Orthopedic surgery in a patient affected by von Willebrand disease Type 3: Benefits of continuous infusion - WFH 2016

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▪ Introduction

Von Willebrand disease (VWD) Type 3 is the most rare and severe form of the disease. It is characterized by a bleeding disorder caused by an almost complete quantitative deficiency in von Willebrand factor (vWF), leading to undetectable levels of VWF in plasma and in platelets, and very low levels of factor VIII (FVIII).

This form, representing 5% of patients affected by VWD, has an annual incidence which shows variations depending on the countries or ethnicities. In fact, it is ranging from 0,5 to 3 per million in Europe and North America, and next to 2 per million in countries associated with a frequent consanguinity.

The hemorrhagic episodes are appearing in the neonatal phase or in the early childhood, typically with severe mucocutaneous bleedings (epistaxis, menorrhagia, bleeding after birth, gastrointestinal bleed, …), extended postoperative bleedings, but also hematoma and haemarthrosis (as in hemophilia A, due to the FVIII defect), or even life-threatened hemorrhage like intracranial bleed.

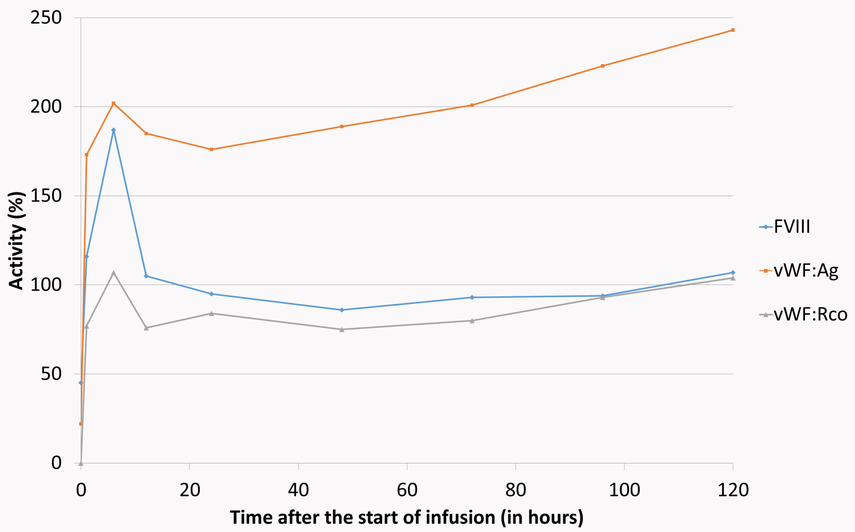
Regarding the joint bleedings, they are leading to an arthropathy with a painful cartilage and bone destruction. In approximately 15% of cases this affection concerns the ankle, which actually suffers from the most important load pressure per surface unit in the human body. Moreover, this articulation is only protected by few muscle mass. When the arthropathy progress until a stage where there is no more possibility to preserve the joint, ankle arthrodesis is historically the most common surgical option, knowing that this kind of operation needs replacement therapies, consisting on infusion of vWF concentrates.

▪ Case Report

The authors report the case of a 35 year-old patient, suffering from VWD Type 3 under prophylaxis since 5 years, who went to Strasbourg University Hospital in order to undergo a left talocrural arthrodesis. The patient already had a right ankle arthrodesis 7 years ago.

Since the clinicians responsible for the patient had a strong experience in haemophilic surgery, it was decided to perform the arthrodesis under continuous vWF infusion.

After an inital bolus of 40 UI.kg^{-1} of a combined vWF and FVIII concentrate (Wilstart®, LFB), a continuous vWF perfusion was started at a rate of 2,8 UI.kg^{-1} per hour. Regular monitoring of levels of vWF antigen (vWF:Ag), vWF ristocetin cofactor activity (vWF:Rco) and FVIII were daily assessed, in order to adjust the infusion rate. The continuous perfusion was finally stopped after a cover of 5 days, and neither bleeding nor thrombotic complications were reported with this VWD Type 3 patient, who, furthermore, is known for smoking and having hypertension.



	Arthrodesis in 2009	Arthrodesis in 2016
Replacement therapy	Repeated injections	Continuous infusion
Length	5 days	5 days
Number of units of vWF concentrates	33 000 UI (Wilfactin®, LFB)	23 000 UI (Wilfactin®, LFB)

▪ Discussion and Conclusion

Continuous infusion of vWF concentrates, which is for now an infrequent choice for replacement therapy among patients with severe VWD, is an interesting option to the classic protocols of repeated injections. The main goal of the perfusion is to avoid plasmatic level varations of coagulation factors, ensuring consequently optimal haemostatic conditions, which enable a smaller number of VWF units given to the patient.