

Pain and quality of life in adults with sickle cell disease

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Abstract

Aim: Aim of the study was to examine the relationship between pain and quality of life in adult patients with sickle cell disease (SCD) and to examine complications of SCD on pulmonary system.

Material and Methods: A total of 25 individuals (19 male, 6 female) diagnosed with SCD were included in the study. Pulmonary system, quality of life and pain was measured. Pain in rest and activity was questioned with Visual Analog Scale. The quality of life was assessed by the Short Form-36 (SF-36) Scale.

Results: It was determined that the individuals experienced an important level of pain in rest and activity levels, however it was seen that the pain level in activity was higher. It was determined that the oxygen saturation average was 91% and incidence of secretion and the number of coughing individuals was 6 and that 2 individuals used accessory respiration muscles. It was also found out that the SF-36 Scale's parameters' scores were low and that the quality of life of individuals were negatively affected. There was a moderate, negative and significant correlation between pain in rest and SF-36 Scale's physical function, pain, general health, mental health parameters.

Conclusion: It has been concluded that in the treatment of patients with SCD, although it is not mentioned in patient history, pain related problems should be taken into consideration. We believe that pulmonary and pain rehabilitation to patients with SCD will support their wellbeing.

Keywords: Sickle Cell Disease; Quality of Life; Pain; Pulmonary System.

INTRODUCTION

Sickle cell disease (SCD), which is one of the world's most frequently seen hemoglobinopathies, is a disease which displays autosomal recessive inheritance. It is characterized by infection, organ damage, pain seizures with intervals and chronic pain. In Turkey, its frequency is between 0,37 and 0,6%. Ischemia in the connective tissues and muscle-skeleton system becomes evident with acute pain, fever, tachycardia and anxiety. The repetitive seizures which are termed as painful seizures are the most common clinical symptoms. Infections, pain, fever, anxiety and hypoxia are triggers for seizures (1-4). In a study, in which 96 individuals were analyzed, it has been stated that 49,5% patients have rushed to the emergency services in the past one year, due to painful seizures. It is inevitable that painful seizures which are so frequently experienced have negative effects on the quality of life (5).

World Health Organization defines quality of life as the individuals' own perception of his own position in life in terms of his goals, expectations, standards and interests within the culture and system of values he lives in (6).

Given the devastating complications of the disease and other co-morbid factors patients experience that influence quality of life, it is increasingly important to understand quality of life. It has been shown in numerous studies that SCD is effective on quality of life depending on symptoms such as pulmonary regurgitation, strokes, pulmonary hypertension, renal failure, congestive heart failure, osteonecrosis and leg ulcer (3,4,7,8) In a meta-analysis study involving 22 studies, it has been stated that quality of life of individuals with SCD is worse in comparison to the general population (9). The life expectation of these individuals who are subject to multiple symptoms cannot be met; thus, their quality of life is affected negatively. When we take a look at literature, it can be seen that quality of life of patients with SCD is rarely analyzed in Hatay, even the rate of SCD is high.

Pulmonary complaints with cause mortality and morbidity in different levels appear acutely and chronically in SCD and more than 20% of these individuals state that they have pulmonary problems. Many pulmonary diseases such as air tract hyperactivity, nocturnal oxyhemoglobin desaturation, thromboembolism, acute chest syndrome and chronic lung disease are seen in patients with SCD in

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quite large numbers (10,11).

Aim of this study was to examine the relationship between pain and quality of life in adult patients with SCD in Turkey and to examine complications of SCD on pulmonary system.

MATERIAL and METHODS

Individuals who have been diagnosed with SCD at the Mustafa Kemal University Hospital's Hematology Department between January 2015 and May 2015 were included in the study. The study was approved by Medical Ethics Committee of Mustafa Kemal University's and also written consents were taken from the patients.

Individuals younger than 18 years of age, who have a physical disease or cognitive inability which prevents them from participating in interviews or filling out scales, currently receiving treatment due to a psychiatric disease were excluded from the study.

Measurements

The demographic information of the patients was recorded. Arterial oxygen saturation was evaluated with pulse oximetry; pain level was evaluated with the 10 cm Visual Analog Scale (VAS) during rest and activity.

The patients were asked to mark the point where they felt pain between 0-10 in VAS on the 10 cm. Horizontal bars (12). Within the scope of pulmonary evaluation, accessory respiration muscle activity, existence of dyspnea, secretion, coughing states were analyzed. The frequency of getting exhausted quickly, weakness, dizziness, headache, dyspnea, palpitation, being cold and having infections were analyzed with Likert type Scale.

Quality of life was evaluated with the Short Form-36 (SF-36) Scale. The SF-36 Scale has eight sub-parameters as physical function, physical role limitation, emotional role limitation, pain, social function, mental health, vitality and general health. Each parameter's score ranges between 0-100 and the score and quality of life are directly proportional. While 0 means bad health, 100 points out to good health (13).

Statistical Analysis

In the evaluation of data, the SPSS version 20.0 software was used (Statistical Package for Social Sciences Inc.; Chicago, IL, ABD). The categorical variables were presented numerically and the continuous variables were presented as mean±standard deviation. The normal distribution of data was analyzed with the Shapiro Wilk Test. The correlation between data was analyzed with Spearman's Correlation Test and $r < 0.40$ was defined as weak correlation; $r = 0.40 - 0.75$ as medium level correlation and $r > 0.75$ as high correlation. The level of significance was accepted as 5%.

RESULTS

The study included a total 25 individuals, 19 of whom were male and 6 of whom were female, aged 19-84. It was

determined that 7 of the individuals were relatives and 9 were married (Table 1).

When the frequency of getting exhausted quickly, weakness, dizziness, headache, and dyspnea, palpitation, being cold and having infections were questioned, it was seen that a majority of the patients marked the "sometimes" option (Table 2).

Age (mean ± SD)	35.64±16.45	
Parental consanguinity: Yes n(%) / No n (%)	7(28)/18(72)	
Marital status: Married n(%) / Single n(%)	9(36)/16(64)	
Length (cm) (mean ± SD): Female / Male	163.33±5.12/ 174.42±7.55	
Weight (kg) (mean ± SD): Female / Male	67.16±5.94/ 69.36±10.21	
Job	n	%
Free	7	28
Teacher	3	12
Officer	7	28
Unemployed	6	24
Student	2	8
Level of education	n	%
Primary School	8	32
Secondary School	8	32
University	9	36

SD: Standard deviation

	Never		Sometimes		Often		Always	
	n	%	n	%	n	%	n	%
Rapid fatigue (n = 25)	1	4	17	68	7	28	0	0
Weakness (n = 25)	2	8	16	64	7	28	0	0
Dizziness (n = 25)	4	16	14	56	7	28	0	0
Headache (n = 25)	3	12	11	44	11	44	0	0
Dyspnea during daily activity, (n = 25)	7	28	10	40	8	32	0	0
Sensation of palpitation (n = 25)	5	20	13	52	7	28	0	0
Chills (n = 25)	5	20	14	56	6	24	0	0
Frequency of infection (n = 25)	8	32	12	48	5	20	0	0

It was determined that the individuals experienced an important level of pain in rest and activity levels, however it was seen that the pain level in activity was higher. It was determined that the oxygen saturation average was 91% and incidence of secretion and the number of coughing individuals was 6 and that 2 individuals used accessory respiration muscles. It was also found out that SF-36 Quality of life survey parameters' scores was low and that the quality of life of individuals was negatively affected (Table 3).

When the relationship between activity pain, rest pain and SF-36 was analyzed, it was seen that while there was no significant relationship between SF-36 and pain which began during activity, there was a moderate, negative and significant correlation between rest pain and SF-36's physical function, pain, general health, mental health parameters (Table 4).

Table 3. Results of pain, pulmonary system evaluation and SF-36 Quality of Life Scale

Pain assessment	Mean±SD
VAS in rest	3.76±1.71
VAS in activity	4.76±1.50
Respiratory evaluation	
Oxygen saturation (Mean ± SD)	91.0±4.17
Coughing (n = 25)	6/19
Presence of secretion (n = 25)	6/19
Accessory respiratory muscles activity (n = 25)	2/23
SF-36 Scale	Mean±SD
Physical Function	62.80±18.14
Physical Role Limitation	22.00±33.32
Pain	34.92±18.71
General Health	45.56±19.65
Vitality	54.80±15.97
Social Function	51.33±21.57
Emotional Role Limitation	29.33±35.10
Mental Health	55.04±17.25

SD: Standart Deviation, VAS: Visual Analog Scale

Table 4. Relationship between resting-activity pain with SF-36

SF-36 Sclae	VAS in activity		VAS in rest	
	r	p	r	p
Physical Function	-0.034	0.870	-0.559	0.004*
Physical Role Limitation	-0.202	0.332	-0.017	0.937
Pain	-0.154	0.462	-0.465	0.019*
General Health	-0.153	0.465	-0.698	0.001*
Vitality	0.037	0.862	-0.342	0.094
Social Function	-0.254	0.221	-0.228	0.273
Emotional Role Limitation	-0.244	0.239	-0.266	0.199
Mental Health	-0.061	0.772	-0.499	0.011*

Spearman Correlation Test, * p<0.05

VAS: Visual Analog Scale

DISCUSSION

We have planned this study with the purpose of evaluating quality of life, pulmonary complaints and pain of individuals with SCD and determining the relationship between quality of life and pain, it was determined that quality of life of these individuals are low, they experience a significant amount of pain in rest and during the activity,

their pulmonary problems are at a level which should be given consideration and that pain in rest affect quality of life negatively.

It is stated that low quality of life causes life expectations related to the disease's complications to decrease and that it may be associated with the individuals' feelings of depression and anxiety which influence their quality of life (14). We think that due to the pain they experience, along with the frequent rate of getting exhausted quickly, weakness, dizziness, headache, dyspnea, palpitation and feeling cold and getting infections, their quality of life gets affected negatively.

In McClish et. al's study, it is stated that all the parameters of the SF-36 survey except mental health parameter were lower compared to normal individuals and that this can be caused by the pain they experienced (3). In our study, it was determined that all of the parameters of SF-36 are lower compared to the norms and that all the parameters of quality of life got affected negatively. Similar to McClish et. al's study, it was seen that the mental health status of the individuals in our study have a higher mean compared to all the other parameters except for physical health. In comparison to the general population, quality of life of individuals with SCD was found to be lower and quality of life should be taken into consideration when the patients are being treated.

Individuals with SCD experience pain in two manners, as chronic and acute. Acute pain begins in a sudden and intense manner as a result of damage in the skeleton system or soft tissue injuries, whereas chronic pain is caused as a result of the avascular lesion of the bones. Acute pain is not continuous and ends after the seizure, whereas chronic pain is the state of continuous pain (15). In Conner's study, 30 children with SCD aged 4-18 years of age were evaluated with the Oucher Scale and their pain levels were analyzed (16). It has been stated that 43% of the children experienced intense pain and that 60% defined their pain as similar to pain experienced during a small surgery or injury. In another study in which 47 individuals with SCD were included, the intensity of pain was stated to be 5.5 over 10 (17). In our study, it was seen that both activity pain and rest pain levels were similar to the averages indicated in the literature.

Anwar et. al declared that the pain parameter of SF-36 of individuals with SCD was 47.9, while this parameter was expressed as 47.4 in another study. In our study, we found that the pain average was 34.9 and considering the results of these studies, it was determined that the pain average of the individuals in our study was worse compared to the literature (3,18).

In Anie et. al's study, in which they analyzed the relationship between pain and quality of life of 96 individuals, it was determined that pain affected quality of life (5). Similarly, it was seen in our study that there is a moderate, negative and significant correlation between the physical function, pain, general health, mental health parameters of SF-36,

and rest pain level as well. It is an inevitable consequence that the increasing pain level negatively influences quality of life. It is apparent that quality of life of patients with SCD will increase by teaching them methods of coping with the pain. We also think that it would be appropriate for the health personnel to provide psychological support to the patients as well as medical treatment and to guide the patients in a manner to support their participation in life.

It was found in our study that the pain which begins during activity has not correlation with quality of life and that only rest pain affects quality of life. Since these individuals who experience symptoms related to SCD prefer resting and inactivity rather than being active during seizures and their normal daily life, the pain which begins during activity is temporary and controllable, whereas rest pain is continuous and uncontrollable. Accordingly, it was concluded that the pain which begins during activity does not affect quality of life.

Although life expectancy of individuals with SCD has increased, average life of men was determined as 42 and average life of women was determined as 48. Among the causes of death, pulmonary reasons are important. In a study in which 3764 individuals with SCD were included, it was stated that 20% of the patients had fatal pulmonary complications related to SCD (11,19-21). It is known that individuals with SCD experience problems such as chronic dyspnea, asthma, pulmonary hypertension and pulmonary fibrosis (21). Oxygen saturation is also stated to be lower compared to normal individuals (22). It is known that the oxygen saturation of healthy individuals should be 95-100% and that they need medical support when it falls below 90% (23). It was determined that the oxygen saturation of the individuals in our study was 91% and that it was lower compared to normal individuals in line with literature. It was observed that they had pulmonary problems at a level which caused indications for them to receive rehabilitation support. In our study, it was determined that due to the oxygen carrying capacity was low, these patients' pulmonary system was affected negatively and pulmonary complications such as oxygen saturation, coughing, secretion, accessory respiratory muscles activity occurred. We believe that pulmonary rehabilitation and teaching respiration techniques to patients with SCD and increasing their cardiovascular endurance will support their wellbeing.

CONCLUSION

It has been concluded that pain affects quality of life negatively; so the treatment of patients with SCD, although it is not mentioned in patient history, pain related problems should be taken into consideration, it would be appropriate to plan the treatment with providing the patients psychological support along with medical treatment.

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