HEMOPHILIA IN A CHROMOSOMAL FEMALE - AN EXTREME EXAMPLE OF LYONIZATION? J.M. Lusher, J.A. Penner, I.Warrier and R.K. Evans. Wayne State University School of Medicine, Detroit and the University of Michigan Medical Center, Ann Arbor, Michigan, U.S.A.

Severe Factor (F.) VIII deficiency is rare in females. We have previously reported two (unrelated) females with 1-3% F. VIII activity (Jour. Pediatr. 74:265-271, 1969). This report concerns another Michigan female with Hemophilia A. Although this 12 year old girl's F. VIII activity has ranged from 1-3% she has had only occasional hemarthroses. Her bleeding time is normal, her platelets aggregate normally with ristocetin and F. VIII antigen quantitation has varied from 1.02-1.40 μ/ml. In addition, platelet retention in a glass bead column has been normal. Her father has classical hemophilia A (F. VIII activity ≤1%, F. VIII antigen 1.61 μ/ml., normal bleeding time and normal platelet aggregation with ristocetin), while her mother appears to be normal (F. VIII activity 90-105% with corresponding values for F. VIII antigen, normal bleeding time and normal platelet aggregation with ristocetin). The child's only sibling, a 9 year old sister has a F. VIII activity of 40-46%, F. VIII antigen of 1.40 μ/ml., normal bleeding time and normal platelet aggregation with ristocetin. While both girls are obligate carriers, one can only speculate on the reasons for the very low levels of F. VIII activity in the propositus. Although it is possible that there was a spontaneous mutation in the propositus' maternal X chromosome, it seems likely that her 1-3% F. VIII activity reflects an extreme degree of lyonization.

REPLACEMENT THERAPY WITH A FACTOR VII CONCENTRATE IN SEVERE FACTOR VII DEFICIENCY. Kl. Schimpf, and K. Zimmermann. Rehabilitation Hospital and Hemophilia Center Heidelberg, Rehabilitation Foundation, Heidelberg, West-Germany.

A 2o year old man, factor VII activity < 1%, was confined to bed since 1 1/2 years because of recurrent joint bleedings. Mobilisation became possible by means of a substitution therapy with a factor (f) VII concentrate (Immuno). The f VII content of the preparation was 25 units (u) per ml. After injection of 4o u of f VII per kg bodyweight (bw), the halflife time was 1oo minutes. With injections of 33 u of f VII per kg bw every 6 hours, minimum levels of 5o% prior to the next injection could be maintained. A steady perfusion with 1,6 u of f VII per kg bw and hour resulted in a level of about 35% f VII. Therefore, under these modes of administration an operation should be possible. Thus, we could perform a gastroscopia with several biopsies without any complication. An intensive physiotherapy was possible by giving 4o u of f VII per kg bw twice a day during 5 weeks. After changing to injections on need, we observed 5 elbow-bleedings within 4 months, which could be controlled with 2 times 4o u per kg bw at the day of bleeding. 5 months later, the patient could be discharged in home-treatment. He was now able to climb steps, to walk for about 5oo m without a cane and to continue his professional education.

THE ALL-AMERICAN BLOOD BUDDY: DONOR MOTIVATION FOR HEMOPHILIA AND OTHER CHRONIC DISORDERS. D. VanHarlingen, D. Child, J. Dzuback, and F. Thompson. Hemophilia Association of Northern New Jersey, East Brunswick, New Jersey, U.S.A.

A novel method of enlisting donors for hemophilia from among high school and college students will be presented. This method has been successfully used in New Jersey and in less than two years of operation has recruited thousands of donors specifically for hemophilia. The program is built on a particular philosophy of responsibility for blood replacement which enhances the importance of the donor in the process of giving blood. The program is aimed at those who would not ordinarily donate blood under conventional recriutment programs.

This system is applicable to other regions and other chronic blood disorders. It has also been proven successful in motivating donors at ages other than the originally intended group. The importance of this approach to both the hemophiliac and the community at large is discussed. Statistics supporting the success and necessity of the program will be presented. We shall also present arguments which refute common criticisms of our system.