



Freiburg Medical Laboratory ME LLC, P.O.Box 3068, Dubai

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Protein C

General:

Protein C is a glycoprotein with a molecular weight of 62,000 which circulates as an inactive precursor in plasma. Its formation is vitamin K dependent. Protein C is activated by splitting the heavy chain of protein C by thrombin. In its active form, protein C has an anticoagulatory and profibrinolytic effect by impeding the coagulation cascade through proteolytic splitting of factors Va and VIIIa. Plasminogen activator inhibitor is inhibited as well. Active protein C needs protein S as a cofactor. Deficiency of protein C (or protein S) is associated with thrombotic events, however please note that factor V (APC resistance) and factor II mutations are the most common cause of inherited thrombosis risks

Protein C deficiency: The Protein C gene is located on chromosome 2q13- q14. To date over 200 different pathogenic mutations are described which were found to be associated with protein C deficiency. Carriers of homozygous mutations are severely affected and suffer from a high intrauterine or postnatal mortality. There are mainly two types of protein C deficiency:

Type-I: Activity and concentration of protein C are decreased.

Type-II: Despite normal or slightly decreased protein C concentration, the activity is reduced.

The classification requires – additionally to the protein C determination – a determination of the antigen concentration of the protein C. Protein C deficiency can either be hereditary or acquired e.g. through liver disease, vitamin K deficiency, asparaginase therapy, consumption coagulopathy or renal insufficiency.

Hereditary protein C deficiency:

Homozygous form (protein C: <1%): purpura fulminans, heavy thromboses – also arterial, peripartal infarction.

Heterozygous form (protein C: <60%)

Acquired protein C deficiency:

In oral anticoagulation, vitamin K deficiency, alimentary, recent thromboembolism, liver dysfunction, liver cirrhosis, acute and chronic hepatitis, disseminated intravascular coagulation, meningococcal, pneumococcal sepsis, salmonella, chickenpox infection, Crohn's disease, acute ulcerative colitis.

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The following tests are available:

 Indication: screening for thrombosis risk (intake of contraceptives, unclear thromboembolism), suspicion of protein C deficiency Material: 3 ml citrate blood, frozen (send immediately) Preanalytics: citrate plasma: 1 part citrate + 9 parts blood. Protein C is unstable. Plasma supernatant must be dispatched frozen! TAT: 7-10 days* Method: PHO Units: % Ref range: see report Note: In order to exclude a protein C deficiency the examinations should be carried out approx. 2 weeks after stopping coumarin (or warfarin) therapy Protein C antigen, total Indication: clarification of protein C deficiency type 1 Material: 3 ml citrate plasma, Frozen TAT: 7-10 days* Method: IMTD Units: % Ref range: 70 -127 Protein C, immunological assay Indication: Differentiation between type I and type II of a hereditary protein C deficiency Material: 2 ml citrate plasma, Frozen TAT: 7-10 days* Method: COAG Units: mg/I Ref range: 2.0 - 3.75 	Protein C activity	
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• Protein C activated resistance, APC

General:

APC resistance is associated with an increased risk of thrombosis. The common factor V Leiden mutation is the main cause (>97%) of APC resistance as it modifies the factor V splitting site of activated protein C.

- Indication: Differentiation of venous thromboembolism, familial thrombosis
 - Material: 3 ml citrated plasma, Frozen
 - TAT: 7-10 days*
 - Method: Coagulation
 - Units: ratio
- Ref.- range: >3.0
 - Note: We strongly recommend to always perform Factor V mutation testing as it is more independent of pre-analytical influences. PTT prolongations can falsify the result, a clarification of aPTT prolongations must be clarified prior to APC testing. Coumarin and anticoagulatory treatment influence the test. In case of pathological values the detection of the factor V "Leiden" mutation should be carried out additionally (*see* Factor 5 Leiden mutation, Proaccelerin)

For complete list of laboratory test offered at Freiburg Medical Laboratory, please visit http://www.fml-dubai.com/parameter-listings/

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