949 LATE DEVELOPMENT OF FOCAL GLOMERULOSCLEROSIS IN TWO PATIENTS FOLLOWING NEPHRECTOMY FOR WILMS TUMOR. Thomas R. Welch and A. James McAdams, Univ. of Cinti., Children's Hospital Medical Center, Divisions of Nephrology and Pathology, Cincinnati.

Proteinuria at the time of diagnosis of Wilms tumor (WT) is occasionally seen in patients who may also have other manifestations of the nephrotic syndrome. In such instances of what has been termed the Drash Syndrome there are morphologic changes of focal glomerulosclerosis (FGS).

We have recently evaluated 2 children who were not proteinuric at the time of unilateral nephrectomy for Wilms tumor and who first developed signs of biopsy documented FGS 17 and 11.5 yrs later. One patient, a boy undergoing nephrectomy at 3.5 months of age currently has a 24 hour urinary protein excretion of 7.4 gm, hypoproteinemia, and hypercholesterolemia; his serum creatinine is 1.3 mg/dl. The other patient, a girl undergoing nephrectomy at 1.5 yrs, underwent renal transplantation at 21 yrs. Neither had evidence of recurrent tumor or a radiation or immune complex mediated injury. Thus, although FGS may be present at the time of diagnosis of WT in some patients, we have found that it may occur after a decade or more.

FGS can be produced experimentally by removal of 3/4 total renal mass. Glomerular abnormalities (microglomeruli) are frequent in the non-tumorous kidney of some WT patients and, combined with the contralateral nephrectomy, contribute to the reduction in renal mass. Increased perfusion of the remnant nephrons may explain the subsequent development of FGS in these patients.

Patients with WT should be monitored well into adulthood. Modification of dietary protein intake may affect the evolution of this glomerulopathy.

950 ADENOVIRUS ASSOCIATED HEMOPHAGOCYTIC SYNDROME IN A BONE MARROW TRANSPLANT PATIENT. R. Wodell, R.Gupta, E.Bayever, C.August, S.Plotkin. U.of Pa. Sch. of Med., Dept. of Peds., Children's Hosp. of Phila.

Virus-associated hemophagocytic syndrome (VAHS) has been linked most commonly to herpes group viruses. It has not previously been reported in a bone marrow transplant (BMT). VAHS occurred in a 5 y/o girl who underwent an autologous BMT for metastatic Wilms tumor. Prior to BMT she received doxorubicin, high dose melphalan, and local radiation to her chest and abdomen. Adenovirus was isolated consistently throughout the course of the transplant from urine and stool, but not from buffy coats or respiratory tract secretions. Bone marrow exam (BME) 2 wks post BMT showed hematopoietic recovery with all three cell lines present. At +4 wks she developed hepatomegaly with elevated liver enzymes, intermittent fever and pancytopenia. BME at that time was hypocellular with marked histiocyte phagocytosis of immature and mature red cells, white cells and platelets. The histiocytes were mature with abundant vacuolated cytoplasm and inconspicuous nucleoli. A BME at +7 wks showed prominent histiocytes and hemophagocytosis and aplasia of all hematopoietic cells. A culture of this marrow grew adenovirus. Nine wks after BMT she developed fatal interstitial pneumonia. Cultures of pleural fluid and lung tissue from open lung biopsy grew adenovirus. Post mortem cultures from lung, heart, spleen and GI tract grew adenovirus. This is the first case of culture proven adenovirus VAHS in a BMT patient.

IMMUNOLOGY

951 ANTIAMOEBIC PROPERTIES OF HUMAN COLOSTRAL MACROPHAGES Gustavo Acosta, Luz M. Rocha, Rocio Reyes, José I. Santos, Armando Isibasi, and Jesus Kumate. Centro Medico Nal. IMSS, Mexico City, Mexico. (Sponsored by Harry R. Hill)

A number of specific and non-specific soluble and cellular antimicrobial factors have been demonstrated in human colostrum. Although macrophages are the most abundant cellular elements in human colostrum, the role of these cells against intestinal parasites has not been explored. We assesed the activity of human colostral macrophages against trophozoites of axenically cultured Entamoeba histolytica. Colostrum samples from 50 Mexican women were obtained by manual expresion into sterile plastic tubes. Macrophages were isolated, washed in HBSS and adjusted to $2x10^6$ cells/ml in RPMI containing 20% fetal calf serum. 1 ml of this cell suspension was dispensed onto sterile coverslips and incubated at 37°C in 5% CO_2 for 24 hrs and then washed 2X with HBSS and observed for viability. Monolayers of adherent cells were then overlayed with trophozoites of E. histolytica to give a macrophage to amoeba ratio of 30:1 and incubated at 37°C in 5% CO_2 and examined by light microscopy at 15', 30', 1 hr, and 3 hr intervals. A progressive diminution in the number of trophozoites was observed when compared to control coverslips containing trophozoites alone. These findings suggest that colostral macrophages and/or their secretagogues are cytotoxic for trophozoites of E. histolytica.

† **952** A NEW GENETIC DISORDER OF NEUTROPHIL OXIDATIVE METABOLISM. Donald C. Anderson, Frank C. Schmalstieg, Neal A. Halsey, Irwin P. Cohen, & Robert S. Daum, Depts. of Ped., Baylor, Houston, TX, & U of TX, Galveston, TX & Tulane U., New Orleans, LA.

Investigations of neutrophil function in a 7 y/o M with recurrent Pseudomonas cepacia pneumonia demonstrated impaired intracellular killing of catalase - positive bacteria; other functions including directed migration, phagocytosis, adherence & secretion (1° & 2° granule) were normal. Quantitative NBT reduction and chemiluminesence (CL) evolution by patient neutrophils (PN) were profoundly diminished (0.1-9% of normal) in response to soluble stimulants (PMA, A23187, NaF) (p<.001 to each), but were in a low normal range (31-77% nl) during phagocytosis or during adherence to plastic substrates. Similarly, superoxide (O_2^-) generation and hexose monophosphate shunt activity of PN were profoundly diminished (0-3% nl) in response to soluble stimulants (p<.001), but demonstrated low normal activities (25-51% nl) during particle ingestion. Maternal neutrophils (MN) demonstrated intermediate abnormalities of NBT reduction, CL evolution and O_2^- generation in response to soluble stimulants, but responses during phagocytosis were normal. Further studies to determine the mechanism of impaired oxidative activity showed elevated levels of cytochrome b_{245} on the surface of MN (470% nl), those of a half brother (550% nl) & PN (200% nl). However, during recovery from Pseudomonas cepacia pneumonia, cytochrome b_{245} was undetectable on PN. These findings indicate that defective oxidative metabolic activity in this disorder is related to abnormal regulation of cytochrome b_{245} expression on the neutrophil surface.

953 NUCLEOLAR ANTI-HISTONES (H) ANTIBODIES (Ab) IN JUVENILE DERMATOMYOSITIS. Carlos M. Arroyave, Lauren M. Pachman, Kenneth C. Rich, Murray Passo, Jeffrey L. Kaine, Charles H. Spencer, Robert W. Nickeson, Jr. Northwestern University Medical School, Children's Memorial Hospital, Department of Pediatrics, Chicago; and Departments of Pediatrics, Indiana University Medical Center, Indianapolis; Washington University School of Medicine, St. Louis; Louisiana State University Medical Center, New Orleans; University of Oklahoma Health Science Center, Oklahoma City.

Ten positive antinuclear antibody (ANA) sera from JDMS patients obtained within 4 months of disease onset were examined by indirect immunofluorescence (IF), radial immunodiffusion, hemmaglutination, and enzyme immunoassays for the specificity of the Ab. Antihistone Ab was identified by IF using HEP_2-HCl washed and H reconstituted slides. No sera had Ab to Sm, RNP, SS-A, SS-B, DNA, and Sc170. Seven of ten showed the presence of IgG, IgA-H Ab, and three IgM-H Ab. All patients with anti-H Ab were reconstituted with fraction H2b and H3 and three with H2a; after reconstitution, the initial IF homogeneous-speckled pattern changed to speckled-nucleolar in 5/7 H-positive sera. In vitro complement fixation with HEP2-H reconstituted slides and JDMS sera showed that Clq but not C3 and C4 fixed this immune complex.

We conclude that JDMS sera with positive ANA has more than one Ab and the H Ab may account for only a portion of the Abs, and immune complexes formed in the presence of JDMS-H sera fixed Clq only.

† **954** STIMULATION OF CORD AND ADULT NK CELLS. J Baley & B Schacter. (Spon. A.Fanaroff), CWRU, RB&C, Cleve, OH.

Cord blood NK cells have decreased activity but are normal in number as identified by antibody B73.1. To determine if they could be stimulated normally, we studied lymphokine and mitogen stimulation of 11 cord and 10 adult overnight cultures. Binding(%B) and killing(%K) of target cell K562 in a single cell assay were compared with % lysis(%L) in a ^{51}Cr release assay. Cord %B was less than adult($p<.05$) in unstimulated (UN) cultures but was no different from adult with IL-2 or α or γ interferon(IFN). Adult cells significantly responded only to IL-2 ($p<.05$). IL-2 increased the %K of the bound cells but the cord %K was still less than adult ($p<.05$). IFN had no effect on %K of adult or cord cells or the %B of adult cells. In the ^{51}Cr release assay, IL-2 and IFN increased both cord and adult %L ($p<.05$). PHA, ConA or PWM stimulation of endogenous IL-2 production also increased %L ($p<.05$) but cord activity always remained less than adult ($p<.01$). There was no synergy with IL-2 and γ IFN or α and γ IFN. Since small increases in %B and %K do not explain large increases in %L, lymphokines may stimulate NK activity by increasing cycling which is not measured in the single cell assay. While cord %B responds to lymphokines, the defective %K remains. Thus, cord NK cells may be unable to cycle effectively because of deficient killing.

	Cord-UN	Cord-IFN	Cord-IL-2	Adult-UN	Adult-IFN	Adult-IL-2
%B	10.0±2.9	12.0±3.0	17.0±3.7	12.9±2.5	14.9± 4.2	17.4± 2.8
%K	18.6±4.2	22.0±4.5	28.1±5.9	30.1±3.9	30.8± 5.6	34.9± 5.1
%L	6.1±2.8	16.5±5.1	29.5±9.9	16.1±7.5	33.5±10.2	46.9±11.9