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Original Article

Experiences and Problems Encountered by Families of Children with Sickle Cell Anemia

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ABSTRACT

Introduction: Sickle Cell Anemia is a disease that has a high level of morbidity and early mortality for patients that are not followed and controlled properly. Study was conducted the aim of determining experiences and problems of families whose children with sickle cell anemia.

Methods: Descriptive study was conducted the aim of determining experiences and problems of families (n= 206) whose children with sickle cell anemia. Before conducting this study, a written permission from the related institution and research ethics committee approval from Gaziantep University were obtained. Questionnaire is made up of two sections (10 questions), socio-demographic of families and data about their problems (15 questions and 11 statements). Data were evaluated SPSS (21.0), number and percentage calculations.

Results: It was determined that 96.1% of participants knew nothing about disease before their children were diagnosed, 92.7% of them are aware the disease was genetically inherited, all participants were a disease carrier themselves, and 93.7% of them had no blood tests before marriage. 97.1% of participants have no support from their spouses, It was determined that 98.5% of children suffer from pain, 60.7% suffer from weakness 51.5% of the participants apply to hospitals to decrease the problems and 48.5% use medications at home.

Conclusion: A great number of families have problems regarding fear of losing their children, lack of social aid and support. Majority of children suffer from pain, weakness, exhaustion, they stay at hospital between at least 1 and 5 times a year, they need blood transfusion.

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Introduction

Sickle cell disease (SCD) is a group of autosomal recessive hemoglobinopathies characterized by the presence of sickle hemoglobin in the red blood cells.[1,2] Sickle Cell Anemia (SCA) is a disease with high morbidity and premature mortality in patients who cannot be followed adequately.[1,3-5] Sickle cell disease is a group of genetic diseases which is especially prevalent in tropical and subtropical regions; however, forced migration and ongoing population movement have spread it throughout the world, with estimated birth rates reaching 0.49 per 1000 in the Americas, 0.07 per 1000 in Europe, 0.68 per 1000 in South and Southeast Asia, and 10.68 per 1000 in Africa.[6,7] It is estimated that around 305.000 SCA babies in the world come to the world annually.[2,8] Although SCA is seen in all races, prevalence is higher in African-origin individuals.[9,10]

The most common type of hemoglobin in hemoglobinopathies in Turkey is hemoglobin S. Especially seen in Hatay, Cukurova region on the Mediterranean coast of Turkey, sickle cell anemia is considered as a public health problem.[11] While the nationwide frequency is between 0.3% and 0.6% in Turkey, it is between 3% and 44% especially in some parts of Mediterranean region.[9-12] Particularly in southern provinces like Adana, Mersin and Hatay, the number of SCA patients and carriers is high.[11,13] According to the latest data from the Ministry of Health and the National Hemoglobinopathy Council, there are currently around 1200 SCA patients in Turkey with a carrier frequency of 10% in Adana, 10.5% in Hatay and 13.6% in Mersin cities.[13]

In chronic diseases such as SCA, home care responsibilities, unpredictable medical costs and uncertainties about the future of children cause problems physical, economic and mental problems for the sick children and the family as well as disruptions in their social and educational lives. The social and psychological problems that the chronic illness creates in the child vary according to the child as an individual, the family, the type of disease, the social environment and the medical care that the child receives, and they makes it easy or difficult for the child to adapt to the disease.[14-17]

This research was conducted because the prevalence of SCA is higher in Hatay than the nationwide average in Turkey and there is limited research on the experiences and problems of individuals with SCA and their families.

Materials and methods

This descriptive study was conducted to determine the experiences and problems encountered by families of children with sickle cell anemia who were hospitalized in the Hematology department of Mustafa Kemal University Research and Practice Hospital between September 7 and November 27, 2015. The study population consisted of all the families of children with sickle cell anemia who were hospitalized in the Hematology department of the same hospital. The study

involved a random sample of families of children with sickle cell anemia who were hospitalized in the Hematology department on the date of research and who agreed to participate in the study and to fill in the questionnaire (n= 206). Prior to data collection process, the study was authorized by the Office of Chief Physician at Hospital and by the Ethical Board of Gaziantep University permission. After the families were informed with a brief description of the research, those who gave their verbal consent and agreed to participate in the study were included in the study.

In literature there are no scales for the problems patients with SCA and their family's experience. Data were collected with structured questionnaire form was prepared by the researchers based on the literature expert opinion and preliminary assessment. The structured questionnaire contained of a total of 25 questions, 11 statements and two parts: (I) the socio-demographic characteristics of the families and (II) their experiences and problems regarding the disease.[1,2,15,17,18] Socio-demographic data were collected through 10 questions on the participants' sex, age, education and marital status. Data about the children with SCA and SCA-related data of their families were collected through 15 questions about number of children with SCA and their ages, level of knowledge about SCA, being a disease carrier, and need for blood transfusion; and 11 statements about challenges in making time for spouses and children, social and financial support, hospital services, health problems of children with SCA, and actions to solve these problems. Expert opinion was received for the validity of the questions on the form and 7 participants were preapplied for the clarity of the questions. The necessary arrangements were made to expert suggestion on form. The reliability of the questionnaire form Cronbach's Alpha was found to be 0.68 (n: 206). While data were collected, the researcher was present in the hall and the participants were assured that their responses would be kept confidential. The researcher encouraged them to fill in self report survey on their own. The researcher made sure that the participants filled in the questionnaire forms individually and completely. The form took about 10-15 minutes to fill in. The obtained data were evaluated by computer, number and percentage calculations.

Results

The results showed that 88.3% of the participants (family or parents) were women, 36.4% were 36-45 years old, 69.9% were primary school graduates, 82% were married and 95.1% had social security. Among the families of children with SCA, 96.1% knew nothing about SCA before their children were diagnosed with this disease, 92.7% were aware that SCA is genetically inherited, 61.2% learnt that their children had SCA when they were 1-5 years old, all the participants were disease carriers themselves, 93.7% had no blood tests before marriage, all of them (100%) had a fear of losing their children with SCA, and 48.1% regretted getting married due to SCA (Table 1).

Table 1. Socio-demographic characteristics of families

Characteristics	N (%)
Parent	
Gender	
Female	182(88.3)
Male	24(11.7)
Average of age	
25-35	5(2.4)
36-45	75(36.4)
46-55	68(33.0)
56-70	58(28.2)
Educational status	
Illiterate	55(26.7)
Primary school	144(69.9)
Secondary school	7(3.4)
Marital status	
Married	169(82.0)
Divorced	37(18.0)
Social security	
Yes	196(95.1)
No	10(4.9)
Knowing about SCA before having a child with SCA	
Yes	8(3.9)
No	198(96.1)
Being aware that SCA is genetically inherited	
Yes	191(92.7)
No	15(7.3)
Time of learning that their children have SCA	
0 to <1 years old	60(29.1)
1 to 5 years old	126(61.2)
5< years old or older	20 (9.7)
Having a fear of losing their children with SCA	
Yes	206(100.0)
Having blood tests before marriage	
Yes	13(6.3)
No	193(93.7)
Regretting getting married due to SCA	
Yes	99(48.1)
No	79 (38.3)
Partially	28(13.6)
Child	
Child with SCA going to school	
Yes	57 (27.7)
No	149 (72.3)
Grade level of the child with SCA	
1^{st} – 4^{th} grade	3 (1.5)
5^{th} – 8^{th} grade	4 (1.9)
9^{th} – 12^{th} grade	37 (18.0)
University	13 (6.3)
Staying at hospital	
Yes	206 (100.0)
Yearly frequency of staying at hospital	
1-5 times	142 (68.9)
6 – 10 times	60 (29.1)
11 times – or more	4 (1.9)
Child with SCA having regular blood transfusion	
Yes	159 (77.2)
No	47 (22.8)

Also, among the children with SCA, 72.3% of could not go to school, 68.9% stayed in hospital at least 1 – 5 times a year and 77.2% needed regular blood transfusion. Out of the families of children with SCA, 66.5% had no difficulty in finding blood, 75.2% were satisfied with the care provided by hospital, 30.1% were blamed by their children for the disease, 62.1% had concerns about their future and their social lives,

60.2% knew other families of children with SCA but 61.2% had no communication with those families, 22.8% needed psychological counseling but none of them received psychological support and counseling, 59.2% recommended couples with SCA or those who were carriers of SCA to avoid getting married, and the most common problem experienced by the families (31.1 %) was not being able to continue their education. Among the families of children with SCA, 98.1% had no support from their spouses, 99.0% were not excluded from their social lives, 20.4% had no support from friends and relatives, 63.1% had financial problems, 69.9% did not have enough time for their spouses and 70.4% could not make time for their other children (Table 2).

Finally, out of the children with SCA, 98.5% suffered from pain, 60.7% suffered from weakness and exhaustion, 20.9% suffered from respiratory distress, 29.1% had partial difficulty in walking, 22.8% had partial difficulty in meeting their own needs and, to relieve their health problems, 51.5% went to hospitals to seek care while 48.5% used medications at home (Table 3).

Discussion

Due to SCA, which is a chronic disease, families may experience feelings such as guilt, helplessness, anxiety and anger, and they may have difficulty in coping with these negative feelings induced by caregiver responsibilities, frequent hospitalization of children, economic burdens caused by medical expenses, and uncertainties about the future of their children. For this reason, families may need psychological support at times.[18,19]In a study by Çakan et al.,[20] half of the participants wanted to receive psychological support, but only very few of them (5.2%) had already received psychological support. In our study, 22.8% of the participants stated that they needed psychological support, but none of them received professional support until the time of the study.

Sickle cell disease (SCD) is a hemolytic anemia, characterized by abnormal hemoglobin production of autosomal recessive inheritance.[3] In our study, 92.7% of the mothers knew that SCA was a hereditary disease, but 93.7% of them stated that they did not have a blood test before marriage. In a study by Daak et al.,[21] 68.5% of the participants knew this disease is genetically inherited and most of them thought that it was important to have a blood test before marriage.

Evidence showed that parents of children with SCA reported that the disease greatly impacts them personally and within their families including through disruptions friendships, family activities, regular routines, and relationships.[6] Due to some persistent illnesses such as SCA, children often have to visit and stay in hospital for treatment. If the child is in hospital, it is usually the mother that usually accompanies the child. Due to frequent hospitalization of the sick child, mothers may not be able to adequately care for their other children, they may reflect their fears and troubles related to the disease to their husbands and other children, and

Table 2. Problems of families with children with SCA

Problems	N (%)
Having difficulty in finding blood	
Yes	42 (20.4)
No	137 (66.5)
Partially	27 (13.1)
Satisfied with the care given by the hospital	
Yes	155 (75.2)
No	23 (11.2)
Partially	28 (13.6)
Child with SCA having information about his or her disease	
Yes	199 (96.6)
No	7 (3.4)
Child with SCA blaming parents for their disease	
Yes	62 (30.1)
No	103 (50.0)
Partially	41 (19.9)
Having problems with the child due to SCA about social life and the future	
Yes	128 (62.1)
No	78 (37.9)
Type of problems experienced by the family due to SCA	
No problem at all	78 (37.9)
Deteriorated social relations	10 (4.8)
Finding a partner for marriage	15 (7.3)
Finding a job	39 (18.9)
Not being able to continue education	64 (31.1)
Knowing other families of children with SCA	
Yes	124 (60.2)
No	82 (39.8)
Communicating with these families	
Yes	80 (38.8)
No	126(61.2)
Need for psychological counseling	
Yes	47(22.8)
No	159(77.2)
Recommendations for couples who are carriers of SCA and about to get married	
They should get married but have no children	6(2.9)
They should not get married	122(59.2)
They should get married, but have children under the control of a doctor	78(37.9)
Having problems with wife/husband due to SCA	
Yes	4 (1.9)
No	202 (98.1)
Receiving support from wife/husband	
Yes	4(1.9)
No	202 (98.1)
Feeling excluded from social life	
Yes	2 (1.0)
No	204 (99.0)
Receiving no help from friends and relatives	
Yes	42 (20.4)
No	164 (79.6)
Having financial problems	
Yes	130 (63.1)
No	76 (36.9)
Not being able to make time for the other child (Ren)	
Yes	145 (70.4)
Partially	32 (15.5)
No	29 (14.1)

Table 3. Health problems experienced by children with SCA

Problems	N (%)
Pain	
Yes	203(98.5)
Partially	3(1.5)
Unable to meet one's own needs	
Yes	15(7.3)
Partially	47(22.8)
No	144(69.9)
Walking difficulty	
Yes	22(10.7)
Partially	60(29.1)
No	124(60.2)
Respiratory distress	
Yes	43(20.9)
Partially	79(38.3)
No	84(40.8)
Weakness and exhaustion	
Yes	125(60.7)
Partially	52(25.2)
No	29(14.1)
Solutions for relieving health problems	
Using medications at home	100(48.5)
Going to hospitals to seek care	106(51.5)

their relations with their husbands may also deteriorate. In our study, 68.9% of the children with SCA stayed in hospital at least 1-5 times during the year, and 70.4% of the mothers stated that they did not make enough time for their other children.[20] Moreover, in our study, 98.1% of the participants did not receive support from their spouses, 20.4% of them did not receive support from their families and relatives, and 63.1% had financial difficulties. Similarly, in a study of Wonkam et al.,[18] the participants did not receive adequate support from the other members of the family (26.7%) and their spouses (15.7%) for their children with SCA.

Major causes of morbidity and mortality in sickle cell anemia include recurrent vaso-occlusive crises, severe anemia, infections, acute chest syndrome and multiple organ failure.[22] Blood transfusion is one of the treatment methods used both for the treatment and prevention of the complications due to the disease.[22] In our study, regular blood transfusions were performed in 77.2% of the children with SCA. In similar studies by Ferreira et al.,[23] and Aloni and Nkee[2] blood transfusion rates of their patients were 68.5% and 74.0% respectively.Although the pain of sickle cell anemic patients is primarily nociceptive pain due to tissue damage; inflammable, throbbing, stabbing, tingling, numbing and tingling neuropathic pain can also be observed.[1,11,24] In our study, the parents reported that their children with SCA had complaints of complications related to the disease such as pain (98.5%), weakness and fatigue (60.7%) and respiratory failure (20.9%). On the other hand, 51.5% of our participants stated that they went to hospitals to seek care while the rest of them used medications at home. In Garadah et al., study, most of the patients with SCA experienced severe bone pain.[25] In a study by Adzika et al.,[26] almost half of the SCA patients went to the hospital for general body pain and back pain.

Conclusion

As a result of this study, we found that the families of children with SCA had financial difficulties, they were unable to make quality time for their other children, their social lives deteriorated, they did not receive support from their spouses and they had concerns about their children's future. Also, the children with SCA suffered from pain and weakness/fatigue and they used medications at home or went to the hospital to reduce their health problems. In the light of these results, it is essential that family counseling centers for parents of children with SCA should be established, family education programs should be designed so as to ensure more attendance by families to these centers, and parents should be encouraged to communicate with each other more often by means of parental courses, seminars, brochures, television programs and radio programs to be offered in these centers. Also, nurses should be trained about supportive communication so that they can help children with SCA and their families deal with the undesired symptoms such as disease-related pain and fatigue. Finally, in order to reduce families' feelings of uncertainty and loneliness, their mental status should closely be monitored and they should be provided with psychosocial support to be offered by nurses, social workers and psychologists when they need. This research conduct on small size of populations and only a hospital is limitations of this study.

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Ethical issues

None to be declared.

Conflict of interest

The authors declare no conflict of interest in this study.

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