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# The effect of on demand and prophylactic therapeutic methods on quality of life among hemophilic children

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## Abstract

**Background:** Treating a chronic disease such as hemophilia is to improve the symptoms and quality of life (QOL) of the patients. This study aimed to study the quality of life among hemophilic children and compare the quality of life between patients receiving prophylactic or on demand treatments.

**Materials and Methods:** In this descriptive-comparative study, we enrolled 60 patients from three main hemophilia care centers in Tehran. All patients were 4-7 year old. Half of the patients were receiving prophylactic and half were receiving on-demand treatment. The assessment tool was Heamo-QoL questionnaire which assesses the quality of life in different dimensions (physical, feeling, family, friends, others, attitude, treatment and behavior). In this instrument, higher points correspond to lower quality of life. The mean quality of life in each dimension and also the total score were determined. Quality of life was compared between prophylactic and on demand treatments.

**Results:** The mean quality of life in groups receiving prophylactic and on-demand treatments were 2.6±0.3 and 3.33±0.4 respectively (P<0.001). All dimensions except "treatment" and "feeling" were different between groups. The highest impairments among patients, regardless of their treatment regimen, were in family and physical dimensions.

**Conclusion:** It is necessary to pay more attention to prophylactic treatment in hemophilic children as it seems to provide a higher quality of life among patients.

**Keywords:** Hemophilia, quality of life, prophylactic, treatment, on demand, Heamo-QoL.

## Introduction

Hemophilia is a hereditary bleeding disorder which results from deficiency or lack of certain coagulation factors in blood and is divided into different types. Among hemophilia subtypes type A and type B are more common and has been classified based on the plasma level of involved coagulation factor into severe (<1% of normal level), moderate (1% to 5% of normal level) and mild (>5% of normal level) categories 1. According to the last global report of the World Federation of Hemophilia in 2011, about 5000 hemophilia A and B patients are living in IRAN 2.

Hemophilia leads to bleeding episodes most often in knee, elbow and wrist joints which may progress to severe arthropathy. This can result in joint pain and its limitation in range of motion, lower self confidence among patients as well as repeated absences in school and can also affect patients' quality of life (QOL). Hemophilia not only affects routine daily relations and activities but also influences person's strategic planning for building the future 3.

Two approaches for controlling the bleeding episodes are replacing coagulation factors or plasma products in each episode (after bleeding), or prophylactic replacement of coagulation factors (before bleeding). The goal of treatment is to both to control symptoms and improve the quality of life

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among patients 4.

Prophylactic therapeutic method, by reducing the bleeding episodes, may improve the QOL. On the other hand, prophylactic treatment needs repeated injections from one to three times a week. These injections usually require referring to therapeutic centers. As a result in this method the prevention of bleeding episodes is achieved in expense of losing time, energy and finance.

Health related quality of life is selected as the key element in evaluating different therapeutic methods in chronic diseases such as hemophilia. Most studies determining the QOL in hemophilic patients have been performed on adults. Data regarding the quality of life in Iranian hemophilic children and the effect of prophylactic and on demand therapeutic methods on their QOL is scarce. This study aimed to assess the health status of hemophilic children in Tehran and to compare the on demand and prophylactic therapeutic methods regarding their effect on the quality of life among patients.

## Material and Methods

This descriptive-comparative study was conducted in three university affiliated hospitals in Tehran (Mofid Children's Hospital, Hemophilia Center, and hemophilia clinic of Imam Khomeini Hospital). We enrolled 60 patients from 4 to7 year old. Half of the patients were receiving prophylactic and the other half were receiving on-demand treatment. All patients suffered either from hemophilia A or B and had no accompanying physical disorders. Patients on prophylactic treatment must have received at least one year of prophylactic treatment.

We used Heamo-QoL questionnaire as the disease specific QOL assessment tool 5. The Questionnaire can be used to gather information from a child, parents or both. For age group of 4-7 years old it contains 16 questions in 8 dimensions in a 3 level Likert scale: physical (2 questions), feelings (2 questions), attitude (1 question), family (3 questions), friends (2 questions), treatment (2 questions) sport (2 questions) and others (2 questions).

Heamo-QoL questionnaire was filled by questioning the child. If the child could not understand the questions, the questionnaire was filled by the parents.

Chronbach alpha of the original questionnaire for all dimensions was 0.84. After translation

**Table 1:** Quality of life among patients entering the study.

| QOL Dimensions | On-Demand | Prophylaxis | P value |
|---|---|---|---|
| Physical | 3.6±0.6 | 2.47±0.9 | <0.001 |
| Feeling | 2.29±0.9 | 2.2 ±1 | 0.06 |
| Attitude | 2.7±1.1 | 1.9±1.1 | 0.003 |
| Family | 3.4±0.6 | 3.8±0.7 | 0.015 |
| Friends | 3.08±1 | 2.52±0.7 | 0.018 |
| Sport | 3.05±0.8 | 2.29±0.9 | 0.018 |
| Treatment | 2.8±1.2 | 2.22±1.2 | 0.06 |
| Other | 2.8±0.9 | 2.03±0.9 | <0.001 |
| ALL | 3.33±0.4 | 2.6±0.3 | <0.001 |

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into Persian, content validity was approved by an expert group consisting of a hematologist, a nurse and an epidemiologist. The internal reliability of the questionnaire in all dimensions which was determined by Chronbach alpha method was 0.74. Test-retest correlation was evaluated on 15 children within 10 days and showed a 0.72 correlation.

Descriptive statistical tables, paired t-test, Mann-Whitney and Fisher's exact tests were used for analyzing the data. P-values less than 0.05 were considered significant. SPSS software (version 16.0, SPSS Co, Chicago IL) was used for the statistical analysis.

## Results

The mean age of the children was 5 years. The age of parents was in the range of 31 to 35. Among 60 children, 30 children received prophylactic treatment and other 30 children received on demand treatment. In both groups, more than 85% of the children suffered from Hemophilia A and about 15% suffered from hemophilia B. In all of the cases the titer of factor (VIII in hemophilia A and IX in hemophilia B) was less than 1%. Inhibitory antibody titer was negative in all children receiving prophylactic treatment and positive in two evaluated children receiving on demand treatment. The age of the first bleeding episode in both groups was in the range of 6-12 months. Families noticed unusual bleeding of the child mostly at the time of circumcision or during the period that the child began to walk.

About, 36% of all children experienced four or more episodes of bleeding in a month and the mean frequency of bleeding was 2.45 per month. Bleeding episodes per month were in average 1.4 times among patients who received prophylactic treatment and 3.8 times in those patients who received on demand treatment. The difference was statistically significant (P=0.001).

Most of the children on prophylactic treatment (66.7%) were injected factor once a week followed by two (20%) and three (13.3%) injections per week. The average times of injection per month were 5.7 times in patients receiving prophylactic treatment method. There was no documented data in this regard in patients who received on demand treatment. Overall, nurses performed 70% of injections and remaining 30% was performed by parents.

The mean QOL in patients receiving prophylactic and on-demand treatments were 2.6±0.3 and 3.33±0.4 respectively (P<0.001). All dimensions except "treatment" and "feeling" were different between treatment groups. The highest impairment in patients, regardless of their treatment regimen, was found in family and physical dimensions respectively (Table 1).

Children who received prophylactic treatment had the most impairment in family dimension but the children who received on demand treatment had the most impairment in the physical dimension followed by the family dimension.

Twenty seven children who experienced bleeding episodes for less than 3 times a month had better quality of life almost in all dimensions (except treatment and "other" dimensions) than the 33 patients with bleeding episodes equal or more than three times a month.

## Discussion

This study showed that most patients suffering from hemophilia experienced bleeding in the first year of their life. First bleeding in joint was reported in the second year of life which supports the suggestion to start prophylactic treatment in the second year of life in hemophilic patients 6,7. Early onset long time prophylaxis has been shown in our study to achieve a better QOL among patients compared to on demand protocol.

The history of prophylactic treatment for hemophilia in European countries dates back to 1950, however, the primary prophylactic treatment in Iran (starting the long-term prophylactic treatment following the first joint or soft tissue bleeding) has began in recent years. Due to economic problems 8, and the shortage of resources prophylactic treatment mostly stops in the age of primary school in Iran. More than half of the prophylactic group in our study received factor once a week and bleeding frequency was considerably less in patient receiving prophylactic treatment.

Patients who receive prophylactic treatment had the most impairment in the family dimension of quality of life indicating the compassion of families in taking care of their children. This was in spite of the fact that nurses performed 70% of the treatments. Gringery et al. in a study on 4-7 years old hemophilic children showed that the family

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dimension with an average score of 34.38±24.8 was the most impaired dimension and this was attributed to excessive support of families 6.

In this study patients on prophylactic treatment method had better QOL than the patients who received on demand treatment which is consistent with other studies. Hartel et al. concluded that QOL in hemophilic patients receiving factor by prophylactic method was higher than the group receiving on demand treatment 9. They attributed this to decreased frequency of bleeding and decreased side effects following regular receipt of the factor. Results obtained from the research performed in 2004 in Europe also showed significant difference between hemophilic patients on prophylactic treatment compared to those receiving on demand treatment 10.

In our study bleeding episodes per month were in average 1.4 among patients who received prophylactic treatment and 3.8 in those who received episodic treatment. Manco-Johnson et al. showed that bleeding in the children who receive prophylactic treatment decreased considerably from 4.9 in on demand group to 0.6 in prophylactic group 7.

Finally, overall QOL score in prophylactic protocol was better than on demand regimen that similar to other studies10.

## Conclusion

It is necessary to pay more attention to prophylactic treatment in hemophilic children as it seems to provide a higher quality of life among patients.

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