March is Hemophilia Awareness Month

see how the IHTC is helping patients

The IHTC offers the “Thrive” website, www.workin.org, and it features hemophilia facts and a series of videos highlighting the stories of people in Indiana who live with hemophilia. The videos and website were created to emphasize that with the right treatment and management, those affected with hemophilia are living productive, normal lives.

Hemophilia is one of the most expensive chronic medical conditions largely due to the cost of treatment that has been found to be anywhere from $100,000 to $300,000 per year. There are a variety of treatments available, including factor replacement therapy, cryoprecipitate, desmopressin (DDAVP), and pharmacological agents like anti-thrombin, Xa inhibitors, and direct thrombin inhibitors. Each of these treatments requires medical care from dedicated experts who have extensive experience and a wide range of available services to address the many aspects of life that are impacted by hemophilia; these include individually tailored medical needs, physical and social functioning, and the impact on their family, work, and school.

In Indiana’s only integrated hemophilia treatment center, the IHTC offers lifelong, comprehensive, patient-centered, multidisciplinary healthcare that goes beyond what we often consider as event or disease based interventions. The IHTC’s multidisciplinary team includes hematologists, nurses, physical therapists, social workers, dietitians, pharmacists, and occupational therapists, and the team is assisted by integrated case managers. All of these are focused on serving the patient and family; coordinating and supporting the patient’s family; and providing education to patient caregivers in order to help the bleeding disorder community thrive.

To learn more about Hemophilia Awareness Month and how the IHTC helps our patients thrive, visit www.workin.org. For more information about the IHTC, visit www.ihtc.org.

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Clinical

Anti-thrombin

Life-threatening...

45-50 IU/kg

mg Protamine/
PCC, rFVIIa, FFP; repeat

10

10 IU/kg

35 IU/kg

15%

Alone

20%

40-50

Adults

Dose of rFVIIa in

5%

K intravenously; the risk is minimized through a slow infusion

Consequently, physicians may be reluctant to administer vitamin

duction of factors II, VII, IX and X. Intravenous administration of

due to decreased synthetic capacity and resultant abnormal pro-

effect observed in 4-6 hours, and the full effect in 12-24 hours. If

held and vitamin K is administered, either intravenously or orally.

in the target range.

surgery or on a subsequent postoperative day depending on the

Warfarin may be resumed 24 hours

warfarin should be discontinued five days before surgery, and

studies should be repeated every few hours to assess response

underlying clinical thromboembolic condition requiring fibri-

In cases of life-threatening intracranial or intracavitary bleed-

hemorrhage, but soft tissue hemorrhage is less common. Desmopressin

therapy is intracranial bleeding; however, soft tissue hemorrhage, and

have very short half-lives, ranging from 5-20 minutes, and are usu-

sal. It is preferable to avoid use of rFVIIa as these agents are

inhibitor, and direct thrombin inhibitors. rFVIIa in a dose of

aspirin and non-steroidal anti-inflammatory medications.

disease. Antiplatelet agents are divided into categories based

upon their mechanism of action. Cox-2 inhibitors include

Warfarin reversal is a prothrombin complex concentrate

PCCs contain low concentrations of factor VII; therefore,

administered ranges between 25-50 international units/kg.

Intravenous administration of PCC is associated with a

response to warfarin. In comparison with heparin, LMWH

dose:

1-2

Renal

system (faster

hemoglobin,

thrombotic

5

Majority

& factor Xa

inhibition of

mediated

VII, IX, X

respectively. The pharmacology of warfarin is regarded

within a therapeutic range of 2.2-3.0. In an attempt to

International Normalized Ratio (INR)

Extracorporeal circuits should be inspected and blood

time since last heparin dose

Warfarin

for removal of anticoagulant therapy may be required include

thrombus. Scientific publications and reviews that address

of the Heparin Association, and THROMBO have partnered with

Considering the different pharmacokinetic and pharmacody-

inhibitor, and direct thrombin inhibitors. rFVIIa in a dose of

60% of the anticoagulant activity; nonetheless, protamine

long acting form combined with aspirin. The thienopyridines

aspirin and non-steroidal anti-inflammatory medications.

reversal of anticoagulant treatment may be required include

The last group of patients to be considered, for whom acute

the mechanism underlying thrombolytic dosage requirement,

Reversal of UFH is achieved with protamine sulfate or a partial

Further, the half-life of UFH is very short and the agent must be

compared with rFVIIa. Protamine sulfate is expensive and

been reserved for patients with heparin-induced thrombocytopenia

have failed to reach therapeutic anticoagulant levels; the

The parenteral drugs have short half-lives; therefore discon-

tichloroacetic acid (TCA).

PCCs are associated with a lower risk of bleeding compared to

Parenteral anticoagulants are the mainstay of treatment for

inhibitor, and direct thrombin inhibitors. rFVIIa in a dose of

may be utilized; however, an infusion of 15-20 mg/kg may require 4-6 units in a 70 kg adult.

INR reversal with recombinant factor VIIa (rFVIIa) is partial,

the factor vitamin K-dependent coagulation factors II, VII, IX,

Necessity for treatment may be predicted by the level of the

WHO states that warfarin is the preferred anticoagulant.

Many clinical scenarios may require reversal of anticoagulant

Intravenous administration of rFVIIa results in a rapid rise

factor Xa. Consequently, patients on warfarin need anticoagulant

of warfarin anticoagulation. The usual dosage of warfarin

cofactor for a specific common clotting factor, the duration of the

anticoagulation

INR...