Learn simply

Antiphospholipid Antibody Syndrome

Suspect the diagnosis of antiphospholipid antibody syndrome (APLAS)[1]

Confirm the diagnosis[2]

Correct clinical setting[3]
- Recurrent pregnancy loss
- Unexplained venous or arterial thrombosis
- Autoimmune thrombocytopenia
- Unexplained IUGR
- Placental abruption
- Recurrent early severe preeclampsia

Confirmatory serologic test[4,5]
- Anticardiolipin antibody (ACA)
- Lupus anticoagulant (LAC)
- Anti-β2-glycoprotein I antibodies

No / Yes

Yes / No

AND

Diagnosis of APLAS cannot be confirmed; consider an alternative diagnosis

The diagnosis of APLAS is confirmed

Diagnosis of APLAS cannot be confirmed; consider an alternative diagnosis

Treatment depends on the clinical features[6]

Recurrent pregnancy loss	Autoimmune thrombocytopenia	Unexplained venous or arterial thrombosis
Anticoagulate during pregnancy (not postpartum or lifelong)	*Recommendations unclear (consider anticoagulation in pregnancy)*	*Anticoagulate during pregnancy and for at least 6 weeks postpartum; consider lifelong anticoagulation*

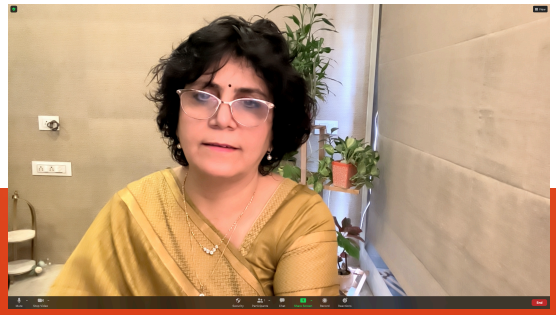
1. Antiphospholipid antibody syndrome (APLAS) is an autoimmune disease characterized by the presence in the maternal circulation of one or more autoantibodies against membrane phospholipid as well as one or more specific clinical syndromes. It is an acquired rather than an inherited condition. As such, it cannot explain a family history of venous thromboembolism (VTE). A significant family history of VTE should prompt testing to exclude inherited thrombophilias, including factor V Leiden mutation, prothrombin gene mutation, and protein S, protein C, and antithrombin deficiency.
2. The diagnosis of APLAS requires two distinct elements: (i) the correct clinical setting; and (ii) confirmatory serologic testing. Approximately 2–4% of healthy pregnant women will have circulating antiphospholipid antibodies in the absence of any clinical symptoms. As such, routine screening for these antibodies in all pregnant women is strongly discouraged.
3. Clinical manifestations of APLAS include: (i) recurrent pregnancy loss (defined as ≥ 3 unexplained first-trimester pregnancy losses or ≥ 1 unexplained second-trimester pregnancy loss); (ii) unexplained thrombosis (venous, arterial, cerebrovascular accident or myocardial infarction); and/or (iii) autoimmune thrombocytopenia (platelets <100,000/mm3). Recent consensus opinions suggest that such clinical conditions as unexplained intrauterine growth restriction (IUGR), massive placental abruption, and recurrent early-onset severe pre-eclampsia be included.
4. At least one of three serologic tests confirming the presence of circulating antiphospholipid antibodies (below) is required to make the diagnosis of APLAS. Moreover, the diagnosis requires the persistence of such antibodies as confirmed by two or more positive tests at least 12 weeks apart.
5. Lupus anticoagulant (LAC) is an unidentified antiphospholipid antibody (or antibodies) that causes prolongation of phospholipid-dependent coagulation tests in vitro by binding to the prothrombin–activator complex. Examples of tests that can confirm the presence of LAC include the activated PTT test, dilute Russel viper venom test, kaolin clotting time, and recalcification time. In vivo, however, LAC causes thrombosis. LAC results are reported as present or absent (no titers are given). The term LAC is a misnomer: it is not specific to lupus (SLE) and it acts in vivo as a procoagulant and not an anticoagulant.

1. Treatment for APLAS depends on the clinical features:

2. For women with thrombosis (such as stroke or pulmonary embolism), therapeutic anticoagulation is indicated with either unfractionated heparin (UFH) or low molecular weight heparin (LMWH) during pregnancy followed by oral anticoagulation (coumadin) postpartum because of a 5–15% risk of recurrence. In pregnancy, regular blood tests are required 4 hours after administration of the drug to ensure that anticoagulation is therapeutic: the PTT should be 1.5- to 2.5-fold normal and anti-Xa activity levels should be 0.6–1.0 U/mL. Side-effects include hemorrhage, thrombocytopenia, and osteopenia and fractures. Such women may need lifelong treatment.

3. For women with recurrent pregnancy loss, treatment should include prophylactic UFH (5000–10,000 units sc bid) or LMWH (enoxaparin (Lovenox) 30–40 mg sc daily or dalteparin (Fragmin) 2500–5000 U sc daily) starting in the first trimester of pregnancy. Although prophylactic dosing does not change PTT, it will increase anti-Xa activity to 0.1–0.2 U/mL. However, it is not necessary to follow serial anti-Xa activity in such patients. The goal of this treatment is to prevent pregnancy loss and to prevent VTE, which is possible in women with APLAS in pregnancy even if they have not had a VTE in the past. Therefore, anticoagulation should be administered throughout pregnancy and typically for 6–12 weeks after delivery.

4. For women with autoimmune thrombocytopenia or a history of severe pre-eclampsia, IUGR or placental abruption, the optimal treatment is unknown. Consider treating as for recurrent pregnancy loss. Postpartum anticoagulation is probably not necessary.



KAWITA BAPAT