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Combined Congenital Deficiency of Factor V and Factor VIII

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To the Editor, References

I am writing this letter to inform Drs. Mazzone and Fichera (Acta haemat. 68: 337–338, 1982) that combined congenital deficiency of factor V and factor VIII has been described in childhood by Dr. Jones et al. in 1962 [1]. 1 of their patients was an 11-year-old girl. We have also observed this combination in 2 boys, 6 and 11 years of age. In addition, I would like to bring to the attention that this entity is most likely related to the decreased activity of natural inhibitor of activated protein C which is an important natural anticoagulant neutralizing the action of the activated forms of factor V and factor VIII [2].

In our 2 unrelated cases, in addition to factor V (8 and 12%) and factor VIII (4 and 8%) deficiencies, considerably decreased factor X activity (20 and 29%) was found. Although combined factor VII and factor VIII deficiencies have been reported [3], I would like to stress that factor X activity should be assayed separately in combined factor V and factor VIII deficiency cases. Because of clinical mildness of factor X deficiency in these patients, it could easily be missed without determining its activity.

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Machin, S.J.; Miller, B.R.: Congenital combined factor VII and factor VIII deficiency. Acta haemat. 63: 167–169(1980).

The above letter has been submitted to Dr. Mazzone and coworkers who offer the following reply: