

CARRIER DETECTION IN HEMOPHILIA B. E. Briët, R.M. Bertina, N.H. van Tilburg, J.J. Veltkamp, Hemostasis and Thrombosis Research Lab, Dept. of Medicine, University Hospital, Leiden, The Netherlands.

We examined 37 obligatory carriers of hemophilia B and 40 normal women. The levels of both factor IX activity and factor IX antigen were determined. The factor IX antigen levels were assayed in a neutralization test in which a rabbit antiserum was used. 12 out of the 37 carriers had oral contraceptives and so did 20 out of the 40 normals.

Higher levels of both factor IX activity and factor IX antigen were found in the women, carriers as well as normals, using oral contraceptives. In carrier detection programs, therefore, a distinction should be made between women using the pill and those who do not. The attempt to discriminate between carriers and normals on basis of factor IX activity and antigen levels proved to be as unsatisfactory as it was on the basis of factor IX activity levels only. The overlapping area of normals and carriers was smaller in the group of women on oral contraceptive medication. On the basis of our findings we expect that we can detect with a 90% confidence 17 carriers and 17 normals in a group of 100 potential carriers with an a priori chance on carriership of 50%. The detection rate is somewhat higher for women using oral contraception. The assay of factor IX antigen by means of the inhibitor neutralization assay does not appear to improve carrier detection.

Results with an immuno-electrophoretic assay of factor IX antigen will be discussed.

THE ROLE OF THE CONSUMER-PATIENT IN THE GROWTH OF A COMPREHENSIVE HEMOPHILIA CARE UNIT. Dorothy Child, John Kopicki, and David VanHarlingen. Hemophilia Association of Northern New Jersey, East Brunswick, New Jersey, U.S.A.

The most efficient means for dealing with the myriad problems of hemophilia is through a comprehensive hemophilia care center involving medical and paramedical personnel, as well as numerous non-medical supportive services. As in all consumer services, however, problems arise when the care center team and the individuals who comprise it are unaware of the true needs of the patients they serve. To reduce such problems, an active and vocal group of hemophilia families and friends can provide insights otherwise unattainable. Working in cooperation with the center, they can provide guidance and seek funding to reduce the cost paid by the patient.

We describe the role of consumer-patients in the initiation and development of New Jersey's first and only comprehensive care center for hemophilia. We also present a detailed examination of the kinds of assistance which has been provided and which is being provided in the growth and expansion of the center's team and its activities. A plan for a statewide treatment network centered around this treatment facility will be presented.

AN EVALUATION OF HOSPITAL FACILITIES FOR CANADIAN HEMOPHILIACS. R. George and M.J. Inwood. Canadian Hemophilia Society, Hamilton, Ontario, Canada.

Wide differences exist in the standard of facilities currently available to Canadian hemophiliacs. In order to categorize these facilities a survey was conducted by members of the Canadian Hemophilia Society of 106 hospitals currently treating hemophiliacs in the 10 provinces of Canada. Two of the provinces (Quebec and British Columbia) had incomplete surveys performed. A scoring system was devised to grade the hospitals from level I to level V, with the following results. Eleven level I (comprehensive treatment centres as defined by the World Federation of Hemophilia) were identified in seven provinces. Seventeen level II (facilities considered acceptable for regularly attending hemophiliacs) were present in 8 provinces. Thirty five level III hospitals (suitable for occasional and uncomplicated therapy) were situated in 8 provinces. Thirty five level IV facilities in 7 provinces and eight level V facilities in 4 provinces were considered to have inadequate facilities for routine or sporadic therapy. Only 63 of the 106 (level I and II and III) had suitable facilities for the reception of hemophiliacs. Of these 63 hospitals only 28 were providing materials and services for home infusion programs. In order that Canada can achieve a standard of facilities comparable to other countries actively involved in hemophilia care, hospitals in all provinces must be upgraded, along with an increased number and distribution of comprehensive treatment centres.