631 NASOPHARYNGEAL LYMPHOEPITHELIOMA. Nancy B. McWilliams, Tapan A. Hazra, Nancy L. Dunn, Edward C. Russell and Harold M. Maurer. Medical College of Virginia, Depts. of Pediatrics and Radiation Therapy, Richmond, Virginia.

Nine previously unreported cases of lymphoepithelioma of the nasopharynx are presented. Mean age at diagnosis was 13.6 yrs. (range 9-19 yrs). Seven patients were black and 5 were male. Symptoms were present from 6 wks.-4 mos. ($\bar{X}$ 2.7 mos.) prior to diagnosis and included painful adenopathy, earache or trismus in 7 patients. Only one had metastases beyond the cervical nodes (pulmonary) at diagnosis.

Treatment consisted of radiation to the nasopharynx (5290-7000 rad) and neck (4780-6500 rad). Chemotherapy regimens varied, but all included adriamycin and cyclophosphamide. The patient with pulmonary metastases also received lung irradiation.

Follow-up has ranged from 2-36 mos. ($\bar{X}$ 20.3 mos.). Two patients have died, 22 and 14 mos. post-diagnosis. The first developed local recurrence and distant metastases at 20 mos. The second, who presented with distant metastases, died of progressive pulmonary disease despite good local control. One patient had cervical and orbital metastases at 8 mos., received an additional 3000 rads, and is disease-free at 32 mos. Six patients remain disease-free from 2-36 mos. ($\bar{X}$ 19 mos.).

We conclude that: 1. Although a delayed diagnosis is common, the prognosis is favorable when disease is localized. 2. The primary treatment is radiation to the tumor and regional lymphatics bilaterally. The benefit of adjunct chemotherapy is in doubt.

632 LATENT DISEASE DETECTION IN HEREDITARY SIDEROBLASTIC ANEMIA (HSA) BY WHOLE/FRACTIONAL ANALYSIS OF ERYTHROCYTE UROPORPHRYNOGEN-1-SYNTHETASE (U-1-S) IN RELATION TO SERUM FERRITIN LEVELS. Thomas D. Miale, P.Robert N. Hast, Peter G. Reizenstein, Spon. by Elia M. Ayoub, University of Florida, Shands Teaching Hospital, Department of Pediatrics, Gainesville and Karolinska Hospital, Hematology Section, Stockholm.

U-1-S and serum ferritin levels were performed on 7 children and 6 adults of a Swedish kindred with either HSA or Christmas disease, including two members with both disorders, to detect latent disease and determine the possibility of genetic linkage between these rare diseases. Frozen erythrocytes from all 13 pts. and simultaneous controls, were thawed, de-glycerolated, and separated on Ficoll-Isopaque gradients. One pt. with clinically severe disease had a broad, heterogenous band in the specific gravity 1.064 layer which consisted of markedly deformed erythrocytes. A similar analysis in two patients with latent disease disclosed a wide, but more buoyant band of hypochromic, less deformed erythrocytes. Simultaneous analysis of frozen and fresh control erythrocytes together with erythrocytes of the remaining pts. yielded a normal band. However, all patients manifested elevated U-1-S levels, despite the presence of normal to low reticulocyte counts. U-1-S levels correlated directly with serum ferritin levels. HLA-D typing was compatible with the lineage with no association of a given HLA-D type with either HSA or Christmas disease. No linkage on X chromosome between these two diseases was revealed by Xg blood group determinations. U-1-S analysis may provide a sensitive screening procedure in certain HSA pedigrees.

633 NEW PROGNOSTIC FACTORS IN CHILDHOOD LEUKEMIA (ALL). Denis R. Miller, Sanford Leiken, Vincent Albo, Leonard Vitale, Robert Hittle, Harland Sather and Denman Hammond, Childrens Cancer Study Group, Los Angeles, California.

Since Feb. 1975, 883 previously untreated children with ALL have been entered on a protocol designed to evaluate prognostic factors which influence induction, remission duration and survival and to identify subsets of patients with high risk of early failure. Complete marrow remission (M_1) rates ranged between 91-95%. The combination of older age (>10yrs), high initial WBC (>20,000/μl) and low serum IgG are associated with a significantly lower M_1 rate. Skin test response to recall antigens, serum inhibition of blastogenesis, HLA type or PAS positivity were not prognostic of induction or remission duration. Patients in poor prognostic group (any age, initial WBC>50,000/μl) with L1 blasts (FAB classification, Br.J.Hem 33:451,1976) had higher M_1 rate than those with L2 blasts (80 vs 67%).

Using the Mantel-Peto-Cox Chi square statistic for life table data, significant factors associated with poor prognosis include: WBC>20,000/μl; age >7yrs; M_3 marrow day 14; CNS disease at diagnosis; L_2 lymphoblasts; decreased serum IgG and IgA; black race. Mediastinal mass is not an independent factor. None of 41 patients in low risk group (age 3-7yrs, WBC<10,000/μl) with L1 blasts, M_1 marrow on day 14, and normal Ig has sustained an adverse event (relapse or death). Identification of prognostic factors should permit more efficient and effective design of future protocols particularly for patients with good and poor prognosis.

634 SPONTANEOUS REMISSION OF ACQUIRED VON WILLEBRAND'S DISEASE (VWD) FOLLOWING RESECTION OF WILM'S TUMOR. Robert R Montgomery, Taru Hays, David G Tubergen. U of Colo Med Ctr and Denver Children's Hospital, Dept of Ped, Denver. (Sponsored by John H. Githens).

This report concerns a child with a Wilm's tumor and laboratory evidence of vWD. The vWD was characterized by a prolonged PTT, low factor VIII (VIII_c), low ristocetin cofactor activity (VIII_r), low factor VIII antigen (VIII_{ag}) and a prolonged bleeding time. In addition, she had low von Willebrand's disease antigen II (vWagII) that has been shown by Montgomery and Zimmerman (Am Soc Hematol Plenary Session, Dec 1977) to be decreased in patients with classical vWD.

The following studies were obtained:

	VIII_c	VIII_r	VIII_{ag}	vWagII	VIII_c inhib	VIII_r inhib
pre op 2	18%	0.28U	<6%	<0.06U	neg.	neg.
pre op 0	23%	0.30U	<6%	<0.06U	neg.	neg.
post op +30	114%	1.40U	160%	---	---	---
post op +92	280%	---	256%	1.40U	---	---

Since this child underwent an earlier uneventful tonsillectomy and both parents and a sibling have normal VIII_c, VIII_r, VIII_{ag} and vWagII, this is felt to represent a spontaneous remission of an acquired vWD without VIII_c or VIII_r inhibitors. This picture of vWD with an VIII_{ag} that is decreased more than the VIII_c and VIII_r has not previously been reported. Since VIII_{ag} and vWagII are physically and immunologically distinct, the finding of decreased VIII_{ag} and vWagII in this case of acquired vWD, as previously found in hereditary classical vWD, suggests that both are under similar genetic and/or synthetic control.

635 CORRELATION BETWEEN FACTOR VIII ANTIGEN ($VIIIag$) AND VON WILLEBRAND'S DISEASE ANTIGEN II (vWagII) IN NORMAL INDIVIDUALS AND PERSONS WITH VON WILLEBRAND'S DISEASE (vWD). Robert R Montgomery, Janet W Johnson, Linda J Jacobson. U of Colo Med Ctr, Dept of Ped, Denver. (Spon. by John H. Githens).

Classical vWD has been characterized by a deficiency of $VIIIag$ as well as a new antigen-vWagII characterized by Montgomery and Zimmerman (Plenary Session, Am Soc Hematol, 1977). Patients with severe vWD have undetectible $VIIIag$ and vWagII. This report concerns the correlation between $VIIIag$ and vWagII in 38 normal individuals, 9 patients with severe vWD, and 16 patients with moderate vWD.

$VIIIag$ and vWagII concentrations were determined by quantitative immunoelectrophoresis using monospecific rabbit antibody to purified factor VIII and vWagII.

	#	$VIIIag$	vWagII	vWagII/$VIIIag$
Normal	38	0.95±.29	0.87±.34	0.93±.33
Mod vWD	16	0.21±.10*	0.55±.23*	3.1 ±1.66*
Severe vWD	9	<0.06*	<0.06*	---- (* p<0.01)

Thus, individuals with vWD have significantly decreased vWagII as well as $VIIIag$ although the degree of decreased vWagII is significantly different from the degree of decreased $VIIIag$.

In normal individuals there is a significant ($p<.001$) linear correlation between vWagII and $VIIIag$ with a correlation coefficient of 0.65. Although individuals with severe vWD have no detectible vWagII or $VIIIag$, individuals with moderate vWD have no correlation between their vWagII and $VIIIag$ levels.

636 NEUTROPHIL MIGRATION DEFECT DURING EXPERIMENTAL NUTRITIONAL IRON DEFICIENCY IN POST-WEANING RATS. Linda L. Moore, James R. Humbert, Avanelle Kirksey, and Josephine T. Lee, State University of New York at Buffalo, The Children's Hospital of Buffalo, Department of Pediatrics, Buffalo and Purdue University, Department of Foods and Nutrition, West Lafayette, Indiana.

The study of neutrophil (PMN) functions in children with iron deficiency (ID) has yielded controversial results, partly because of the uncontrollable association of other nutritional deficiencies with that condition. We have studied the effect of isolated iron deficiency on in vivo PMN migration in six rats fed an iron-poor diet until 70 days post-weaning. The same diet was fed to 12 control animals, either as paired feedings (PF) or ad libitum feedings (AL); these controls were made iron sufficient with intramuscular iron dextran at weaning. Seventy days post-weaning weights (mean ± SE, in g) were essentially identical in ID (205 ± 13), PF (188 ± 6), and AL (225 ± 15) rats. Anemia was evident in ID rats (Hb. 9.7 ± 0.7 g/dl, MCV 47 ± 1.5 fl.) but not present in PF or AL controls (Hb. 17 ± 1.7 and 14.9 ± 0.3, MCV 54 ± 1.3 and 57 ± 1.3 respectively). PMN migration into the peritoneal cavity was induced by 18-hour stimulation with Na caseinate and expressed as total numbers of PMNs X 10^5/100 g body weight. PMN migration was significantly lower in ID animals (49.3 ± 6.2) than in PF (125 ± 28) and AL (111 ± 18) animals ($P < 0.025$). In post-weaning rats, isolated iron deficiency is associated with the development of a pronounced decrease in PMN migration, in vivo. This decrease might contribute to the increased tendency toward infections observed in some ID subjects.