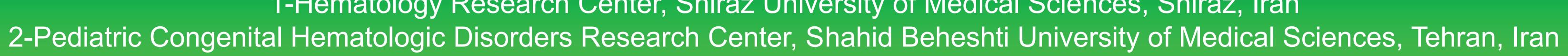


Modified primary prophylaxis in previously untreated patients with severe hemophilia A in Iran

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Introduction

Hemophilia common most high financial burden. coagulopathy Recently the low dose tailoring method of primary prophylaxis has been introduced for Previously Untreated Patients (PUP). (1, 2) The aim of the current study is to evaluate the efficacy and safety of low dose tailoring method of primary prophylaxis in PUPs with severe hemophilia A in Iran.

Methods

In this pre-post interventional study 33 patients with severe hemophilia A (factor level <1%) who were previously untreated and affiliated to universities in capital city (Shahid Beheshti University of Medical Sciences) and southern Iran (Shiraz University of Medical Sciences) were evaluated during 2014-2015. Primary continuous modified prophylaxis was used for patients with age less than 3 year old, after first and before the second episodes of obvious clinical bleeding in large joints including knee, elbow, hip and shoulder

or large soft tissue hematoma or large amount of bleeding. Prophylaxis was started by 25 IU/Kg once per week and increased to twice or three times a week according to defined bleeding events.

Results

The mean age at diagnosis and age of starting prophylaxis were: 4.9 ± 4.8 months (range: at birth-22months) and 13 ± 8.2 months (range: 1-35months) respectively. The most common bleeding types before getting prophylaxis was hematoma and ecchymosis in 11 patient (33.3%).Mean annual bleeding rate of patients after prophylaxis was to 1.08 ± 2.21 episodes per year. None of the patients had inhibitors prior to therapy. Inhibitor was present in 5 patients (15.1%) after prophylaxis that three of them were receiving recombinant factor VIII concentrate.

Discussion

In the current study untreated hemophilic patients who were younger than 3 year got prophylactic factor replacement with modified prophylaxis regimens (dosage of 25IU/Kg) once a week in 31 patients and twice per week in only two patients for a period of one year. It seems primary continuous modified prophylaxis has been successful in reducing bleeding episodes in children with severe hemophilia A and in the texture of lower cost it can improve quality of life of these patients compared to on-demand therapy represented in several studies (10,6,2).So it seems modified prophylaxis, once per week is costbenefit in countries where they are facing with shortage of factor.

References

- 1- Berntorp E, Fischer K, Miners A. Models of prophylaxis. Haemophilia. 2012;18(s4):136-40.
- 2- Schwarz R, Ljung R, Tedgård U. Various regimens for prophylactic treatment of patients haemophilia. European journal haematology. 2015;94(s77):11-6.

