

GEORGIAN MEDICAL NEWS

ISSN 1512-0112

NO 2 (359) Февраль 2025

ТБИЛИСИ - NEW YORK



ЕЖЕМЕСЯЧНЫЙ НАУЧНЫЙ ЖУРНАЛ

Медицинские новости Грузии
საქართველოს სამედიცინო სიახლენი

GEORGIAN MEDICAL NEWS

Monthly Georgia-US joint scientific journal published both in electronic and paper formats of the Agency of Medical Information of the Georgian Association of Business Press.
Published since 1994. Distributed in NIS, EU and USA.

GMN: Georgian Medical News is peer-reviewed, published monthly journal committed to promoting the science and art of medicine and the betterment of public health, published by the GMN Editorial Board since 1994. GMN carries original scientific articles on medicine, biology and pharmacy, which are of experimental, theoretical and practical character; publishes original research, reviews, commentaries, editorials, essays, medical news, and correspondence in English and Russian.

GMN is indexed in MEDLINE, SCOPUS, PubMed and VINITI Russian Academy of Sciences. The full text content is available through EBSCO databases.

GMN: Медицинские новости Грузии - ежемесячный рецензируемый научный журнал, издаётся Редакционной коллегией с 1994 года на русском и английском языках в целях поддержки медицинской науки и улучшения здравоохранения. В журнале публикуются оригинальные научные статьи в области медицины, биологии и фармации, статьи обзорного характера, научные сообщения, новости медицины и здравоохранения. Журнал индексируется в MEDLINE, отражён в базе данных SCOPUS, PubMed и ВИНТИ РАН. Полнотекстовые статьи журнала доступны через БД EBSCO.

GMN: Georgian Medical News – საქართველოს სამედიცინო სიახლენი – არის ყოველთვიური სამეცნიერო სამედიცინო რეცენზირებადი ჟურნალი, გამოიცემა 1994 წლიდან, წარმოადგენს სარედაქციო კოლეგიისა და აშშ-ის მეცნიერების, განათლების, ინდუსტრიის, ხელოვნებისა და ბუნებისმეტყველების საერთაშორისო აკადემიის ერთობლივ გამოცემას. GMN-ში რუსულ და ინგლისურ ენებზე ქვეყნდება ექსპერიმენტული, თეორიული და პრაქტიკული ხასიათის ორიგინალური სამეცნიერო სტატიები მედიცინის, ბიოლოგიისა და ფარმაციის სფეროში, მიმოხილვითი ხასიათის სტატიები.

ჟურნალი ინდექსირებულია MEDLINE-ის საერთაშორისო სისტემაში, ასახულია SCOPUS-ის, PubMed-ის და ВИНТИ РАН-ის მონაცემთა ბაზებში. სტატიების სრული ტექსტი ხელმისაწვდომია EBSCO-ს მონაცემთა ბაზებიდან.

WEBSITE

www.geomednews.com

К СВЕДЕНИЮ АВТОРОВ!

При направлении статьи в редакцию необходимо соблюдать следующие правила:

1. Статья должна быть представлена в двух экземплярах, на русском или английском языках, напечатанная через **полтора интервала на одной стороне стандартного листа с шириной левого поля в три сантиметра**. Используемый компьютерный шрифт для текста на русском и английском языках - **Times New Roman (Кириллица)**, для текста на грузинском языке следует использовать **AcadNusx**. Размер шрифта - **12**. К рукописи, напечатанной на компьютере, должен быть приложен CD со статьей.

2. Размер статьи должен быть не менее десяти и не более двадцати страниц машинописи, включая указатель литературы и резюме на английском, русском и грузинском языках.

3. В статье должны быть освещены актуальность данного материала, методы и результаты исследования и их обсуждение.

При представлении в печать научных экспериментальных работ авторы должны указывать вид и количество экспериментальных животных, применявшиеся методы обезболивания и усыпления (в ходе острых опытов).

4. К статье должны быть приложены краткое (на полстраницы) резюме на английском, русском и грузинском языках (включающее следующие разделы: цель исследования, материал и методы, результаты и заключение) и список ключевых слов (key words).

5. Таблицы необходимо представлять в печатной форме. Фотокопии не принимаются. **Все цифровые, итоговые и процентные данные в таблицах должны соответствовать таковым в тексте статьи**. Таблицы и графики должны быть озаглавлены.

6. Фотографии должны быть контрастными, фотокопии с рентгенограмм - в позитивном изображении. Рисунки, чертежи и диаграммы следует озаглавить, пронумеровать и вставить в соответствующее место текста **в tiff формате**.

В подписях к микрофотографиям следует указывать степень увеличения через окуляр или объектив и метод окраски или импрегнации срезов.

7. Фамилии отечественных авторов приводятся в оригинальной транскрипции.

8. При оформлении и направлении статей в журнал МНГ просим авторов соблюдать правила, изложенные в «Единых требованиях к рукописям, представляемым в биомедицинские журналы», принятых Международным комитетом редакторов медицинских журналов - <http://www.spinesurgery.ru/files/publish.pdf> и http://www.nlm.nih.gov/bsd/uniform_requirements.html В конце каждой оригинальной статьи приводится библиографический список. В список литературы включаются все материалы, на которые имеются ссылки в тексте. Список составляется в алфавитном порядке и нумеруется. Литературный источник приводится на языке оригинала. В списке литературы сначала приводятся работы, написанные знаками грузинского алфавита, затем кириллицей и латиницей. Ссылки на цитируемые работы в тексте статьи даются в квадратных скобках в виде номера, соответствующего номеру данной работы в списке литературы. Большинство цитированных источников должны быть за последние 5-7 лет.

9. Для получения права на публикацию статья должна иметь от руководителя работы или учреждения визу и сопроводительное отношение, написанные или напечатанные на бланке и заверенные подписью и печатью.

10. В конце статьи должны быть подписи всех авторов, полностью приведены их фамилии, имена и отчества, указаны служебный и домашний номера телефонов и адреса или иные координаты. Количество авторов (соавторов) не должно превышать пяти человек.

11. Редакция оставляет за собой право сокращать и исправлять статьи. Корректур авторам не высылаются, вся работа и сверка проводится по авторскому оригиналу.

12. Недопустимо направление в редакцию работ, представленных к печати в иных издательствах или опубликованных в других изданиях.

При нарушении указанных правил статьи не рассматриваются.

REQUIREMENTS

Please note, materials submitted to the Editorial Office Staff are supposed to meet the following requirements:

1. Articles must be provided with a double copy, in English or Russian languages and typed or computer-printed on a single side of standard typing paper, with the left margin of 3 centimeters width, and 1.5 spacing between the lines, typeface - **Times New Roman (Cyrillic)**, print size - 12 (referring to Georgian and Russian materials). With computer-printed texts please enclose a CD carrying the same file titled with Latin symbols.

2. Size of the article, including index and resume in English, Russian and Georgian languages must be at least 10 pages and not exceed the limit of 20 pages of typed or computer-printed text.

3. Submitted material must include a coverage of a topical subject, research methods, results, and review.

Authors of the scientific-research works must indicate the number of experimental biological species drawn in, list the employed methods of anesthetization and soporific means used during acute tests.

4. Articles must have a short (half page) abstract in English, Russian and Georgian (including the following sections: aim of study, material and methods, results and conclusions) and a list of key words.

5. Tables must be presented in an original typed or computer-printed form, instead of a photocopied version. **Numbers, totals, percentile data on the tables must coincide with those in the texts of the articles.** Tables and graphs must be headed.

6. Photographs are required to be contrasted and must be submitted with doubles. Please number each photograph with a pencil on its back, indicate author's name, title of the article (short version), and mark out its top and bottom parts. Drawings must be accurate, drafts and diagrams drawn in Indian ink (or black ink). Photocopies of the X-ray photographs must be presented in a positive image in **tiff format**.

Accurately numbered subtitles for each illustration must be listed on a separate sheet of paper. In the subtitles for the microphotographs please indicate the ocular and objective lens magnification power, method of coloring or impregnation of the microscopic sections (preparations).

7. Please indicate last names, first and middle initials of the native authors, present names and initials of the foreign authors in the transcription of the original language, enclose in parenthesis corresponding number under which the author is listed in the reference materials.

8. Please follow guidance offered to authors by The International Committee of Medical Journal Editors guidance in its Uniform Requirements for Manuscripts Submitted to Biomedical Journals publication available online at: http://www.nlm.nih.gov/bsd/uniform_requirements.html
http://www.icmje.org/urm_full.pdf

In GMN style for each work cited in the text, a bibliographic reference is given, and this is located at the end of the article under the title "References". All references cited in the text must be listed. The list of references should be arranged alphabetically and then numbered. References are numbered in the text [numbers in square brackets] and in the reference list and numbers are repeated throughout the text as needed. The bibliographic description is given in the language of publication (citations in Georgian script are followed by Cyrillic and Latin).

9. To obtain the rights of publication articles must be accompanied by a visa from the project instructor or the establishment, where the work has been performed, and a reference letter, both written or typed on a special signed form, certified by a stamp or a seal.

10. Articles must be signed by all of the authors at the end, and they must be provided with a list of full names, office and home phone numbers and addresses or other non-office locations where the authors could be reached. The number of the authors (co-authors) must not exceed the limit of 5 people.

11. Editorial Staff reserves the rights to cut down in size and correct the articles. Proof-sheets are not sent out to the authors. The entire editorial and collation work is performed according to the author's original text.

12. Sending in the works that have already been assigned to the press by other Editorial Staffs or have been printed by other publishers is not permissible.

**Articles that Fail to Meet the Aforementioned
Requirements are not Assigned to be Reviewed.**

ავტორთა საქურაღებოლ!

რედაქციაში სტატიის წარმოდგენისას საჭიროა დაიცვათ შემდეგი წესები:

1. სტატია უნდა წარმოადგინოთ 2 ცალად, რუსულ ან ინგლისურ ენებზე დაბეჭდილი სტანდარტული ფურცლის 1 გვერდზე, 3 სმ სიგანის მარცხენა ველისა და სტრიქონებს შორის 1,5 ინტერვალის დაცვით. გამოყენებული კომპიუტერული შრიფტი რუსულ და ინგლისურენოვან ტექსტებში - **Times New Roman (Кириллица)**, ხოლო ქართულენოვან ტექსტში საჭიროა გამოვიყენოთ **AcadNusx**. შრიფტის ზომა – 12. სტატიას თან უნდა ახლდეს CD სტატიით.

2. სტატიის მოცულობა არ უნდა შეადგენდეს 10 გვერდზე ნაკლებს და 20 გვერდზე მეტს ლიტერატურის სიის და რეზიუმეების (ინგლისურ, რუსულ და ქართულ ენებზე) ჩათვლით.

3. სტატიაში საჭიროა გაშუქდეს: საკითხის აქტუალობა; კვლევის მიზანი; საკვლევი მასალა და გამოყენებული მეთოდები; მიღებული შედეგები და მათი განსჯა. ექსპერიმენტული ხასიათის სტატიების წარმოდგენისას ავტორებმა უნდა მიუთითონ საექსპერიმენტო ცხოველების სახეობა და რაოდენობა; გაუტკივარებისა და დაძინების მეთოდები (მწვავე ცდების პირობებში).

4. სტატიას თან უნდა ახლდეს რეზიუმე ინგლისურ, რუსულ და ქართულ ენებზე არანაკლებ ნახევარი გვერდის მოცულობისა (სათაურის, ავტორების, დაწესებულების მითითებით და უნდა შეიცავდეს შემდეგ განყოფილებებს: მიზანი, მასალა და მეთოდები, შედეგები და დასკვნები; ტექსტუალური ნაწილი არ უნდა იყოს 15 სტრიქონზე ნაკლები) და საკვანძო სიტყვების ჩამონათვალი (key words).

5. ცხრილები საჭიროა წარმოადგინოთ ნაბეჭდი სახით. ყველა ციფრული, შემაჯამებელი და პროცენტული მონაცემები უნდა შეესაბამებოდეს ტექსტში მოყვანილს.

6. ფოტოსურათები უნდა იყოს კონტრასტული; სურათები, ნახაზები, დიაგრამები - დასათაურებული, დანომრილი და სათანადო ადგილას ჩასმული. რენტგენოგრაფიების ფოტოასლები წარმოადგინეთ პოზიტიური გამოსახულებით **tiff** ფორმატში. მიკროფოტოსურათების წარწერებში საჭიროა მიუთითოთ ოკულარის ან ობიექტივის საშუალებით გადიდების ხარისხი, ანათალების შედეგების ან იმპრეგნაციის მეთოდი და აღნიშნოთ სურათის ზედა და ქვედა ნაწილები.

7. სამამულო ავტორების გვარები სტატიაში აღინიშნება ინიციალების თანდართვით, უცხოურისა – უცხოური ტრანსკრიპციით.

8. სტატიას თან უნდა ახლდეს ავტორის მიერ გამოყენებული სამამულო და უცხოური შრომების ბიბლიოგრაფიული სია (ბოლო 5-8 წლის სიღრმით). ანბანური წყობით წარმოდგენილ ბიბლიოგრაფიულ სიაში მიუთითეთ ჯერ სამამულო, შემდეგ უცხოელი ავტორები (გვარი, ინიციალები, სტატიის სათაური, ჟურნალის დასახელება, გამოცემის ადგილი, წელი, ჟურნალის №, პირველი და ბოლო გვერდები). მონოგრაფიის შემთხვევაში მიუთითეთ გამოცემის წელი, ადგილი და გვერდების საერთო რაოდენობა. ტექსტში კვადრატულ ფხიხლებში უნდა მიუთითოთ ავტორის შესაბამისი N ლიტერატურის სიის მიხედვით. მიზანშეწონილია, რომ ციტირებული წყაროების უმეტესი ნაწილი იყოს 5-6 წლის სიღრმის.

9. სტატიას თან უნდა ახლდეს: ა) დაწესებულების ან სამეცნიერო ხელმძღვანელის წარდგინება, დამოწმებული ხელმოწერითა და ბეჭდით; ბ) დარგის სპეციალისტის დამოწმებული რეცენზია, რომელშიც მითითებული იქნება საკითხის აქტუალობა, მასალის საკმაობა, მეთოდის სანდოობა, შედეგების სამეცნიერო-პრაქტიკული მნიშვნელობა.

10. სტატიის ბოლოს საჭიროა ყველა ავტორის ხელმოწერა, რომელთა რაოდენობა არ უნდა აღემატებოდეს 5-ს.

11. რედაქცია იტოვებს უფლებას შეასწოროს სტატია. ტექსტზე მუშაობა და შეჯერება ხდება საავტორო ორიგინალის მიხედვით.

12. დაუშვებელია რედაქციაში ისეთი სტატიის წარდგენა, რომელიც დასაბეჭდად წარდგენილი იყო სხვა რედაქციაში ან გამოქვეყნებული იყო სხვა გამოცემებში.

აღნიშნული წესების დარღვევის შემთხვევაში სტატიები არ განიხილება.

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PSYCHOLOGICAL FEATURES OF THE SUBJECTIVE PERCEPTION OF THE QUALITY OF LIFE OF PATIENTS WITH HEPATOCEREBRAL DYSTROPHY

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Abstract.

Introduction: Wilson's disease (WD) is a rare autosomal recessive disorder of copper metabolism. The clinical features associated with WD are highly varied. However, subtypes generally reflect neurological, hepatic, and psychiatric symptoms. Cognitive dysfunction is often reported, and patients suffer from additional depressive symptoms. The goal of our work was detection of psychological features of the subjective perception of the quality of life of patients with Wilson's disease

Material and methods: Psychodiagnostic examination was performed for 33 patients with WD. All patients were presented as General group (GG), trembling form (TF), trembling-rigid form (TRF), rigid-arrhythmic-hyperkinetic form (RAHF), extrapyramidal-cortical form (ECF) and abdominal form (AF). To determine the characteristics of the subjective perception of quality of life (QOL) by patients with WD and the effectiveness of their social functioning in general, a method for assessing the integrative indicator of quality of life according to recommendations J. Mezzich. Quality of Life Index is a concise instrument for comprehensive, culture-informed, and self-rated assessment of health-related quality of life. It is composed of 10 items (from physical well-being to global perception of quality of life). Each item is rated on a 10-point scale. The subjective perception was eliminated with general perception, physical well-being, psychological well-being, self-care and independence in actions, working capacity, interpersonal interaction, socio-emotional support, civil and service support, personal realization, spiritual realization. Detection of degree of voluntary-regulatory, cognitive, speech, motor and emotional disorders were performed also for all patients.

Results: The results of the study showed that a decrease in the assessments of the general perception of QOL ($\beta=-0.64$, $p=0.00$) and the nature of interpersonal interaction ($\beta=-0.56$, $p=0.01$) in patients with WD is associated with an increase in the level of depression. Satisfaction with their physical well-being in them is associated with the level of depression ($\beta=-0.55$, $p=0.00$) and movement disorders ($\beta=-0.30$, $p=0.03$). Subjective perception of psychological well-being in these patients is associated with the degree of anxiety ($\beta=-0.48$, $p=0.00$), depression ($\beta=0.55$, $p=0.00$) and the level of voluntary regulation disorders ($\beta=-0.65$, $p=0.00$). Satisfaction with the level of self-care and independence in actions is associated with the degree of anxiety ($\beta=-0.50$, $p=0.00$), rigidity ($\beta=-0.50$, $p=0.00$) and movement

disorders ($\beta=-0.47$, $p=0.00$). Subjective assessments of patients' ability to work are associated with the level of movement disorders ($\beta=-0.81$, $p=0.00$) and depression ($\beta=-0.34$, $p=0.04$). Satisfaction with socio-emotional support is associated with rigidity ($\beta=-0.87$, $p=0.00$) and anxiety ($\beta=-0.43$, $p=0.00$). Subjective perception of personal fulfillment in these patients is associated with the degree of voluntary-regulatory disorders ($\beta=-0.61$, $p=0.00$) and the level of anxiety ($\beta=-0.43$, $p=0.00$). Patients' satisfaction with their spiritual fulfillment is combined with rigidity of behavioral reactions ($\beta=-0.75$, $p=0.00$).

Thus, the features of the subjective perception of QOL in patients with WD are primarily associated with the characteristics of their emotional sphere, namely, with the level of expression of anxiety-depressive symptoms.

Conclusions: The level of subjective perception of quality of life of patients with hepatocerebral dystrophy is associated with a number of clinical and psychological factors, such as the form of the disease, its duration, and the severity of motor and emotional disorders. Thus, the highest assessments of almost all QOL parameters are noted in patients with the abdominal form of WD. Patients with extrapyramidal-cortical and rigid-arrhythmohyperkinetic forms assess their level of physical and psychological well-being significantly lower than other patients.

Key words. Cognitive impairment, hepatocerebral degeneration, psychology, prevention.

Introduction.

Wilson's disease (WD) (or hepatocerebral dystrophy (HCD)) is a rare autosomal recessive disorder of copper metabolism [1,2]. The clinical features associated with WD are highly varied. However, subtypes generally reflect neurological, hepatic, and psychiatric symptoms [3,4]. Cognitive dysfunction is often reported, and patients suffer from additional depressive symptoms [5] as in some other hepar pathology [6].

Quality of life (QOL) is currently one of the most important criteria for assessing the results of treatment of various diseases along with traditional clinical criteria. This criterion evaluates components associated and unassociated with the disease and allows for a differentiated determination of the impact of the disease itself (its symptoms and signs), as well as the treatment being carried out, on changes in the functional state and psychology of behavior, and limitation of the patient's social activity [7,8]. Quality of life is considered as an individual relationship between the individual's position in the life of

society in the context of culture, the value systems of this society and the goals of the individual himself, his plans, opportunities, and the degree of general instability. Quality of life has emerged as a crucial concept for the assessment of health and the planning of health care. Desirable features for the evaluation of quality of life include comprehensiveness, self-relatedness, cultural sensitivity, practicality and psychometric soundness. An attempt to meet these challenges led to the development of a brief multicultural quality of life instrument and to the appraisal of its applicability, reliability and validity [7,9].

Today, there are two main approaches to determining quality of life. One of them is based only on subjective assessments of the patient, or medical personnel, or the patient's relatives. The second approach involves a comprehensive assessment of the patient's quality of life, including both his own subjective picture and the assessments of his relatives and medical specialists. It should be emphasized that for the successful adaptation of patients, it is extremely important to take into account their subjective picture of quality of life, which largely reflects the compensatory resources of patients [9,10].

It has been shown that the quality of life of various patient groups is closely related to both the severity of somatic, neurological and mental pathology and the characteristics of their socio-demographic and individual psychological characteristics. Therefore, when assessing the quality of life in patients with WD, it is necessary to take into account the characteristics of their cognitive and voluntary-regulatory functions, changes in the emotional sphere, which can significantly affect the subjective perception of various aspects of their lives [11].

Concerning of said above goal of our work was detection of psychological features of the subjective perception of the quality of life of patients with Wilson's disease.

Materials and Methods.

The study was performed in accordance with the principles of the Helsinki Declaration of the World Medical Association "Ethical Principles of Medical Research Concerning Human Subjects" (2013) with written informed consent. Psychodiagnostic examination was performed for 33 patients with WD. All patients were presented as General group (GG). They were divided for trembling form (TF, n=11), trembling-rigid form (TRF, n=14), rigid-arrhythmic-hyperkinetic form (RAHF, n=3), extrapyramidal-cortical form (ECF, n=3) and abdominal form (AF, n=2). To determine the characteristics of the subjective perception of quality of life (QOL) by patients with WD and the effectiveness of their social functioning in general, a method for assessing the integrative indicator of quality of life according to recommendations J. Mezzich [12]. Quality of Life Index is a concise instrument for comprehensive, culture-informed, and self-rated assessment of health-related quality of life. It is composed of 10 items (from physical well-being to global perception of quality of life). Each item is rated on a 10-point scale. The subjective perception was eliminated with general perception, physical well-being, psychological well-being, self-care and independence in actions, working capacity, interpersonal interaction, socio-emotional support, civil and service support, personal realization, spiritual realization [12]. Detection of degree of voluntary-regulatory, cognitive, speech,

motor and emotional disorders were performed also for all patients. The age of the patients ranged from 17 to 55 years and averaged 30.06 ± 7.53 years. The average age of disease onset was 23.71 ± 7.52 years.

Statistical processing of the data was performed using the Statistica for Windows 8.0 software package. Methods of descriptive statistics (determination of numerical characteristics of variables - arithmetic mean (M), mean sampling error (m), determination of the reliability of differences (p), which were tested via the Student-Fisher t-test in representative samples) were used. Correlation between indicators was assessed using Spearman's correlation coefficient (r). The difference in values between comparative indicators was considered significant at $p < 0.05$.

Results.

Results of the study of the subjective perception of quality of life and the effectiveness of their social functioning in general are presented in Table 1. It was indicated that patients with WD assess their overall quality of life quite highly (7.1 ± 2.2). Analysis of the data taking into account the form of the disease showed that patients with the abdominal form of WD are more satisfied with their quality of life than other patients (GG - 8.5 ± 2.1 and TF - 7.0 ± 1.8 ; TRF - 6.8 ± 2.8 ; RAHF - 6.6 ± 1.3 , ECF - 8.0 ± 0.1 , respectively).

Patients with WD rate the nature of interpersonal interaction (8.0 ± 2.0), the level of spiritual fulfilment (7.9 ± 1.9) and socio-emotional support (7.7 ± 2.1) the highest. They are least satisfied with their physical (6.4 ± 2.6) and psychological (6.4 ± 2.5) condition. There are certain features of the perception of QOL in patients with WD depending on the form of the disease. Thus, patients with extrapyramidal-cortical and rigid-arrhythmohyperkinetic forms rate their level of physical well-being lower than other patients (ECF - 4.0 ± 1.4 , RAHF - 4.1 ± 2.1 and TF - 6.7 ± 1.9 ; TRF - 5.8 ± 3.1 ; GG - 9.5 ± 1.7).

In patients with tremor and tremor-rigid forms, no significant differences in QOL assessments were observed. These patients were most satisfied with the level of their spiritual fulfilment (TF - 7.7 ± 1.5 and TRF - 8.3 ± 2.3), interpersonal interaction (TF - 7.8 ± 1.1 and TRF - 7.7 ± 2.8) and socioemotional support (TF - 7.8 ± 1.7 and TRF - 7.7 ± 2.3). These patients were less satisfied with their physical (TF - 6.7 ± 1.9 and TRF - 5.8 ± 3.1) and psychological (TF - 6.6 ± 1.7 and TRF - 5.9 ± 3.2) condition than with other aspects of life. Compared with other patients, patients with the abdominal form of WD are characterized by the highest QOL scores for almost all parameters. They are most satisfied with their level of performance (10.0 ± 0.0), physical (9.5 ± 1.7) and psychological (9.0 ± 1.4) well-being, the nature of interpersonal interaction (10.0 ± 0.0) and personal fulfilment (9.5 ± 0.7). They estimate the level of their spiritual fulfilment (7.0 ± 4.2) and socio-emotional support (7.0 ± 4.2) somewhat lower. Patients with the abdominal form of WD assess their level of physical (GG - 9.5 ± 1.7 and TF - 6.7 ± 1.9 , $p < 0.05$) and psychological (GG - 9.0 ± 1.4 and TF - 6.6 ± 1.7 , $p < 0.05$) well-being, ability to work (GG - 10.0 ± 0.0 and TF - 6.8 ± 2.0 , $p < 0.05$), personal fulfilment (GG - 7.3 ± 1.5 and TF - 7.7 ± 1.1 , $p < 0.05$) and interpersonal interaction (GG - 10.0 ± 0.0 and TF - 7.8 ± 1.1 , $p < 0.05$) significantly higher than patients with the tremor form.

Table 1. Results of the study of subjective perception of quality of life in patients with WD (in points).

Quality of life indicator	Average values of the indicator in groups, points					
	General group (n=33)	Trembling form (n=11)	Trembling-rigid form (n=14)	Rigid-arrhythmic-hyperkinetic form (n=3)	Extrapyramidal-cortical form (n=3)	Abdominal form (n=2)
1	2	3	4	5	6	7
General perception of QOL	7.1±2.2	7.0±1.8	6.8±2.8	6.6±1.3	8.0±0.1*	8.5±2.1
Physical well-being	6.4±2.6	6.7±1.9	5.8±3.1	4.1±2.1	4.0±1.4	9.5±1.7*
Psychological well-being	6.4±2.5	6.6±1.7	5.9±3.2	6.1±1.3	6.0±1.4	9.0±1.4
Self-care and independence in actions	7.3±2.5	7.5±1.7	6.8±3.5	6.3±1.5	8.0±1.4	8.0±2.8*
Working capacity	6.9±2.5	6.8±2.0	6.4±3.2	6.4±2.3	7.0±0.1	10.0±0.0*
Interpersonal interaction	8.0±2.0	7.8±1.1	7.7±2.8	7.6±1.9	8.5±2.1	10.0±0.0*
Socio-emotional support	7.7±2.1	7.8±1.7	7.7±2.3	7.7±2.1	7.5±2.1	7.0±4.2
Civil and service support	7.4±2.3	6.5±2.3	8.1±2.3	8.0±2.4	7.5±0.7	9.0±1.4
Personal realization	7.5±2.2	7.3±1.5	7.5±3.1	6.1±2.3	6.5±0.7	9.5±0.7*
Spiritual realization	7.9±1.9	7.7±1.5	8.3±2.3	7.2±2.3	7.5±0.7	7.0±4.2

Note: * - The reliability of differences $p < 0.05$ according to the Mann-Whitney U criterion.

Table 2. The relationship between QOL parameters and the degree of voluntary-regulatory, cognitive, speech, motor and emotional disorders in patients with GCD.

Parameter of multiple regression model	β -coefficient ¹⁾	p- level of significance	MCC ²⁾	CMD ³⁾
Overall quality of life (const.)				
Depression level	-0.64	0.00	0.65 (p=0.00)	0.41
Physical well-being (const.)				
Depression level	-0.55	0.00	0.85 (p=0.00)	0.72
Degree of motor impairment	-0.30	0.03		
Psychological well-being (const.)				
Anxiety level	-0.48	0.00	0.85 (p=0.00)	0.72
Depression level	-0.55	0.00		
Degree of voluntary regulation disorders	-0.65	0.00		
Self-service and independence in actions (const.)				
Anxiety level	-0.50	0.00	0.90 (p=0.00)	0.81
Rigidity of behavioral reactions	-0.50	0.00		
Degree of motor impairment	-0.47	0.00		
Working capacity (const.)				
Depression level	-0.34	0.04	0.78 (p=0.00)	0.60
Degree of motor impairment	-0.81	0.00		
Interpersonal interaction (const.)				
Depression level	-0.56	0.01	0.56 (p=0.05)	0.31
Socio-emotional support (const.)				
Anxiety level	-0.43	0.00	0.93 (p=0.00)	0.86
Rigidity of behavioral reactions	-0.87	0.00		
Civil and service support (const.)				
Anxiety level	-0.62	0.04	0.63 (p=0.04)	0.40
Personal realization (const.)				
Anxiety level	-0.43	0.00	0.85 (p=0.00)	0.72
Degree of voluntary regulation disorders	-0.60	0.00		
Spiritual realization (const.)				
Rigidity of behavioral reactions	-0.75	0.00	0.63 (p=0.01)	0.39

Notes.

1) – The table presents only significant regression coefficients.

2) – MCC – multiple correlation coefficient for the corresponding regression models.

3) – CMD – coefficient of multiple determination for the corresponding regression models.

It should be noted that patients with WD experience a certain restructuring of the socio-psychological space and the value-semantic sphere of the individual. The narrowing of the usual circle of everyday communication is often combined with an expansion of the spectrum of virtual communications on the Internet, which to a certain extent can contribute to the socio-psychological adaptation of these patients.

The relationship between the patients' QOL assessments and their gender, age, age at disease onset and duration was analyzed. The results of the study showed that the subjective perception of their quality of life in men (7.7 ± 1.1) and women (6.1 ± 3.2) did not differ significantly. No relationship was found between the QOL assessments and the patients' age ($r_{xy} = 0.12$, $p = 0.56$), or their age at disease onset ($r_{xy} = -0.38$, $p = 0.08$). At the same time, the examined patients had a positive correlation between the overall perception of QOL and the duration of the disease ($r_{xy} = 0.45$, $p = 0.04$). These data may indicate that some patients are adapting to their disease, which is accompanied by the development of constructive coping strategies and the formation of an adequate internal picture of the disease. At the same time, most patients show a decrease in criticism of their condition, which increases as the disease progresses and can be expressed in relatively high assessments of the quality of their life. In addition, an increase in quality of life assessments to a certain extent can indicate the effectiveness of the treatment, as a result of which the severity of many symptoms of the disease is reduced to one degree or another and the social functioning of patients improves.

To determine the relationship between the subjective perception of certain parameters of QOL and the degree of voluntary-regulatory, cognitive, speech, motor and emotional disorders in patients with WD, a multiple regression analysis was conducted (Table 2).

The results of the study showed that a decrease in the assessments of the general perception of QOL ($\beta = -0.64$, $p = 0.00$) and the nature of interpersonal interaction ($\beta = -0.56$, $p = 0.01$) in patients with WD is associated with an increase in the level of depression. Satisfaction with their physical well-being in them is associated with the level of depression ($\beta = -0.55$, $p = 0.00$) and movement disorders ($\beta = -0.30$, $p = 0.03$). Subjective perception of psychological well-being in these patients is associated with the degree of anxiety ($\beta = -0.48$, $p = 0.00$), depression ($\beta = 0.55$, $p = 0.00$) and the level of voluntary regulation disorders ($\beta = -0.65$, $p = 0.00$). Satisfaction with the level of self-care and independence in actions is associated with the degree of anxiety ($\beta = -0.50$, $p = 0.00$), rigidity ($\beta = -0.50$, $p = 0.00$) and movement disorders ($\beta = -0.47$, $p = 0.00$). Subjective assessments of patients' ability to work are associated with the level of movement disorders ($\beta = -0.81$, $p = 0.00$) and depression ($\beta = -0.34$, $p = 0.04$). Satisfaction with socio-emotional support is associated with rigidity ($\beta = -0.87$, $p = 0.00$) and anxiety ($\beta = -0.43$, $p = 0.00$). Subjective perception of personal fulfillment in these patients is associated with the degree of voluntary-regulatory disorders ($\beta = -0.61$, $p = 0.00$) and the level of anxiety ($\beta = -0.43$, $p = 0.00$). Patients' satisfaction with their spiritual fulfillment is combined with rigidity of behavioral reactions ($\beta = -0.75$, $p = 0.00$).

Thus, the features of the subjective perception of QOL in patients with WD are primarily associated with the characteristics

of their emotional sphere, namely, with the level of expression of anxiety-depressive symptoms.

Discussion.

The quality of life is a broad-based scale and was originally developed in response to a dearth of culturally relevant measures to assess quality of life [7]. That is a 10-item, self-report measure with items rated on a scale from one to ten (poor to excellent). It takes about 3 min to complete and is used to assess ten dimensions of life satisfaction: physical well-being; psychological/emotional well-being; self-care and independent functioning; occupational functioning; interpersonal functioning; social emotional support; community and services support; personal fulfilment; spiritual fulfilment; and overall quality of life [14]. The ten items were developed based on thematic analysis of identified dimensions in relevant international literature. In the literature it is described as one of the most comprehensive measures in its scope as it includes inquiries that range from physical well-being to spirituality for WD and hepar injury [15,16]. Experience of other pathological condition with involvement of cerebral tissue could be useful also [17-19] with using of non-invasive methods [20,21] that could be implemented in educational, practical and scientific activity [22-25].

Patients with movement disorders, despite a fairly high overall perception of QOL, assess their physical well-being, level of work capacity and self-care worse. In this study, no linear relationships were found between the subjective picture of QOL of patients and the level of their speech disorders. This result may indicate the functioning of the corresponding defense mechanisms and coping strategies of patients, a certain restructuring of their socio-psychological space and the development of such forms of communication that minimize verbal communication (Internet, SMS messages). Also, no direct relationship was determined between subjective assessments of QOL and the degree of cognitive impairment in these patients. It can be assumed that significant relationships between the severity of cognitive impairment and the subjective picture of QOL exist mainly in the presence of gross cognitive deficit in patients, including a decrease in criticism of their condition.

Thus, the obtained data may indicate that it is the specificity of the emotional sphere of patients that is one of the most significant factors involved in the formation of the subjective picture of their quality of life.

The level of subjective perception of QOL in patients with WD is associated with a number of clinical and psychological factors, such as the form of the disease, its duration, and the severity of motor and emotional disorders. Thus, the highest assessments of almost all QOL parameters are noted in patients with the abdominal form of WD. Patients with extrapyramidal-cortical and rigid-arrhythmohyperkinetic forms assess their level of physical and psychological well-being significantly lower than other patients. It was found that overall satisfaction with QOL in patients with WD significantly correlates with the duration of the disease, which may indicate that some patients are adapting to the disease, which is accompanied by the development of constructive coping strategies and the formation of an adequate internal picture of the disease and

improvement mental health finally [26]. At the same time, most patients experience a decrease in criticism of their condition, which increases as the disease progresses and can be expressed in relatively high assessments of the quality of their life.

Conclusion.

The level of subjective perception of quality of life of patients with hepatocerebral dystrophy is associated with a number of clinical and psychological factors, such as the form of the disease, its duration, and the severity of motor and emotional disorders. Thus, the highest assessments of almost all QOL parameters are noted in patients with the abdominal form of WD. Patients with extrapyramidal-cortical and rigid-arrhythmohyperkinetic forms assess their level of physical and psychological well-being significantly lower than other patients.

Funding: This research received no external funding

Conflict of interest statement: The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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