

Epidemiology of Protein C Deficiency:

Protein C deficiency is present in approximately 0.2% of the general population.

Risks of Protein C Deficiency:

The overall estimated incidence (annual occurrence) of deep venous thrombosis is 1 episode for every 1000 persons. This figure does not separate patients who had predisposing conditions from those who do not.

For patients that are born with both of the copies of the protein C gene abnormal (called homozygous), the result is often a severe form of thrombosis called purpura fulminans. Purpura fulminans involves severe clotting throughout much of the body, ultimately causing death to the tissues. This is a life-threatening condition.

At this time, the data available do not suggest any role between protein C deficiency and arterial thrombosis (stroke, heart attack).

Treatment of Protein C Deficiency:

Treatment of a patient with protein C deficiency depends upon the individual patient's risk of thromboembolic disease. When a patient has a venous clot, regardless of what thrombophilic state(s) they may have, they will receive anticoagulation. This is accomplished by several different medications: 1) heparin, 2) warfarin and 3) low-molecular-weight heparins. These medications are generally continued for 3-6 months.

Patients that have had multiple thromboembolic episodes or are at high risk of further episodes (for example, multiple deficiencies) may be considered for long-term oral anticoagulation (warfarin). Because studies have demonstrated an increased risk of recurrent venous thromboembolic disease in patients with protein C deficiency, long-term oral anticoagulation is recommended. Long-term anticoagulation has risks associated with it (approximately a 3% chance per year of having a major hemorrhage, of which approximately 1/5 are fatal). Beginning long-term anticoagulation is influenced by the patient's overall risk of recurrent thrombosis balanced against the risks associated with long-term anticoagulation on an individual basis.

In patients who are homozygous, the risk of death from thrombosis is imminent. As a result, treatment necessitates providing a source of protein C. This can be done through blood products such as fresh frozen plasma (FFP). In addition, a form of human protein C concentrates has become available in the United States. This has been used previously in

Europe. At this time, there are no studies comparing the efficacy of FFP versus the protein C concentrates in severe protein C deficiency related thrombosis.

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