



DISORDINI EMORRAGICI ACQUISITI: ASPETTI
DIAGNOSTICI E CLINICI

TRANSIENT IDIOPATHIC ACQUIRED VON WILLEBRAND SYNDROME: A RARE CHALLENGING CONDITION WITH A POSSIBLE RAPIDLY AND FAVOURABLE EVOLUTION

G. Murante^{1|2|4}, S. Bergonzi^{1|2|4}, M. Capetta^{1|2|4}, F. Valeri^{1|2}, C. Dainese^{1|2}, A. Valpreda³, R. Pastorino^{1|4}, F. Bocchi^{1|4}, B. Bruno^{2|4}, A. Borchellini^{1|2}

¹Regional Centre for Hemorrhagic and Thrombotic Diseases; ²Division of Hematology; ³Central Lab. Baldi and Riberi, AOU Città della Salute e della Scienza di Torino, Italy; ⁴Dep. of Molecular Biotechnology and Health Sciences, University of Turin, Italy

Background and aims. The acquired von Willebrand syndrome (AvWS) is a rare but probably underestimated bleeding disorder characterized by laboratory findings and clinical presentations similar to those of inherited von Willebrand disease. However, unlike the inherited disease, AvWS occurs in adults with no personal or family history of bleeding diathesis. Although AvWS is particularly frequent in lymphoproliferative or myeloproliferative disorders, it can also be associated with solid tumors, immunologic and cardiovascular disorders including congenital heart defects, aortic stenosis, and the use of left ventricular assist devices, and other miscellaneous conditions. Various mechanisms are implied in the pathophysiology of AvWS, the majority of them leading to the increased degradation or clearance of circulating vWF. The aim of this abstract is to demonstrate the significance of follow-up in cases of AvWS without a recognised underlying cause, given the possibility of spontaneous clinical and laboratory resolution of the haemorrhagic condition.

Case report. Considering the aforementioned points, we present a case of a 56 years-old woman that was referred to our centre in 2024 due to multiple episodes of epistaxis and easy bruising. She exhibited negative bleeding histories even in trigger conditions (surgery, childbirth) and no family history of bleeding diathesis. No comorbidities were present at diagnosis and both autoimmune evaluation and cancer screening were negative. One year after diagnosis, a complete clinical and laboratory resolution was observed. As rare disease, the diagnostic process is complex and involves multiple steps, requiring an experienced and specialized lab-

oratory. Despite pathological laboratory results, confirming the acquired nature of this disorder remains a significant diagnostic challenge. The first step routine coagulation tests and vWF screening were performed: factor VIII (FVIII) dosage, VWF Antigen (vWF:Ag), VWF activity (VWF:Ac), VWF ristocetin cofactor activity (VWF:RCO) and collagen binding assay (CBA). Afterwards, dilution and subsequent in vivo recovery test with plasma derived von Willebrand factor concentrate were performed. A genetic test was performed. Dilution and in vivo recovery tests strongly suggested an acquired form. Genetic analysis was inconclusive. One year after diagnosis, no more antibodies were found.

Conclusions. The AvWS is a rare bleeding disorder with a diagnostic work-up and optimal treatment highly individualized. The management remains challenging, particularly when discrepancies arise between laboratory findings and the patient's medical history. For this reason, especially in cases without an underlying cause, it is important to perform close follow-up for the possibility of rapid favorable resolution of the condition.

Parameter	2024	2025
FVIII:C	35%	120%
vWF Antigen (vWF Ag)	33%	90%
vWF Ristocetin Cofactor (vWF RCo)	8%	75%
vWF Collagen Binding Activity (vWF CBA)	22%	70%