Antiphospholipid syndrome

- Antiphospholipid antibodies are a heterogeneous group of autoantibodies directed against phospholipid binding proteins.
- Antiphospholipid antibodies can be broadly categorized into:
  1. those antibodies that prolong phospholipid-dependent coagulation assays, known as lupus anticoagulants (LA), or
  2. anticardiolipin antibodies (aCL).
- The presence of these antibodies in patients with arterial or venous thrombosis or pregnancy morbidity comprises the antiphospholipid antibody syndrome (APS).
- This syndrome is referred to as:
  1. primary APS when it occurs alone and
  2. secondary APS when it occurs in association with other conditions, such as systemic lupus erythematosus (SLE).
- Antiphospholipid antibodies are also found in patients with infections such as human immunodeficiency virus (HIV) and may develop during therapy with medications such as chlorpromazine. Their clinical importance in these settings is unknown.

**General**

**Lupus anticoagulants**

- Lupus anticoagulants are antibodies that block phospholipid surfaces important for coagulation.
  1. increase APTT
  2. prolonged APTT does not correct with a 1:1 mix with normal platelet-free plasma
  3. correction of the clotting time after addition of excess phospholipids confirms the presence of LA.

- Consensus guidelines recommend screening for LA with 2 or more phospholipid-dependent coagulation tests, including the:
  1. activated partial thromboplastin time
  2. dilute Russell viper venom time
  3. kaolin clotting time
  4. dilute prothrombin time
  5. textarin time
  6. talpin time

**Anticardiolipin antibodies**

- Anticardiolipin antibodies share a common in vitro binding affinity for cardiolipin and can be detected using ELISA.

**Diagnostic criteria for antiphospholipid syndrome**

- According to the Sapporo criteria, APS is present in patients with 1 clinical and 1 laboratory criterion.
- Clinical criteria include:
  1. objectively confirmed arterial, venous, or small-vessel thrombosis,
  2. pregnancy morbidity consisting of recurrent fetal loss before the 10th week of gestation, 1 or more unexplained fetal death at or beyond the 10th week of gestation, or premature birth due to placental insufficiency, eclampsia, or preeclampsia.
- Laboratory criteria include:
  1. medium or high titer IgG or IgM aCL or
  2. the presence of LA on 2 or more occasions at least 6 weeks apart.

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**Treatment**

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