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Outcomes of Congenital Bleeding Disorders

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Background: Hemophilia is an x-linked inherited bleeding disorder that requires lifelong medical support.

Objective: To evaluatethe prevalence, presentation and management of inherited bleeding disorders.

Setting: Hematology Department, King Abdulaziz University, Saudi Arabia.

Design: Retrospective study.

Method:Sixty-fourpatients withinherited bleeding disorders were included in the study from January 2000 to January 2011.Clinical, baseline coagulation screen, factor assay and serological testing were collected.

Result: Thirty-one patients (48.4%) were diagnosed with hemophilia A,eighteen patients (28.1%) had von Willebrand disease, eleven patients (17.9%) had Hemophilia B and four patients had other factor deficiencies. Elevenpatients with Hemophilia A and one patient with Hemophilia B had severe disease. The prevalence of hepatitis C virus infection was 5%. All patients were negative for hepatitis B S Ag and human immune deficiency virus antibodies.

Conclusion: The distribution of hereditary bleeding disorders reported in this study is similar to other studies; however, the prevalence of von Willebrand disease was lower than expected.We believe there is under-representation of bleeding disorders in our referral population due to lack of awareness and diagnostic expertise in remote areas of the Western region.

Furthermore, the implementation of rigorous donor screening and the adoption of nation-wide hepatitis B vaccination have successfully reduced transfusion-transmitted viral infections among Saudi patients.

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