Pregnancy and delivery in a woman with von Willebrand Disease type 2M: successfull treatment with Wilfactin®

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Von Willebrand Disease (vWD) is a bleeding disorder that results from inherited defects in the concentration, structure and function of the glycoprotein von Willebrand factor (vWF

vWD classification helps us to identify clinically important subgroups of patterns, denoting characteristics that imply a pattern of attributes, including clinical signs and treatment response.

For example, it has been shown that the hemorrhagic symptoms seems to be more severe in type 2A respect to the type 2M, with a ten times greater chance of gastrointestinal bleeding, related to an angiodysplasia, caused by the loss of high molecular weight multimers.

Pregnancy in a woman with vWD may carry a significant risk of bleeding. In order to correct the primary vWF defect and avoid supra-phyiologic plasma levels of Factor VIII, a pure concentrate devoid of FVIII was developed and used (Wilfactin®)

We here report the management of pregnancy and delivery of a female with vWD type 2M.
The patient, before becoming pregnant, had used Wilfactin for the control of menometrorrhagias, with excellent response and hemoglobin values of 11 gr/dl.
Before treatment, the patient, because of bleeding and despite the use of a product containing both FVIII and vWF, not maintained hemoglobin values exceeding 8 gr/dl.

In march the patient becomes pregnant and not practice more replacement therapy with Wilfactin until the time of delivery.

During pregnancy the patient was monitored with regular checks of blood count and coagulation, not presenting symptoms of bleeding.

During the seven month of pregnancy the values were as follows:
Hb: 11 gr/dl, PLTS: 240.000/mmc, A.P.: 109% INR: 0.99, aPTT: 51".5, fibrinogen: 669 mg/dl FVIII:C 8.7%, vWRiCoF: 8.4%, vWFAg: 11.3%

During the eight month of pregnancy we had the following values:
Hb: 10.5 gr/dl, PLTS: 210.000/mmc, A.P.: 94% INR: 1.01, aPTT: 48".2, fibrinogen: 1.243 mg/dl, FVIII:C 18%, vWRiCoF: 7.7%, vWFAg: 18.2%

It was decided to give birth with cesarean section for a better control of bleeding and was carried out the following therapy:
- **Plasma concentrated containing both FVIII and vWF, 25 I.U./Kg b.w. (2.000 I.U.) e.v. to be administered one hour before the delivery**
- **Wilfactin, 1.000 I.U. e.v. 30 minutes before the delivery**
- **Saline solution 500 cc with tranexamic acid, 2 gr e.v. starting 30 minutes before the delivery**
- **Wilfactin, 1.000 I.U. e.v. 2 hours after the delivery**
- **Wlfactin, 1.000 I.U. e.v. the day after the delivery**

Cesarean section took place normally, a male was borne; there were no bleeding complications during and after the delivery.

Immediately after the delivery, sample controls were performed, with the following results:
Hb: 10 gr/dl, PLTS: 257.000/mmc, A.P.: 100% INR: 1,00, aPTT: 31".8, fibrinogen: 876 mg/dl, FVIII:C 50%, vWRiCoF: 32%, vWFAg: 46%.

This case report provides, in our opinion, suggestions on monitoring and management of pregnant patients with vWD, by using a product containing vWF and only small amounts of FVIII.