal-pelvic sarcoma is a particularly lethal form of childhood cancer, because it is advanced at diagnosis; local control is difficult due to large tumor size.

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908 HEMATOLOGIC FINDINGS IN PERTUSSIS. Sreedhar Rao and Kenneth Bromberg. (Spon. by Audrey Brown) SUNY/Downstate Med. Ctr/Kings County Hosp., Brooklyn.

Patients infected with *Bordetella pertussis* frequently exhibit leukocytosis and lymphocytosis. Eosinophilia has been noted less frequently. Peripheral blood counts were analyzed in 17 patients with laboratory confirmed pertussis (culture ± 14 , fluorescence ± 3) to look for other hematological abnormalities. Fifteen of 17 patients were below the age of one year; others were 4 and 35 years old.

Leukocytosis ($>15,000/\text{cmm}$) was noted in 12 patients; 11 of these also had lymphocytosis ($>10,000/\text{cmm}$). Five patients were neutropenic ($\text{ANC} < 1500/\text{cmm}$). Eosinophilia ($>500/\text{cmm}$) was noted in 6 patients. No patient had a hemoglobin $< 10 \text{ gm/dl}$. The platelet count was normal ($150,000-400,000/\text{cmm}$) in 15 patients and elevated in 2. Previous investigators have described the lymphocytes in pertussis as being small and mature. In contrast, 5 patients in this study had immature lymphocytes that were medium sized with scanty cytoplasm, loose nuclear chromatin and one or more nucleoli on light microscopy. While the previously described lymphocytosis and eosinophilia were seen, the hematologic findings in pertussis should be expanded to include neutropenia and the presence of immature lymphocytes.

909 BONE INFARCTION, OR OSTEOMYELITIS IN CHILDREN WITH SICKLE CELL DISEASE: BONE AND BONE MARROW SCAN FINDINGS. S.P. RAO, N. SOLOMON, S. MILLER, and

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Bone infarction (INF) occurs commonly in patients with sickle cell disease (SCD), and is frequently associated with infarction of the bone marrow (INF). Clinically and radiographically it is difficult to distinguish INF from Osteomyelitis (OSTEO). Bone scan using Tc99 diphosphonate &/or bone marrow scan using Tc99 Sulfur Colloid were done in 33 children (1-16 yrs.) with SCD (SS 32, S-C 1) who had localized bone pain and fever. Twenty-six of 33 patients had INF and 7 had bacteriologically proven OSTEO (*Salmonella* 5, *S. Pneumo* 2).

	BONE SCAN UPTAKE			MARROW SCAN UPTAKE		
	$\uparrow$	$\downarrow$	N	$\uparrow$	$\downarrow$	N
INF	13/19	3/19	3/19	0	10/11	1/11
OSTEO	7/7	0	0	0	0	4/4

As shown in table, while bone scan uniformly showed increased uptake in OSTEO, uptake was also frequently increased in INF. In contrast, marrow scan demonstrated normal uptake in all four OSTEO patients evaluable; uptake was diminished or absent in 10/11 patients with INF. Thus, in a patient with SCD who has localized bone pain and fever, a normal marrow uptake at the site of pain strongly suggests OSTEO and the need for diagnostic bone aspiration and appropriate therapy. Bone scan appears to be less useful than marrow scan in distinguishing between INF and OSTEO.

† 910 VITAMIN B-12 (Cb) AND FOLATE INTERRELATIONSHIP IN A CASE OF HOMOCYSTEINURIA-METHYLMALONIC ACID (HC-MMA) - URIA DUE TO GENETIC CO-FACTOR DEFICIENCY. Yaddanapudi Ravindranath, Ingeborg Krieger. Children's Hospital of MI., Det.

We observed the 9th known case of HC-MMAuria and the 4th with associated megaloblastic anemia. Treatment accomplished for the first time recovery from biochemical and clinical manifestations in a 2 month old female. The response provided insights into the mechanism of the "folate trap" and the pathogenesis of symptoms. Prior to treatment, plasma folate and Cb were elevated. Oral folate (0.5 mg/d) was given for the first 2 weeks. Reticulocytes rose to 7.6; urine HC dropped from 3.01 to 1.44 $\mu\text{mol/mg Cr}$; but MMA did not change (5.70 mg/mg Cr). Subsequent treatment with (HO)Cb (1000 $\mu\text{g q.o.d.}$) over 3 months caused these changes: Hgb: 7.2 to 12.8 g/dl; MCV: 117 to 102; RBC 2.05 to 3.68 Mill; WBC: 3200 to 8000, head growth from 0.20 to 0.42 cm/wk; but there was persistent HCUria (1.44-0.76 $\mu\text{mol/mg Cr}$), although MMAuria remained absent. Plasma methionine was low. Oral folate (2 mg/d) was therefore added. HC disappeared and head growth accelerated further to 0.5 cm/wk. Methionine supplements caused reappearance of HC (0.4 $\mu\text{mol/mg Cr}$). After oral betaine HC was minimal (0.10 $\mu\text{mol/mg Cr}$) and disappeared when folate was increased to 4mg/d. These observations speak against the existence of a biochemical folate trap, but do not rule out physical trapping of methyltetrahydrofolate due to Cb deficiency. Differences in the resolution of symptoms is attributable to differences in sensitivity of Cb requiring functions to the deficiency. Cb dependant folate transport into cells for demethylation appears to be most sensitive.

911 CLINICAL AND ULTRASTRUCTURAL CORRELATES OF RECEPTORS FOR PEANUT AGGLUTININ (PNA) IN ACUTE LYMPHOBLASTIC LEUKEMIA (ALL). E.C. Russell, J.L. Haar, T. Mohana-kumar, (Spon. by H.M. Maurer). Medical College of Virginia, Department of Pediatrics, Children's Medical Center, Richmond.

Receptors for the lectin PNA have been identified on blast cells from a subpopulation of children with ALL and early data indicated an association between PNA receptors and poor prognosis. Analysis of 174 children with ALL of the non T, non B, non pre-B phenotype reveals that 27.6% have receptors for PNA. There is correlation between presence of PNA receptors and initial WBC $> 100,000$ (50% PNA+), age < 2 years (40% PNA+) and absence of the common ALL antigen (48% PNA+, cALLa-), all features known to predict poor prognosis in childhood ALL.

We used electron microscopy (EM) to characterize the ultrastructure of lymphoblasts. PNA+ blasts reveal an electron dense reaction product on the cell surface when agglutinated with PNA-conjugated HRP and reacted with DAB. Staining of the cells with uranyl acetate and lead citrate reveals blunt pseudopodia, surface pinocytotic vesicles and irregularly shaped nuclei with lobulated euchromatic chromatin. These features are characteristic of undifferentiated cells and are distinctly different from those EM features of identically prepared blasts lacking the PNA receptor.

We conclude that PNA receptors on blasts correlate with other clinical and immunological predictors of poor prognosis in ALL and that PNA+ blasts have unique ultrastructural features seen in immature, undifferentiated lymphoid cells.