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CASE 1:

A 14 month old male is rushed to the ER by his parents , because he fell about 15 mins ago while trying to walk by himself and hit his head on the floor. Immediately after the fall, he cried out loudly but became lethargic short after. He had one episode of emesis, on the way to the hospital. Parents report that their son was diagnosed with Hemophilia A soon after birth when he had prolonged bleeding from the circumcision site. He has not had any trauma or bleeding from other sites since then and is not on any medications at home. In the ER, vitals are T 37.7, HR 130, RR 36, BP 84/68.On physical examination, he is lethargic but arousable, pupils are equal and reactive to light, no focal neurological deficit, lungs CTAB ,heart RRR, no murmurs. What is the most appropriate next step in the management of this baby?

A. Obtain labs: CBC, PT, aPTT, Bleeding time, factor VIII assay .
B. Order a stat CT brain without contrast.
C. Administer recombinant factor VIII with a target of achieving > 50% factor VIII level.
D. Administer fresh frozen plasma.
E. Administer recombinant factor VIII with target of >100% factor VIII level.

A. Obtain labs:CBC, PT, aPTT,Bleeding time.
B. Order a stat CT scan of the brain.
C. Start recombinant factor VIII infusion with a target of > 50% factor VIII level.
D. Start fresh frozen plasma infusion.
E. Start recombinant factor VIII infusion with a target of >100% factor VIII level.

Given the history of Hemophilia A and trauma to the head along with the clinical presentation of being lethargic, it most likely points to an intracranial bleed.

In a known hemophiliac presenting with a life threatening bleed as in this case, appropriate treatment should initiated before imaging. Hence CT scan is not the next most appropriate step.

Treatment of acute life threatening bleeding is as follows:

- *Goal is to achieve plasma levels of factor VIII > 100%.*
- 50-75 IU/kg factor VIII concentrate, then initiate continuous infusion of 2-4 IU/kg/hr to maintain factor VIII >100 IU/dL for 24 hr‘ then give 2-3 IU/kg/hr continuously for 5-7 days to maintain the level at >50 IU/dL and an additional 5-7 days to maintain the level at >30 IU/dL.

- Formula to calculate the dose of factor VIII is as follows:

Dose of F VIII = wt (in kgs) x desired % increase in the level x 0.5.

Labs are not necessary at this time since it will not change the management of the acute bleeding in hemophilia.

Laboratory studies for suscepted hemophilia include CBC, Coagulation studies , Factor VIII assay.

We can expect to see normal to low H & H, *normal platelet count* and *elevated aPTT.*
PT and bleeding time are normal.

Factor VIII Assay:

Levels are compared with a normal pooled plasma standard which is designated as having 100% activity or equivalent. Normal values are 50-150%.

Hemophilia is classified as follows based on the level of activity:

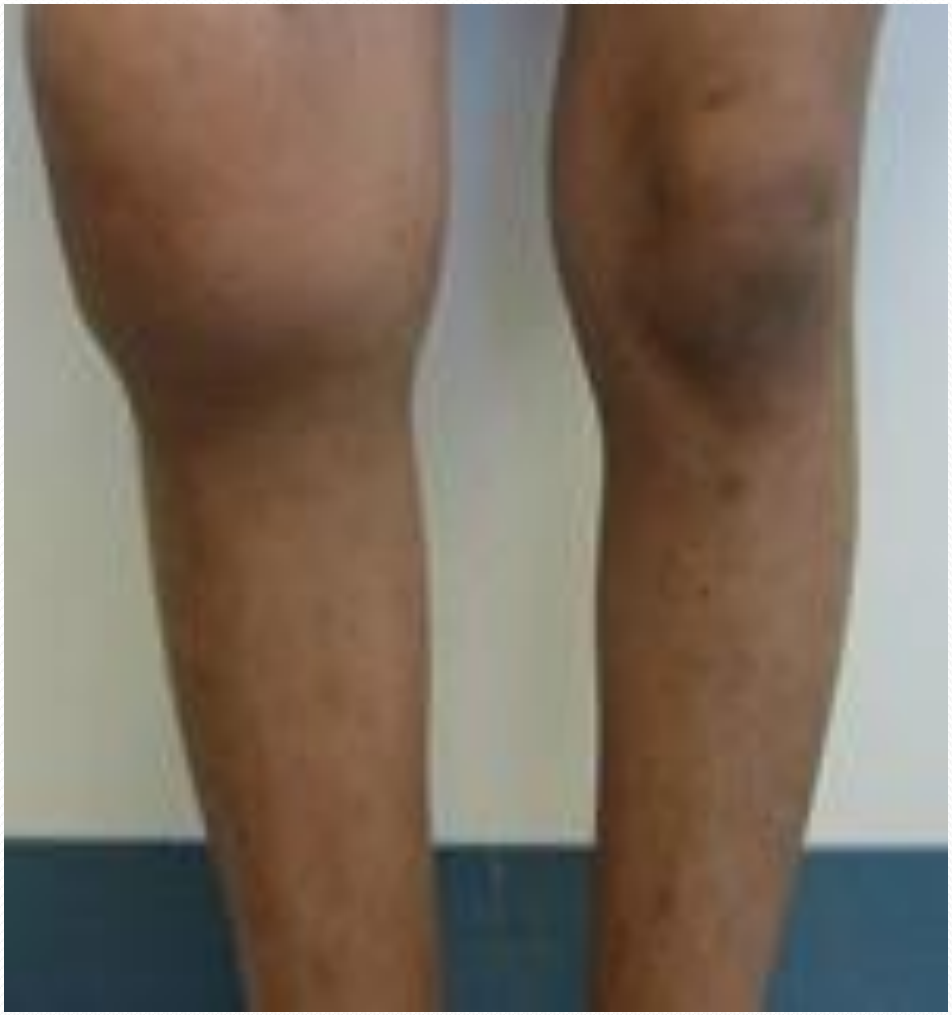
- Mild : >5 % but <40 %
- Moderate: > 1% but <5 %
- Severe: <1% . These individuals tend to have more frequent and spontaneous bleeding episodes.

Plasma level of > 50 % of factor VIII is desirable in other types of major hemorrhages like hemarthroses, muscle bleeds etc. However, intracranial bleeding requires higher plasma levels to prevent long term complications.

Fresh frozen plasma and cryoprecipitate are no longer the treatment of choice for treating acute bleeding episodes in hemophilia due to the increased risk of transmission of transfusion related diseases and high volume overload.

CASE 2 :

A 10 year old boy presents to the ER with pain, swelling and a warm tingling sensation in the right knee since 2 days. He denies any trauma to the knee joint. His mom states that he has Hemophilia A and has had similar episodes 4-5 times before in the same knee joint . On physical exam, vitals are stable, the knee joint is swollen, red ,tender to palpation with restriction of the movement . Which of the following statements is true?



A. No further testing is required and it should be treated with recombinant factor VIII.
B. Short course of corticosteroids should be considered.
C. He should be put on short time prophylaxis with factor VIII.
D. Rest and splinting for the joint and referral for Hemophilia Treatment Center after acute symptoms resolve.
E. All of the above.

A. No further testing is required and it should be treated with recombinant factor VIII.
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C. He should be put on short time prophylaxis with factor VIII.
D. Rest and splinting for the joint and referral for Hemophilia Treatment Center after acute symptoms resolve.
E. All of the above.

Hemarthrosis or bleeding into a joint is one of the most common presentation of hemophilia. The knee is the most common joint followed by elbow and ankle. Bleeding may occur spontaneously or following a minor trauma, depending on the severity of the disease .

When the same joint undergoes repeated hemorrhage, it becomes a target joint and is predisposed to developing hemophiliac arthropathy.

The main stay of treatment remains replacement of factor VIII with a goal of achieving 40-50% level of the factor on day 1, then 20IU/kg on days 2, 3, 5 until joint function is normal or back to baseline. Consider additional treatment every other day for 7-10 days.

In addition, a short course of prednisone at a dose of 60-80 mg/day for 3 days and then 40 mg/day for 2 days can be given to reduce the pain and swelling from the synovial inflammation.

After the treatment of the acute bleeding episode, the patient should be placed on prophylaxis with factor administration 3-4 times/week for several weeks to months. This approach helps to maintain a higher trough level of the factor in the plasma and reduces the rate of deterioration of the joints from repeated hemorrhage.

Rest and splinting is necessary for the faster recovery of the acute episode. All hemophilia individuals should be enrolled in a Hemophilia Treatment Center where multiple subspecialty physicians are involved in providing more comprehensive care to prevent long term complications.

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THANK YOU.