Liver diseases

Clotting-Guide
vom Labor zur Diagnose

Description:

Almost all coagulation factors are synthesized in the liver, except VWF and factor VIII, which are also synthesized in endothelial cells. For the synthesis of some factors (factors VII, IX, X, II, protein C, protein S), vitamin K is necessary to enable the gamma-carboxylation of these proteins. The gamma-carboxyl groups are necessary for the binding to phospholipids via calcium ions. If they are missing, the hemostatic function is significantly impaired.

To distinguish synthesis disorder from vitamin K deficiency, either a diagnostic administration of vitamin K or single coagulation factor analysis including non-vitamin K dependent factors (e.g. factor V or antithrombin) can be used.

A synthesis disorder can be detected with PT, because this assay is primarily dependent on factor VII activity (due to the short half-life of factor VII - 6h). If the synthesis disorder is more pronounced, the APTT can also be prolonged and fibrinogen reduced.

Treatment options:

If necessary (e.g. in the case of acute bleeding), substitution can be made with prothrombin complex concentrates (PCC; various manufacturers). A dose of 50 U/kg/d is usually sufficient. One unit of PCC per kg of body weight increases the PT by 1%.

Fibrinogen deficiency can be treated with fibrinogen concentrates (Hemocomplettan®, Fibryga®) in a dose of 1-2 g/d.

If necessary, antithrombin can be replaced with antithrombin concentrates (various manufacturers). Antithrombin substitution should only be carried out in the case of relative antithrombin deficiency (antithrombin significantly lower than the PT), DIC, thrombosis or heparin resistancy.

In severe liver failure, replacing all proteins is best done with plasma infusions. Since large amounts of plasma are necessary to achieve relevant coagulation effects (20-40 mL/kg), plasma exchange often necessary.

The information in the product information must be observed!

Surveillance:

Measurement of the PT, in acute bleeding this should be over 50%.
The fibrinogen level should be kept above 1 g/L, and in acute bleeding above 1.5-2 g/L.
Antithrombin should be kept in balance with the PT (%).
If you have any questions or are unclear, you should contact a specialist coagulation department.

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