HEMATO-ONCOLOGY

CLOTTING FACTOR REPLACEMENT THERAPY

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Abstract: *Inherited disorders of clotting factor deficiency are known to occur with all coagulation factors. Of these, Von Willebrand disease, hemophilia A and B are the commoner conditions. Fresh frozen plasma contains all coagulation factors and hence in the past it was used as the major therapy for all inherited clotting factor deficiencies presenting with bleeds. Later cryoprecipitate was discovered and used for deficiency of fibrinogen, factorVIII, factor XIII and Von Willebrand disease. Both these blood products have to be administered in large volumes and they also carry a high risk of transfusion transmitted infections. This led to the discovery of clotting factor concentrates. Good manufacturing practices have resulted in the availability of products with high degree of purity and safety. Plasma derived single factor concentrates are available for all factors except for factor II and factor V. Advances in genetic engineering led to the discovery of recombinant factors which have very high safety profile. Currently recombinant forms of factor VIIa, factor VIII, factor IX and factorXIII are available. The standard of care for factor deficiencies is to replace the missing factor using clotting factor concentrates to enable patients to lead a completely normal life. This article deals with factor replacement therapy for the common and rare bleeding diatheses.*

Keywords: *Factor replacement therapy, Clotting factor concentrates, Fresh frozen plasma,Cryoprecipitate.*

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Points to Remember

- ***Clotting factor concentrates are available for almost all factor deficiencies except FV and they are the drug of choice for congenital factor deficiencies.***
- ***FFP contains all coagulation factors and hence can be used in a coagulopathic child with bleeds if specific factor concentrates are not available.***
- ***Cryoprecipitate contains fibrinogen, FVIII, FXIII and von Willebrand factor and can be used in deficiencies if specific factor is not available.***
- ***Recombinant FVIIa and activated prothrombin complex concentrate (aPCC) are useful in arresting bleeding in hemophilia children with inhibitors.***
- ***Prophylaxis using continuous factor replacement is recommended as the standard of care in haemophilia patients.***
- ***Tranexamic acid should be avoided in patients receiving prothrombin complex concentrates (PCC).***

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