876 BONE MARROW TRANSPLANTATION (BMT) IN CHRONIC GRANULOMATOUS DISEASE (CGD). Naynesh Kamani, Charles S. August, Steven D. Douglas, Edith Burkey, Harold W. Lischner, U. of Penn. Sch. of Med., Children's Hosp. of Phila., Dept. of Pediatrics, Temple U. Sch. of Med., St. Christopher's Hosp. for Children, Dept. of Pediatrics, Philadelphia, PA.

A 5 month-old Amish male infant with CGD underwent BMT from his 5 year-old, histocompatible brother after a preconditioning regimen of busulfan 8 mg/kg and cyclophosphamide 200 mg/kg. At the time of BMT, he was free of infection and remained so through the course of his transplant. He engrafted promptly with complete reversal of the neutrophil function defect (Table), and no sign of graft vs. host disease. This was followed by loss of the erythroid graft and gradual deterioration in neutrophil function (Table). Now, 16 months after BMT, he is free of infection, growing normally with essentially no evidence for neutrophil engraftment.

TIME	RBC TYPE	QUANTITATIVE NBT REDUCTION ΔO.D.		QUALITATIVE % NBT+ PHAGOCYTES	
		Patient	Control	Patient	Control
Donor	A	0.39	0.41	98	--
Patient					
Pre-BMT	AB	0.03	0.32	<0.5	98
+ 2 MONTHS	A-AB	0.72	0.31	n.d.	n.d.
+ 4 MONTHS	A-AB	0.21	0.37	21	97
+ 6 MONTHS	n.d.	0.08	0.69	19	99
+ 9 MONTHS	AB	0.03	0.35	1.5	96
+12 MONTHS	n.d.	0.03	0.31	2	88

879 FEVER IN INFANTS AND YOUNG CHILDREN WITH SICKLE CELL DISEASE (SCD). Gungor Karayalcin, Lorry Rubin, David Chung and Philip Lanzkowsky. Sch of Med, Health Sci Ctr, SUNY at Stony Brook, Children's Hospital of LIJ-HMC, Dept of Peds, New Hyde Park, NY.

Forty-four patients with SCD (39 SS and 5 SC disease), aged ≤ 5 yrs (mean 28 ± 17 mths) were hospitalized for 140 febrile episodes (temperature ≥ 38.4°C.) from Jan. 1978 to July 1983. All received continuous oral penicillin prophylaxis (OPP) from time of diagnosis and polyvalent pneumococcal vaccine (PPV) at 2 yrs of age. At least 2 blood cultures were obtained from all patients during hospitalization. All patients (except those with temperature < 38.9°C and vaso-occlusive syndrome-VOS) were treated with IV ampicillin for a minimum of 48-72 hrs. VOS accounted for 52 episodes (37%) (37 abdominal crises; 12 hand-foot syndrome and 3 aseptic necrosis of bones); fever of unknown cause in 18 episodes (13%) and fever with an infectious source in 70 episodes (50%). There were 28 episodes of pneumonia, 16 otitis media, 13 URI, 5 pharyngitis, 4 UTI, 1 osteomyelitis. The 6 episodes of bacteremia were associated with temperature ≥ 39.8°C. The bloodstream isolates were Streptococcus pneumoniea (2), Haemophilus influenzae, type b (1), Group B salmonella sp (1), Klebsiella sp (1), and Staphlococcus aureus (1). The 2 patients with pneumococcal bacteremia (age 16 and 22 mths) were non-compliant with OPP and had not received PPV because of their age. All patients responded to treatment and recovered from the febrile episode. The low incidence of pneumococcal bacteremia (2 cases per 147 patient-years) may be attributable to OPP and/or PPV.

● **877** THE BIOCHEMICAL BASIS OF THE CLINICAL INTERACTION OF Fe DEFICIENCY AND Pb INTOXICATION. S. Kapoor, C. Seaman, D. Hurst, S. Matos, S. Piomelli, College of Physicians and Surgeons, Columbia U., Pediatric Hematology, NY.

In children, iron (Fe) deficiency and lead (Pb) intoxication often coexist and aggravate each other; the elevation of erythrocyte porphyrin induced by Pb is much made prominent by Fe deficiency. We studied the interaction of Pb and Fe on ferrochelatase, the mitochondrial enzyme that catalyzes the incorporation of Fe into protoporphyrin. Mitochondria were isolated from reticulocytes of patients with sickle-cell syndromes undergoing therapeutic exchange transfusions. Enzyme kinetics of ferrochelatase were then studied on freeze-thawed preparations by measuring the incorporation of 59Fe into heme. In the absence of Pb, the KmFe was 13 μM, and Vmax was .021 μM/min/mg protein. In the presence of varying concentrations of Pb, the Ki was estimated at 50 μM. As concentrations of Pb were progressively decreased from 100 μM to 1 μM, in the presence of various Fe concentrations, the effect of Pb became much more pronounced as the Fe concentration decreased. At Pb concentrations between 10 μM and 1 μM (corresponding to 200 → 20 μg/dl) a more marked inhibition could be demonstrated when the Fe concentration was below 10 μM.

These data demonstrate that, when Fe deficiency is present, human mitochondrial ferrochelatase is more sensitive to the effect of low doses of Pb. These results indicate a possible biochemical mechanism for the clinical interactions of Fe deficiency and Pb poisoning in children.

880 HEAD TRAUMA IN CHILDREN WITH HEMOPHILIA. Gungor Karayalcin, Ashok Shende, Robert Festa, Vishwa Kapoor, Rhina Rodriquez and Philip Lanzkowsky. Sch of Medicine, Health Sci Ctr, SUNY at Stony Brook, Children's Hospital of LIJ-HMC, Dept of Peds, New Hyde Park, NY.

Central nervous system bleeding is the main cause of death in hemophilia accounting for 34% of all deaths. Between January 1981 and December 1983, 26 children with hemophilia ranging in age from 1 to 17 years (mean of 7.5 + 4.5) had 67 hospital admissions for head trauma. Eighteen had Factor VIII, seven Factor IX deficiency and one Von Willebrand's Disease. Of the 26 children, 17 had severe hemophilia and 2 had Factor VIII inhibitors. All patients had scalp hematoma; 3 had lacerations at the site of the hematoma; 1 had severe cerebral concussion; 3 had a history of possible loss of consciousness; 12 had headache; 1 had vomiting and 4 had dizziness on admission. All patients received factor replacement therapy within 12 hours of trauma and factor levels were maintained over 50% for 2 to 9 days (mean 2.7 + 1.1) and all recovered without sequelae. Eighteen patients had skull x-rays on 38 occasions, all of which were negative. Six patients had EEGs, all of which were normal. Of 20 patients who had CT scans on 25 occasions, all were negative. Hemophilia patients treated with adequate factor replacement following head trauma have an excellent prognosis and in the absence of persistent neurological signs or symptoms probably do not require CT examination.

† **878** PLASMA ANTITHROMBIN III LEVELS IN CHILDREN WITH HOMOZYGOUS SICKLE CELL DISEASE (SCD). Gungor Karayalcin, David Chung, Pretti Pinto and Philip Lanzkowsky. Sch of Med, Health Sci Ctr, SUNY at Stony Brook, Children's Hospital of LIJ-HMC, Dept of Peds, New Hyde Park, NY.

Plasma Antithrombin III activity (AT-IIIAc) and antigen (AT-IIIAgn) levels were determined in 75 children with SCD in the steady state aged 2 to 19 yrs (mean of 9.6 ± 5.6) and in matched controls. Thirty-three of these patients were also studied during steady state (SS) and during vaso-occlusive crisis (VOC).

	Control	SS	SS	VOC
	(N=75)		(N=33)	
AT-IIIAc(%)	112.7±8.9	88.8±12.3	93.6±11.9	78.2±10.8
(p value)	< 0.0005		< 0.0005	
AT-IIIAgn (mg/dl)	12.0±1.7	11.7±1.5	11.8±1.9	10.0±2.0
(p value)	NS		< 0.0005	

In the steady state, AT-IIIAc was below control values whereas AT-IIIAgn levels were normal. During VOC there was decrease in both AT-IIIAc and AT-IIIAgn as compared to the steady state. Of the 33, fifteen patients had daily AT-IIIAc and AT-IIIAgn determinations. Decreased levels of AT-IIIAc and AT-IIIAgn rose above the initial levels with clinical improvement (68.6±13.9;8.1±2.7 and 88.4±11.2;12.0±2.8, respectively, P < 0.001) 24-48 hours after the symptoms had disappeared. Thrombotic phenomena appear to play an important role in VOC of SCD and sub-normal AT-IIIAc is found in patients in steady state. The further decline in functional AT-III is probably related to increased consumption and decreased production of AT-IIIAgn due to altered liver function during VOC.

881 PARTIAL EXCHANGE TRANSFUSION (PET) FOR TREATMENT OF PRIAPISM IN CHILDREN WITH SICKLE CELL DISEASE (SCD) Gungor Karayalcin, Ashok Shende, Robert Festa and Philip Lanzkowsky. School of Medicine, Health Sciences Centre, SUNY at Stony Brook, Children's Hospital of LIJ-HMC, Department of Pediatrics, New Hyde Park, N.Y. 11042.

Sixteen episodes of priapism in 6 children with SCD, aged from 9 to 15 years (mean 12.0 ± 2.5) were treated with 2 or 3 PET. In 14 episodes (4 patients) PET was started within 6 to 8 hours from the onset. Marked clinical improvement occurred within 1.3 ± 0.5 days and complete recovery followed each episode. In 2 episodes (2 patients) PET was started at 17 and 20 hours from the onset. In these 2 cases there was only partial response to PET and needle aspiration of corpora cavernosum and glandular A-V fistula were performed at the 3rd and 5th day of the onset, respectively.

	PRE-PET		POST-PET I		POST-PET II		POST-PET III	
	HbS	Hct	HbS	Hct	HbS	Hct	HbS	Hct
N	16		16		16		4	
Range	84-100	18-26	40-62	26-31	27-43	31-36	11-26	34-37
MEAN	93.8	22.3	51.6	28.9	32.4	33.4	19.5	35.5
SD	+3.9	+2.7	+6.5	+1.7	+6.5	+1.5	+6.2	+1.3

Priapism is a serious complication of SCD and may result in impotence. Conservative measures generally are not effective and impotence (partial or complete) is a frequent sequal to surgical procedures. Treatment with PET is successful alternative therapy at the early onset of priapism.