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## RESEARCH OF THE QUALITY OF LIFE AND STRESS LEVEL OF PEOPLE WITH HEMOPHILIA IN KAZAKHSTAN

This article is devoted to the study of quality of life and stress levels in patients diagnosed with hemophilia. Hemophilia is a hereditary disease characterized by a sharp decrease in blood coagulation ability due to the low level of factor 8 or factor 9 in the blood, which manifests itself in the first months or years of life in the form of external bleeding and internal hemorrhage. Early disability, physical disabilities limit the adaptation of hemophilia patients in all age groups. Constant fear, anxiety, uncertainty in the future change the structure of the individual. The hallmarks of patients with hemophilia are the presence of an increased level of personal anxiety, depression, low self-esteem, low social activity. The purpose of the study is to study the features of quality of life and the level of stress resistance in patients with hemophilia. The scientific novelty lies in the fact that for the first time in Kazakhstan, the study of the psychological characteristics of patients with hemophilia was carried out, in particular the degree of quality of life and the level of stress. As a result, the quality of life of patients with hemophilia was revealed in such areas as physical and psychological well-being, self-perception, microsocial support, social well-being, as well as subjective self-assessment of their quality of life and their health.

**Key words:** hemophilia, hemophilia patients, coping behavior, stress, quality of life, stress tolerance, resilience.

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### Қазақстандағы гемофилияға шалдыққан адамдардың стресс деңгейі мен өмір сапасын зерттеу

Бұл мақала гемофилия диагнозы бар емделушілердің стресс деңгейі мен өмір сапасын зерттеуге арналған. Гемофилия – қанда 8 фактор немесе фактор 9 деңгейі аздығынан қан ұюының күрт төмендеуімен сипатталатын және адам өмірінің алғашқы айларынан немесе жылдарынан сыртқы қан ағуы мен ішкі қан ағулармен көрініс табатын тұқым қуалайтын ауру. Ерте жастағы мүгедектік, физикалық кемістіктер гемофилия ауруына шалдыққандардың бейімделуін үнемі шектейді. Тұрақты қорқыныш, үрей, болашақтағы сенімсіздік тұлға құрылымын өзгертеді. Гемофилияға шалдыққан емделушілердің ерекше белгілері, жеке тұлғаның үрейленуі, депрессия деңгейінің жоғары болуы, өзін-өзі төмен бағалауы, төмен әлеуметтік белсенділік болып табылады.

**Түйін сөздер:** гемофилия, гемофилия диагнозы бар емделушілер, копинг-жүріс-тұрысы, стресске төзімділік, стресс, өмір сапасы, резильенттілік.

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### Исследование качества жизни и уровня стресса у лиц с гемофилией в Казахстане

Данная статья посвящена изучению исследований качества жизни и уровня стресса у пациентов с диагнозом гемофилия. Гемофилия – это наследственное заболевание, характеризующееся резким снижением способности крови к свертыванию в силу низкого уровня 8 фактора или фактора 9 в крови, проявляющееся в первые месяцы или годы жизни в виде наружных кровотечений и внутренних кровоизлияний. Ранняя инвалидизация, физические недостатки ограничивают адаптацию больных гемофилией во всех возрастных группах. Постоянный страх, тревога, неуверенность в будущем изменяют структуру личности. Вследствие этого, отличительными признаками пациентов с гемофилией являются наличие повышенного уровня

личностной тревожности, депрессии, низкая самооценка, низкая социальная активность. Цель данного исследования – изучение особенностей качества жизни и уровня стрессоустойчивости у пациентов с гемофилией. Научная новизна исследования состоит в том, что впервые в Казахстане проводилось изучение психологических особенностей у пациентов с гемофилией. В результате были выявлены качество жизни пациентов с гемофилией по таким сферам, как физическое и психологическое благополучие, самовосприятие, микросоциальная поддержка, социальное благополучие, а также субъективная самооценка своего качества жизни и своего здоровья.

**Ключевые слова:** гемофилия, пациенты с гемофилией, копинг-поведение, стресс, качество жизни, стрессоустойчивость, резильентность.

## Introduction

Currently, there is a tendency of reevaluation of value orientations and beliefs, corresponding to the new social reality in Kazakhstan. At the same time, Kazakhstan also faced socio-economic, organizational and medical problems, in which changes in coping behavior, resilience and resistance to stress were the most important difficulties for people with hemophilia. Since, in addition to general difficulties, limitations are imposed due to the characteristics of the disease. Social factors that caused various changes in the structure of coping behavior, resilience and resistance to stress, primarily affected persons with hemophilia susceptible to social and domestic changes. Coping – strategies, resilience and stress resistance of people with hemophilia are the first to undergo changes caused by changes in society and the current status of hemophilia disease. In this regard, the problem of studying the resilience and stress tolerance, in revealing the psychological aspects of the formation, development, transformation, refraction of the socio-psychological characteristics of people with hemophilia, is of particular relevance. Hemophilia is a hereditary disease characterized by a sharp decrease in the ability of blood to clot due to the low level of factor 8 in the blood, manifested in the first months or years of life in the form of external bleeding and internal hemorrhages (Guidelines, 2012:7). In most cases, it has a severe course and is accompanied by frequent relapses. Early disability, physical defects limit the adaptation of patients with hemophilia in all age groups. Constant fear, anxiety, uncertainty in the future change the personality structure, its reactivity, and worsen the adaptation of patients with hemophilia sometimes contribute to the emergence of mental disorders. Russian studies interested us because they have a similar socio-economic situation. Western countries studies differed in their research on adults with hemophilia, the presence of longitudinal studies. In the scientific literature there are studies of the medical aspects of persons with hemophilia, but at the same time, the problem of the psychological characteris-

tics of patients with hemophilia in Kazakhstan has not been studied to date.

## Main part

Any chronic disease limits the patient's social adaptation in all age periods, primarily due to a violation of the personality-environment interaction and changes in the system of personality relations in connection with the disease. Despite the formal provision of drugs for the treatment of persons with hemophilia, there is no timely provision of them, there is no socio-psychological support for such persons, which violates the holistic approach to the general support of individuals in Kazakhstan. Nor were scientific studies conducted on the socio-psychological conditions and characteristics of persons with hemophilia. In Kazakhstan, about one and half thousand patients suffer from various forms of hemophilia.

Ryabov V.I. studied 35 children with hemophilia, all boys, aged from 9 years to 15 years. And he obtained the following results:

1. Psychological features of people with hemophilia consist in emotional instability of 78.6% of patients with a high proportion of extrovert orientation / 43.8% of people with hemophilia patients, with introverted orientation- 38.6.

2. For patients with hemophilia, there is a high level of personal anxiety, including high of anxiety -56.12%, and a lack of low personal anxiety.

3. Response to the disease of children suffering from hemophilia. The most frequent response types are anxious -13.6 and ergopathic type of response-10,6% . The high occurrence of anxiety-neurasthenic (20%) and anxiety-hypochondriac (16%) types was revealed, which indicates the personal maladjustment of hemophilia patients ( Ryabov ,1993:16).

The research of the personality characteristics of patients with hemophilia showed that the duration of the disease and the appearance of a complication (hemophilic hemorrhages) naturally changed the personality attitude of the disease in most patients. In

56.1% of children in the process of the disease, there appeared a feeling of inferiority, irritability due to the presence of visible physical defects. Strengthening the concerns of patients with regard to their disease and the future, along with the emergence of a more adequate attitude to the disease, is characteristic of puberty with its high demands on its physical condition and appearance (80.0% of patients). The study of the internal picture of the disease in patients with hemophilia, in particular their response to the disease, showed a heterogeneous relationship to the disease in children and adolescents. Along with a harmonious attitude towards the disease, the leading ones turned out to be “disturbing” (37.9% of patients) and “neurasthenic” (21.2% of patients) variants, manifested by increased anxiety, emotional lability and excessive mood swings (Bagaev, 2002:33).

Nalesnaya I.M. conducted research of adolescents with hemophilia and came to specific results. 1. Due to the lack of close relationships of trust with significant adults. Adolescents with hemophilia have unsatisfied family situation in general. In the perception of adolescents, these groups have the status of hostile and autonomous, which is reflected in the implementation of authoritative style in terms of decision making and the lowest level of encouragement of autonomy of teenager.

2. For teenagers with diagnosis of hemophilia, emotionally negative attitudes toward peers are characteristic, as well as in extreme cases, the absence of friendships. In the group of healthy adolescents, there is identification with the rollers of informal groups. Thus, “hemophilia” hampers the process of developing relations between men and women, and also hampers the process of development of relations and cooperation with peers, which are characterized by the absence of identifications with the roles of reference micro-level groups.

3. For adolescents with diagnosis of hemophilia, there is a lack of time perspectives, uncertainty and continuity of ideas about the future, the subjective significance of the past and, at the same time, negative attitude towards it. In this regard, the future is subjectively important, the presence of specific goals.

4. Adolescents with diagnosis of hemophilia have lower self-esteem on the happiness scale compared with their healthy peers associated with the disease, experiencing their illness. Characteristically high differences between real and ideal self-esteem.

5. Adolescents with diagnosis of hemophilia have a low level of reflection, unattainable lifestyle, and discrepancy between the feelings of adult, other people, and slower development of self-

consciousness compared to healthy peers. Lack of causal relationships with adults, the absence of any projections in the future, underdevelopment of personal reflections, undifferentiated lifestyle, lack of identification with the roles of reference micro-level groups, propensity to insults caused by the need to acquire identity - the central task of adolescence.

6. The solution of some problems for children with diagnosis of hemophilia requires a later time (for example, release from parental care), difficult in terms of resources to solve them (the process of acquiring identity) (Nalesnaya 2009:23).

In DuTreil S. opinion, social problems associated with employment, disability and absences in the workplace also negatively affect the patient’s perception of self-respect and self-esteem. Low self-esteem correlates with depression, anxiety, and low social expectations; hemophilia patients have lower self-esteem than healthy people. In addition to these problems, patients with inhibitors may experience bleeding that cannot be stopped, and embarrassment that they cannot compete with other men in sports, in personal relationships and in physical appearance. The appearance of an inhibitor to factor 8/9 is considered the most serious complication associated with the treatment of hemophilia because inhibitors inhibit medication treatment. The QoL study included 50 patients with hemophilia and 78 healthy adults. Patients with inhibitors experience depression, social isolation, fatigue, loss of interest, and irritability, and depression worsens health. DuTreil S. offers a list of measures to improve the psychological state of patients with inhibitors: referral to psychological counseling, using the strategy of strong qualities, cognitive-behavioral method, encouraging new social contacts and openness, communication with family and medical workers, meditation (DuTreil 2014:118).

Abroad in western countries, where high levels of medicine and social assistance may cause negative psychological signs, such as feelings of depression and frustration associated with the limitations and pain caused by illness in everyday life. But such phenomena are are and do not have a common tendency (Laura Palareti, Silvia Pot, Frederica Cassisi & Francesca Emiliani 2015).

In Kazakhstan, quality of life research in medicine was used, but questionnaires adapted for medicine were used – PedsQL (Brimzhanova 2017). There are different definitions of resilience, but we take the definition of the American Psychological Association, which defines sustainability and a process of good adaptation in the face of disasters, injuries, tragedies, threats and sources of stress

(Southwick, S. M., Bonanno, G. A., Masten, A. S., Panter-Brick, C., & Yehuda, R. 2014).

Stress tolerance is resistance to stress without undesirable and irrational changes (Bengt Schager 2009:3). The problem of stress and socio-psychological assistance in stressful situations requires an integrated approach, precisely from the standpoint of a new area of scientific knowledge - "stress medicine" ( Reshke , Schröder 2010 , Khaurand, S,Ulrikh, G., Veniger, M. 2015). The use of the theory of steps of destabilization by Schreder is a conceptually sound and relevant practice of the basis for the regulation of behavior, including patients with hemophilia (Shredder 2016 ).

The theoretical significance of the study is that a socio-psychological research of patients with hemophilia in Kazakhstan will be conducted for the first time. Based on the results obtained, approaches to the socio-psychological support of people with hemophilia in Kazakhstan, including socio-psychological correction, can be developed. It is necessary to open special hematological centers. It is important to ensure psychosocial support for patient care and well-being hemophilia patient (and their families) into the hematology center. It is important that in developed countries there are hematological centers with the position of a social worker or a psychologist (Massimo, Gary , Jiménez-Yuste, Rolf Ljung , Pier Mannuccio, Gianluigi, Eduardo & Silva 2015), (Masoume Rambod, Farkhondeh Sharif, Zahra Molazem, Kate Khair & Sylvia von Mackensen 2018).

Moreover, there are perennial studies abroad that emphasize the effectiveness of social and psychological intervention in patients with hemophilia. Good social and psychological support for people with hemophilia is effective even with the presence of concomitant diseases associated with HIV, hepatitis C. Research in Germany shows good adaptation of people with hemophilia, professional employment, social activity, orientation to the future, positive family relations. Psychotherapeutic influence have a positive effect on medical indicators (Alexandra Stefanie 2017:98).

The effectiveness of the socio-psychological correction was noted in countries where there is no high level of medical care, but there is socio-psychological support for patients with hemophilia. (Frederica Cassis 2007). Socio-psychological support, we understand is social and psychological assistance, including assistance in finding a job, helping choose of future profession, motivating social activity, supporting extensive interpersonal contacts, etc.

## Materials and Methods

16 adult patients with hemophilia in Kazakhstan were interviewed in May 2020, all males were from 23 to 41 years old. The World health organization quality of life (WHOQOL) – BREF and the Leipzig Express Chronic Stress Test , consisting of 7 statements and 4 answer choices were used. The study was conducted using the Googleforms application, the Whatsup individual and specialized group mailing list for people with hemophilia. Also included were several points for collecting socio-anamnestic information in patients with hemophilia. The control group of subjects consisted of 9 adult men from 23 to 42 years old.

## Results

All respondents in their place of residence have healthcare organizations - a clinic and (or) a hospital. Patients with hemophilia were hard of hemophilia level 14 people, or with light hemophilia-2, with a moderate degree were absent. Most (87.5 percent-14 people) of people with hemophilia live in the city, the rest in villages. By marital status, the majority are married — 13 people, or 81.2%, and single people — 3 or 18.8%. Of those with hemophilia, only one is without a disability group, the rest are with disabilities: 1 has a first group, 7 people with a second group and 7 people with a third group. Generalized social data for people with hemophilia are presented in table 1.

**Table 1** – Social data of people with hemophilia

	marital status		social status		type of settlement		severity of hemophilia			disability group			
	married	single	worker	unemployed	city	village	hard	meddle	light	1	2	3	abcent
numbers	13	3	12	4	14	2	14	0	2	1	7	7	1
percents	81,2	18,8	75	25	87,5	12,5	87,5	0	12,5	6	44	44	6

43.8% of respondents – 7 people rated their quality of life of patients with hemophilia as good, 1 person rated it as bad, and half of the respondents (8 people) were not determined in assessing their quality of life. At the same time, 4 people or 25%, a patient with hemophilia rated their health as poor, 2 people or 12.5% rated it as good, more than half of patients with hemophilia did not determine their health assessment, namely 10 people or 62.5%. The interpretation of the quality of life questionnaire in 4 areas was as follows: Physical and psychological well-being-18.38;

Self-perception-17.25; Microsocial support-11, Social well-being-24.6875. In the control group of “healthy” subjects there were 9 adult men, from 23 to 42 years old. Also, most of them live in the city -77.8%, all have access to a medical organization. Only 1 person noted a chronic disease, no one has a disability group, 77.8% -7 people work. 6 of them were single 66.7%, and married 3 people, 33.3%. According to the Leipzig rapid test for detecting chronic stress, the level of stress in patients with

hemophilia is higher than in the control group of healthy subjects.

### Conclusion

The revealed results and the set of methods performed are intermediate studies as a pilot study. In the future, it is planned to increase the number of studied and the inclusion of used techniques for resilience, stress resistance and coping strategies. While maintaining the identified trends in patients with hemophilia, corrective socio-psychological programs can be introduced to improve the quality of life. It is necessary to open special hematological centers, where complex intervention for hemophilia patients, both medical services, social and psychological services, would be carried out. It is necessary to implement a hematological center in the position of a social worker or psychologist. The inclusion of psychological services in multidisciplinary teams can improve the psychosocial health of patients and their results.

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