Abstract

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Critical Analysis of the patients with Congenital Prekallikre in Deficiency and a Purported Bleeding Tendency

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Abstract

Objective: To analyse the bleeding manifestations presented by some patients with congenital prekallikrein deficiency. Patients and methods: 106 papers dealing with patients with prekallikrein deficiency were obtained by a time unlimited PubMed Search and by the revaluation of personal files dealing with patients with prekallikrein deficiency. Only proven or highly probable cases of congenital deficiency were included. Acquired defects were excluded. Results: Out of the 106 papers examined, eleven patients were reported to manifest a variable bleeding tendency. Bleeding was usually mild (epistaxis, easy bruising, bleeding after oral or nasal surgery). It was severe in one infant (hematomas, hemarthrosis). Transfusion therapy with whole blood or fresh frozen plasma was used in 4 patients. In only two instances there are the demonstrations of a reduction or stoppage of bleeding after administration of Fresh Frozen Plasma. The rest of these purported bleeders underwent tonsillectomy, delivery and other surgical procedures without undue bleeding. Local factors or transient coagulation defects are the likely explanation for the occasional bleeding manifestations observed. The mutations found in the three patients who were investigated by molecular biology techniques were Gly401Glu (two cases) and Cys529Tyr (one case). Conclusion: An analysis of the cases of Prekallikrein deficiency and bleeding suggests that these patients need no replacement therapy but in the case of surgery requiring cardio-pulmonary bypass procedures and heparin administration. In this case the basic prolongation of the clotting tests may complicate the evaluation of the effect of heparin

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