

Immune-Coagulopathy



Description:

In immun-coagulopathies, autoantibodies influence the function of one or more coagulation factors. Depending on the clotting factor inhibited, its residual activity and the strength of the binding, there can be varying degrees of bleeding:

Factor XII, XI: common, after infections; no tendency to bleed

Factor VIII, IX: rare, see text for acquired hemophilia; severe bleeding possible, dependent on residual factor activity

Factor X: very rare; severe bleeding possible, dependent on residual factor activity

Factor V: very rare; severe bleeding possible, dependent on residual factor activity

Factor II: very rare; severe bleeding possible, dependent on residual factor activity

Factor VII: very rare; no? bleeding tendency

Factor XIII: very rare; variable bleeding tendency

Factor VWF: very rare; variable bleeding tendency

Treatment options:

- *Reduction of bleeding tendency:* coagulation factor concentrates are usually ineffective at high inhibitor titres. Therefore, the only option is often rhFVIIa (Novoseven®). Dose 45-90 mcg/kg every 2-4 hours, according to the product information!

- *Prevention of antibody formation:* today almost only steroids and (off-label) rituximab are used as immunosuppression.

Surveillance:

Determination of the activity of the respective coagulation factor

Quantification of inhibitor titer using the Bethesda assay (and modifications thereof).

For questions contact a coagulation specialist.

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>