607 RADIATION-INDUCED GANGLIONEUROBLASTOMA AND NEURAL TUMORS. Bangaru Jayalakshmamma, Robert Chilcote, Wendy Recant, Martin Colman, (Spon. by Eddie Moore), Pritzker School of Medicine, University of Chicago, Michael Reese Hospital and Medical Center, Departments of Pediatrics, Pathology, Radiation Oncology, Chicago

Radiation therapy induces thyroid neoplasia, but the frequency and nature of other tumors are incompletely defined in man. A 7-year-old who presented with supraclavicular ganglioneuroblastoma had received gamma radiation for a cutaneous hemangioma in the same area during infancy. A review of institutional records of patients receiving moderate dose radiation therapy between 1938 and 1951 revealed an additional 4 cases of neural tumors (2 neurofibroma, 2 neurilemmoma) diagnosed 6-31 years (median: 25 years) following localized radiation therapy for enlarged tonsils (3) and keloid (1). The age at the time of radiation therapy ranged from 1-13 years (median: 4 years) and at the time of diagnosis 7-34 years (median: 30 years). The three with enlarged tonsils had received 750 rads, while the others had received unknown doses of "contact" gamma radiation. All tumors occurred within the radiation field, and no patient had a family history of congenital malformations or neurofibromatosis. We estimate that less than one such tumor should have occurred among the 5000 patients similarly irradiated. Our observations suggest a previously unreported etiologic relationship between moderate dose radiation therapy and neurogenic tumors with an extremely long latency period.

608 THROMBOCYTOSIS IN HISTIOCYTOSIS X. Peri Kamalakar, James R. Humbert, and John E. Fitzpatrick. State University of New York at Buffalo, The Children's Hospital of Buffalo, Department of Pediatrics, Buffalo.

Thrombocytosis as an initial feature of histiocytosis X has never been reported. In 8 of 12 patients with histiocytosis X seen at Children's Hospital of Buffalo in the past twenty years, platelet counts were available; seven of them were greater than 400 X 10^9/ l at diagnosis ($\bar{x} \pm SE = \pm 54 \times 10^9$/ l). All seven patients, who had an initial Lahey-Lucaya score averaging 3.1 (range: 2-4), are in disease-free remission ranging from 1 to 12 years. The eighth patient, who was thrombopenic at presentation and had a score of 7, is lost to follow-up. Following onset of therapy, platelet counts decreased progressively; they averaged 540 X 10^9/ l in treated but still symptomatic patients; 430 X 10^9/ l in treated, disease-free patients; and 375 X 10^9/ l in patients in complete remission and off all drugs. The difference between platelet counts at diagnosis and during remission is significant ($P < 0.01$). In the only patient who relapsed, thrombocytosis also recurred and disappeared again with subsequent remission. The etiology of the thrombocytosis is unknown, although in two cases, iron deficiency may have been contributory. Whether or not the heretofore undescribed initial thrombocytosis indicates a favorable prognosis is open to question.

609 GALL BLADDER DISEASE (GBD) IN CHILDREN WITH SICKLE CELL DISEASE (SS). Gungor Karayalcin, Anju Khanijou, Arturo J. Aballi, Phillip Lanzkowsky. Sch. of Med. Health Sciences Ctr., State Univ. of N.Y. at Stony Brook and Long Island Jewish-Hillside Med. Ctr., Dept. of Pediatrics, New Hyde Park, New York.

The present study was carried out to determine the incidence and clinical manifestations of GBD in children with SS. Thirty-two SS patients ranging in age from 2 to 17 years (mean 9.3) had oral cholecystography (OC) performed and in addition 6 of these had sonography. On OC 6 patients aged 17, 16, 16, 13, 6 and 3 years had evidence of GBD. Two had non-opaque and one opaque stones and in 3 there was no visualization after repeat OC with a double dose of telepaque. Of the 6 patients who had sonography 2 were normal and 4 had abnormal gall bladders consistent with chronic choleystitis without demonstrating cholelithiasis. OC on these 6 patients revealed that 2 had normal gall bladders, 3 had non-functioning gall bladders and 1 had non-opaque stones.

GBD in these patients did not seem to correlate with the incidence of painful abdominal crises. The incidence of GBD in this series was 18% which is higher than that previously reported in children with SS. Since the incidence of severe symptoms and/or complications of cholelithiasis is about 50% with an overall mortality of 2.7% within the first five years in unoperated patients (Lund, J.: Ann. Surg. 151:153, 1960) the presence of gallstones in children with SS is an indication for elective cholecystectomy.

610 SCINTIGRAPHIC DETECTION OF BONE AND BONE MARROW INFARCTS IN SICKLE CELL DISORDERS. Haewon C. Kim, Abass Alavi, Marie O. Russell, and Elias Schwartz. Depts. of Pediatrics and Nuclear Medicine, Univ. of Pa. Sch. Med.

Differentiation of osteomyelitis (OSTEO) from bone infarction (BI) in patients with sickling disorders is a difficult clinical problem. In patients with OSTEO there is increased uptake of isotope on bone scan (BS) soon after onset of symptoms. We have used ^{99m}Tc sulfur colloid for bone marrow scan (BMS) and ^{99m}Tc diphosphonate for BS to evaluate acute bone pain in 26 patients with sickling disorders. ^{99m}Tc tagged agents are readily available, and radiation is within the acceptable range for repeated diagnostic tests. In all 22 patients whose course subsequently indicated BI, the area of infarction had decreased or absent uptake on BMS. The corresponding area on BS varied in appearance depending on the interval between onset of pain and scanning. Of 7 patients studied within 5 days of onset, 6 had decreased uptake and 1 had increased uptake on BS in the area of the marrow infarct. Of 15 patients evaluated 5 days or more after onset, 4 had decreased and 11 had increased uptake on BS in the area of infarct due to reactive bone formation. One patient had diffuse increased uptake on BS with normal BMS, ruling out infarction and consistent with the clinical diagnosis of septic arthritis. Three patients with cellulitis had normal BS and BMS. In patients with BI extensive abnormality on BMS with minimal abnormality on BS indicates primary involvement of the marrow. Since these typical patterns are present at any stage of BI, the combination of BS and BMS may be useful in differentiating BI from early OSTEO.

611 SERUM IRON CONCENTRATION (SI) AND TRANSFERRIN SATURATION (SAT) ARE LOWER IN NORMAL CHILDREN THAN IN ADULTS. Marion A. Koerper and Peter R. Dallman. University of California Medical Center, Department of Pediatrics, San Francisco.

Low SI or low Sat without anemia is often equated with latent iron deficiency. Several studies have suggested that the SI and Sat may be lower in children than in adults. However, in the absence of systematically derived standards, adult values are commonly used. We studied a healthy group of 286 children and 39 adults between 9 a.m. and 2 p.m. and excluded 129 with any result that might suggest iron deficiency (either serum ferritin (SF) <12 ng/ml, free erythrocyte protoporphyrin (FEP) >3.0 µg/g Hgb, or low MCV or Hgb for age) or hemoglobinopathy (abnormal Hgb electrophoresis). Means ± SEM and 95% confidence limits were:

Age, yr	n	SI, µg/dl	Sat, %
0.5-2	52	68±3.6 (16-120)*	22±1.1 (6-38)*
2-6	56	72±3.4 (20-124)*	25±1.2 (7-43)*
6-12	55	73±3.4 (23-123)*	25±1.2 (7-43)*
18+	33	92±3.8 (48-136)	30±1.1 (18-46)

The SI and Sat were significantly lower in children than in adults (*$p<0.01$). Of the 163 presumptively normal children, 17% would have been designated as subnormal by usual criteria for adults (Sat<16%). There are also wide variations in SI and Sat independent of age, due to diurnal, dietary, and other factors, that are reflected in a large normal range. For better accuracy of diagnosis SI and Sat should be used in conjunction with at least one other test of iron status, and developmental norms for SI and Sat must be applied.

612 IRON DEFICIENCY ANEMIA IN CHILDREN WITH JUVENILE RHEUMATOID ARTHRITIS (JRA). Marion A. Koerper, David A. Stempel, John J. Miller III and Peter R. Dallman. University of California Medical Center, Department of Pediatrics, San Francisco and Children's Hospital at Stanford, Department of Pediatrics, Stanford.

Children with JRA often have anemia that may be due either to chronic inflammatory disease or to iron deficiency (possibly related to blood loss with aspirin therapy) or both. The two causes may not be distinguishable by mean corpuscular volume (MCV), free erythrocyte protoporphyrin (FEP), or serum iron/iron binding capacity (Sat). In order to determine whether serum ferritin (SF) would predict response to iron therapy, we measured hemoglobin (Hgb), MCV, FEP, Sat and SF in 48 children with JRA aged 22 mo to 20 yr. Of 26 with active polyarticular or systemic JRA or ankylosing spondylitis, 17 had anemia, 18 had low MCV for age, and 21 had elevated FEP. Thirteen children with anemia were treated with iron (2 mg/kg/d for 3 mo). Eleven raised their Hgb by at least 1.0 gm/dl (7 reversed anemia completely); MCV and FEP were also corrected in a similar number. All 6 with SF <30 ng/ml responded, but 2/4 with SF >90 ng/ml also responded. Of the 22 with pauciarticular JRA, only 2 were anemic. We conclude that iron deficiency is likely to be a major component of anemia in children with active polyarticular and systemic JRA. Low SF is a reliable predictor of response to iron therapy, but a response is also seen in some patients with high values. Consequently, a 3-month trial of iron (2 mg/kg/d) is justified on the basis of anemia alone in patients with active JRA.