Disturbance in the intrinsic coagulation system (contact activation)

Clotting-Guide
vom Labor zur Diagnose

Description:

In addition to high molecular weight kininogen (HMWK) and prekallikrein (PKK), contact activation includes the coagulation factors XII, XI, VIII and IX. All of these factors are captured by the APTT, but not by the PT. A deficiency of HMWK, PKK or factor XII never causes bleeding, even if the APTT is sometimes very prolonged. A deficiency of factor XI can cause mild bleeding. Deficiencies of factor VIII (hemophilia A) or IX (hemophilia B) can cause mild to severe spontaneous and postoperative bleeding, depending on the severity (see text on hemophilia).

The causes of an intrinsic factor deficiency are either genetic (e.g. congenital hemophilia) or acquired, e.g. through blocking autoantibodies (see also text on acquired hemophilia or immune coagulopathies). Antibodies against factors XII and XI are very common in response to viral infections and have no clinical relevance.

Treatment options:

No therapy necessary for HMWK, PKK or factor XII deficiency.

In the case of congenital deficiency of factors XI, VIII, IX: substitution with recombinant or plasmatic factor concentrates (see text on hemophilia) or plasma (F XI).

For autoimmune-related factor deficiencies: no need for therapy for factor XI deficiency. For treatment of IX and VIII deficiency, see text on acquired hemophilia.

Surveillance:

APTT
specific coagulation factor activities
In case of questions please 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