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Review Article

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FACTOR 5 LEIDEN DEFICIENCY AND PREGNANCY

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ABSTRACT

Factor 5 leiden deficiency or activated protein C resistance is a thrombophilia characterized by blood hypercoagulability and constitutes a risk of venous thromboembolic disease.Nowadays this disorder does not constitute an obstacle to pregnancy but a diagnosis and an adapted monitoring allows to carry a pregnancy to term.

KEYWORDS: factor 5 leiden; C-reactive protein ; pregnancy; monitoring.

INTRODUCTION

Coagulant factor 5, also called proaccelerin or labile factor, is the plasma cofactor of prothrombinase, which converts prothrombin to thrombin. It also participates in physiological anticoagulation by deactivating activated factor VIII. VF deficiency, also called parahaemophilia, was first described by Owren in 1943.

It is a very rare hereditary (autosomal recessive) coagulation disorder, with a prevalence of 1 in 1,000,000.Nearly 200 cases have been reported in the literature,with available data indicating that this deficiency manifests itself by bleeding of the skin and mucous membranes,unlike hemophilias A and B, which most often manifest themselves by hemarthroses.Data in the literature on VF deficiency during pregnancy are few.

REVIEWS OF THE LITTERATURE

Factor V deficiency is often suspected in the presence of bleeding symptoms associated with a decrease in PT and a prolongation of APTT.[1,2,3] if the FV level is low, a consumption coagulopathy, a hepatopathy, a combined FV and FVIII deficiency, and the presence of acquired anti-FV antibodies must be eliminated before the diagnosis is retained. This deficiency is due to a mutation in the gene coding for human VF: to date, 56 mutations have been described. Childbirth and the post-partum period are described as situations with a high risk of haemorrhage in patients with congenital VF deficiency.[5]

There are few data in the literature concerning the management of pregnancy and delivery in these patients.[4] Concerning the route of delivery, some authors prefer programmed caesarean section at term, bearing in mind that this is a surgical procedure with a risk of haemorrhage, while others prefer spontaneous or induced vaginal delivery, if obstetric conditions allow, but on condition that blood products are available at all times,[7,8,9]

Regarding replacement therapy, there are no VF concentrates, and the only source for VF replacement is FFP at a dose of 20 ml/kg before vaginal delivery or caesarean section, which would allow a VF rate >20%, the minimum necessary for satisfactory haemostasis, and suggest maintaining FFP transfusions every 12 to 24 hours in the postpartum period until healing.[10,11,12] In our patient, the transfusion of 20 ml/kg resulted in an increase in the VF rate to 26%, and the maintenance of the transfusion of 5 ml/kg per 12 hours allowed the VF rate to be maintained at approximately 15%, which prevented bleeding complications.[13,14] The possible complications of FFP transfusions are water overload, the theoretical risk of viral transmission, which leads to a preference for viroinactivated products despite a lower concentration of VF, and the development of anti-VF antibodies. In the latter case, the therapeutic options are activated prothrombin complex concentrates, recombinant activated FVII, or platelet transfusions.[15]

CONCLUSION

Delivery and postpartum are described as high bleeding risk situations in patients with congenital VF deficiency. Transfusion of FFP with monitoring of PT and VF levels in the peripartum period can prevent this risk.

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