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Reports on AIDS

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This publication includes all the articles related to AIDS that have appeared in the *Morbidity and Mortality Weekly Report*, published by the Centers for Disease Control. These articles, arranged in chronological order, track the reporting of information on AIDS from 1981, when CDC first published information on Kaposi's sarcoma and *Pneumocystis carinii* pneumonia occurring in young homosexual men. In 1981, CDC formed a task force to establish risk factors, carry out laboratory studies, and disseminate timely information on the disease now known as the acquired immunodeficiency syndrome (AIDS).

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Pneumocystis carinii Pneumonia among Persons with Hemophilia A

CDC recently received reports of three cases of *Pneumocystis carinii* pneumonia among patients with hemophilia A and without other underlying disease. Two have died, one remains critically ill. All three were heterosexual males; none had a history of intravenous (IV) drug abuse. All had lymphopenia, and the two patients who were specifically tested have had *in vitro* laboratory evidence of cellular immune deficiency. The case reports follow.

Patient 1: A 62-year-old resident of Westchester County, New York, with a history of chronic hepatitis had received frequent injections of Factor VIII concentrate for severe hemophilia for many years. In February 1981, he began to experience weight loss and vague right upper quadrant abdominal discomfort associated with laboratory evidence of increasing hepatic dysfunction. In December 1981, while hospitalized in Miami, Florida, for elective knee surgery, he complained of cough and fever. He was lymphopenic, and chest X-ray revealed interstitial infiltrates compatible with viral pneumonia. He was discharged in late December after a brief course of corticosteroids associated with overall clinical improvement. He returned in severe respiratory distress a few days later. Open lung biopsy on January 5 revealed *P. carinii*, for which he received sulfamethoxazole/trimethoprim (SMZ/TMP) during the 2 weeks before death. *P. carinii* pneumonia and micronodular cirrhosis were documented at post-mortem examination.

Patient 2: A 59-year-old lifelong resident of Denver, Colorado, noted the onset of gradual weight loss, dysphagia associated with pharyngitis, aphthous-like ulcers, and anterior cervical adenopathy beginning in October 1980. As a patient with severe hemophilia, he had received frequent injections of Factor VIII concentrate for several years. Weight loss continued over a period of months. Oropharyngeal candidiasis was diagnosed in February 1982. He was hospitalized in May 1982 with symptoms including nausea, vomiting, and recurrent fever. Pneumonia was diagnosed, and *P. carinii* and cytomegalovirus (CMV) were repeatedly identified from lung tissue or bronchial secretions using histopathologic and culture techniques. Therapy with SMZ/TMP and pentamidine isethionate continued until death on July 5, 1982. Laboratory evidence for cellular immune dysfunction included absent mitogen responses and depletion of the T-helper lymphocyte cell population, relative increase in T-suppressor cells, and resultant inverted T-helper/T-suppressor ratio.

Patient 3: A previously healthy 27-year-old lifelong resident of northeastern Ohio developed fever, urinary frequency and urgency, and extreme lassitude in July 1981. He had frequently received parenteral Factor VIII concentrate for severe hemophilia. Bilateral pneumonia was diagnosed in October 1981, and open lung biopsy revealed *P. carinii*. He responded successfully to a 3-week course of SMZ/TMP. In February 1982, he received ketoconazole to suppress repeated episodes of oral candidiasis. He was hospitalized again in April with fever, splenomegaly, anemia, and lymphopenia. An extensive tumor work-up (including laparotomy) did not uncover an underlying malignancy. Cultures of bone marrow, liver, mesenteric lymph nodes, and blood grew *Mycobacterium avium*. *In vitro* immunological testing in March indicated a reduction in absolute number of circulating T-cells. Subsequent, more extensive testing documented the lack of lymphocyte responsiveness to mitogens, absolute and relative decrease in T-helper cells, relative increase in T-suppressor cells, and resultant inverted T-helper/T-suppressor ratio.

For each patient, records of the administration of Factor VIII concentrate were reviewed to determine manufacturer and lot numbers. No two of the patients are known to have received concentrate from the same lots.

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Editorial Note: *Pneumocystis carinii* pneumonia has not been previously reported among hemophilia patients who have had no other underlying diseases and have not had therapy commonly associated with immunosuppression. A review of the Parasitic Disease Drug Service's records of requests for pentamidine isethionate for 1980-1982 failed to identify hemophilia among the underlying disorders of patients for whom pentamidine was requested for *Pneumocystis carinii* therapy.