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Accepted Manuscript

Title: Health-related quality of life of teen/adult patients with cystic fibrosis in the Republic of North Macedonia

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Health-related quality of life of teen/adult patients with cystic fibrosis in the Republic of North Macedonia

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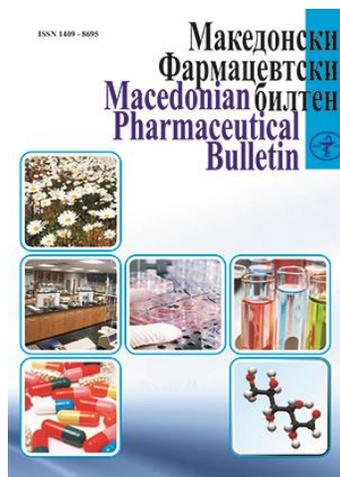*str. Mother Tereza 47, 1000 Skopje, R.N. Macedonia*

Abstract

The present study investigates the Health-related quality of life of teen/adults patients with cystic fibrosis in the Republic of N. Macedonia for the first time. The main objective was to describe the health-related quality of life status of these patients with cystic fibrosis and the second objective was to investigate the connection of the current medical treatment with the analysed parameters.

The survey was conducted on 31 patients by using the Cystic Fibrosis Questionnaire Revised for Health-related quality of live measurement and questions for current medical treatment.

All patients scored their digestive condition with the highest score, while the lower score was reported for social activity. The gender, the nationality and the educational level of the patients with Cystic Fibrosis had no significant impact on their perception of Health-Related Quality of Live.

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The highest score for digestive condition was in positive correlation with the fact that all patients continuously used enzyme therapy. Patients who practiced physical therapy and physical activities reported high scores for physical conditions and body images. This finding point out that patient with Cystic Fibrosis should be encouraged to practice physical activity more often.

The lowest scores for life activities and treatment burden shows that this medical condition has a negative impact on the patients' self-perception and in the execution of their daily activities.

Keywords: cystic fibrosis, Health-Related Quality of Live, digest, daily activities

Introduction

Cystic fibrosis (CF) is genetic disorder, the most common inherited disease in the white population, with autosomal recessive manner of inheritance (Tsui and Buchwald, 1991). The incidence of CF in the white population is about 1:2500 live births (Bobadilla et al., 2002). As a genetic disorder, CF affects mostly the lungs, but it also affects the pancreas, liver, kidneys and intestine too. The disease is characterized by different expression and at different rates of progression in different organs (www.cff.org). The clinical picture is dominated by symptoms of impaired functioning of the respiratory and intestinal tract (Naceva Fushtikj, 2012).

The median survival of CF patients has doubled since 1969 to 1990 from 14 to 28 years and in 2007 it was 37.4 years (Fitz Simmons, 1993; Zemanick et al., 2010). It is estimated that children born in the last decade, using the current standards of treatment in specialized centers for CF, could live for 40-45 years, even without further treatment progression (Elborn, 1993; Lewis et al., 2007). According to available data in 33 EU countries, more than 42000 patients with CF have been registered (www.ecfs.eu). In Republic of North Macedonia a total of 112 patients with CF have been registered at the beginning of 2017 (www.cf.mk).

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The treatment of the Macedonian teen/adults patients with CF is carried out at the Centre for Cystic Fibrosis, which is an integral part of the University Clinic for Children's Diseases - Skopje. This centre is also part of the Register of patients with cystic fibrosis at EU level - the European Cystic Fibrosis Society Patient Registry (ECFSPR). The Centre for Cystic Fibrosis makes efforts related to the implementation of the European consensus on standards of care for patients with CF (Kerem et al., 2005).

Health-related quality of life (HRQoL) is a parameter that is examined in the area of clinical effectiveness. HRQoL can be defined as a patient's general subjective perception of the effect of illness and intervention on physical, psychological and social aspects of daily life (www.ispor.org/workpaper/emea-hrql-guidance.pdf5; www.fda.gov/downloads/drugs/guidances/ucm193282.pdf 2009).

Physical functioning refers to mobility, self-care, usual activities and other functional abilities. Psychological health includes elements like cognitive functioning, emotional distress and anxiety. Finally, social health refers to the quantity and quality of social contacts and interactions (Goodman, 1998). A single domain, e.g. physical functioning, is insufficient to cover HRQoL, even though it is an endpoint relevant to patients (Jackowski and Guyatt, 2003).

The measurement of HRQoL is particularly important in chronic diseases where an assessment of the impact of the intervention should be made in order to improve the functional capabilities of the patients. HRQoL measurements could be conducted using generic HRQoL instruments, disease-specific HRQoL instruments or population-specific HRQoL instruments (www.eunethta.eu).

In Republic of North Macedonia, the HRQoL survey of patients with CF has not been conducted before. The main goal of this survey was to describe the current health status of patients with CF in R. of North Macedonia. The second goal was to investigate the connection of the current medical treatment with the HRQoL parameters. This study covered adult patients with different ethnic backgrounds.

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Material and methods

The participants included in this research were adolescents and adult patients (≥ 14 years of age). The treatment is carried out at the Center for Cystic Fibrosis, University Clinic for Children's Diseases - Skopje.

The survey of the patients was conducted by using the available Macedonian version of Cystic Fibrosis Questionnaire Revised (CFQ-R) as a disease-specific HRQoL instrument. (Henry et al., 2017). Previously, consent to use this questionnaire was provided. Additional questions for current medical treatment/therapy were prepared by the physician (Macedonian national leader for treatment of CF), who is responsible for the medical treatment of these patients.

The questions given in this diseases-specific HRQoL instrument covered three modules: HRQoL, symptoms and overall health perception

The CFQ-R Adults cover eight domains from HRQoL module (Physical Functioning, Vitality, Emotional state, Social limitations, Role Limitations/School Performance, Body Image, Eating Disturbances, Treatment burden), three domains from symptoms module (Respiratory, Digestive and Weight) and one domain from health perception scale (Health Status, present/evolution).

The survey was conducted for a period of nine months. The questionnaires were administrated as a self-administered. CFQ-R questionnaires were distributed during the regular control process of patients and after their fill at home, patients were asked to return the questionnaires at the next control.

In total 31 patients were cover with these CFQ-R Teen/Adults questionnaire. All involved patients were informed about the study objectives and data confidentiality, and were asked to indicate their agreement to participate.

The answers of the questions from HRQoL module were given as a 5 distinct 4-point Likert scales (always/often/sometime/never). The score was ranged between 0 to 100, whereas higher scores representing a better health.

The statistical analysis was performed using SPSS statistical software (available with the questionnaire, SPSS statistical software), Student t-test for independent samples,

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Mann-Whitney nonparametric test, Analysis of Variance-post hoc Tukey test, Kruskal-Wallis ANOVA-post hoc Mann-Whitney test, Pearson test and Spearman test.

Results

Thirty-one patients (adults and adolescents) were involved in HRQoL survey and 12 domains (HRQoL, symptoms and health status) were analyzed. Among them there were 20 men and boys (64.52%) and 11 women and girls (35.48%). The ethnic structure of the patients was dominated by ethnic Macedonians. According to the level of education, the most prevalent were patients with completed secondary school.

The socio-demographic profile (gender, age, nationality and level of education) of all included teen/adults patients with CF patients is presented in Table 1.

Table 1

The descriptive parameters for all 12 domains are presented in Table 2. All patients (adults and adolescents) scored their digestive condition with the highest score (mean 87.81±16.6; med 100; IQR 22.2), while the lower score was reported for social activity (mean 49.64±8.6; med 50; IQR 11.1).

Table 2

The male patients scored their physical condition ($p=0.39$), vitality ($p=0.9$), social activity ($p=0.43$) and role ($p=0.66$) not significantly higher compared to female patients. The female patients scored higher their emotional condition ($p=0.55$), eating ($p=0.77$), treatment burden ($p=0.98$), health perception ($p=0.64$), body image ($p=0.39$), weight ($p=0.84$) and respiratory function ($p=0.41$), but these differences were not significant. The identical scores, from male and female patients, were given for their digestive condition (Table 3).

Table 3

The results showed that there were not statistical significantly difference in HRQoL perception between Albanian and Macedonian patients with CF (Table 4)

Table 4

The education level of the patients with CF has no statistically significant influence on the self-perception of physical condition ($p=0.355$), vitality ($p=0.71$), emotional condition ($p=0.45$), eating ($p=0.79$), treatment burden ($p=0.26$), health perception ($p=0.5$), social activity ($p=0.24$), body image ($p=0.44$), role ($p=0.92$), weight ($p=0.79$) and respiratory function ($p=0.38$). The education level was found significant only for the perception of the digestive condition as a part of HRQoL ($p=0.012$) (Table 5).

Table 5

At the time of the study, all analyzed patients received enzyme therapy and inhaled mucolytic. 29 patients (93.55%) practiced physical therapy, 25 patients (80.64%) received an antibiotic through inhalation, 22 patients (70.97%) took vitamins, while 9 patients (29.03%) received an antibiotic per os.

18 patients (58.06%) received antibiotic treatment per os during the last year and 11 of them (35.48%), due to exacerbation of the disease and worsening of the health condition, were hospitalized. High scores of 87.81, 80.64 and 79.93 were given for digestive condition, weight and eating condition, respectively.

The majority of the patients in this group believe that physical exercises have a positive impact on their health status (Table 6).

Table 6

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Discussion

The gender of the patients with CF at the age of 14 and older had no significant impact on their perception of HRQoL. The review of the literature data showed that Dębska et.al. (2003) also reported that there is no statistically significant difference between the genders. The only statistically significant difference in HRQoL parameters between women and men, was found for the Physical parameter. However, the study conducted by Gee et al. (2003), refers differences in the assessment of HQoL between men and female.

Finding that males have a poorer body image than females is in line with previous research by Abbott et al. (2000). The available literature data show that some findings (Gee et al., 2003) are not in accordance with the study results obtained in this research.

In one study was referred that there is a differences in the clinical manifestation of CF's symptoms between black and white patients (Hamosh et al., 1998). Taking into account the fact that Macedonian and Albanian patients belong to the white race, differences in the manifestation of symptoms of CF are not expected. Reference this, the similar perception in HRQoL parameters (without statistical significance) between patients from these two ethnics was expected. The Macedonian patients reported higher score for emotion, eating, social activity, role and digestive condition, but there was no statistically significant difference. The physical condition, vitality, treatment burden, health perception body image, weight and respiratory function were scored not significantly higher by the patients with Albanian nationality in comparison with the patients from Macedonian nationality.

All seven patients with faculty degree, scored their digestive function with the highest score 100. The lowest score for digestive function was obtained from patients with partially completed secondary education, with mean score 69.44 and mediana 66.67 (*p=0.012*). Contrary to our finding, the education level of Polish and Greek patients had a significant influence for scoring of all HRQoL parameters (Borawska-Kowalczyk, 2015; Stofa et al., 2016). The reason why the education level of the patients with CF in R. of North Macedonia has no statistical impact on the HRQoL scoring could be explained by the

fact that the Macedonian health care givers pay more attention on the patients with lower education.

In terms of frequency of practicing physical therapy, most of the answers were "at least twice" and "at least once a day". Half of the patients (48.39%) answered that they are physically active for more than 30 minutes per day. Walking is the most common type of practiced physical activity by the patients with CF older than 14 years. Almost 71% of the patients practice walking and about 50% of patients used cycling. After a certain physical activity, 16.13% of the patients did not feel any change, 38.71% answered that the physical activity acts favorably on their condition (more easily breathed and less clogged), while 35.48% of the patients answered that they were coughing more secret. Patients who practiced physical therapy and physical activities reported high scores for physical conditions and body images. Previous reports also have indicated that regular physical activity had a positive impact on lung function in patients with CF (Schneiderman et al., 2014; Swisher and Erickson, 2008).

The lowest score for social activity (49.64) and treatment burden was reported within our previous study (Nakov et al., 2020). This fact could be explained by the negative impact of their medical condition in their self-perception and in the execution of their daily social activities.

The findings of the obtained high scores for digestive condition, weight and eating condition, are in a positive correlation with the fact that all patients included in this survey (N=31) took enzyme therapy. Most of surveyed patients (N=22) used vitamin therapy too. All surveyed patients used mucolytic through inhalation and majority of the patients (80.64%) also used antibiotic through inhalation. These results are in a positive correlation with the fact that the respiratory function is scored with relatively high score of 73.65. The benefit of the enzyme, antibiotic and physical therapy on the digestive condition and the respiratory function was also observed for pediatric CF patients in R. North Macedonia (Nakov et al., 2019).

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Conclusion

In conclusion, the HRQoL study of teen/adult patients with CF in the R. of North Macedonia was conducted for the first time.

The findings of our study revealed that the nationality (Macedonian or Albanian) and the gender have no statistically significant impact on the perception for HRQoL.

The highest score for digestive condition in all analyzed subjects was in positive correlation with the fact that all patients continuously used enzyme therapy, thereby indicating the important role of this kind of therapy in patients with CF. This study also revealed that, in contrary to other countries, the level of education in teen/adult patients with CF in R. of North Macedonia has no statistical impact on the HRQoL scoring on the perception of their digestive function. The obtained results could be explained by the fact that the Macedonian health care givers pay more attention on the patients with lower education.

Patients who practiced physical therapy and different physical activities (waking, cycling) reported relatively high scores for physical conditions and body images. This finding underline the importance of the physical activity and the need of encouraging the patients with CF to practice physical activity more often.

Acknowledgments

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Резиме

Испитување на квалитет на живот во однос на здравјето кај адолесценти/возрасни пациенти со цистична фиброза во Република Северна Македонија

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Клучни зборови: цистична фиброза, квалитет на живот во однос на здравјето, дигестивна функција, дневни активности

Во ова студија се презентирани резултати за квалитетот на живот во однос на здравјето кај адолесценти/возрасни пациенти со цистична фиброза во Република Северна Македонија. Ваков тип на истражување кај нас е спроведен за прв пат. Примарна цел на истражувањето беше да се направи процена на квалитетот на живот во однос на здравјето кај оваа група пациенти а секундарна цел беше овие добиени резултати да се поврзат со моментална терапија која ја добиваат пациентите вклучени во ова истражување.

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Во истражувањето беа вклучени вкупно 31 пациент, со користење на ревидираниот прашалник за испитување на квалитет на живот во однос на здравјето за пациенти со цистична фиброза и дополнителни прашања за тековниот медицински третман на оваа група испитаници.

Дигестивната функција беше оценета со највисока оцена од страна на сите пациенти а најниска оцена беше добиена за нивните дневни / социјални активности. Полот на пациентите, нивната национална припадност и степенот на нивното образование немаа статистичко значење во нивната перцепција за квалитетот на живот во однос на здравјето.

Највисоката оцена добиена за дигестивната состојба е во позитивна корелација со податокот дека сите испитаници континуирано користат ензимска терапија. Пациентите кои се на физикална терапија и практикуваат физичка активност, со повисока оцена ја имаат оценето нивната физичка кондиција. Со овој податок се наметнува заклучокот дека оваа група на пациенти треба да бидат поттикнати за редовна физичка активност, на дневна база.

Најниските оцени за нивните дневни активности и ограничувањата на третманот се потврда на фактот дека оваа здравствена состојба има негативно влијаните во личната перцепција на овие пациенти и во спроведувањето на нивните дневни активности.

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Table 1. The socio-demographic profile of these patients

Gender	
Male	20 (64.52 %)
Female	11 (35.48 %)
Age	
38 - 47	2 (6.45 %)
27 - 37	7 (22.58 %)
17 - 26	10 (32.26 %)
14 - 16	12 (38.71 %)
Nationality	
Ethnic Macedonians	23 (74.19 %)
Ethnic Albanians	5 (16.13 %)
Other	1 (3.23 %)
I prefer not to response	2 (6.45 %)
Level of education	
Primary education	9 (29.03 %)
Part of secondary education	4 (12.9 %)
Secondary education	11 (35.48 %)
High education	7 (22.58 &)

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Table 2. Descriptive parameters for analyses of HRQoL domains

Variable	N	Mean ± SD	Median (IQR)	min – max
Physical	31	73.52 ± 18.3	75.0 (33.33)	
Vitality	31	68.01 ± 19.0		33.3 – 100
Emotion	31	74.19 ± 20.9	80.0 (33.33)	
Eating	31	79.93 ± 20.2	88.89 (33.33)	
Treatment Burden	31	59.86 ± 21.6	66.67 (33.33)	
Health Perceptions	31	70.25 ± 21.2		11.11 - 100
Social	31	49.64 ± 8.6	50.0 (11.11)	
Body Image	31	77.42 ± 22.9	77.78 (44.4)	
Role	31	78.76 ± 18.4	83.33 (25)	
Weight	31	80.64 ± 29.5	100.0 (33.33)	
Respiratory	31	73.65 ± 15.3		33.33 – 94.44
Digest	31	87.81 ± 16.6	100.0 (22.22)	

Standard deviation (SD), Interquartile range (IQR), Number of patients (N), no statistical significance (ns)

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Table 3. Descriptive statistics for all analyzed HRQoL domains presented by gender

Descriptive Statistics	Gender		p value
mean ± SD *(min-max) median(IQR)	male N=20	female N=11	
Physical	74.37 ± 20.6 79.16 (29.7)	71.97 ± 14.1 70.83 (29.7)	[b]p=0.39 ns
Vitality	68.33 ± 18.6 66.67 – 83.33	67.42 ± 20.6 66.67 – 75.0	[a]p=0.9 ns
Emotion	72.33 ± 22.2 73.33 (45)	77.58 ± 19.1 86.67 (26.7)	[b]p=0.55 ns
Eating	79.44 ± 19.5 83.33 (41.7)	80.81 ± 22.3 88.89 (33.3)	[b]p=0.77 ns
Treatment Burden	59.44 ± 22.9 61.11 (41.7)	60.61 ± 20.1 66.67 (33.3)	[b]p=0.98 ns
Health Perceptions	68.89 ± 22.7 11.11 – 100	72.73 ± 18.8 33.33 – 100	[a]p=0.64 ns
Social	50.56 ± 8.2 51.0 (5.6)	47.98 ± 9.4 50.0 (11.1)	[b]p=0.43 ns
Body Image	73.89 ± 26.1 77.78 (41.7)	83.84 ± 14.4 88.89 (22.2)	[b]p=0.39 ns
Role	80.0 ± 17.6 83.33 (22.9)	76.51 ± 20.3 75.0 (33.3)	[b]p=0.66 ns
Weight	80.0 ± 29.4 100 (33.3)	81.82 ± 31.1 100 (33.3)	[b]p=0.84 ns
Respiratory	71.94 ± 16.4 33.33 – 94.44	76.77 ± 13.1 55.56 – 94.44	[a]p=0.41 ns
Digest	87.78 ± 17.2 100 (22.2)	87.88 ± 16.1 100 (22.2)	[b]p=0.97 ns

[a]p(Student t-test for independent samples); [b]p(Mann-Whitney test); Standard deviation (SD), Interquartile range (IQR), Number of patients (N), no statistical significance (ns)

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Table 4. Descriptive statistics for all analyzed HRQoL domains presented by ethnicity

Descriptive Statistics mean ± SD *(min-max) median(IQR)	Ethnicity				p value
	ethnic Macedonian N=23	ethnic Albanian N=5	other N=1	prefer not to answer N=2	
Physical	72.83 ± 19.9 75.0 (33.33)	83.33 ± 8.8 83.333 (16.7)	70.83	58.33 ± 5.9 58.333	[b]p=0.26 ns
Vitality	68.84 ± 19.2 33.33 – 100	70.0 ± 19.2 41.67 – 91.67	66.67	54.17 ± 29.5 33.33 – 75.0	[a]p=0.93 ns
Emotion	75.36 ± 22.4 80.0 (40)	68.0 ± 19.7 66.67 (36.7)	86.67	70.0 ± 14.1 70.0	[b]p=0.5 ns
Eating	81.64 ± 19.7 88.89 (33.3)	71.11 ± 25.6 77.78 (50)	66.67	88.89 ± 15.7 88.89	[b]p=0.31 ns
Treatment Burden	58.45 ± 22.1 55.56 (33.3)	68.89 ± 21.4 77.78 (33.3)	55.55	55.56 ± 31.4 55.56	[b]p=0.34 ns
Health Perceptions	70.05 ± 22.8 11.11 – 100	77.78 ± 7.9 66.67 – 88.89	66.67	55.56 ± 31.4 33.33 – 77.78	[a]p=0.43 ns
Social	50.24 ± 8.3 51 (11.1)	48.89 ± 12.7 50 (19.4)	50.0	44.44 ± 0 44.44	[b]p=0.77 ns
Body Image	74.39 ± 24.0 77.78 (33.3)	93.33 ± 9.9 100 (16.7)	55.56	83.33 ± 23.6 83.33	[b]p=0.099ns
Role	79.71 ± 20.7 83.33 33.3()	78.33 ± 9.5 75.0 (16.7)	66.67	75.0 ± 11.8 75.0	[b]p=0.89 ns
Weight	78.26 ± 32.7 100 (33.3)	93.33 ± 14.9 100 (16.7)	66.67	83.33 ± 23.6 83.33	[b]p=0.33 ns
Respiratory	72.71 ± 16.6 33.33 – 94.44	81.11 ± 8.4 72.22 – 88.89	77.78	63.89 ± 11.8 55.56 – 72.22	[a]p=0.34 ns
Digest	87.92 ± 16.7 100 (22.2)	86.67 ± 18.3 88.89 (27.8)	100	83.33 ± 23.6 83.33	[b]p=0.88 ns

[a]p(Student t-test for independent samples); [b]p(Mann-Whitney test); Standard deviation (SD), Interquartile range (IQR), Number of patients (N), no statistical significance (ns)

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Table 5. Descriptive statistics for all analyzed HRQoL domains presented by level of completed education

Descriptive Statistics mean ± SD *(min-max) median(IQR)	Level of completed education				
	Primary school N=9	Part of secondary school N=4	Secondary school N=11	Faculty N=7	**p value**
Physical	75.93 ± 23.1 87.50 (33.3)	78.12 ± 16.8 83.33 (30.2)	76.14 ± 15.5 79.17 (20.8)	63.69 ± 16.8 70.83 (16.7)	[b]p=0.35 ns
Vitality	70.37 ± 21.7 33.33 – 100	72.92 ± 29.93 33.33 – 100	68.94 ± 14.9 41.7 – 91.7	60.71 ± 16.5 33.33 – 75	[a]p=0.71 ns
Emotion	65.92 ± 19.8 73.33 (33.3)	76.67 ± 20.0 73.33 (36.7)	78.18 ± 23.7 86.67 (40)	77.14 ± 19.9 80.0 (40)	[b]p=0.45 ns
Eating	75.31 ± 20.6 66.67 (38.9)	83.33 ± 21.9 88.88 (38.9)	83.84 ± 21.9 88.89 (33.3)	77.78 ± 19.2 77.78 (44.4)	[b]p=0.79 ns
Treatment Burden	70.37 ± 16.7 77.78 (27.8)	55.56 ± 20.3 55.56 (38.9)	59.59 ± 23.9 55.56 (44.4)	49.21 ± 22.1 44.44 (44.4)	[b]p=0.27 ns
Health Perceptions	74.07 ± 16.7 44.44 – 100	66.67 ± 28.7 33.33 – 100	74.75 ± 15.8 44.44 – 100	60.32 ± 29.3 11.11 – 100	[a]p=0.5 ns
Social	48.15 ± 11.1 50.0 (19.4)	44.44 ± 12.0 47.22 (22.2)	53.54 ± 4.5 55.56 (5.6)	48.41 ± 6.9 50.0 (5.6)	[b]p=0.24 ns
Body Image	82.72 ± 17.7 88.89 (33.3)	63.89 ± 18.9 61.11 (36.1)	81.82 ± 15.1 88.89 (11.1)	71.43 ± 37.3 88.89 (44.4)	[b]p=0.44 ns
Role	78.71 ± 21.3 83.33 (37.5)	79.17 ± 14.4 75.0 (25)	79.54 ± 21.8 83.33 (33.3)	77.38 ± 13.4 83.33 (25)	[b]p=0.92 ns
Weight	77.78 ± 33.3 100 (33.3)	75.0 ± 31.9 83.33 (58.3)	87.88 ± 22.5 100 (33.3)	76.19 ± 37.1 100 (33.3)	[b]p=0.79 ns
Respiratory	75.31 ± 15.9 50.0 – 94.44	63.89 ± 13.2 55.56 – 83.33	78.28 ± 12.5 50.0 – 94.44	69.84 ± 18.7 33.33 – 94.4	[a]p=0.38 ns
Digest	83.95 ± 19.3 88.89 (27.8)	69.44 ± 13.9 66.67 (25)	89.89 ± 14.4 100 (22.2)	100 ± 0 100	[b]p=0.012 sig

[a]p (Analysis of Variance; post hoc Tukey test), [b]p(Kruskal-Wallis ANOVA; post hoc Mann-Whitney test), Standard deviation (SD), Interquartile range (IQR), Number of patients (N), no statistical significance (ns)

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Table 6. Impact of physical exercises of patients' health

Do you think that physical exercises and sports are important for your health?	
Yes, I feel better	22 (70.97%)
Rarely exercise	8 (25.81%)
I do not have a time to practice	0
I feel tired	1 (3.23%)

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