Quality of Life in Patients with Sickle Cell Anemia

Thesis
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Naglaa Fathy Mahmoud

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List of Abbreviations

Abb.	Full term
ACS	*Acute Chest Syndrome*
CBC	*Complete Blood Count*
G6PD	*Glucose-6-phosphate Dehydrogenase*
HB	*Hemoglubin*
HBA1	*Hemoglobin A1*
HBA2	*Hemoglobin A2*
HBF	*Hemoglobin F*
HBS	*Hemoglobin S*
HCT	*Hematocrit*
HRQoL	*Health-Related Quality of Life*
OOL	*Quality of Life*
PH	*Pulmonary Hypertension*
PLT	*Platelets*
RBCs	*Red Blood Cells*
RCADS	*Revised Child Anxiety and Depression Scale*
RHC	*Right Heart Catheterization*
SCD	*Sickle Cell Disease*
SCT	*Sickle Cell Trait*
TLC	*Total Leukocytic Count*
VOC	*Vaso Occlusive Crises*
WHO	*World Health Organization*

ABSTRACT

Background: Sickle cell disease impacts the physical, emotional and psychological aspects of life of the affected persons and alters the health related quality of life. Our aim to study the pediatric quality of life score in patients with sickle cell anemia in relations to social, emotional and disease variables.

Methods: A cross sectional study was conducted in hematology clinic –Ain Shams University pediatric hospital. It included 40 patients with sickle cell disease between 5 and 18 years. Details of diagnosis, sickle cell disease complications, socioeconomic and assessment of disease severity were revised. Psychological assessment using children depression inventory (CDI), revised Children's Manifest anxiety scale (RCMAS). Health related quality of life was done using the sickle cell module

Results: The mean age of studied patients 15.35±5.53 years, (22 55%) males and 18(45%) females, 12 (30%) had illiterate fathers and 12 (12.8%) had college degree, nineteen (48.7%) of fathers were manual workers. As regards maternal education, 20(50%) were illiterate and 39 (39.5%) were housewives. The mean age at diagnosis was 20.56±21.9 months, with a mean frequency of 0.88±1.76 vasoocclusive crisis per year, and a mean frequency of hospitalization of 3.85±3.55 per year. Severe anxiety was present in 2(5%) of patients and severe depression in 36 (90%) of patients.

We found no significant difference in total QOL score in the three studied age group(5-8, >8-13 years and>13-18 years) (P=0.440) ; however there was higher pain score in 8-13 years compared with 13-18 years age group .A significant higher scores for worrying in males 59.06±21.20 compared to females4 4.31±20.73, P=0.033.There was significant higher scores for communication problems in mothers with College degree 93.50±8.19 compared with the illiterate mothers 56.76±19.20, P=.001 and mothers with Diplome /secondary schools /institute 67.46±16.50, P=.025 with no significant difference in quality of life in relation to father education. There was significant higher quality of life scores with less than 5 hospitalization/year 53.51±14.03, compare to more than 542.99±12.75, P=.032 in addition to increased communication problems with high rate of hospitalization (P=.021). There was significant higher scores for emotions in patients on regular transfusion 58.68 ±32.98, compared to those who are not transfusion dependent.There was better scores of quality of life in patients compared with their corresponding parents.

Conclusion: The main determinants of quality of life were the frequency of hospitalization and the blood transfusion therapy. Advanced mother education improves quality of life in patients especially in communications issues.

Keywords: Acute Chest Syndrome - Quality of Life - Pulmonary Hypertension.

INTRODUCTION

Sickle cell disease (SCD) is a genetic blood disorder caused by abnormal hemoglobin that damages and deforms red blood cells (RBCs). The abnormal red cells break down, causing anemia, and obstruct blood vessels, leading to recurrent episodes of severe pain and multiorgan ischemic damage. SCD affects millions of people throughout the world and is particularly common among people whose ancestors come from sub-Saharan Africa ***(Williamson, 2007).***

According to the World Health Organization (WHO), the quality of life (QOL) is defined as "the individuals' perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns" ***(WHOQOL, 1995).***

Sickle cell disease impacts the physical, emotional and psychological aspects of life of the affected persons, exposing them to disease associated stigma from the society and alters the health related quality of life ***(Adeyemo, 2015).***

Adolescents with SCD have significantly worse HRQoL than their peers in all of the most important dimensions of HRQoL (physical functioning, physical roles limitation, emotional roles limitation, social functioning, bodily pain, vitality and general health perception) except mental health. Recent hospital admission and SCD related complication

further lowered HRQoL scores. In Nigeria over seventy percent of adolescents with SCD have moderate to high level of perception of stigmatisation. Hospitalisation, SCD complication, SCD stigma were inversely, and significantly associated with HRQoL ***(Ayinde, 2015).***

Aim of the Work

To study the pediatric quality of life score in patients with sickle cell anemia in relation to the clinical severity of the disease, hospitalization rates, complications and laboratory variables.

Chapter 1

SICKLE CELL ANEMIA

Sickle cell anemia is the best known hereditary hematological disorder in human beings.. Estimates suggest that 250, 000 children are born annually with sickle cell anemia worldwide and thus it is among the most important epidemiological genetic diseases in Brazil and the world ***(Diniz et al., 2009).***

Disease prevalence worldwide

- In the USA, according to the Centers for Disease Control and Prevention (CDC). SCD affects approximately 100, 000 Americans, it occurs among about 1 out of every 365 Black or African-American births,.about 1 out of every 16, 300 Hispanic-American births.And about 1 in 13 Black or African-American babies is born with sickle cell trait ***(www.cdc.gov).***

- In the United Kingdom (UK) it is thought that between 12,000 and 15,000 people have sickle cell disease with an estimate of 250,000 carriers of the condition in England alone. As the number of carriers is only estimated, all newborn babies in the UK receive a routine blood test to screen for the condition ***(www.cdc.gov).***

Of the 330,000 babies born with a major hemoglobinopathy worldwide, 275,000 have SCD, making it the major global hemoglobinopathy ***(Weatherall, 2011).*** SCD patients in the developed world account for only 10% of the

world's SCD patient population ***(Aygun & Odame, 2012).*** **In 2008** reported United Nations estimates that there are between 20 and 25 million people worldwide living with SCD, of which 12–15 million live in Africa ***(Aliyu et al. 2008).*** It is estimated that 75–85% of children born with SCD are born in Africa, where mortality rates for those under age 5 range from 50% to 80% ***(Makani et al., 2011).***

The incidence of the SCT in Cameroon, the Democratic Republic of Congo, Gabon, Ghana, and Nigeria ranges from 20% to 30%, and in some parts of Uganda, the prevalence is 45% ***(Agasa et al., 2010).***

The prevalence of SCD in Saudi Arabia varies significantly in different parts of the country, with the highest prevalence is in the Eastern province, followed by the southwestern provinces. The reported prevalence for sickle-cell trait ranges from 2% to 27%, and up to 2.6% will have SCD in some areas. Clinical and hematological variability exists in SCD in Saudi Arabia with two major phenotypes: a mild phenotype and a severe phenotype ***(Jastaniah, 2011).***

In Egypt: Some researchers hypothesized that HbS gene was present among the predynastic Egyptian and they showed the presence of HbS in mummies (about 3200 BC) ***(Marin, 1999)***.

The first case of SCA in Egypt was reported in 1951 by Abbasy ***(Abbasy, 1951).*** Since then, several studies have been