

# HIGHLY PURIFIED VWF/FVIII COMPLEX CONCENTRATE AS FIRST LINE THERAPY IN FOUR PATIENTS AFFECTED BY ACQUIRED HAEMOPHILIA A AND CARDIOVASCULAR DISEASE

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## BACKGROUND

Bleeding control is the first priority in acquired haemophilia A (AHA). International guidelines recommend bypassing agents (BA) as first line therapy. However, literature shows that BA use may be associated with thromboembolic complications, more likely in patients with underlying cardiovascular diseases.

Here we describe four cases of bleeding in AHA patients with high cardiovascular risk successfully treated with highly purified (HP) VWF/FVIII concentrate.

### Patient 1

A 69 years old man, with a recent history of myocardial infarction treated with percutaneous transluminal coronary angioplasty and stenting, presented with anaemia (Hb 79 g/L) due to rectus abdominis haematoma. Other laboratory findings showed aPTT 102 sec, FVIII:C 0.3%, inhibitor 6 BU.



Patient was transfused with packed red blood cells (PRBC) and treated with a bolus of HP VWF/FVIII (20000 IU; 263 IU/kg) followed by continuous infusion (240 IU/kg/day, adjusted to attain FVIII levels of 60-80%) for 11 days.

Considering ischaemic heart disease, therapy with clopidogrel was restarted at discharge.

### Patient 2

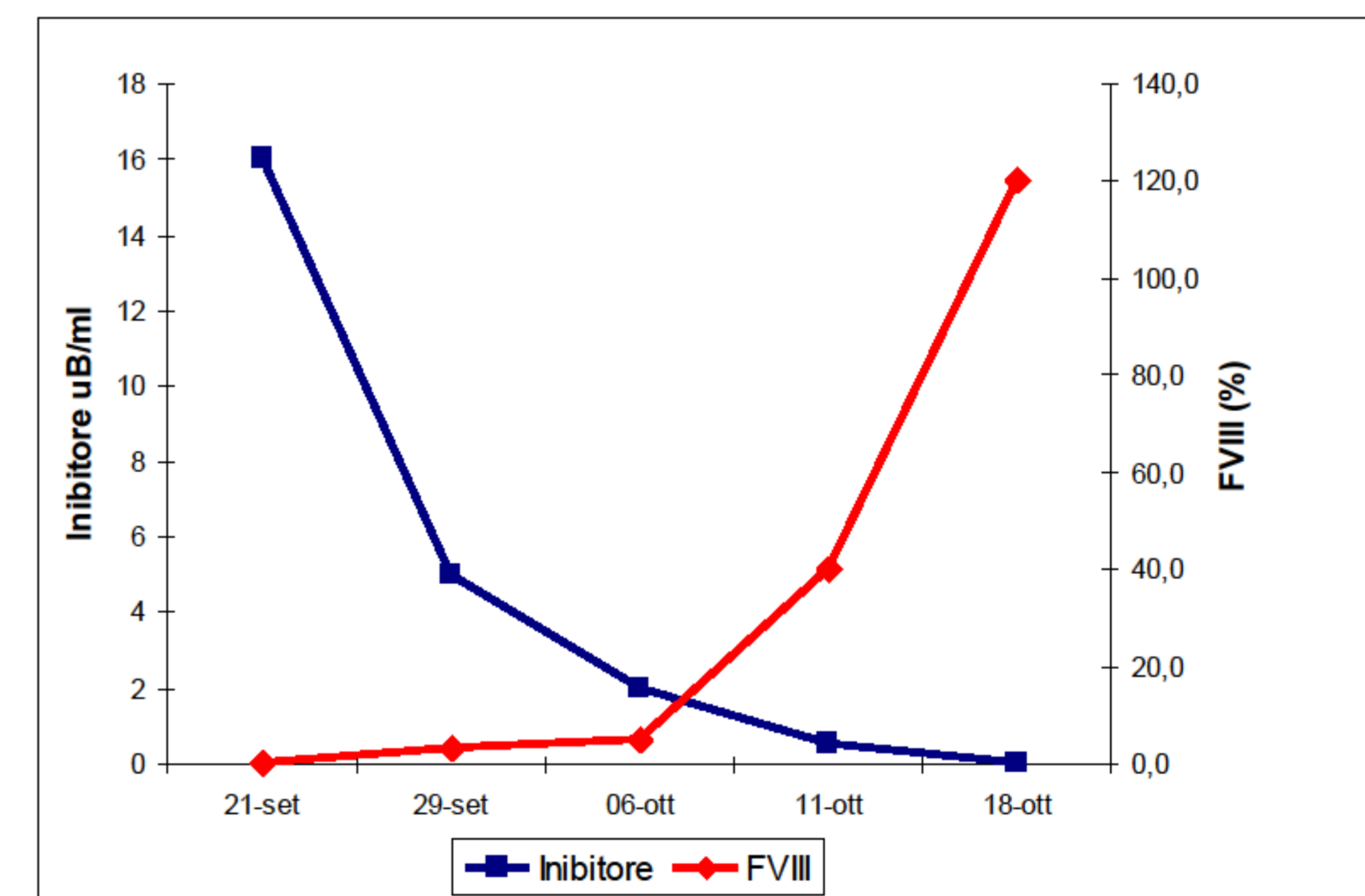
A 65 years old man, with severe carotid artery disease and previous mesenteric stenting, was admitted with severe anaemia (Hb 46 g/L) due to large bilateral haematoma of upper limbs, aPTT 60s, FVIII 10.4%, inhibitor 1 BU.

The patient was transfused with PRBC and treated with 100 IU/kg of HP VWF/FVIII for 14 days.

Three months after discharge, a malignant cancer of the lung with bone's metastasis was diagnosed.

### Patient 3

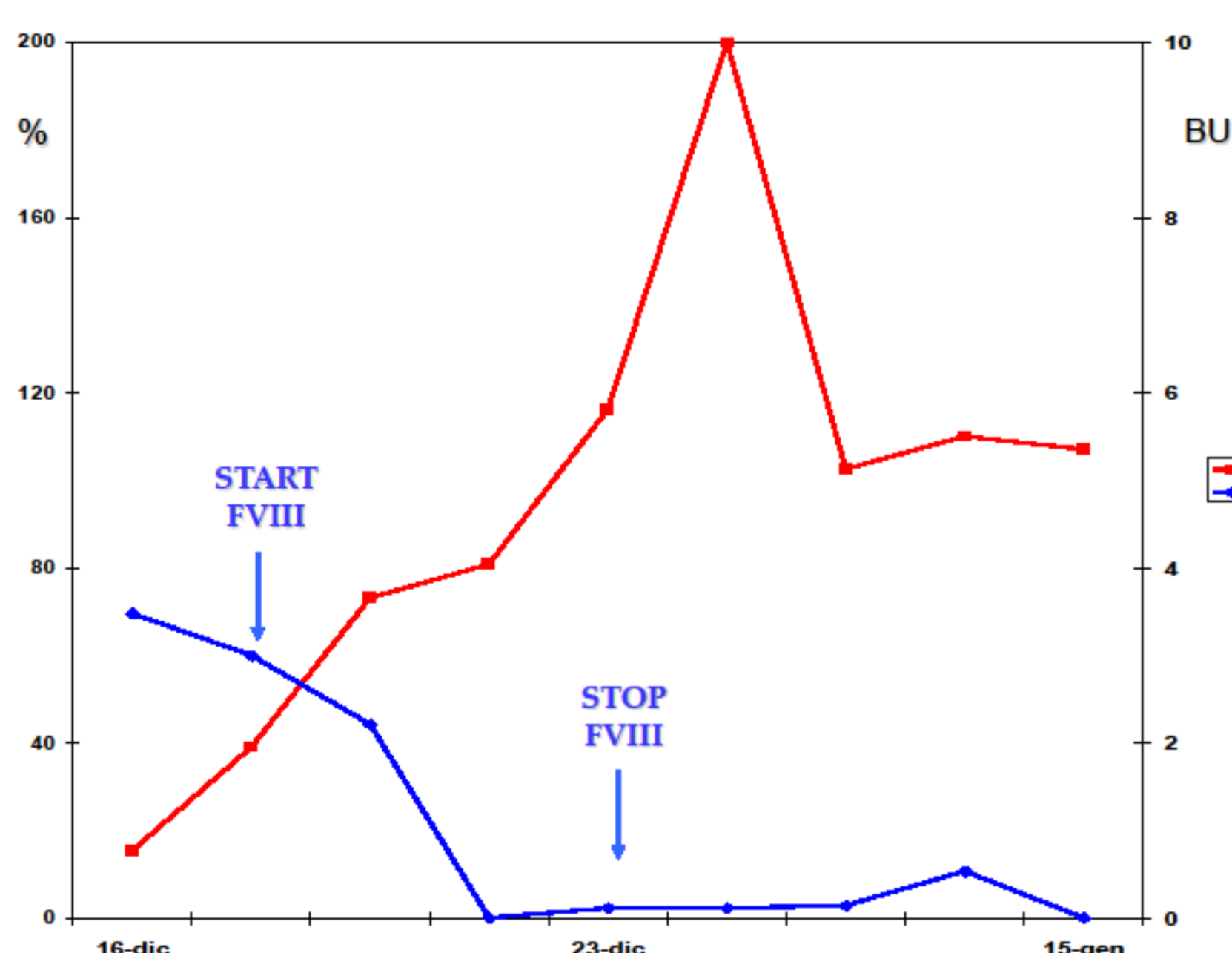
A 78 years old male, affected by severe coronary heart disease and previous carotid endarterectomy, presented for syncope and anaemia (Hb 90 g/L) due to retroperitoneal haematoma. FVIII:C was 2.4% and inhibitor 10.5 BU



The patient was treated with 3 PRBC and FVIII concentrate 300 U/kg bolus, followed by 80 U/kg/day for 7 days.

### Patient 4

A 75 years old man was admitted for a large haematoma on left calf; he had a history of severe coronary heart disease and left carotid stenting, on secondary prevention with aspirin 75 mg/day. Haemoglobin was normal, FVIII:C was 15.3% and inhibitor 3.48 BU/ml.



The patient was treated with FVIII concentrate 120 U/kg bolus, followed by 240 U/kg/day for 13 days.

## CONCLUSIONS

In all cases **haemorrhage stopped at once**, followed by progressive hematomas reabsorption. Therapy with prednisone and cyclophosphamide was started, obtaining inhibitor disappearance in approximately three weeks. **No thrombotic complications** occurred during follow-up.

Our clinical experience shows that the highly purified VWF/FVIII concentrates are effective in controlling hemorrhage in AHA patients, particularly if inhibitor titre is low, and could be considered the first line therapy in AHA patients with cardiovascular comorbidity.

## REFERENCES.

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