

PERINATAL MANAGEMENT IN WOMAN WITH MILD HEMOPHILIA B: A TWIN PREGNANCY CASE

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Background. Women affected by haemophilia, and haemophilia carriers, may face bleeding challenges during pregnancy and delivery, particularly for haemophilia B (HB) given that factor IX levels do not increase in pregnancy. Evidence on the use of extended half-life (EHL) factor IX concentrates in this setting remains very limited.

Case Report. This case describes the perinatal management of a 29-year-old woman with mild haemophilia B (c.707G>A p.Gly236Asp factor IX gene variant; baseline factor IX 37%; International Society of Thrombosis and Haemostasis Bleeding Assessment Tool: 0) carrying monochorionic diamniotic twin male pregnancy. Family history included severe HB in maternal uncle, cousins and great-uncle; maternal grandmother and great-aunt were HB carriers. Pregnancy course was regular. Maternal factor IX levels remained stable: 15 weeks GA (gestational age) 41%, 32 weeks GA 49%. Genetic counselling offered; invasive prenatal diagnosis declined. Elective caesarean section performed at 35 weeks GA for obstetric indication. Preoperative Eftrenonacog Alfa (Alprolix®) 3000 IU (33 IU/kg) infused 30 minutes prior, achieving post-infusion factor IX 55%; tranexamic acid 10 mg/kg IV 1h pre-procedure then every 8 hours; strict no-forceps/vacuum policy. Estimated blood loss: 500 ml. Atraumatic cord blood sampling showed neonatal factor

IX levels 37%. Postpartum course was unremarkable: normal lochia and wound healing, maternal factor IX 43% at 24 hours, LMWH (Low Molecular Weight Heparin) 4000 IU/day thromboprophylaxis was started at day 1 to day 8; the patient was discharged at day 4 without bleeding. No primary/secondary postpartum haemorrhage occurred; no additional factor IX infusions were required: a single EHL factor IX infusion provided effective haemostatic coverage for caesarean delivery with minimal blood loss and excellent peripartum safety. A multidisciplinary team (haematologist, obstetrician, anaesthesiologist, neonatologist, coagulation laboratory physician) defined antenatal strategy minimizing maternal bleeding risk and neonatal trauma through coordinated replacement therapy timing, antifibrinolytics, obstetric planning and postpartum surveillance.

Conclusions. Published experience on EHL factor IX use in pregnant HB women remains scarce, particularly twin gestations where maternal-foetal management complexity increases. Our case outcome supports individualized Eftrenonacog Alfa prophylaxis as effective option for delivery management in mild HB women, making feasible a tailored peripartum care that balances maternal haemostasis, neonatal assessment and thromboprophylaxis needs in high-risk coagulation disorders.