Dysfibrinogenemia or Afibrinogenemia

Description:

Fibrinogen dysfunction or fibrinogen deficiency can occur due to mutations in the fibrinogen genes. Depending on the mutation, different functionalities of fibrinogen can be disturbed. As a result, the functional fibrinogen concentration (measurement according to Clauss) is reduced, but an immunological determination of the fibrinogen concentration can be normal.

Clinically, there is a more or less pronounced risk of bleeding. However, some forms are also associated with an increased risk of arterial and/or venous thrombosis and embolism.

Treatment options:

Fibrinogen deficiency with bleeding can be treated with fibrinogen concentrates (Hemocomplettan®, Fibryga®) in a dose of 1-4 g/d.

In milder forms, symptomatic improvement of bleeding can also be achieved with tranexamic acid (Cyklokapron®, 1000 mg po or iv 3 times daily).

The informations in the product information must be observed!

Surveillance:

Measurement of fibrinogen according to Clauss and with immunological methods.

For 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