



A case of a combined commonly inherited bleeding and clotting disorder

To the Editor: We report the uncomplicated outcome of pregnancy in a patient with a combined bleeding and clotting disorder managed with ante- and postpartum thromboprophylaxis.

A 30-year-old woman was referred for screening for an inherited thrombophilia. Her brother had developed a life-threatening deep-vein thrombosis after a mild sports injury. A noteworthy finding on personal history was easy bruising. Thrombotic screen testing revealed a heterozygous factor V Leiden mutation and a bleeding screen type I von Willebrand disease (vWD). She later presented at 12 weeks' gestation with her first pregnancy. Factor VIII, von Willebrand factor (VWF) antigen and activity levels measured at presentation and in the

third trimester were in the normal range. Thromboprophylaxis with enoxaparin 20 mg daily (weight 56 kg) was prescribed from 32 weeks' gestation until 4 weeks before delivery. Anti-Xa activity and platelet counts were monitored. She went into preterm labour and delivered a male baby by caesarean section.

The factor V Leiden mutation is the most frequent cause of familial thrombosis and vWD is the most common inherited bleeding disorder.^{1,2} Concurrence of these disorders is estimated to occur in 0.25% of Caucasians and poses therapeutic challenges, particularly in pregnancy.

Pregnancy is physiologically associated with a hypercoagulable state and an increased risk of venous thrombo-embolism (VTE), with a risk of 1/1 000 deliveries.³ In pregnancy FVIII and VWF levels normalise, thereby ameliorating the bleeding risk. The presence of the factor V Leiden mutation increases the risk of VTE and pregnancy-related morbidity to 1/500.⁴ Pregnancy-related complications include recurrent miscarriages, pre-eclampsia, placental abruption and intra-uterine growth restriction.⁵ However, the use of thromboprophylaxis in the management of pregnant women with a known thrombophilia and no prior VTE is based on case-control studies and a small number of prospective studies.

Management of these patients therefore remains controversial. The American College of Chest Physicians 2008 guidelines recommends postpartum prophylaxis for 4 - 6 weeks (grade 2C). After delivery the patient's risk should be individually assessed to determine whether prophylaxis is indicated (grade 1C).³ In view of this patient's significant family history she was managed with both ante- and postpartum prophylaxis.

Heparins are the preferred drug during pregnancy because they do not cross the placenta,³ and low-molecular-weight heparin (LMWH) is preferred to unfractionated heparin.⁶ Monitoring of LMWH anti-Xa levels is recommended 3 hours after injection, with adjustment of the dose to maintain a level of 0.2 - 0.6 IU.³

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