

BLOOD COAGULATION & FIBRINOLYSIS

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The spectrum of inherited bleeding disorders in pediatrics.

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Abstract

Inherited bleeding disorders (IBDs) are caused by quantitative and qualitative alterations of either platelets or plasma proteins involved in coagulation and fibrinolysis. Hemophilias are the most frequent IBDs; however, accumulated data from various studies reported that von Willebrand disease (VWD) is the most common cause of IBD, with an increased incidence of platelet function defects, mostly due to the increased rate of consanguinity in some communities. VWD is an inherited disorder of homeostasis due to quantitative or qualitative defect of von Willebrand factor. Data on its epidemiology and impact in developing countries are limited. The objective of this study was to assess the local prevalence of some IBD and establish the clinical and historical variables that are predictive for those bleeding disorders in pediatrics. The study involved 43 children with various bleeding manifestations and 15 age- and sex-matched controls, recruited from the Pediatrics Hematology Clinic at the National Research Centre, Sausan Mubarek children's hospital in Cairo, Egypt and the King Abdulaziz University Hospital, Jeddah, Kingdom of Saudi Arabia. Hematological profile included platelet counts and function, prothrombin time, partial thromboplastin time, factor VIII antigen and its activity, factor IX antigen and its activity, von Willebrand factor and its activity assayed with multimeric analysis. A total of 12 (27.9%) children had VWD, 11 (25.5%) had hemophilia A, three (7%) had hemophilia B, seven (16.3%) had platelet dysfunction and 10 (23.3%) had bleeding with undiagnosed cause. Two of the VWD cases had type I, three had type II, four had type III and one case appeared to have type IIM and another to have IIB VWD. Bruising and epistaxis were the main symptoms in all children with VWD. The majority of platelet dysfunction disorders were diagnosed as Glanzmann's thrombasthenia. VWD and Glanzmann's thrombasthenia should be considered not uncommon causes of IBDs in children in Egypt and Kingdom of Saudi Arabia. Routine hematological screening should be mandatory in children with positive family history of bruising and bleeding as a predictor for IBD.

