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A special thanks to Lori Laudenbach, APN Bleeding Disorders Program for writing this month's Transfusion Tales.

Patient Case: Marty is a 17 year old young man with severe hemophilia A. He presents to the emergency room with a two day history of pain in his right calf. He has no recollection of injury. On physical examination his right calf is warm to the touch and measures 2 cm larger when compared to his left. Marty is registered with the Home Infusion Program through the Bleeding Disorders Program (BDP); he last self-administered factor VIII three days ago.

QUESTION 1 What is the first course of action in Marty's emergency care?

- Obtain a group & reserve
- Fast track to radiology
- Obtain factor VIII from Blood Transfusion Services or use patient's own supply and infuse as per Marty's Factor First Card or BDP Treatment Protocol
- Give analgesia
- R.I.C.E (Rest, Ice, Compression, Elevation)

QUESTION 2 Classify the following statements as True or False

- Factor VIII is supplied by only one manufacturer
- Factor VIII is reconstituted by the Blood Transfusion Services (BTS)
- Administration of factor VIII requires completion of a "Consent to Treat" form
- Factor VIII is best administered through IV direct push
- Group and reserve is required prior to obtaining factor VIII from BTS

QUESTION 3 What are the potential consequences of failing to treat the cause of pain in Marty's right calf? (choose all that apply)

- Decreased capillary refill in Marty's toes
- Decreased sensation in Marty's foot
- Increased tightness and intense pain
- Loss of limb
- Lost days from work or school

ANSWERS:

- c
- (a)False, (b)False, (c)False, (d)True, (e)False
- a, b, c, d, e

DISCUSSION: Hemophilia A & B are x-linked inherited conditions causing a deficiency in factor VIII or IX respectively. The hallmark of hemophilic bleeding is internal in nature. Normal circulating factor VIII and factor IX levels are 0.50 u/mL to 2.0 u/mL. Individuals with severe hemophilia (<0.01 u/mL) have no circulating factor VIII or IX causing them to bleed spontaneously. Those with mild (0.05-0.30 u/mL) or moderate hemophilia (0.01-0.05 u/mL) bleed as a result of injury and or invasive procedure. The most common bleeding sites are into joints and muscles, which left untreated, lead to early arthritis and disability. Standard of care in the developed world is for individuals affected by severe hemophilia to self-administer factor two to three times each week to ensure that their baseline factor levels do not fall below 0.01 u/mL. All severely affected patients followed by the BDP are certified in the preparation and administration of factor. From time to time, a patient may run out of home infusion supplies and encounters bleed situations that can be limb or life threatening necessitating an emergency room encounter. Those with mild or moderate hemophilia treat on demand and rely on the expertise of the emergency room staff for factor administration following injury or prior to invasive procedure.

WHEN IN DOUBT, TREAT.

If a bleed is suspected factor replacement should be done prior to any investigations. Delay in treatment leads to increased morbidity resulting in lost days from work or school. All patients registered with the Bleeding Disorders Program (BDP) are issued a [Factor First](#) wallet card that provides treatment guidelines that are also available in Power Chart under the Blood Information tab or in the BDP Treatment Protocol binder located in the ER and BTS.

TREAT FIRST, INVESTIGATE LATER

Bleeds into the muscles of the forearm and calf can lead to compartment syndrome, a limb threatening condition where the blood supply and nerve conduction are compromised to the hand and foot respectively. Aggressive medical management is critical to prevent need for emergency fasciotomy. Admission to hospital and regular boluses of factor replacement are required.

There are a number of recombinant factor VIII replacement products licensed for use in Canada. Advate, Kogenate FS and Xyntha are manufactured by Baxter, Bayer and Pfizer respectively. Patients are informed shortly after diagnosis about their treatment options and decide, in consultation with the BDP Medical Director, which factor concentrate they will use for the prevention and treatment of bleeding. Factor VIII is obtained through the Canadian Blood Services and made available in the hospital setting through the Blood Transfusion Laboratory (BTL). Cross matching is not required. Reconstitution instructions and administration guidelines are available on the [BDP website](#). Factor replacement products are reconstituted at the patient's bedside, not in the BTL. Given the small volume once reconstituted, factor replacement products are best delivered by intravenous direct push through the butterfly infusion set that comes with the product or through the closest port to the IV insertion site. A self-directed module for IV direct medication administration is available on the LHSC and SJHC intranet site. [Competency Development Program: IV Direct Medication Administration](#)