

Is the oral contraceptive or hormone replacement safe?

The use of the oral contraceptive pill (OCP) or hormone replacement therapy (HRT) is associated with a small increase in the risk of thrombosis in everybody. This risk is increased if you have Protein C deficiency. We don't advise the use of the pill or HRT if you have Protein C deficiency.

What is the risk in pregnancy?

Pregnancy itself carries a small increase in the risk of thrombosis. If you have Protein C deficiency we recommend treatment with low dose heparin during and for a few weeks after pregnancy.

Royal United Hospital Bath

Department of Haematology

Protein C Deficiency Information for Patients

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What is Protein C deficiency?

Protein C is a natural anticoagulant. This is part of the body's safety mechanism to stop clots forming where they are not needed. When there is a deficiency of Protein C, the natural balance between clotting and protective anti clotting is changed. This increases the risk of thrombosis. Inheritance of Protein C deficiency is rare but important to recognise as it is one of the thrombophilias with the greatest risk of thrombosis.

How is Protein C deficiency inherited?

We have two Protein C genes – one from our mother and one from our father. Therefore there are three possibilities:

- We inherit normal Protein C genes from each parent (no increased risk of thrombosis)
- We inherit one normal gene from one parent and one abnormal from the other. This is known as being *heterozygous* and the Protein C levels in our body will be approximately half normal.
- If we inherit the abnormal gene from both our parents we are *homozygous* and our Protein C levels will be very low.

Therefore if a parent is *heterozygous* they will have one abnormal and one normal gene. This means that their children will have a 50% chance of inheriting the abnormal gene.

If both Protein C genes are abnormal the patient is *homozygous* and one copy of the abnormal gene will be passed on to all their children. This is associated with a significant risk of thrombosis and presents at birth or soon after.

Why is Protein C deficiency important?

Protein C deficiency is associated with an increase in the risk of venous thrombosis (clots in the veins), especially in the legs. There is also a lesser association with arterial clots at a young age. Low levels of protein C can be found in association with some disorders such as liver disease and diabetes. Treatment with the anti clotting drug Warfarin can also give low results on testing and so we need to know about your medical history and medications when looking at the results of Protein C tests.

Treatment

Your doctor can advise you about ways to reduce the risk of thrombosis, such as during long haul flights. Should you require a surgical procedure or long stay in hospital you may need low dose heparin injections to prevent clots. If you develop a thrombosis with Protein C deficiency we may recommend lifelong Warfarin.

Should my Family be tested?

Since Protein C deficiency has a tendency to run in families, we usually recommend that if one member of a family is found to have Protein C deficiency, other members of the immediate family should be tested. Although specific treatment is not required in affected family members, simple precautions in situations of increased risk are advised.