Original Article

Access this article online
Quick Response Code:
Website: www.jahjournal.org
DOI: 10.4103/jaah.jaah_7_21

Health-Related Quality of Life of Adolescents with Sickle Cell Disease on Hydroxyurea: A Case-Control Study

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Abstract:

BACKGROUND: Sickle cell disease (SCD) is a chronic multisystem disorder associated with acute and chronic complications that may negatively impact the quality of life (QoL). The study aimed to assess the health-related QoL (HRQoL) of patients with SCD on hydroxyurea (HU) and the factors affecting HRQoL domains.

MATERIALS AND METHODS: This case-control study included 174 patients with SCD (12–18-year-old); 87 were on HU for at least 1 year and 87 were not receiving HU. It also included 174 healthy adolescents of the same age group. The HRQoL was assessed using the Short Form 36 Health Survey version 2 (SF-36v2). A multivariate linear regression analysis was performed to assess the independent effect of studied variables on HRQoL dimensions.

RESULTS: Patients with SCD who were not on HU were found to have significantly lower SF-36 v2 scores (all domains) compared to those on HU and control group, $P < 0.001$. While patients on HU had significantly lower SF-36 v2 scores in physical functioning, role physical (RP), general health, and physical health component score only compared to the control group, $P < 0.001$. Multivariate linear regression analysis revealed significant associations between duration of HU therapy and RP ($R^2 = 0.208$, $P = 0.021$) and mental health component score ($R^2 = 0.389$, $P = 0.047$) and between hemoglobin levels with social functioning ($R^2 = 0.870$, $P = 0.023$).

CONCLUSIONS: HU has improved the HRQoL of SCD patients, in almost all domains, in addition to the improvement in many disease-related complications, mainly painful episodes. The positive impact of HU on HRQoL was significantly associated with the duration of therapy.

Keywords:

Hydroxyurea, quality of life, sickle cell disease

Introduction

Sickle cell disease (SCD) is a chronic and complex multisystem disorder associated with multiple acute and chronic complications including painful vasoocclusive events, cerebral vasculopathy, priapism, and renal or lung disease. SCD, as a chronic incurable disease, requires comprehensive care that includes screening, prevention, health education, and management of acute and chronic complications.[1-3]

Health-related quality of life (HRQoL) is an important patient-reported outcome measure for children and aids in the understanding of the well-being of children with SCD.[4] It provides an assessment of how an illness, its complications, and its treatment are experienced by a patient.

The wide clinical variability of the disease and frequent hospitalizations for SCD complications, mainly the pain, may negatively impact the quality of life (QoL) of patients,[5] beside that increased life expectancy in patients with SCD due to recent medical advances has increased

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Submitted: 29-Jan-2021
Revised: 20-Apr-2021
Accepted: 16-May-2021
Published: 28-Apr-2022

How to cite this article: Mones HM, Hassan MK, Al Hussein Ahmed BA. Health-related quality of life of adolescents with sickle cell disease on hydroxyurea: A case-control study. J Appl Hematol 2022;13:13-21.