
We have treated a 21 year old white male with classic hemophilia A for acute lymphoblastic leukemia. At presentation, the patient had a white blood count of 30,000 with 90 per cent blasts. The bone marrow aspirate revealed 80 per cent blasts which were periodic acid-schiff positive and peroxidase negative. Remission was induced with a course of methotrexate, vincristine and prednisone. Factor VIII activity was measured by the method of kaolin activated partial thromboplastin time using congenitally deficient plasma at presentation and while in remission. Factor VIII activity was less than one per cent at presentation and the same while in complete remission. At no time during the course of the disease did the measured factor VIII level spontaneously rise above one per cent concentration. Bleeds felt to be typical of hemophilia occurred at occasional intervals during the patients two year course of treatment. These did not appear to occur with any less frequency than prior to the onset of the acute leukemia. These findings are in contrast to previously published reports of increased factor VIII levels during the uncontrolled phase of acute lymphocytic leukemia.

LEUKEMIA IN FAMILIES WITH ISOLATED CASES OF SEVERE HEMOPHILIA A.

EUGERD, O.: The Institute for Thrombosis Research, Rikshospitalet, Oslo, Norway.

In 80 Norwegian families with severe hemophilia A (factor VIII<1%) 119 living cases are under study. In 40 of these families only one hemophiliac is known, and in 2 families only 2 hemophiliac brothers; investigations showed that of 36 of the mothers 30 were carriers, 6 not carriers or uncertain. In 7 of the 42 families, 5 cases of leukemia, most of them myeloid type, were recorded, and one case of myeloma. In 6 of these families low blood factor XII, about half of normal average activity, was demonstrated in the hemophiliacs and/or in his mother and/or in one of her parents. Information from Norwegian families with non-isolated severe hemophilia A, have given no records of leukemia. The high incidence of leukemia in families with isolated cases of severe hemophilia A indicates a common predisposing cause to leukemia and to mutations to hemophilia A.

NITRO BLUE TETRASULFON REDUCTION TEST IN HEMOPHILIA. P. BIBARDI, M. MURFFINI, F. SALVATI A. SERI and P. ROSSI FERRINI. Hematology Department, Florence, Italy.

Nitro Blue Tetrasulfon (NBT) reduction by neutrophils has been reported in many patients with bacterial infections or other inflammatory states. Occasionally it has been observed that hemophilic patients frequently displayed high NBT-reduction values beyond infectious diseases. NBT histochemical reduction by leukocytes was investigated in 47 hemophilic as 20 apparently health volunteers. Leukocytes were isolated by sedimentation from heparinized blood (10 000 rpm). The buffy coat is incubated with 0.5% NBT solution v/v for 30 min at 37°C and then for 15 min at room temperature. Thin smears were stained with Pappenheimer's stain. A low score (NBT-positive neutrophils<10%) in 17 normal subjects whereas in 34 hemophilia A significant (X^2=7.21; p<0.05) high score (NBT-positive neutrophils >10%) was observed. Correlation with factor VIII or IX levels, frequency of hemorrhhosis or transfusions is not significant (p>0.5). Nevertheless in 3 young hemophilic patients, unaffected from hemorrhhosis, the NBT-test is constantly negative. The findings suggest that in hemophiliacs the phagocytic function by neutrophils is frequently enhanced in course of the chronic synovitis.