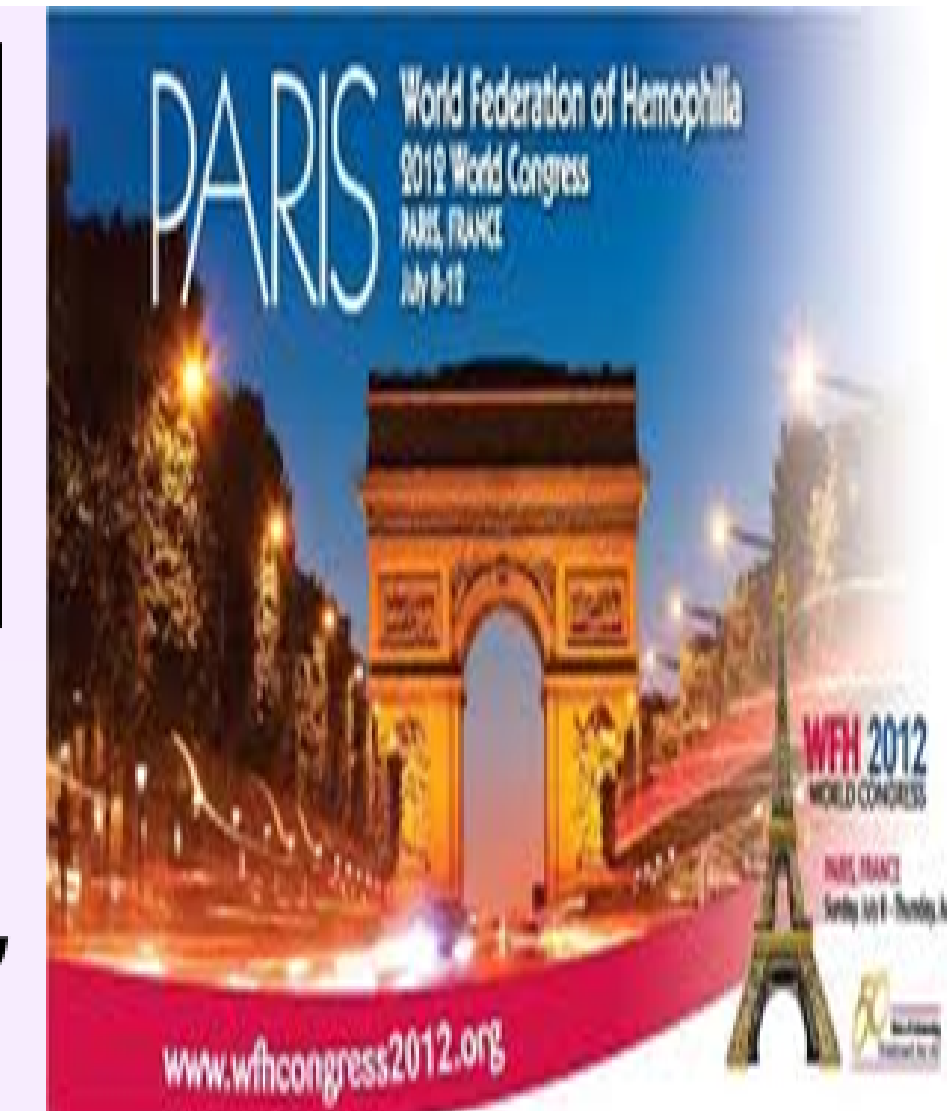




Bleeding Patterns In Severe Hemophilia A Infants and Toddlers On Prophylaxis Vs On Demand Therapy: A Prospective Randomized Observational Study

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INTRODUCTION

Hemophilia is characterized by frequent and lifelong bleeding, either spontaneously or following trauma. The majority of bleeding episodes occur in the large hinge-like synovial joints of the knee, ankle and elbow. The regular administration of factor VIII (FVIII) concentrate, usually three times per week greatly reduces the incidence of spontaneous bleeding into joints and muscles, thereby reducing arthropathy, changing lifestyle and improving long term outcome .

AIM OF THE WORK

The purpose of this prospective study was to compare prospectively the bleeding patterns in severe hemophilia A children younger than six years on prophylaxis Vs on demand therapy.

SUBJECTS AND METHODS

Patients

A total of 75 patients with severe haemophilia A (FVIII level <1%) were enrolled in the study . After 1st joint bleed; patients were randomized into two groups in (1:2) ratio and followed for 2 years.

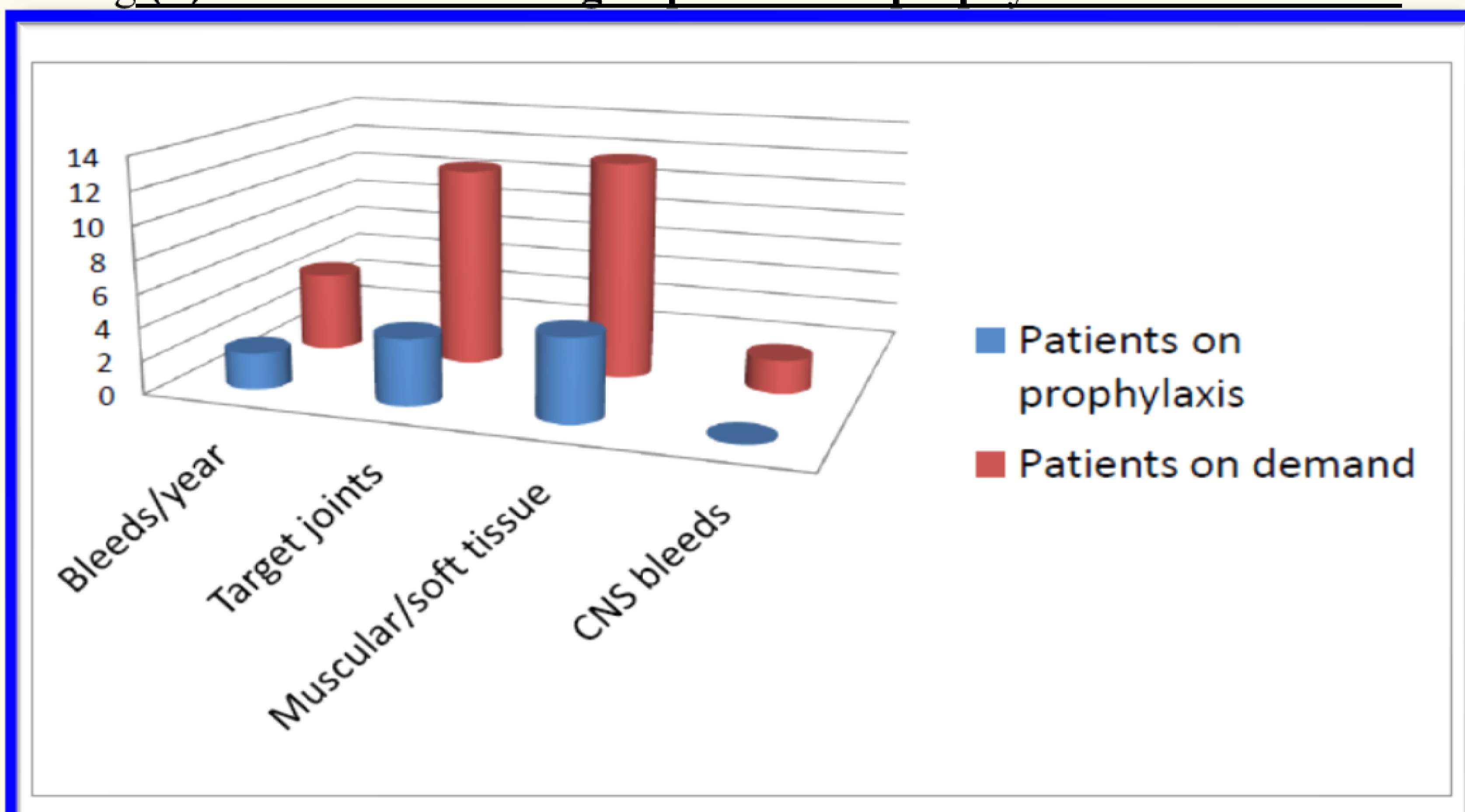
Methods:

Twenty five children received prophylaxis therapy with initial dose of (50 IU/kg/week) then escalated if needed, while the rest (n=50 patients) were treated on demand on a dose of (15-25 IU kg for 1-3 doses /joint bleed).

All bleeding episodes from birth on reported from patients with severe hemophilia A who were seen at our hemophilia center were recorded , confirmed and followed up by hemophilia clinicians. Data on number and location of bleeding episodes were reported and identified . Bleeds were defined as events requiring FVIII treatment and were described by date, time, site (joint or non-joint) and cause (traumatic or a traumatic). The site of the bleed could be recorded as joint, soft tissue-muscle, soft tissue-other, bleeding within a body cavity or intracranial.

Aggregated data were analyzed using descriptive statistics.

Fig (2):Patterns of bleeding in patients on prophylaxis Vs on demand



Patients who participated in this study attended interactive session in Hemophilia work shop 14-16 Jan 2012 at Ain Shams University that was sponsored by **Novo Nordisk Haemophilia Foundation**

RESULTS

- Median dose of FVIII consumption in prophylaxis therapy was (57 IU/ kg/ week); while those on demand median dose was (18 IU/kg/week).
- Overall; a total of 67 bleeds/year were observed in 22 /25 patients on prophylaxis Vs 282 bleeds/year in 50/50 patients on demand.
- Out of all children; 28(37%) were first diagnosed during the first 40 days due to post circumcision bleeding; all of them had a negative family history of hemophilia.
- Patients on prophylaxis 6-12 months old had a median of 2.2 bleeds/year; predominantly oral cavity bleeding; mostly related to tooth eruption (89%), none had intracranial hemorrhage (ICH), nor bleeding orifices, four develop target joints (8%)
- Number of bleeds were 4.8 per year for those on demand therapy; predominantly knee Joint bleeding (63%) as well as muscle bleeding into the buttocks, thigh, and forearm, one intracranial hemorrhage (2%), 12(24%) children had target joints (40% knees, 33% ankles and 27% had elbows or more than one joint) and six muscle bleeds during 2 years follow up.

Fig(1):Comparison of Knee joint image for patient on demand therapy Vs prophylaxis



CONCLUSION

- Both frequency and pattern of bleeding were different among patients either on primary prophylaxis or on demand therapy; in favour of prophylaxis; it seemed appropriate to begin primary prophylaxis in children with severe hemophilia A after 1st joint bleed at an early age to minimize the development of target joints.

