



EXPRESSION THE CONCEPT OF PAIN IN PATIENTS WITH SICKLE CELL ANEMIA (A QUALITATIVE STUDY)

^{1*}HOSSEINI NIK AMMAR, ²ABDEYAZDAN GHOLAM HOSSEIN
³D.R HEIDAR ALI ABEDI

^{1*}Medical-Surgical Nursing graduate student education,
Isfahan(Khorasgan) Branch, Islamic Azad University, Isfahan, Iran
^{2,3}Department of Nursing and Midwifery, Isfahan (Khorasgan) Branch,
Islamic Azad University, Isfahan, Iran

ABSTRACT

Sickle Cell Anemia is the commonest inherited disease in world which is caused by a mutation in genes that leads to abnormal hemoglobin and defectivered blood cells. Red blood cells become rigid and sticky and deforms into sickles or crescent moons which are unable to move in vessels. They, therefore, block blood flow, decrease tissue oxygen, and finally lead to ischemia. Each year 300 thousand infants are born with SCD. 300 million people in world and 2.5 million in the U.S. are affected with SCD. The disease has several symptoms and signs. Periodic episodes of pain, called crises, are a major symptom of sickle cell anemia. The recurrence of pain is completely unpredictable. This study aims to investigate the pain experience in patients with SCD. Considering the nature of the study a philosophical research method, descriptive phenomenology, was used. Participants consisted of 11 patients (4 females and 7 males) with Sickle Cell Disease in Kohkiloye-Boyerahmad Province, Iran. Sampling was done on the basis of the aim of the study. Interviews were unstructured and a 7-stage Colaizzi Method was used to analyze and codify the collected data. 100% of participants complained of pain and described it as its worst experience. 40 first-level codes and 3 second-level codes were extracted including the cause of pain recurrence, pain consequences, and the type of pain which altogether created the main concept of pain. Participants complain about irregular severe pains which disturb their normal life. It is suggested their needs and problems be addressed specifically. Particular programs are required to support these patients, meet their needs, and improve the quality of their lives.

Keywords: Sickle Cell Anemia, PATIENTS, hemoglobin and defectivered blood cells

INTRODUCTION

Sickle-shaped Cell Disease (DSC) is the commonest haemoglobinopathy which influences a specific type of intracellular protein called hemoglobin. DSC is anautosomal disorder of recessive globin β Gene. It is one of the commonest genetic disorders in the United States. Gene mutation leads to formation of abnormal hemoglobin (HBS) in red blood cells. In stickle cell anemia, red blood cells become rigid and sticky and deforms into sickles or crescent moons. These irregularly shaped cells cause RBC to stick to the blood vessels, which can slow or block blood flow and tissue oxygen, ischemia, as well as periodic

episodes of pain, called crises. The disease leads to creation of intense red blood cells with little oxygen through mutated hemoglobin. Polymerized S hemoglobin mutation in red blood cells blocks small block vessels^{1,2, 3,18}. Sickle Cell Anemia (SCA) is the most common genetic diseases in Africa, the Mediterranean Islands, South and Central America, and India. In the United States, it most commonly affects blacks (100,000 people), which is diagnosed by periodic episodes of pain. In Nigeria, with the highest level of SCD morbidity in world, 45 to 90 thousand infants are born with SCD each year. In 2004, 699 adults lost their lives to SCD. In general, Sickle-shaped Cell Disease is a significant contributor to mortality in world. More

than 217 thousand babies are born with SCD each year. In the United States in each 00 African-American babies, one is born with SCD^{3, 4, 5, 6}.

Pain in Sickle Cell Anemia

Sickle Cell Anemia is a chronic disease which affects the quality of life¹⁹. Severe pain attacks are the major symptom of SCA (20). Patients with SCA normally refer to doctors with three types of symptoms: pain, anemia, and infection. Earlier, SCD management was using narcotics to relieve pain during crises. Patients with periodic episodes of pain need to be hospitalized. Medical reports in hospitals and clinics reveal that 10-20 percent of patients experience episodes of pain frequently, 40-50 percent at times, and 30-40 percent of patients never experience crises. Recurrence of crises is not necessarily related to vessels blockage, it can also appear as a result of venous necrosis in bones in hips, shoulders, wrists, and legs. Crises consequences are disturbed sleep, weakness, and reducing hours of activity and work⁷. Episodes of pain in SCD have a direct impact on patients, their families, as well as their caregivers' quality of life. Its Pathophysiology thereof is complicated and varies depending on the place where the pain occurs. There are three types of pain: chronic crises, chronic pain syndrome, and neuropathy pain (8). Pain in SCD is quite unpredictable and recurring. Relevant data in 2001 show that 40-50% of children with SCD were referred to hospitals due to developing such symptoms as pain, fever, increases number of inhales, wheezing, and pale skin¹. Sickle Cell Anemia is diagnosed by venous blockage, hypoxemia, hypoxia, hemolytic anemia, and Icterus. Multiple pathologic processes of SCD include inflammation, damage, hemolysis, and sticking red blood cells to venous endothelium. Patients with HgbSS have such symptoms as recurring crises, organ chronic syndrome, and chronic hemolytic anemia. Chronic crises attack in blocked blood vessels is the most prevalent event. It is also the major reason for which patients with SCD refer to hospitals. Sickle-shaped cells get stuck in small blood vessels which slow the blood flow and cause severe pain. The pain may last for a short period of time or last longer. To improve the condition of patients with sickle Cell Disease and to relieve their pains during crises, special cares from pain specialists and nurses as well as blood treatments may be required⁹. Chronic crises attack in blocked blood vessels is also the commonest reason for hospitalizing children with SCD. The recurrence of crises is unpredictable and may vary

in intensity, the region, and duration. With respect to intensity, the pain is described to be more severe than post-surgery or cancer-related pains. Pain management in SCD is multi-dimensional which consists of medicinal, physical, and psychological management⁷. Crises may occur at the early stages of life with the drastic decrease in the level of fetal hemoglobin. Dactylitis might be the first appearance of the disease. It causes metacarpal and metatarsal adductus which in turn results in pain attacks as well as swelling in hands and feet. Newborn infants and children with dactylitis are irritable. They may avoid walking and cry when they are touched. Attacks can last from 3 to 10 days. Climate changes and dehydration can lead to crises. Children explain the pains as being throbbing, beating or pulsing¹⁰. Chronic pains and crises are the commonest effect of SCD. Crises in DCS can lead to chronic anemia, susceptibility to infections, chronic lung diseases, Thoracic syndrome, stroke risk, delayed growth and puberty in teenagers¹¹. It is worthy to note that the mortality rate due to SCD depends on crises frequency in cells. A study on +20 year old patients with SCD shows that patients who experience crises in three parts of body each year, die sooner in comparison to those who experience fewer times of crises. In 2004 in the United States, 83149 adults were hospitalized due to DCS crises which cost about 488 million dollars. In 1973, the average life span was 14, but thanks to advances in diagnosis and care of SCD patients, they now live into their 40s and 50s⁶. Damage to organs and the consequent pain from sticking the sickle-shaped cell to the walls of small vessels are the distinguishing features of SCD. Pain emerges through several paths like body, neuropath, and venous mechanisms¹². It may lead to depression, solitariness, and mental illnesses (5). Patients with SCD are mostly from low-income families and deprived of receiving good treatment. They also suffer from unpredictable daily problems. Increase in the intensity of pain may cause death. In SCD patients, the neuropathy pain equals to the pain from a gunshot or a knife wound, that is, a severe and throbbing pain. Touching SCA patients causes hurt. Environmental and central nerve mechanisms are the sources of pain in SCA patients. Patients with SCD experience severe pain attacks called venous blockage crises. These painful crises may start at the age of 6 months old and continue to recur unpredictably over a lifetime¹³. SCD can harm a person's lungs, kidneys, bones, spleen, nervous system, and venous diseases as the common basin for simultaneous emergence of the afore-mentioned

problems (4). Problems due to periodic pain attacks, frequent prolonged hospitalization, and isolation from social environment and activities may lead to psychological disorders, learning problems, low self-confidence, changes in relationships, and decreased quality of life. Assessing pain in pre-school children is a challenge, as they are unable to explain the pain. Expressions of pain in children under 6 include crying, moaning, snoring, yelling, motionlessness organs, painful touch (15). Intensity, frequency, and the place of SCD periodic pain attacks as well as the frequency of being hospitalized aggravate with aging. The intensity of SCD pain attacks may be underestimated by caregivers, as pain in SCD is like an "iceberg" from which only the tip is visible and the massive body is hidden under water. Periodic crises have an impact on patients' physical, psychological, and social conditions. Crises aggravate by aging. Local damages may be completed after the first two decades of life, Patients may also experience central neuropathic pain in their 30s. Crises elevate during cold season, changes in temperature, increase in atmospheric pressure, and wind speed (16). Crises occur with no prior sign. Patients are usually hospitalized. They receive dosages of narcotics to relieve their pain and it had unavoidable side effects like nausea, vomiting, and scratch which are additional sources of suffering to the patients. Some patients avoid receiving narcotics due to its side effects and therefore suffer the severe pain longer (17). In spite of genetic and biochemical studies on SCD and the vast advances in DCS pathophysiology, modern treatment is the continuation of symptom one including pain control, oxygen, antibiotics, red blood cell transfusion, and Hydroxyurea that prevents painful episodes in SCD (14).

Methodology

Considering the nature of the study and the aim of describing life events the way they have happened, a philosophical research method, descriptive phenomenology, was used. Qualitative research method is a comprehensive term which refers to techniques and approaches of inquiry employed to gain insights into patients' understanding, view point, experience, and their interpretation of social environment. This exploratory method aim is used to provide an in-depth investigation on answers to questions which is not attainable through quantitative research methods. It is mostly used in social sciences and humanities. However it has

been employed widely in medical sciences recently (21). The current study was carried out in 2014 on patients with Sickle-shaped Cell Disease in Kohkiluyeh-Boyer-Ahmad Province, Iran. Having selected the participants on the basis of inclusion criteria, required instructions were given to the participants and the time and place of the interview were declared. We got written consent from each participant. As the goal of qualitative sampling is to gain an understanding of the event, sampling was done on the basis of the aim of the study. In-depth interviews were carried out to collect data. This method has no specific structure in which researcher seeks for interviewees' experienced/perceived concepts on the main question of the study. The duration of interviews varied between 25-40 minutes. A second session of interview was held if needed. Interviews were pursued until required data was elicited and no new code was obtained. Having carried out 11 interviews we reached to a point where data were repetitive and interviewing process was stopped. A 7-stage Colaizzi Method was used to analyze and codify the collected data. In order to review the data by the interviewees, the interview (partial or complete) transcription along with the primary codes were available to them. It was to compare the extracted ideas by the researchers with interviewees' opinions. Interviewees' comments to correct the researcher's ideas and understanding were included. In case of ambiguity in codes and data, interviewees were asked to provide more explanations.

RESULTS

Participants consisted of 11 patients (4 females and 7 males) with Sickle Cell Disease. Their education level varied from elementary to high school diploma. Participants' age ranged from 18 to 44. 100% of participants complained of pain and described it as its worst experience. 40 first-level codes and 3 second-level codes were extracted including the cause of pain recurrence, pain consequences, and the type of pain which altogether created the main concept of pain.

The cause of pain recurrence

Participants mentioned a variety of causes of pain attacks including physical reasons such as dehydration, infection, cold, hot or cold weather, too much physical activities and exercise, immobility, decreased intake of fluids, not taking or missing a dose of medication, consecutive baths,

decreased level of hemoglobin, severe menstrual pain, increased level of mineral iron, or psychological reasons such as anger, tension, stress, being badly treated, or thinking over problems. Participant No 3: "The pain occurs when I get angry. In addition, dehydration and infection during the summer cause the recurrence of crises". Participant No 5: "Pain starts when I run outside in cold weather or get too much exercise. It occurs suddenly in my legs and spread to my chest and my whole body. Stress also causes pain. My doctors strongly suggest I be relaxed and away from stressful situations".

Pain consequences

All participants described the periodic episodes of pain as the worst and unpleasant experience they have had ever which lead to additional mental, physical, and social difficulties. Bones pain, muscles pain, extreme psychological and mental pressure, inability in hiding the pain, invisibility of vessels, feeling hot, sweating, immobility, isolation from people, not being able to tolerate people around, thought of hurting themselves or committing suicide, numbness, disability, cardiovascular and lung problems were consequences of pain mentioned by the participants. Participant No 11: "I sweat a lot when I am having pain. I cannot move and I feel like I am disabled". Participant No 3: "When pains occur I cannot stand anyone. I become short-tempered and I fight with my family. I am depressed and isolated from others and want to die. Bad thoughts haunt me and I think of committing suicide".

The type of pain

Participants believe that pain is their major problem. They stated that the pain is so severe that they are not able to bear it. In addition, they complained of prolonged duration of pain and the variety of problems it has created. Severity, irregularity, unpredictability, consecutiveness, and suddenness of pains were the problems participants complained about. Participant No 1: "pains are irregular. I may have pain for 15 days a month. I may experience no pain for 4 or 5 months as well. There is no specific time for it to occur. At times it may recur when I am sitting down and relaxing". Participant No 5: "Pain starts when I run outside in cold weather or get too much exercise. It occurs suddenly in my legs and spread to my chest and my whole body. Stress also causes pain. My doctors strongly suggest I be relaxed and away from stressful situations. When the pain is so severe I

need to hospitalized or receive narcotics outpatiently".

DISCUSSION

Our findings reveal that Sickle Cell Disease (SCD) has a variety of symptoms. Periodic episodes of pain, called crises, are a major symptom of sickle cell anemia. The pain may vary in intensity and can last for a few hours to a few weeks. In their retrospective study, Boid et al. (2014) investigated the severity and the region of pain recurrence in patients aged 14.3 ± 18.6 referred to hospitals in the west of India and Jamaica over the period from 2006 to 2010. 8 regions of pain were reported in 101 patients (42 females, 59 males). Lower body members were the commonest regions where pains recur (44.6%). In 60.3% of patients one region of pain was reported. 75.2% of patients were diagnosed to have severe pain. All patients with "severe" or "mild to moderate" pains were treated using narcotic medicines. 55.3% of patients with severe pain needed the second line of medicinal treatment, and 14.5% needed the third line of treatments. Painful crises in patients with SCD are accompanied with severe pain which needs a second line of treatment to relieve the pain (22). Episodes of severe pain in SCD result from blockage in vessels or necrosis in bones and tissues. Severity and the duration of pains aggravate by aging. Stimulations like stress, dehydration, infection, being exposed to cold weather, too much exercise, and being at high altitudes cause the recurrence or increase of pain. Episodes of severe pain in patients with SCD provoke stress, anxiety, depression, isolation, aggressiveness, and thought of committing suicide. Using narcotics to relieve the periodic pains in such patients may result in addiction. It is in consistency with Ahmadi (2015) that states in spite of increased life expectancy in patients with SCD, they still suffer from severe pains. Episodes of pain are chronic and unpredictable. Frequent refers to emergency wards and hospitalization lead to decreased self-confidence, joblessness, low quality of life, depression, stress, and anxiety (22). Participants complain about irregular severe pains which disturb their normal life. Signorli (2013) studied the frequency of crises in 47 patients in Brazil. Patients were $++18$ years old and suffering from SCD. They were selected randomly. 78% of patients were black and 59.6% were females with average age of 30. His findings revealed that the average recurrence of SCD pain attacks is 7 episodes per

year. Findings show that pain needs to be diagnosed, measured, and treated properly in patients with SCD (8).

CONCLUSION

Considering the fact that, patients with SCD

experience severe and unbearable episodes of pain which create a wide variety of problems in their daily lives, their needs and problems must be addressed specifically. Particular programs are required to support these patients, meet their needs, and improve the quality of their lives.

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