FACTOR 8 ACTIVITY IN A HEMOPHILIAC PATIENT WITH ACUTE LYMPHOCYTIC LEUKEMIA. S.C. Meyer, M.B. Troner, E.C.Y. Lian, and M.A. Silverman. Department of Oncology, University of Miami, Miami, Florida, U.S.A.

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 lymphocytes. Bone marrow asperate revealed 80 per cent blasts which were periodic acid-schiff positive and peroxidose negative. Remission was induced with a course of methotrexate, vincristine and prednisone. Factor 8 activity was measured by the method of kaolin activated partial thromboplastin time using congenitally deficient plasma at presentation and while in remission. Factor 8 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 8 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 8 levels during the uncontrolled phase of acute lymphocytic leukemia.

LEUKEMIA IN FAMILIES WITH ISOLATED CASES OF SEVERE HEMOPHILIA A.

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In 80 Norwegian families with severe hemophilia A (f.VIII≤1%) 119 living cases are under study. In 40 of these families only one hemophiliac is known, and in 2 families only 2 hemophilic brothers; investigations showed that of 38 of the mothers 30 were carriers, 8 not carriers or uncertain. In 7 of the 42 families, 9 cases of leukemia, most of them myeloid type, were recorded, and one case of myelomatosis. In 6 of these families low blood f.XII, about half of normal average activity, was demonstrated in the hemophiliac and/or in his mother and/or in one of her parents. Informations 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 BLEU TETRAZOLIUM REDUCTION TEST IN HEMOPHILIA. F.BIRARDI, M.MORFINI, F.SALTI A.SERI and P.ROSSI FERRINI. Hematology Department, Florence, Italy.

Nitro Bleu Tetrazolium (NBT) reduction by neutrophils has been reported in many patients with bacterial infections or other inflammatory states. Occasionally it has been observed that hemophiliacs frequently displayed high NBT-reduction values beyond infectious diseases. NBT histochemical reduction by leukocytes was investigated in 47 hemophiliacs and 20 apparently health volunteers. Leukocytes were isolated by sedimentation from heparinized blood (10 U./ml). The buffy coat is incubated with 0,2% NBT solution v/v for 30 min. at 37°C and therefore 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 hemophiliacs 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 hemarthrosis or transfusions is not significant (p>0,5). Nevertheless in 3 young hemophiliacs, unaffected from hemarthrosis, the NBT-test is costantly negative. The findings suggest that in hemophiliacs the phagocytic function by neutrophils is frequently enhanced in course of the chronic synovitis.