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The impact of inhibitors on the quality of life in patients with hemophilia

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Abstract

Objective: To investigate the association of health-related quality of life in hemophilia patients with inhibitor and clinical and demographic characteristics.

Methods: In this multi-center cross-sectional study, 41 male patients with hemophilia A were investigated from May to October 2021. All patients were registered at the Hemophilia Clinic affiliated with Shiraz and Zahedan Universities of Medical Sciences in Iran. Health-related quality of life of the patients was evaluated by the Short Form-36 questionnaire.

Results: The patients' mean ± SD of age was 36.9 ± 13.2 (range: 18–76) years. Eleven patients (26.8%) were inhibitor positive. In univariate analysis, physical function, mental health dimension, and total Short Form-36 scores were significantly lower in the inhibitor-positive patients ($p < 0.001$, $p = 0.045$, and $p = 0.035$, respectively). Moreover, patients with severe disease showed significantly lower scores in physical function ($p < 0.001$), physical health dimension ($p = 0.018$), and total Short Form-36 ($p = 0.031$) than those with mild and moderate hemophilia. Also, blood-borne infections showed a significant association with lower score in physical health dimension ($p = 0.038$). In addition, annual bleeding rate showed significant negative correlations with physical health dimension ($r_s = -0.609$, $p < 0.001$), mental health dimension ($r = -0.317$, $p = 0.044$), and total Short Form-36 ($r = -0.455$, $p = 0.003$) scores. In multiple linear regression analysis, disease severity revealed a significant negative relationship with scores in physical function ($p = 0.001$), role physical (RP) ($p = 0.015$), general health (GH) ($p = 0.006$), physical health dimension ($p = 0.006$), and marginally in total Short Form-36 score ($p = 0.054$). Also, age of the patients showed a significant negative association with physical function and GH scores ($p < 0.001$ and $p = 0.015$, respectively).

Conclusion: Disease severity and age were shown as independent factors affecting health-related quality of life, but inhibitor alone was not an independent influencing factor. Reduced health-related quality of life was also observed in hemophilia patients with higher annual bleeding rate and blood-borne infections. Therefore, it is necessary to pay more attention to these subgroups. Further studies with larger sample size are needed for more accurate results.

Keywords

hemophilia, inhibitor, quality of life, SF-36 questionnaire

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Introduction

Hemophilia is an X-linked congenital bleeding disorder caused by a deficiency of coagulation factor VIII (FVIII) (in hemophilia A) or factor IX (in hemophilia B), with a frequency of 1 in 10,000 births which often affects males. Hemophilia usually presents with easy bruising in early childhood, spontaneous bleeding into joints, muscles, and soft tissues, or excessive bleeding after trauma or surgery.[1] Hemophilia A accounts for 80%–85% of all hemophilia patients. The disease severity is classified into three subtypes: severe (<1%), moderate (between 1% and 5%), and mild (up to 40%).[2]

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hemophilia patients.[38] This issue emphasizes paying more attention to close monitoring of hemophilia patients and the use of primary prophylaxis in patients with higher risk of bleeding.[39]

The employment status is one of the other factors that can affect QoL. There is a close relationship between education, employment, and QoL. Employment can affect the QoL not only through increasing life satisfaction and mental well-being of people, but also through increasing income.[40] Various disabilities in hemophilia patients result in a higher rate of unemployment in hemophiliacs than general population. Thus, the unemployment of young people with hemophilia compared to the general population is a major concern from a social point of view.[41] Based on the results of our study, significantly higher QoL scores in the PH and MH dimensions were obtained in employed patients and students compared to the unemployed and retired patients despite the comparable average age in both groups.

This study was limited due to the lack of a control group and the small number of hemophilia patients with inhibitor because inhibitor development is a rare complication. In addition, QoL may also depend on several factors (social determinants of health) that can vary between countries, making accurate comparisons between different countries difficult.

Conclusion

Disease severity and age were determined as independent factors influencing the HRQoL. Reduced HRQoL was also observed in hemophilia patients with higher annual bleeding rate, blood-borne infections, and in unemployed patients. It warrants further attention to these subgroups of patients. Precise attention should be considered in the prevention and management of bleeding symptoms in these patients. On the other hand, improving the social environment of these patients by providing educational facilities and creating a suitable job situation can improve the physical and MH of these patients. Based on the results of this study, the inhibitor alone had no significant impact on HRQoL in hemophilia patients. However, further studies with larger sample sizes are needed for better evaluation and more accurate results.

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Author contributions

S.H. designed the study, performed the data analysis, and edited the manuscript. M.J. edited the manuscript and collected the data. S.K. prepared the original draft and data collection. H.T. and N.J. collected the data. S.P. collected the data and edited the manuscript. M.K. had the concept of article and edited the manuscript.

Declaration of conflicting interests

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Ethics approval

Ethical approval for this study was obtained from the Ethics Committee of Shiraz University of Medical Sciences (IR.SUMS.MED.REC.1399.422).

Informed consent

Written informed consent was obtained from all subjects before the study.

Trial registration

Not applicable.

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Supplemental material

Supplemental material for this article is available online.

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