

Managing antiphospholipid syndrome

This is the lay version of the EULAR recommendations for the management of antiphospholipid syndrome in adults. The original publication can be downloaded from the EULAR website: www.eular.org.

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Introduction

EULAR recommendations give advice to doctors, nurses and patients about the best way to treat and manage diseases. For the first time, EULAR has developed recommendations on the management of adults with antiphospholipid syndrome.

Expert physicians, health professionals and patients worked together to develop these recommendations. Patients in the team ensured that the patient point of view was integrated in the recommendations. The authors performed a systematic literature review of the evidence for different treatments in adults with antiphospholipid syndrome.

What do we already know?

Antiphospholipid syndrome is a rare autoimmune disorder that causes inflammation and blood clots. The syndrome affects women more frequently than men. It is associated with the presence of at least one of three antiphospholipid antibodies in the blood. These three types of antibodies are called 1) lupus anticoagulant, 2) anticardiolipin antibodies and 3) antiphospholipid antibodies. Antiphospholipid syndrome can be split into different risk categories. These are based on the levels of antibodies in your blood. Having high levels – or having two or three types of the different antiphospholipid antibodies – would put you in the high-risk category.

People with antiphospholipid syndrome are at higher risk of having a stroke or heart attack, or suffering from deep vein thrombosis (often shortened to DVT). Pregnant women with antiphospholipid syndrome have an increased risk of pregnancy complications. These complications include early miscarriage, loss of the baby in later stages of pregnancy, and preterm birth.

What do the recommendations say?

Overall, there are three general principles and 12 recommendations. The general principles say that it is important to identify factors that put people at high risk of having blood clots or pregnancy complications. They also suggest that people with antiphospholipid syndrome should follow general guidelines to prevent cardiovascular disease, and that everyone should receive education and counselling about the importance of taking their medicine as prescribed, as well as advice to help them stay fit and healthy.

Each recommendation is based on available scientific evidence or expert opinion. The more stars a recommendation has the stronger the evidence is.

One star (*) means it is a weak recommendation with limited scientific evidence.

Two stars (**) means it is a weak recommendation with some scientific evidence.

Three stars (***) means it is a strong recommendation with quite a lot of scientific evidence.

Four stars (****) means it is a strong recommendation supported with a lot of scientific evidence.

- People with a high-risk antiphospholipid antibody profile should take low-dose aspirin even if they do not have any symptoms.*****

In people with a high-risk antiphospholipid antibody profile, the risk of having a first blood clot is halved by taking low-dose aspirin (75–100 mg daily).
- In people with Lupus and no history of blood clots or pregnancy complications, use of low-dose aspirin depends on their risk profile.**/*****

High-risk profile: Low-dose aspirin (75–100 mg daily) is recommended for people with Lupus and a high-risk antiphospholipid antibody profile.

Low-risk profile: Low-dose aspirin (75–100 mg daily) can also be considered in people with Lupus and a low-risk antiphospholipid antibody profile.
- For women with a history of antiphospholipid syndrome during pregnancy, low-dose aspirin treatment is recommended once the pregnancy has come to an end.*****

The use of low-dose aspirin (75–100 mg daily) will depend on your antiphospholipid antibody profile, and your general cardiovascular risk.
- In people with definite antiphospholipid syndrome and a first blood clot in a vein, treatment with an anticoagulant called a vitamin K antagonist is recommended.******

Vitamin K antagonists are recommended to treat blood clots in people with antiphospholipid syndrome. In people with clotting problems despite good adherence to a vitamin K antagonist, a class of drugs called *direct oral anticoagulants* can be considered instead. Direct oral anticoagulants can also be used in people with an allergy or intolerance to vitamin K antagonists.

Triple aPL positivity: However, a type of direct oral anticoagulant called rivaroxaban should not be used in people who have all three kinds of antiphospholipid antibodies (often called triple aPL positivity).
- In people with definite antiphospholipid syndrome and repeated blood clots in veins despite treatment with a vitamin K antagonist, monitoring or changing treatment may be needed.***

Monitoring may be needed to make sure that the vitamin K antagonist is being taken properly, and that blood clotting targets are met. If targets are met, adding low-dose aspirin or changing treatment to a low molecular weight heparin (sometimes shortened to LMWH) could also be considered to help prevent blood clots.
- In people with definite antiphospholipid syndrome and a first blood clot in an artery, treatment with a vitamin K antagonist is recommended.****

Clots in an artery are more likely to come back. Vitamin K antagonists are recommended in people with a blood clot in an artery.

Triple aPL positivity: Rivaroxaban should not be used in people with triple aPL positivity and arterial thrombosis.
- In people with blood clots in their arteries that have come back despite taking a vitamin K antagonist, adding low-dose aspirin or changing to LMWH may be considered.***

Other drugs such as anti-malarials or statins could also be considered.
- In women with a high-risk antiphospholipid antibody profile but no history of blood clots or pregnancy complications, low-dose aspirin during pregnancy should be considered.***

This recommendation applies to women with or without Lupus. Low-dose aspirin can help to prevent pregnancy complications and should be given at a dose of 75–100 mg per day.

- In women with a history of antiphospholipid syndrome during pregnancy only (no prior clots), low-dose aspirin on its own or in combination with heparin might be needed depending on previous pregnancy complications.*****

Three or more miscarriages: Women who have had three or more spontaneous miscarriages before the 10th week should receive low-dose aspirin and heparin at a prophylactic dose during pregnancy.

Fetal loss at or after the 10th week: Low-dose aspirin and heparin at a prophylactic dose during pregnancy is recommended.

Preterm birth: Low-dose aspirin on its own or in combination with heparin is recommended for women who have had a baby before the 34th week of pregnancy because of eclampsia or severe pre-eclampsia, or placental insufficiency.

Two miscarriages, or delivery after 34 weeks due to severe pre-eclampsia or eclampsia: Low-dose aspirin either alone or in combination with heparin might be considered for women who have had two recurrent spontaneous miscarriages before the 10th week or delivered a baby after 34 weeks due to eclampsia or severe pre-eclampsia (based mainly on expert's opinion).

After delivery: Women who receive heparin during pregnancy should continue the dose for 6 weeks after the baby is born to reduce the risk of blood clots.
- Women who have repeated pregnancy complications despite treatment with low-dose aspirin and heparin can consider increasing the heparin dose or adding a different medicine.***

An increased heparin dose, the addition of hydroxychloroquine during pregnancy, or addition of low-dose prednisolone in the first trimester might be considered for women who keep having pregnancy complications despite taking low-dose aspirin.
- Women with a history of blood clots due to antiphospholipid syndrome should receive higher dose (therapeutic dose) of heparin during pregnancy.****

Women who are taking a vitamin K antagonist for their antiphospholipid syndrome should switch to heparin as soon as their pregnancy is confirmed, and ideally before week 6.
- In people with antiphospholipid antibodies, any infections should be treated quickly, and interruptions in anticoagulation medicine should be minimised.***

This can help to prevent the development of catastrophic antiphospholipid syndrome (often shortened to CAPS). CAPS can happen when people stop taking their anticoagulation treatment, have infections, or after surgery.

Summary

Overall, the recommendations highlight the general management for people with antiphospholipid syndrome. If you have this condition, these recommendations will give you some guidance on what to expect from your doctor and what treatments you may be offered.

Recommendations with just one or two stars are based mainly on expert opinion and not backed up by appropriate clinical studies. They may be as important as those with three or four stars.

If you have any questions or concerns about your disease or your medication, you should speak to a health professional involved in your care.