

Acquired haemophilia A

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Acquired hemophilia A (AHA) is a rare autoimmune bleeding disorder characterized by inhibitory antibodies directed against circulating coagulation factor (F) VIII. The incidence of AHA has been estimated to be 0.2 to 1.0 per million per year. Typically, patients with no previous personal or family history of a bleeding present with spontaneous bleeds and an isolated prolonged activated partial thromboplastin time. The final diagnosis is made on the basis of low FVIII levels and the presence of FVIII inhibitor. The pattern of bleeding varies from mild bleeding requiring no treatment, to severe life-threatening hemorrhage, with 9 to 22% mortality.

The mainstays of the treatment are the management of acute bleeding and the inhibitor eradication. First-line treatment of bleeding is with a bypassing agent. Two available licensed regimens are recombinant factor VIIa and the activated prothrombin complex concentrate. The risk of severe life-threatening bleeding continues until the FVIII antibody has been eradicated, therefore immunosuppression should be commenced in all patients as soon as the diagnosis has been made. Commonly used regimens are steroids alone or steroids combined with cyclophosphamide. The anti CD20 monoclonal antibody, rituximab, has shown good results in patients who do not respond to standard immunosuppressive agents.

AHA usually presents to clinicians without prior experience of the disease, therefore it is important to increase the awareness of this disorder among health care professionals. Early recognition, rapid diagnosis and prompt referral to a specialized centre are important to optimize the treatment and improve the outcome for this patient group.

Biography

Jelena Roganovic completed her MD and Ph.D. from Rijeka University School of Medicine. She is board-certified in pediatric hematology/oncology and pediatrics. She completed her residency in pediatrics in Croatia, and a fellowship in pediatric hematology/oncology in Padua, Italy. Roganovic is the Chief of the Division of Pediatric Hematology and Oncology at Children's Hospital of Rijeka, and Full Professor of Clinical Pediatrics at the University of Rijeka. She has published numerous papers and proceedings in the field of pediatric hematology and oncology, and serves as an editorial review board member of repute.

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