

Our study shows the clinical progression of HIV disease in a group of haemophilic children infected for between six and 11 years. The data is taken from the standard clinical practices of 10 established northern European haemophilia centres. Further follow up will continue to provide insight into the progression of HIV infection in this age group.

- 1 Pneumocystis carinii pneumonia among persons with hemophilia A. *MMWR* 1989; 31: 365-7.
- 2 Lee CA, Phillips AN, Elford J, et al. The natural history of human immunodeficiency virus infection in haemophilic cohort. *Br J Haematol* 1989; 73: 228-34.
- 3 Brettler DB, Levine PH. Factor concentrates in hemophilia, which one to choose? *Blood* 1989; 73: 2067-73.
- 4 Sullivan JL, Brewster FE, Brettler DB, et al. Hemophilic immunodeficiency: influence of exposure to factor VIII concentrate, LAV/HTLV III and herpes viruses. *J Pediatr* 1986; 108: 504-10.
- 5 Jason JM, Stehr-Green J, Holman RC, Evatt BL. Human immunodeficiency virus infection in hemophilic children. *Pediatrics* 1988; 82: 565-70.
- 6 Wagner N, Bialek R, Radinger H, et al. HIV-1 infection in a cohort of haemophilic patients. *Arch Dis Child* 1990; 65: 1301-4.
- 7 Revision of the CDC surveillance case definition for acquired immunodeficiency syndrome. *MMWR* 1987; 36: 35-155.
- 8 Wagner N, Bialek R, Radinger H, Brackmann HH, Becker M. Intravenous immunoglobulin in HIV-1 infected haemophilic patients. *Arch Dis Child* 1992; 67: 1267-71.
- 9 Lee CA, Phillips AN, Elford J, Janossy G, Griffiths P, Kernoff P. Progression of HIV disease in a haemophilic cohort followed for 11 years and the effect of treatment. *BMJ* 1991; 303: 1093-6.
- 10 Cuthbert RJG, Ludlam CA, Tucker J, et al. Five year prospective study of HIV infection in the Edinburgh haemophilic cohort. *BMJ* 1990; 301: 956-61.
- 11 Goedert JL, Kersler CM, Aledort LM, et al. A prospective study of human immunodeficiency virus type-1 infection and the development of AIDS in subjects with hemophilia. *N Engl J Med* 1989; 321: 1141-8.
- 12 Aledort LM, Hilgartner MW, Pike MC, et al. Variability in serial CD4 counts and relation to progression of HIV-1 infection to AIDS in haemophilic patients. *BMJ* 1992; 304: 212-6.

Epstein-Barr virus and cystic fibrosis

Non-bacterial causes of respiratory exacerbations in cystic fibrosis include respiratory syncytial virus, adenovirus, influenza virus, and rhinovirus. A recent study in Albany, New York state (Glenna Winnie and Robert Cowan, *The Pediatric Infectious Disease Journal* 1992; 11: 722-6) points for the first time to Epstein-Barr virus (EBV) as an important pathogen in this disease.

Thirty five patients with cystic fibrosis and chronic respiratory tract colonisation with *Pseudomonas aeruginosa* were studied retrospectively. All had been seen regularly at the Albany Cystic Fibrosis Center and their ages ranged from 6 to 18 years. Sixteen of them had serological evidence of previous EBV infection but, of the 19 EBV susceptible patients, 12 were admitted to hospital with an exacerbation of chest infection during an 18 month period beginning on 1st July 1987. Five of these 12 had serological evidence of EBV infection acquired shortly before the exacerbation and their clinical features were compared with those of the seven control patients who had an acute exacerbation without EBV infection.

The patients with recent EBV infection were older (mean age 16.8 v 12.5 years) but their preadmission clinical (Shwachmann-Kulczicki) and chest x ray (Brasfield) scores were similar to those of the controls. Infection with EBV was associated with worse scores during the hospital admission and on follow up for six months afterwards. Spirometric tests confirmed a greater deterioration in the EBV infected group. Average percentage falls from preadmission values at the time of hospital admission for EBV infected (control) patients were: clinical score 24.7 (3.9), chest x ray score 20.2 (6.5), forced vital capacity (FVC) % predicted 21.0 (8.2), forced expiratory volume in one second (FEV₁ %) 28.7 (9.4). At six months' follow up the figures were: clinical score 21.9 (-1.7), chest x ray score 17.9 (4.6), FVC 9.0 (0.9), FEV₁ % 18.4 (4.9). During the follow up period readmission to hospital was more than 10 times more frequent in the EBV group. Clinical evidence of infectious mononucleosis was not found in the patients with recent EBV infection and only one developed the characteristic atypical lymphocytosis. Anorexia and weight loss were the best clinical clues to EBV infection.

If these figures can be replicated then clearly the implications are considerable both for understanding deterioration in cystic fibrosis and possibly for preventing it. It is not clear whether the deterioration in lung function with EBV infection was caused by viral infection of the lungs or was a result of abnormal immune mechanisms induced by the virus. Such altered mechanisms might either lead directly to lung damage or cause increased pathogenicity from pseudomonas.

ARCHIVIST