Acquired hemophilia A (AHA)

Clotting-Guide
vom Labor zur Diagnose

Description:

A very rare form of hemophilia A is caused by autoantibodies inhibiting factor VIII (acquired hemophilia). This disease occurs especially in older patients and is associated with the new onset of bleeding, mainly affecting the skin, deep muscles, soft tissues and the retroperitoneum, but hardly any joints. Bleeding can be severe and life-threatening. The management is complex and expensive and requires special knowledge.

Treatment options:

Treatment with human factor VIII concentrates is usually ineffective because they are immediately blocked by the autoantibodies. With bypass preparations, e.g. recombinant activated human factor VII (Novoseven®) or activated prothrombin complex concentrates (FEIBA®) or recombinant porcine factor VIII concentrates (Obizur®), hemostasis can be achieved in these patients, but these therapies are extremely expensive.

Emicizumab (Hemlibra®) has also been used off-label for acquired hemophilia for two years with great success. However, the causal therapy for acquired hemophilia A is immunosuppression to achieve elimination of autoantibodies.

Since the standard immunosuppresssive therapy with cortisone and cyclophosphamide for several weeks causes unacceptable toxicity and high mortality in these older patients, treatment is now mostly done with rituximab (off-label).

Despite these advances, the management of acquired hemophilia remains challenging and should only be undertaken in coordination with experienced centers.

Surveillance:

Determination of factor VIII levels with the appropriate test systems and at the appropriate time.

References:

Thomas L, Laboratory and Diagnosis, 2023, Release 5: https://www.labor-und-diagnose.de/index.html
Parameter catalog of the Clinical Institute for Laboratory Medicine, Med.Univ.Wien and AKH Vienna: https://www.akhwien.at/default.aspx?pid=3982
List of services for clinical chemistry, Univ.Klinikum Ulm: https://www.uniklinik-ulm.de/zentrale-einrichtung-klinische-chemie/leistungskatalog.html