Lupus anticoagulant

Clotting-Guide
vom Labor zur Diagnose

Description:

Lupus anticoagulants are immune disorders in which autoantibodies against phospholipid-bound proteins (annexin V, prothrombin, etc.) cause an apparent prolongation of phospholipid-dependent coagulation tests. They belong to the so-called anti-phospholipid antibodies, as well as anti-cardiolipin antibodies and antibodies against beta-2-glycoprotein 1.

Despite a prolonged APTT, lupus anticoagulants do not cause bleeding. Only when associated with antibodies against prothrombin (lupus anticoagulant-hypoprothrombinemia syndrome, here the PT is also pathological) or against platelets (lupus anticoagulants with immune thrombocytopenia), bleeding can occur.

Lupus anticoagulants often arise after infections or in association with other autoimmune diseases (lupus erythematosus, etc.), pregnancy, malignancies, etc. These “secondary” lupus anticoagulants usually have no clinical relevance and disappear again after the triggering cause has been treated.

However, lupus anticoagulants can also occur spontaneously and can then be associated with a significant risk of arterial and venous thromboembolism, recurrent abortions and pregnancy complications (antiphospholipid antibody syndrome, APLAS). Arterial thromboembolism in particular can cause severe impairment of the affected patient.

Catastrophic APLAS (CAPS), which is associated with extensive thrombosis in small vessels and severe organ damage and has a high mortality, is rare. There is overlap with disseminated intravascular coagulopathy (DIC), heparin-associated thrombocytopenia (HIT), and thrombotic microangiopathies (TMA).

Treatment options:

Secondary lupus anticoagulants do not require treatment.

Spontaneous lupus anticoagulants without thrombosis: no therapy, only monitoring, and conventional thrombosis prophylaxis in risk situations.

Lupus anticoagulants in patients with thrombosis or embolism: anticoagulation with vitamin K antagonists (DOAC only in exceptional cases).

Lupus anticoagulants in women with recurrent abortions: aspirin and/or low molecular weight. Heparin throughout pregnancy (but little evidence for this).

In case of CAPS: immunosuppression, anticoagulation, plasma exchange, complement inhibition, intensive care treatment, as required.

Diagnostics and monitoring:

Lupus-sensitive APTT (depending on the phospholipids used in the test system, some APTT assays are very sensitive to lupus anticoagulants, e.g. APTT-LA, moderately sensitive, e.g. APTT-STA, or insensitive, e.g. APTT-FS).
diluted Russell's Viper Venom Time (dRVVT).

The PT is usually normal (except in the lupus anticoagulant - hypoprothrombinemia syndrome)
The diagnosis of lupus anticoagulants requires the following criteria:

- Prolongation of more than 1 phospholipid-dependent coagulation tests (mostly APTT-LA and dRVVT)
- No therapy with anticoagulants
- No deficiency of clotting factors
- Signs of inhibition in the plasma mixing study
- Confirmation by a confirmatory test (normalization of clotting time after addition of excess phospholipids)
- Persistence of the abnormalities after repeating the laboratory diagnosis after 3 months

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
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