CLINICAL AND BIOLOGICAL STUDY OF OBLIGATE CARRIERS OF HEMOPHILIA A.

MILOUDI A. GUENOUNOU K. FRIGAA I.
Center of Hemobiology and Blood Transfusion, Mustapha University Hospital, Algeria .
ifrigaa@yahoo.fr

Introduction

The levels of factor VIII (FVIII) show considerable variability in women who are carriers of hemophilia A (CHA). A new nomenclature was proposed by the International Society on Thrombosis and Hemostasis (ISTH) in 2021. This nomenclature takes into account personal bleeding history and baseline plasma FVIII levels, and now, CHA with normal factor VIII levels are included in this new classification. The aim of this study is to present the clinical and biological profile of obligate carriers of hemophilia A diagnosed at CHTS Mustapha.

Patients and Methods: The study focused on 14 CHA. The concept of hemophilia was present in the families of all the patients. An ISTH-BAT bleeding score followed by a comprehensive hemostatic assessment was conducted.

Results: The interview revealed the presence of a bleeding syndrome in 6/14 (42.85%) patients, with a median ISTH-BAT bleeding score of 2, ranging from 0 to 16. The bleeding symptoms were predominantly post-traumatic hematomas and oral cavity bleeding in 4/14 (28.57%), followed by menorrhagia in 3/14 (21.42%), with a menstrual duration exceeding 10 days, bruising, post-surgical or post-dental extraction bleeding in 2/14 (14.28%), as well as postpartum hemorrhages, epistaxis, and bleeding from a superficial wound lasting more than 10 minutes in 1/14 (7.14%). The median FVIII level was 68%, with extremes ranging from 41 to 119%. FVIII levels were not correlated with bleeding, as they were normal in 3/14 (21.42%) of symptomatic patients. The study of von Willebrand factor showed levels within the reference ranges. The median VWF level was 116% (68-206%). The FVIII/VWF ratio used for diagnostic orientation was decreased in 12/14 (85.71%), confirming the carrier status.

Conclusion: Most CHA are asymptomatic, and typically, the bleeding risk is related to FVIII levels. Our study, conducted on a small population, showed a minority of symptomatic CHA despite having normal factor VIII levels. Recognizing the CHA status, as specified in the new nomenclature, aims to improve diagnosis and allows these patients to benefit from appropriate

therapeutic management based on antifibrinolytics, desmopressin, and FVIII concentrates in more severe cases.