



Weekly

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Update on Acquired Immune Deficiency Syndrome (AIDS) among Patients with Hemophilia A

In July 1982, three heterosexual hemophilia A patients, who had developed *Pneumocystis carinii* pneumonia and other opportunistic infections, were reported (1). Each had in vitro evidence of lymphopenia and two patients who were specifically tested had evidence of T-lymphocyte abnormalities. All three have since died. In the intervening 4 months, four additional heterosexual hemophilia A patients have developed one or more opportunistic infections accompanied by in-vitro evidence of cellular immune deficiency; these four AIDS cases and one highly suspect case are presented below. Data from inquiries about the patients' sexual activities, drug usage, travel, and residence provide no suggestion that disease could have been acquired through contact with each other, with homosexuals, with illicit drug abusers, or with Haitian immigrants--groups at increased risk for AIDS compared with the general U.S. population. All these patients have received Factor VIII concentrates, and all but one have also received other blood components.

Case 1: A 55-year-old severe hemophiliac from Alabama developed anorexia and progressive weight loss beginning in September 1981. He had developed adult-onset diabetes mellitus in 1973, which had required insulin therapy since 1978. He had had acute hepatitis (type unknown) in 1975. In March 1982, he was hospitalized for herpes zoster and a 17-kg weight loss. Hepatosplenomegaly was noted. The absolute lymphocyte count was 450/mm³. Liver enzymes were elevated; antibodies to hepatitis B core and surface antigens were present. A liver biopsy showed changes consistent with persistent hepatitis. Evaluation for an occult malignancy was negative. The zoster resolved following 5 days of adenosine arabinoside therapy.

In early June, he was readmitted with fever and respiratory symptoms. Chest x-ray showed bibasilar infiltrates. No causative organism was identified, but clinical improvement occurred coincident with administration of broad spectrum antibiotics. Laboratory studies as an outpatient documented transient thrombocytopenia (63,000/mm³) and persistent inversion of his T-helper/T-suppressor ratio (TH/TS =0.2). He was readmitted for the third time in early September with fever, chills and nonproductive cough. His cumulative weight loss was now 47 kg. Chest x-ray demonstrated bilateral pneumonia, and open lung biopsy showed infection with *P. carinii*. He responded to sulfamethoxazole/trimethoprim (SMZ/TMP). His T-cell defects persist.

Case 2: A 10-year-old severe hemophiliac from Pennsylvania had been treated with Factor VIII concentrate on a home care program. He had never required blood transfusion. He had been remarkably healthy until September 1982 when he experienced intermittent episodes of fever and vomiting. Approximately 2 weeks later, he also developed persistent anorexia,

fatigue, sore throat, and nonproductive cough. On October 20, he was admitted to a hospital with a temperature of 38.4 C (101.2 F) and a respiratory rate of 60/min. Physical examination revealed cervical adenopathy but no splenomegaly. The absolute number of circulating lymphocytes was low (580/mm³) and the T-helper/T-suppressor ratio was markedly reduced (TH/TS = 0.1). His platelet count was 171,000/mm³. Serum levels of IgG, IgA, and IgM were markedly elevated. Chest x-rays showed bilateral pneumonia and an open lung biopsy revealed massive infiltration with *P. carinii* and *Cryptococcus neoformans*. Intravenous SMZ/TMP and amphotericin B have led to marked clinical improvement, but the T-cell abnormalities persist.

Case 3: A 49-year-old patient from Ohio with mild hemophilia had been treated relatively infrequently with Factor VIII concentrate. During the summer of 1982, he noted dysphagia and a weight loss of approximately 7 kg. In October, he was treated for cellulitis of the right hand. Two weeks later, he was observed by a close relative to be dyspneic. He was admitted in November with progressive dyspnea and diaphoresis. Chest x-rays suggested diffuse pneumonitis. His WBC count was 11,000/mm³ with 9% lymphocytes (absolute lymphocyte number 990/mm³). The TH/TS ratio was 0.25. Open lung biopsy revealed *P. carinii*. The patient was treated with SMZ/TMP for 6 days with no improvement, and pentamidine isethionate was added. Virus cultures of sputum and chest tube drainage revealed herpes simplex virus. He died on November 22.

Case 4: A 52-year-old severe hemophiliac from Missouri was admitted to a hospital in April 1982 with fever, lymphadenopathy, and abdominal pain. Persistently low numbers of circulating lymphocytes were noted (480/mm³). Granulomata were seen on histopathologic examination of a bone marrow aspirate. Cultures were positive for *Histoplasma capsulatum*. The patient improved after therapy with amphotericin B. During the following summer and early fall, he developed fever, increased weight loss, and difficulty thinking. On readmission in early November, he had esophageal candidiasis. Laboratory tests showed profound leukopenia and lymphopenia. A brain scan showed a left frontal mass, which was found to be an organizing hematoma at the time of craniotomy. A chest x-ray showed "fluffy" pulmonary infiltrates. Therapy with SMZ/TMP was begun. Exploratory laparotomy revealed no malignancy. A splenectomy was performed. Biopsies of liver, spleen, and lymph node tissues were negative for *H. capsulatum* granulomata. The lymphoid tissue including the spleen showed an absence of lymphocytes. His total WBC declined to 400/mm³ and the TH/TS cell ratio was 0.1. He died shortly thereafter.

Suspect Case: Described below is an additional highly suspect case that does not meet the strict criteria defining AIDS. A 7-year-old severe hemophiliac from Los Angeles had mild mediastinal adenopathy on chest x-ray in September 1981. In March 1982, he developed a spontaneous subdural hematoma requiring surgical evacuation. In July, he developed parotitis. In August, he developed pharyngitis and an associated anterior and posterior cervical adenopathy, which has not resolved. In late September, he developed herpes zoster over the right thigh and buttock, and oral candidiasis.

Chest x-rays revealed an increase of the mediastinal adenopathy and the appearance of new perihilar infiltrates. In late October, enlargement of the cervical nodes led to a lymph node biopsy. Architectural features of the node were grossly altered, with depletion of lymphocytes. Heterophile tests were negative. IgG, IgA, and IgM levels were all elevated. He has a marked reduction in T-helper cells and a TH/TS ratio equal to 0.4. Recent progressive adenoid enlargement has caused significant upper airway obstruction and resultant sleep apnea. Reported by M-C Poon, MD, A Landay, PhD, University of Alabama Medical Center, J Alexander, MD, Jefferson County Health Dept, W Birch, MD, State Epidemiologist, Alabama Dept of Health; ME Eyster, MD, H Al-Mondhiry, MD, JO Ballard, MD, Hershey Medical Center, E Witte, VMD, Div of Epidemiology, C Hayes, MD,

State Epidemiologist, Pennsylvania State Dept of Health; LO Pass, MD, JP Myers, MD, J Politis, MD, R Goldberg MD, M Bhatti, MD, M Arnold, MD, J York, MD, Youngstown Hospital Association, T Halpin, MD, State Epidemiologist, Ohio Dept of Health; L Herwaldt, MD, Washington University Medical Center, A Spivack, MD, Jewish Hospital, St. Louis, HD Donnell MD, State Epidemiologist, Missouri Dept of Health; D Powars, MD, Los Angeles County-University of Southern California Medical Center, SL Fannin, MD, Los Angeles County Dept of Health Svcs, J Chin, MD, State Epidemiologist, California State Dept of Health; AIDS Activity, Div of Host Factors, Div of Viral Diseases, Center for Infectious Diseases, Field Svcs Div, Epidemiology Program Office, CDC.

Editorial Note

Editorial Note: These additional cases of AIDS among hemophilia A patients share several features with the three previously reported cases. All but one are severe hemophiliacs, requiring large amounts of Factor VIII concentrate. None had experienced prior opportunistic infections. All have been profoundly lymphopenic (1000 lymphocytes/mm³)) and have had irreversible deficiencies in T-lymphocytes. Clinical improvement of opportunistic infections with medical therapy has been short lived. Two of the five have died.

In most instances, these patients have been the first AIDS cases in their cities, states, or regions. They have had no known common medications, occupations, habits, types of pets, or any uniform antecedent history of personal or family illnesses with immunological relevance.

Although complete information is not available on brands and lot numbers for the Factor VIII concentrate used by these additional five patients during the past few years, efforts to collect and compare these data with information obtained from the earlier three cases are under way. No common lot number has been found among the lots of Factor VIII given to the five patients from whom such information is currently available.

These additional cases provide important perspectives on AIDS in U.S. hemophiliacs. Two of the patients described here are 10 years of age or less, and children with hemophilia must now be considered at risk for the disease. In addition, the number of cases continues to increase, and the illness may pose a significant risk for patients with hemophilia.

The National Hemophilia Foundation and CDC are now conducting a national survey of hemophilia treatment centers to estimate the prevalence of AIDS-associated diseases during the past 5 years and to provide active surveillance of AIDS among patients with hemophilia.

Physicians are encouraged to continue to report AIDS-suspect diseases among hemophilia patients to the CDC through local and state health departments.

Reference

1. CDC. Pneumocystis carinii pneumonia among persons with hemophilia
A. MMWR 1982; 31:365-7.

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