



### Factors Effecting Hospitalization Frequency of Sickle Cell Anemia Patients

Orak Hücreli Anemi Hastalarının Hastaneye Yatış Sıklığını Etkileyen Faktörler

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#### ABSTRACT

**Purpose:** In this study, we aimed to find factors effecting hospitalization frequency of our sickle cell anemia (SCA) patients with painful crisis.

**Material and Methods:** Eighty four SCA patients in steady-state condition participated to the study: Seventy with homozygous, and 14 with sickle cell-beta thalassemia disease. In this study we also reported demographic and clinical data of our patients from February 2010 to October 2012.

**Results and Conclusion:** We found that patients using hydroxyurea and penicillin prophylaxis stayed in hospital more times than the others. No relationship between high hospitalization and other variables have been found. However genetic differences may be present among these patients which warrant further studies.

**Key Words:** Sickle cell anemia, hospitalization frequency

#### ÖZET

**Amaç:** Bu çalışmada ağrılı krizle gelen orak hücreli anemi hastalarımızın hastaneye yatış sıklığını etkileyen faktörleri bulmayı amaçladık.

**Materyal ve Metod:** Yetmiş i homozigot, 14'ü orak-beta talasemi olan 84 orak hücreli anemi hastası stabil durumdayken çalışmamıza katıldı. Bu çalışmada aynı zamanda 2010 ve 2012 yılları arasında hastalarımızın demografik ve klinik verilerini bildirdik.

**Bulgular ve Sonuç:** Hidroksiüre alan hastaların hastaneye daha sık yattıklarını ve hastaneye yatışla diğer değişkenler arasında bir ilişki olmadığı saptanmıştır. Hastaneye yatış açısından diğer değişkenler yönünden fark bulunamadı.

**Anahtar Kelimeler:** Orak hücreli anemi, hastaneye yatış sıklığı

#### INTRODUCTION

Sickle cell disease is the most prevalent hereditary hemoglobinopathy and multiorgan disease. It has serious mortality and morbidity. Pathophysiology of the disease is multifactorial. Mechanical obstruction of vessels by rigid Hemoglobin S, vasoconstriction after intravascular hemolysis, inflammation, endothelial cell activation, increased prothrombotic factors can play role of

the complications of the disease. Multisystem damage can be seen as acute chest syndrome, pulmonary fibrosis, pulmonary hypertension, hypoxemia, cerebral infarction, convulsion, cognitive disorders, congestive heart failure, isosthenuria, hematuria, papillary necrosis, glomerulonephritis, priapism, retinal artery occlusion, proliferative retinopathy, vitreous hemorrhage, leg ulcers and immune system

disorders. Hospital admission causes of these patients are vasoocclusive crisis (painful crisis of abdomen and bones, acute chest syndrome, central nervous system events, priapism), spleen and liver sequestration crisis, aplastic and hyperhemolytic crisis. There are clinical differences among sickle cell anemia patients. In this study we investigated clinical and laboratory features of our SCA patients from hospital records. We aimed to find which patients were hospitalized more frequently.

### MATERIALS and METHODS

In this study, archive records of patients with homozygous-SS disease, and sickle- beta thalassemia were evaluated. Cases were selected among patients admitted to hospital between 2010 and 2012. We presented numeral data by using mean value±standart deviation, and ratio. We accepted frequent hospitalization as hospitalizations for painful crises  $\geq 4$  times in the last 30 months.

### RESULTS

We investigated 84 sickle cell patients' records. Thirty two were male, 52 were female with a mean age of 26 years (16 to 54 years). Clinical features of patients (autosplenectomy, splenomegaly, splenectomy, cholelithiasis, aseptic necrosis, renal disease, cerebrovascular event, leg ulcer, hydroxyurea, folic acid and zinco using) and laboratory findings (hemoglobin, hematocrit, leukocyte platelet, lactat dehydrogenase (LDH), uric acid, vitamin, C reactive protein, alanine aminotransferase, aspartate aminotransferase, total bilirubin, direct bilirubin, thyroid stimulating hormone, vitamin B12, folic acid, Hepatitis B and C positivity) were summarized. We accepted frequent hospitalization number as 4 and more in 2,5 years. In addition we investigated relationship of hospitalization frequency with clinical and laboratory findings (Table 1 and 2). We found that the patients using hydroxyurea and depot penicillin were hydroxyurea was more hospitalized than the others ( $p:0,018$  and  $0,028$  respectively) (Table 3).

**Table 1. Clinical features of SCA patients**

	n/%
Autosplenism/splenectomy	47 (56 %)/ 9(10,7 %)
Splenomegaly	21 (25 %)
Cholelithiasis	55 (65,4 %)
Aseptic necrosis history	10 (11,9 %)
Renal disease	8 (9,5 %)*
Cerebrovascular event history	4 (4,8 %)#
Leg ulcer	4 (4,8 %)
Hydroxyurea using	54 (64,3 %)
Zinco using	26 (31 %)
Folic acid using	50 (59,5 %)
Depot penicilin using	6 (7,1 )

\*4 nephrolithiasis, 1 ureteropelvic junction obstruction, 1 chronic renal failure, 1 hyperkalemia, 1 meduller calcinosis  
#2 ischemic, 2 hemorrhagic stroke

**Table 2. Laboratory features of SCA patients**

	Mean±standart deviation
Hemoglobin (g/dl)	9,1±1,50
Hematocrit (%)	26,59±4,29
Leukocyte (x10 <sup>9</sup> /L)	11±3,82
Platelet (/μL)	399,500±17830
LDH (U/L)	788,7± 305,45
Uric acid (mg/dl)	5,59±1,83
Vitamin D3(nmol/L)	15,4 (3,67-50,2)
CRP (mg/L)	7,48± 5,95
AST (U/L)	35±19,1
ALT (U/L)	28,6±31,2
Total bilirubin (mg/dl)	3,05±2,59
Direct bilirubin (mg/dl)	0,80±1,41
TSH (Mu/L)	2,62±1,77
Viatmin B12 (pg/ml)	343,50± 232,27
Folic acid (ng/ml)	13,89±6,01

## DISCUSSION

Some sickle cell painful crisis can not be managed at home. In cases of severe pain, acute chest syndrome, dehydration, septicemia, patients must be hospitalized. We observed some SCA patients were hospitalized with painful crisis almost every month, some were once a year. In a study conducted with 13 years and older sickle cell patients, high utilization was found associated with severe sickle cell disease. Septisemia and mood disorder were defined as preventable factors<sup>1</sup>. In another study with sickle cell children there was found that older age and secondary diagnoses (especially pneumonia and constipation) were associated with high resource use during vasoocclusive crisis hospitalizations<sup>2</sup>. In a study conducted with 142 sickle cell patients

in Pennsylvania, emergency department was found a common place for utilization of sickle cell patients with a mean of 7,4 visits per patient year. This study showed that females were older and had less emergency department and hospital admission<sup>3</sup>. In a study longed for 5 years, there was found that high hospital utilization was correlated with chronic disease complications such as aseptic necrosis. Neither age or gender effected to admission to hospital<sup>4</sup>. In our study we investigated hospitalization frequencies and found that sickle cell patients with hydroxyurea using was more hospitalized than others. As known hydroxyurea are given to patients with frequent vasoocclusive crisis. In the other words patients with high disease burden. Depot penicillin prophylaxis are given to SCA patients frequent

bacterial infections as well. There may be some genetic factor effecting vasoocclusive crisis. But some of the patients are incompliant to hydroxyurea use and this may be effective on the higher rates of painful crises among hydroxyurea users. High leukocyte counts are associated with severe sickle cell disease<sup>5,6</sup>. But in our study we didn't found leukocyte count related with high hospitalization

rate. This may be due to leukocyte lowering agent hydroxyurea.

### CONCLUSION

We found no relationship between high hospitalization with anything except to hydroxyurea and depot penicillin using. But there may be genetic differences among these patients. Further studies are needed.

**Table 3. Relationship of hospitalization frequency and clinical and laboratory findings**

Hospitalization frequency			
	Low (sayı/%)	High (sayı/%)	p
Sex			
Female	22	30	1
Male	13	19	
Age			
<30	31	18	0,134
≥30	26	6	
Folic acid using			
Yes	29	21	0,493
No	23	11	
Hydroxyurea using			
Yes	28	26	0,018
No	24	6	
Depot penicillin using			
Yes	1	5	0,028
No	51	27	
Hyposplenism			
Yes	36	21	0,811
No	16	11	
LDH level			
High (≥480U/L)	5	45	0,495
Low/Normal( <480 U/L)	5	26	
Leukocyte level			
Low (<10x10 <sup>9</sup> /L)	22	13	1
High(≥10x 10 <sup>9</sup> /L)	30	19	

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