Protein C or S Deficiency

Protein C and Protein S deficiency are autosomal dominant inherited disorders characterized by an inability to inactivate coagulation factors V and VIII. Because normal levels of protein C and S help to inhibit these clotting factors, those with a deficiency are in a hypercoagulable state. Patients display recurrent DVTs, or DVTs that arise at a young age. For long-term anticoagulation, the choice of medication is highly patient-dependent. Options include warfarin, or direct oral anticoagulants such as dabigatran or apixaban. When warfarin is chosen, patients are typically initially started on heparin, and slowly bridged to warfarin. This is because these patients are at risk for warfarin-induced skin necrosis, which is sometimes a presenting feature of the disease.



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Pathophysiology

Autosomal Dominant

Domino

Protein C and Protein S deficiency are inherited in an autosomal dominant pattern.

Inability to Inactivate Factor Va and VIIIa

Inability to kill the 5-rabbit and 8-beaver building clots

Protein C and S normally work to inactivate Factor Va and VIIIa, decreasing coagulability. When patients are born with a deficiency of these proteins (C and S), they are unable to inactivate the clotting pathway effectively.

Symptoms

Hypercoagulable State

Hiker-clog State

Being unable to effectively inhibit clotting factors Va and VIIIa, patients with protein C or S deficiency are in a hypercoagulable state.

Recurrent DVTs or DVTs at Young Age

DVT with redness and swelling

Due to their hypercoagulable state, patients can develop recurrent DVTs (deep vein thromboses). Often, protein C or S deficiency can be diagnosed when a child presents with DVTs at a young age.

Treatment

Begin Heparin

Hippie-heron

In order to prevent warfarin necrosis, patients should be started on heparin initially. Heparin inhibits factor Xa, which is not affected by proteins C or S. Furthermore, heparin is an IV medication which has an immediate effect on anticoagulation, which is why it is recommended to use this drug while "bridging" to warfarin.

Slowly Bridge to Warfarin

Snail Bridge to War-fairy

For long-term anticoagulation, the choice of medication is highly patient-dependent. Options include warfarin, or direct oral anticoagulants such as dabigatran or apixaban. If warfarin is chosen, patients should be very slowly bridged from heparin to warfarin due to the risk of warfarin-induced skin necrosis. Warfarin does not have an immediate effect on anticoagulation, and patients taking this medication transiently develop a hypercoagulable state.

Hemorrhagic Skin Necrosis

Hemorrhage-hammer Necrosis-crow

Warfarin inhibits production of clotting factors II, VII, IX X, as well as protein C and protein S. Protein C and protein S have shorter half-lives than the other factors and are depleted faster. Thus, when patients take warfarin, the initial depletion of protein C and S makes them hypercoagulable for a short period of

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time. It is during this time period that patients with protein C and S deficiency are further depleted, leading to warfarin necrosis. The hypercoagulable state leads to clotting of vessels supplying the skin, causing necrosis.

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