

Management of acquired haemophilia A

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Summary. Acquired haemophilia A is an auto-immune disease caused by an inhibitory antibody to factor VIII. The pattern of bleeding varies but patients remain at risk of life threatening bleeding until the inhibitor has been eradicated. The cornerstones of management are; rapid and accurate diagnosis, control of bleeding, investigation for an underlying cause and eradication of the inhibitor by immunosuppression. Patients should always be managed jointly with a specialist centre even if they present without significant bleeding. Despite an extensive literature, few controlled data are available and treatment guidelines are based on expert opinion. To treat bleeds recombinant factor VIIa and activated prothrombin complex concentrate are equally efficacious but both are superior to factor VIII or desmopressin. Immunosuppression should be started as soon as the diagnosis is made. Commonly used regimens are steroids alone or combined with cytotoxic agents. Rituximab is being used more widely but current evidence does not suggest that it improves outcomes or reduces side effects.

Keywords: acquired haemophilia, bypassing agents, factorVIII, immunosuppression, inhibitor.

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