Protein S Deficiency

What is protein S?
This is the name given to one of the natural anticoagulants found in the blood.

Why is protein S important?
If there is not enough protein S in the blood or it does not work properly then clots may be more likely to form in veins (e.g. Deep vein thrombosis, pulmonary embolism).

How does the deficiency of protein S come about?
Usually, by the inheritance from either your mother or father, although it can rarely occur in association with a number of medical conditions such as kidney disease. Your doctor will exclude other causes before deciding that you have an inherited deficiency. The diagnosis is definite if more than one member of a family is shown to have a low level of protein S.

When is thrombosis likely to occur?
It can occur at any time although it is rare in childhood. The degree of risk increases with age.

What is the risk of thrombosis with protein S deficiency?
There is an increased risk of thrombosis in the veins in individuals with protein S deficiency. However the level of risk is also dependent on whether or not there are additional risk factors present.

Factor which increase the risk include:
- Being overweight
- Smoking
- Prolonged immobilisation
- Having an operation
- After an accident
- The combined oral contraceptive pill
- Hormone replacement therapy
- Severe illness such as cancer
- Presence of another inherited thrombotic tendency.
Pregnancy
There is growing evidence that protein S deficiency may be associated with obstetric complications such as recurrent miscarriages or stillbirth. Your doctor will advise you about this and be able to discuss possible treatments.

Having found out that I have protein S deficiency, what should I do?
The first thing to say is that you should not see yourself as ill or “abnormal”. Your thrombotic tendency may not cause you any problems at all but you can take steps to reduce the risk of thrombosis in the future by doing the following:

- Lead a physically active life.
- Eat a healthy diet and avoid becoming overweight.
- Avoid smoking.
- Avoid prolonged immobility during long journeys.
- Take advice from your doctor if you are planning a long aeroplane flight (longer than 8 hours).
- Avoid certain types of contraceptive pill.
- Take advice before starting hormone replacement therapy.
- Liaise with your doctor if entering a high risk period for clotting such as major surgery, immobility or pregnancy. Your doctor will advise you if you need special hosiery or medication.

Should other family members be tested?
Guidelines recommend that testing for heritable thrombophilia is not indicated in unselected patients presenting with venous thrombosis. Testing of selected patients may be helpful in long term management.

If you have any questions about the information in this leaflet, please contact:

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