A Tentative Classification of Factor XIII Deficiency in Two Groups

To the Editor:

Immunological and immunofluorescent studies were carried out on plasma and platelets of 3 cases of congenital factor XIII deficiency. Two of these patients were originally thought to have normal factor XIII sub-unit S and no subunit A [4]. However, repeated assays carried out using different lots of antiserum showed that the patients lacked in reality both sub-unit S and subunit A (fig. 1). The false positive result was due to the presence of an anti-factor VIII contaminant in the antiserum originally used (Behringwerke lot 2434). That this is so is well demonstrated by the observation that in von Willebrand’s disease no second (factor VIII) peak was evident (fig. 1).

These two patients seem to be the first cases in whom no factor XIII subunit S is detected. The third patient had a normal subunit S and no subunit A. In agreement with the above findings are some immunofluorescent studies (fig. 1). No factor XIII antigen was found in fact by the indirect immunofluorescent technique in normal, factor XIII deficiency and von Willebrand’s disease platelets. On the contrary, using the non-monospecific antiserum (Behringwerke lot 2434), a fluorescent pattern similar to that observable using an anti-factor VIII antiserum had been noted [1].

On the basis of these observations a tentative classification of factor XIII deficiency in two groups is proposed: type I, which appears to be rare, is characterized by the lack of both factor XIII subunits S and A. Type II, which appears to be relatively common, is characterized by normal and near normal subunit S and no subunit A.

The need for a re-evaluation of published cases of factor XIII deficiency by means of monospecific antisera is indicated. This is more so if one takes into account the fact that most research work on the subject was carried out using the antisera supplied by Behringwerke Laboratories [2, 3, 5]. The possibility that lot 2434 or similarly contaminated lots of antiserum might have been used remains open.
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1 2

Fig. 1. Electroimmunoassay. Top (contaminated anti-subunit S antiserum): (1) 1:2 diluted pooled normal plasma; (2) undiluted pooled normal plasma; (3) von Willebrand’s disease; (4) another patient with von Willebrand’s disease; (5) factor XIII deficiency (case 1); (6) factor XIII deficiency (case 2, sister of case 1); (7-10) 1:1, 1:2, 1:4 and 1:8 diluted pooled normal plasma. Two major rockets or peaks are evident in normal plasma; only one rocket is evident in von Willebrand’s disease and in factor XIII deficiency. The rockets seen in von Willebrand’s disease (wells 3 and 4) are taller as compared to those seen in factor XIII deficiency (wells 5 and 6). The plasma of the 3rd patient with factor XIII deficiency is not reported but showed two rockets as normal plasma. Bottom (monospecific anti-subunit S antiserum): (1) von Willebrand’s disease; (2) another patient with von Willebrand’s disease; (3) factor XIII deficiency (case 3); (4) factor XIII deficiency (case 1); (5) factor XIII deficiency (case 2, sister of case 1); (6-9) 1:1, 1:2, 1:4 and 1:8 diluted pooled normal plasma. Note that case 3 (well 3) shows a normal subunit S rocket, whereas case 1 and 2 show no precipitate (wells 4 and 5).

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