

LOW DOSE FACTOR REPLACEMENT IN THE HEMOPHILIAC. P. Kelly, M. Boutaugh, and J. Penner, University of Michigan, Hemophilia Control Center, Ann Arbor, Michigan, U.S.A.

The standard dose of factor administered for treatment of acute bleeding in hemophiliacs is usually 20 units per Kg. To assess the effectiveness of lower doses, 103 hemophilic campers were randomly assigned and treated with 3 different dosages, i.e. 7-9 u/kg, 11-15 u/kg, and 15-17 u/kg, for treatment of soft tissue and joint bleeds. Patients with inhibitors, and those with bleeding into critical areas were excluded from the study. It was found that, when administered early, the 7-9 u/kg dose was as effective as doses in the 15-17 u/kg range for treatment of spontaneous bleeding in the extremities.

In summary, our experience with a large number of hemophilic youngsters attending a summer camp would indicate that a low dose therapeutic approach (7-9 u/kg) is effective, and permits most of the patients to participate fully in camp activities.

DOSE REQUIREMENT FOR SUBSTITUTION THERAPY IN HEMOPHILIA A. J.P. ALLAIN, Centre pour enfants Hémophiles, French Red Cross, La Queue-les-Yvelines, France.

In order to determine the relationship between different doses of factor VIII and their clinical effect, 70 children with severe hemophilia A treated with single doses of cryoprecipitate were studied. Plasma factor VIII levels were tested by a one stage assay on samples taken before, and 30 and 60 min. post-infusion. Clinical failure was defined as an absence of clinical improvement or the occurrence of bleeding in another site within 24 hours post-treatment. The relationship between plasma factor VIII levels or doses injected in u/kg and clinical result followed an exponential curve. Calculated from 153 factor VIII recoveries, plasma factor VIII levels of 0.35 and 0.52 u/ml corresponded to 95 and 99% satisfactory treatment, respectively. The same clinical result was obtained with 20 and 31 u of factor VIII/kg when studied in 1043 infusions of lyophilized cryoprecipitate. The in vivo recovery being 0.015 u factor VIII activity/ml for 1 u/kg injected, these dosages provided plasma factor VIII levels of 0.30 and 0.47 u/ml respectively. Since home treatment is largely based on single infusions of factor VIII, it is suggested that most current bleedings can be adequately treated with a dose of factor VIII providing a plasma factor VIII activity of 0.30-0.35 u/ml.

A CASE OF HENOCCH-SCHOENLEIN PURPURA IN HEMOPHILIA. A.L. Gonzaga, C.B. Azevedo and L.F. Baré, Centro de Hematologia Santa Catarina, Rio de Janeiro, Brazil.

A 25-year-old male, white, with severe hemophilia B (F.IX < 1%) developed a rash of small erythematous macules, purpuric spots and petechia on the lower extremities. These skin lesions were discrete, not pruritic, not papular and with no areas of necrosis. No relationship to previous infection was determined. As the patient did not present any other clinical alteration, he was observed daily on an ambulatory basis in a four days' period, without any medication. On the fifth day parallel to the disappearance of the skin lesions, the patient began to complain of abdominal pain, in crisis of moderate intensity. As the abdominal discomfort increased rapidly on the following 24 hours, we introduced specific therapy on an in-patient basis. The case evolved to an acute abdominal picture that required surgical intervention. The laparotomy resulted in large resections of jejunum and ileum that showed large hemorrhagic and necrotic areas. The post-operative period elapsed without incidents and the patient left the hospital in three weeks. This case that at the beginning could not give us a clear diagnosis of an anaphylactoid purpura shows us once more that in hemophilia, we must transfuse as early as possible.