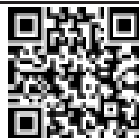


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10.4103/jah.jah\_7\_21

# 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 ( $R^2 = 0.370$ ,  $P = 0.023$ ).

**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.<sup>[1–10]</sup>

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.<sup>[11]</sup> 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,<sup>[12]</sup> beside that increased life expectancy in patients with SCD due to recent medical advances has increased

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Submitted: 20-Jan-2021

Revised: 20-Apr-2021

Accepted: 18-May-2021

Published: 28-Apr-2022

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**How to cite this article:** Monea 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.