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Quality of Life and the Degree of Disease Acceptance In Patients with Spinocerebellar Ataxia

Jakość życia i stopień akceptacji choroby pacjentów chorych na ataksję rdzeniowo-móżdżkową

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Abstract

Introduction. Spinocerebellar ataxias (SCA) belong to a group of rare genetic diseases. They are characterized by the multiplicity of symptoms, of which the most characteristic is limb ataxia and dysarthria. The disease progresses over time, leading to severe disability. So far an effective method of treating SCA has not been invented.
Aim. The main purpose was to analyze the quality of life and the level of disease acceptance in patients with SCA.
Material and Methods. The study included 70 patients with the average duration of SCA of approximately 13 years. The age of patients ranged from 26 to 81 years (mean 50±10 years). All participants were examined using the Acceptance Illness Scale (AIS) and the Health Survey (SF-36). Statistically significant differences in the degree of disease acceptance were observed in patients who differed in terms of education level and gender.
Results. In the physical sphere, women assessed their life quality more favorably than men (p=0.040), however, men declared better scores in bodily pain (p=0.017). It was observed that, in the sphere of mental health, men were more peaceful, and women were believed in deterioration of their health condition (p=0.039). Patients with SCA exhibit a very low level of both, disease acceptance and the quality of life. Women worse than men accept the disease, and they feel useless more often. The higher level of disease acceptance is shown by patients with higher education than those with vocational education. An interpersonal relationships get worse, especially among men, however, women often feel useless. Patients with SCA rate their life quality very low.
Conclusions. It is necessary to educate patients, their caregivers, medical staff in order to develop desirable health behaviors. Understanding this rare disease will help improve the quality of life and therefore will improve the level of disease acceptance. **(JNNN 2018;7(1):12–21)**
Key Words: spinocerebellar ataxia, quality of life, acceptance of disease, patient's self–assessment

Streszczenie

Wstęp. Ataksje rdzeniowo-móżdżkowe (SCA) należą do grupy rzadkich chorób genetycznych. Cechuje je wielość objawów, z których najbardziej charakterystycznym jest ataksja kończyn oraz dysartria. Choroba postępuje w czasie prowadząc do ciężkiej niepełnosprawności. Jak na razie nie wynaleziono skutecznej metody leczenia SCA.
Cel. Celem było zbadanie jakości życia i stopnia akceptacji choroby pacjentów z SCA.
Materiał i metody. W badaniu wzięło udział 70 pacjentów ze średnim czasem trwania SCA rzędu 13 lat. Wiek chorych wahał się w przedziale od 26 lat do 81 (średnio 50±10 lat). Wszyscy uczestnicy zostali przebadaniu kwestionariuszem akceptacji choroby (AIS) oraz jakości życia (SF-36). Istotne statystycznie różnice w poziomie akceptacji choroby zaobserwowano przy podziale chorych ze względu na wykształcenie i płeć.
Wyniki. Kobiety lepiej niż mężczyźni oceniają jakość swojego życia w domenie sprawności fizycznej (p=0,040), jakkolwiek mężczyźni uzyskiwali lepsze wyniki w domenie dolegliwości bólowych (p=0,017). Zaobserwowano, iż w domenie zdrowia psychicznego, mężczyźni w ciągu ostatnich 4 tygodni byli bardziej wyciszeni, a kobiety częściej uważały, iż stan ich zdrowia ulegnie pogorszeniu (p=0,039). Chorzy z SCA wykazują bardzo niski stopień, zarówno akceptacji choroby, jak i jakości życia. Kobiety gorzej niż mężczyźni akceptują chorobę, częściej czują się niepotrzebne. Wyższy stopień akceptacji choroby wykazują pacjenci z wykształceniem wyższym, niż pacjenci z wykształceniem zawodowym. Relacje

interpersonalne chorych ulegają zaburzeniu, szczególnie wyraźnie zjawisko to dotyczy mężczyzn. Pacjenci z SCA oceniają bardzo niską jakość swojego życia.
Wnioski. Konieczna jest edukacja pacjenta, jego opiekunów i personelu medycznego w celu wyrobienia pożądanych zachowań zdrowotnych. Zrozumienie istoty tej rzadkiej choroby, przyczyni się do poprawy jakości życia, a co za tym idzie poprawy stopnia akceptacji choroby. **(PNN 2018;7(1):12–21)**
Słowa kluczowe: ataksja rdzeniowo-móżdżkowa, jakość życia, akceptacja choroby, samoocena chorych

Introduction

Spinocerebellar ataxia (SCA) is a rare, chronic, neurodegenerative genetic disorder that is subject to autosomal dominant inheritance. SCAs fall under various categories. SCA1, SCA2 and SCA8 are autosomal dominant ataxias type 1 (ADCA1). Genes located on various chromosomes have an unstable number of microsatellite repetitions, both in the coding and non-coding parts of genes [1–3].

The disease, causes damage to the cerebellum and its connections [4]. The spinocerebellar ataxia is inherited in an autosomal dominant manner. The risk of transmission of the disease from parents to their children is 50%; the transmission is not gender — linked.

Incidence of SCA in Europe is estimated at between 0.9 and 3.0 per 100000 cases depending on region. Globally, the highest number of people with SCA live in Cuba (6.57 cases/10(5) inhabitants) and in the Azores Island [5,6]. Globally, SCA1, SCA2, SCA3, SCA6 and SCA7 represent the highest numbers of cases [1]. However, the most frequent are SCA3 [4].

So far, in Poland, the occurrence of SCA1, SCA2, SCA8 and SCA17 were reported. The most common cases were SCA1 and SCA2, with no SCA3 — the most common type of SCA in Western Europe. Axial signs in SCA1 and SCA2 include truncal and limb ataxia, dysarthria, choking, gaze palsy and motor-sensory axonal neuropathy [7].

The clinical characteristics of the disease, suggesting a specific SCA genotype, include in the case of SCA1 — pyramidal symptoms, hypermetric saccades; SCA2 — diminished velocity saccades, hyporeflexia, tremor, dementia (rare); and SCA8 — posterior column paresthesia, pyramidal symptoms [8].

Polish comparative studies of SCA1 and SCA2 have shown that SCA1 patients suffer from dysphagia, depression and pyramidal symptoms more frequently than SCA2 patients. However, the latter more often have clinical symptoms of neuropathy as well as extrapyramidal symptoms [9].

Generally ataxia is characterized by inability to coordinate balance, ataxia of upper and lower limbs, eye movement abnormalities, dysphagia or dysarthria. In spinocerebellar ataxia, dysarthria is progressive. The ability and opportunity to communicate with the world, especially with loved ones, is extremely important in human life. To articulate words properly and speak clearly, the cooperation between articulation organs, respiratory and phonatory systems is necessary. Balanced participation of different muscles is also essential. Therefore, the proper articulation of speech sounds depends on the pyramidal and extra pyramidal tracts as well as on the cerebellum (movement coordination). Symptoms of the disease usually appear in the third and fourth decade of life. However, it is hard to diagnose the onset of the disease because of its insidious and slowly progressive nature [10]. The decreased coordination and articulation impairment cause that people with ataxia move and speak like intoxicated persons. Because of this, they are stigmatized, which leads to their depression and psychosocial disorders.

So far, no effective remedy that could improve and prolong life quality has been found, therefore, patients should remain under care of interdisciplinary teams of specialists, including neurologopedists and physiotherapists.

The general objective of the study was to investigate the quality of life and the acceptance level across patients suffering from SCA in Poland. The specific objective in the social sphere was to make patients gain greater self-awareness measured by answers to questions such as: "Who am I? How does it feel?, Do I accept my disease?" as well as to analyze if patients' relatives and friends understand their emotional situation. The specific objective in the practical sphere was to show patients, caregivers and medical staff how best to improve the quality of life in patients with spinocerebellar ataxia.

Material and Methods

Participants' Characteristics

The study group included 70 patients from Poland with confirmed SCA (35 women and 35 men). Patients with confirmed SCA1 outnumbered others — 52 persons (74.3% of the respondents), then patients with SCA2 — 13 persons (18.6%) and with SCA8 (1 person). SCA type was not confirmed in four of the respondents although inheritance and symptoms clearly suggested spinocerebellar ataxia.

Patients aged 26 to 81 with persons 50 years of age (±10) outnumbering others. Persons with secondary education were the largest group (26 persons), next were those with higher education (24 persons) and last with vocational education (20 persons). The disease duration ranged from 2 years (the respondent was 29 years old) to 45 years (the respondent was 81 years old during the study). The average duration of the disease was estimated to be 13 years (with the range oscillating between 2 and 45 years).

The duration of the disease is 10–30 years. Such a big difference between the literature data and the interview data may result from the fact that it is difficult to determine the onset of the disease [11]. The symptoms appear gradually, slowly and are very diverse. Thus, the sick person, after being diagnosed, may attribute to ataxia various symptoms that occurred many years before the actual manifestation of the disease.

Although symptoms that are more characteristic of individual SCA subtypes appear, in the study group no significant differences in the clinical picture have been observed.

Ethical Considerations

The research was approved by the Bioethics Committee of Wrocław Medical University (no. KB-534/2013). The investigation conforms with the principles outlined in the Declaration of Helsinki. The study was conducted in accordance with the principles of good clinical practice. The dignity and rights of participants were respected at all times.

Measurement Tools

The surveys made use of the SF-36 questionnaire to measure the quality of life and the Acceptance Illness Scale (AIS). The scale is designed to measure the level of acceptance of a disease in adults. It also evaluates the respondents' adaptation to a disease and can be applied for any disease entity. An interview questionnaire prepared specifically for the study was also used.

Statistical Analysis

The outcomes of metric characteristics (quantitative) are presented in tables as the arithmetic mean and standard deviation (M±SD). The hypotheses on normal distribution were verified using Shapiro–Wilk and David-Hellwig tests with the significance level $p<0.05$.The significance of differences in mean values in two groups were verified using Student's t-test for the unpaired variables. The significance of differences in mean values in more than two groups were verified using analysis of variance. In the case of statistically significant results ANOVA ($p<0.05$), post-hoc tests were carried out (multiple comparisons by least significant difference test-LSD). For calculations and graphics STATISTICA 10.0 software were used (StatSoft, Dell Inc., USA).

Results

Fifty-one point four percent of the respondents (36 persons) reported inability to coordinate balance as the first symptom of the disease. Thirty-three point four percent of respondents (24 persons) named unsteady walk as the first symptom. Two people reported tendency to stumble while walking, another two people tendency to fall as the first symptom.

Speech impairment was reported by 92.9% of patients. Five of the respondents claimed no change in speech. Eighty-eight percent of the respondents (80% of women and 97.1% of men) defined difficulty with speech as problems with articulation. The problem with clear words articulation was more often reported by men than women. The observed variation in response to this question between men and women was statistically significant ($p=0.027$).

Eating disorder was reported by 62.9% of patients (54.3% of women and 71.4% of men). Thirty percent of the respondents defined those symptoms as difficulties with chewing. Forty-one persons (58.6%) noticed that difficulties with eating intensified with time. Sixty two point nine percent of the respondents complained about difficulties with swallowing liquid — men (80%) outnumbered women (45.7%). This difference was statistically significant ($p=0.003$). Problems with swallowing liquid were defined by 44.3% of patients as choking on liquids, and by 15.7% as choking simultaneously with mouth leak. Forty-seven percent of the respondents noticed that those difficulties intensified with time. The progressive difficulty with swallowing liquids was more often reported by men (74.3%) than women (40.0%). The observed differences between women and men were statistically significant ($p=0.004$). All respondents had difficulties with swallowing saliva when resting. Difficulties with swallowing saliva while speaking were noticed by 37.1% of the respondents. Thirty-four point three percent of the respondents complained about difficulties with unrestricted breathing (Table 1).

When asked about the physical rehabilitation, 64.3% of the respondents confirmed participation in such rehabilitation activities, 42.9% of whom believed that there were too few rehabilitation activities offered. Thirty-one point four percent of patients did not

Table 1. Characteristics of speech and swallowing disorders in the group of women and men and the results of the comparisons

Questions of the survey	Overall N=70	Woman N=35	Man N=35	p
1. Have you encountered any problems with eating?				0.138[a]
No	26 (37.1%)	16 (45.7%)	10 (28.6%)	
Yes	44 (62.9%)	19 (54.3%)	25 (71.4%)	
2. Have you encountered any problems with chewing?				
No	49 (70.0%)	26 (74.3%)	23 (65.7%)	0.434[a]
Yes	21 (30.0%)	9 (25.7%)	12 (34.3%)	
3. Do difficulties with eating intensify?				0.467[a]
No	29 (41.4%)	16 (45.7%)	13 (37.1%)	
Yes	41 (58.6%)	19 (54.3%)	22 (62.9%)	
4. Have you encountered any difficulties when drinking?				0.003[a]
No	26 (37.1%)	19 (54.3%)	7 (20.0%)	
Yes	44 (62.9%)	16 (45.7%)	28 (80.0%)	
5. Do difficulties with drinking intensify?				0.004[a]
No	30 (42.9%)	21 (60.0%)	9 (25.7%)	
Yes	40 (47.1%)	14 (40.0%)	26 (74.3%)	
6. Have you encountered any problems with swallowing saliva when resting?				1.000
No	0 (0%)	0 (0%)	0 (0%)	
Yes	70 (100%)	35 (100%)	35 (100%)	
7. Have you encountered any problems with swallowing saliva when speaking?				0.621[a]
No	44 (62.9%)	21 (60.0%)	23 (65.7%)	
Yes	26 (37.1%)	14 (40.0%)	12 (34.3%)	
8. Have you encountered any tongue movement problems?				0.632[a]
No	34 (48.6%)	16 (45.7%)	18 (51.4%)	
Yes	36 (51.4%)	19 (54.3%)	17 (48.6%)	
9. Have you encountered any lips movement problems?				0.584[a]
No	52 (74.3%)	27 (77.1%)	25 (71.4%)	
Yes	18 (25.7%)	8 (22.9%)	10 (28.6%)	
10. Do you have any difficulties with unrestricted breathing?				1.000[a]
No	46 (65.7%)	23 (65.7%)	23 (65.7%)	
Yes	24 (34.3%)	12 (34.3%)	12 (34.3%)	
11. Are your speech difficulties connected with clear word articulation problems?				0.027[b]
No	8 (11.4%)	7 (20.0%)	1 (2.9%)	
Yes	61 (88.6%)	28 (80.0%)	34 (97.1%)	

[a] — Pearson's chi-square test; [b] — Fisher's exact test

participate in any rehabilitation activities (3 women stated that they did not need such activities). More men (71.4%) than women (57.2%) attended rehabilitation, men also believed those activities should be more extensive. The difference in attending rehabilitation between men and women was statistically significant and was p=0.042.

When filling in the Acceptance Illness Scale questionnaire (AIS), respondents were to answer eight following statements: I have troubles with adjusting to limitations imposed by the disease; my health condition does not allow me to do what I like the most; the disease makes me feel useless; my health condition makes me more dependent on other people than I would want like

to; because of my disease I am a burden to my family and friends; my health condition does not make me feel a valued person; I will never be as self-sufficient as I would like to be; I think that people surrounding me often feel uncomfortable because of my disorder. In the Polish adaptation of AIS, unlike in the US language version, all statements were formulated in an unambiguous way, describing only difficulties connected with the disorder. This is why the lowest scorings referred to lowest degrees of disease acceptance.

An average value of disease acceptance in the responding group was 20.15±7.3. Except for the statement "This disease makes me feel useless sometimes", the results observed in the degree of disease acceptance by women and men were not statistically significant (p>0.05). Women more often than men felt useless (p=0.012) (Figure 1).

Figure 1. Comparing average number of points in women's and men's statement: "The disease makes me feel useless sometimes" and results of Mann–Whitney U test

In the degree of disease acceptance, no statistical significance was observed (p>0.05) in groups of patients of different age, in groups of patients whose first symptoms occurred at different times, or in groups of patients who experienced difficulties with speech at different times.

Statistically significant differences were observed in patients with different education. Patients with higher education showed higher degree of disease acceptance than those with vocational education (p=0.031) (Table 2 and Figure 2). The analysis of the data from SF-36 questionnaire showed that the average life value was pointed 102 per maximum 171.

Table 2. Basic points statistics of disease acceptance in groups of patients with different education

Acceptance Illness Scale AIS From 1 — definitely agree To 5 — do not agree	Education			P
	Vocational N=20	Secondary N=26	Higher N=24	
SUM				
Mean±SD	17.5±6.7	19.7±7.0	22.8±7.5	
Median (Q_1±Q_3)	19 (10÷22)	20 (13÷22)	23.5 (16÷27)	0.031

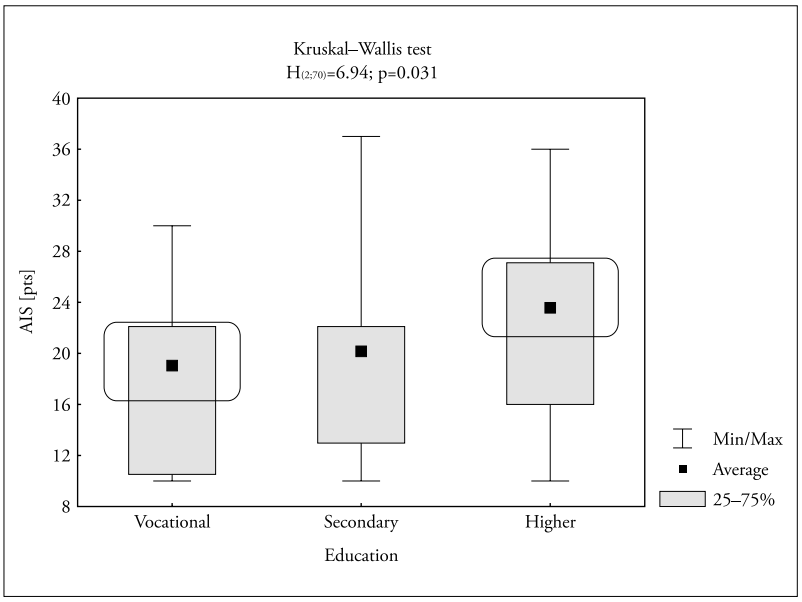

Figure 2. Comparing average number of points in groups of patients with different education (AIS questionnaire) and Kruskal–Wallis test results

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Statistically significant differences between women and men were observed in the sphere of physical functioning. Women could better handle activities of average difficulty, such as: displacing table, hovering, bowling, and golfing (p=0.029), including taking one (p=0.028) or several flights of stairs (p=0.020), walking approx. 100 m (p=0.042), bathing or getting dressed (p=0.035).

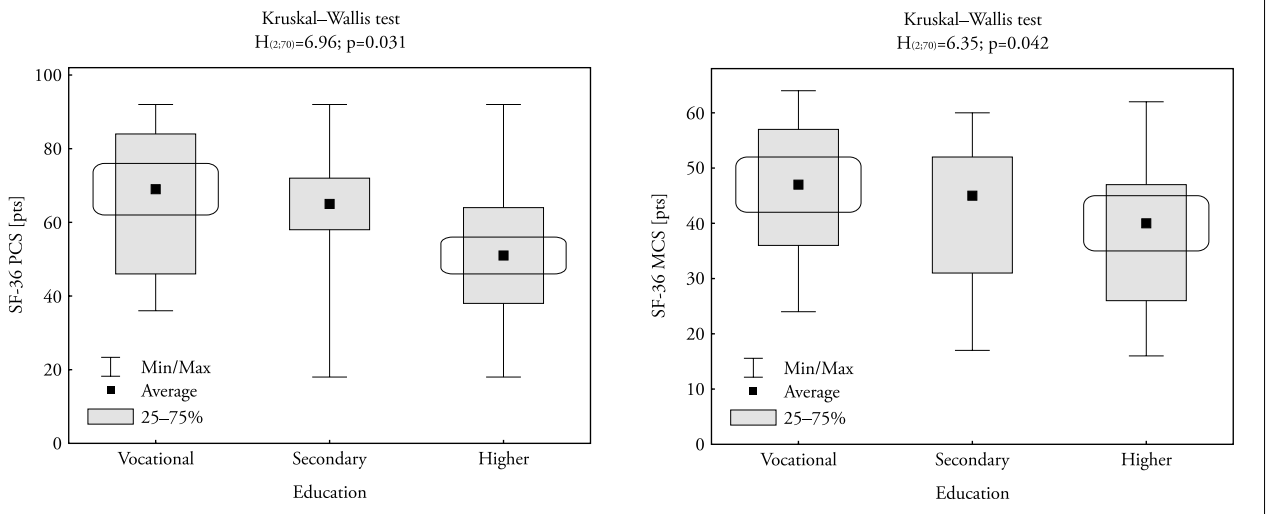
In the sphere of bodily pain, statistically significant differences between women and men were related to the frequency of the intensifying pain. Women more often than men felt pain during the last month (p=0.044); patients complained about limitations in their daily work and home lives (p=0.021).

In the sphere of mental health, it was observed that men were more peaceful and calm than women during the last four weeks (p=0.048). Women more often than men believed their health condition would get worse (p=0.039).

No statistically significant difference was observed in assessment of the quality of life in groups of patients of different age (p>0.05). Statistically significant difference, however, was observed in groups of patients with different education. Patients with higher education reported higher life quality in physical and mental spheres than those with vocational education (p<0.05) (Table 3 and Figure 3).

Table 3. Basic points statistics based on SF-36 questionnaire in groups of patients with different education

SF-36 quality of life Questionnaire results	Education			p
	Vocational N=20	Secondary N=26	Higher N=24	
PCS — physical component summary				
Mean±SD	66.6±19.5	63.4±18.7	52.6±19.8	
Median (Q_1÷Q_3)	70 (46÷84)	65 (58÷72)	52 (38÷64)	0.031
MCS — mental component summary				
Mean±SD	46.8±11.8	42.5±12.3	36.8±13.4	
Median (Q_1÷Q_3)	47 (37÷57)	46 (31÷52)	41 (27÷47)	0.042
QoL — Quality of Life				
Mean±SD	113.3±29.3	105.8±29.4	89.3±30.0	
Median (Q_1÷Q_3)	114 (88÷139)	112 (90÷126)	87 (69÷113)	0.024



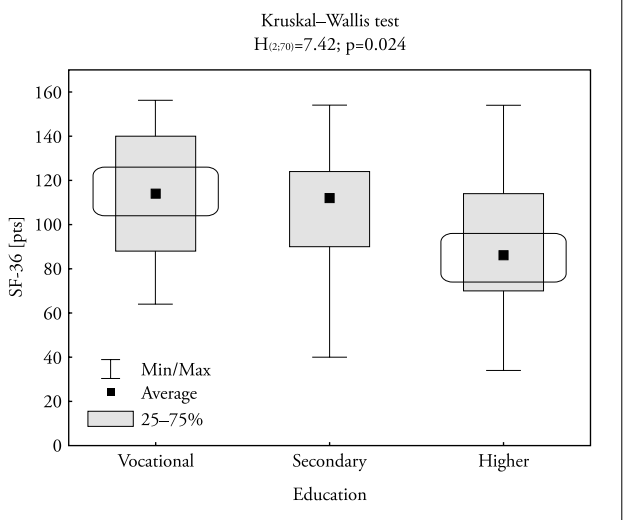


Figure 3. Comparing the average number of points in the assessment of life quality (questionnaire SF-36) in groups with different education and Kruskal–Wallis test results

In groups of patients whose first disease symptoms occurred at different times, level of physical activity (p=0.009) and general life quality level (p=0.021) is worse in patients who observed first symptoms more than 15 years ago (Table 4 and Figure 4).

Results of assessment of life quality in groups of patients whose first speech difficulties occurred at different times showed that patients who observed their speech difficulties 6 or more years before rated their physical condition, mental condition and general quality of their lives low (p<0.05) (Table 5 and Figure 5).

Table 4. Basic points statistics of SF-36 questionnaire in groups of patients who had first disease symptoms at different times

SF-36 quality of life Questionnaire results	Time when first symptoms occurred [years]			p
	<10 N=29	11–15 N=23	15< N=18	
PCS — physical component summary				
Mean±SD	58.0±18.5	54.0±17.0	73.2±20.9	
Median (Q_1÷Q_3)	62 (49÷69)	56 (41÷65)	84 (51÷92)	0.009
MCS — mental component summary				
Mean±SD	41.2±12.2	38.4±12.9	46.7±13.6	
Median (Q_1÷Q_3)	43 (32÷49)	40 (27÷50)	49 (32÷60)	0.126
QoL — Quality of Life				
Mean±SD	99.3±28.1	92.4±28.3	119.9±32.2	
Median (Q_1÷Q_3)	105 (83÷118)	99 (64÷114)	122 (90÷150)	0.021

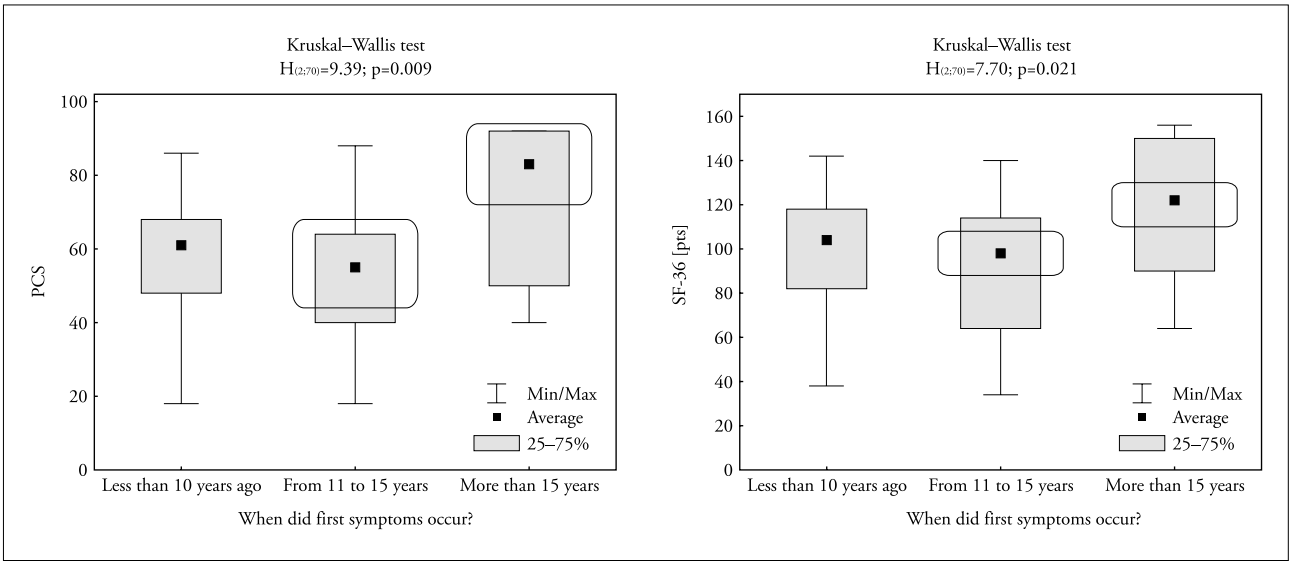


Figure 4. Comparing the average number of points in the assessment of life quality (questionnaire SF-36) in groups of patients whose first disease symptoms occurred at different times and Kruskal–Wallis test results

Table 5. Basic points statistics of the assessment of the quality of life in groups of patients who had first speech difficulties at different times

SF-36 quality of life Questionnaire results	When did the first difficulties with speech occur?		p
	<5 years ago N=54	6 years < N=16	
PCS — physical component summary			
Mean±SD	55.5±18.0	77.9±16.6	
Median (Q_1÷Q_3)	59 (41÷68)	87 (66÷92)	<0.001
MCS — mental component summary			
Mean±SD	39.9±13.1	47.8±11.2	
Median (Q_1÷Q_3)	42 (28÷50)	49 (38÷56)	0.036
QoL — Quality of Life			
Mean±SD	95.4±29.0	125.6±25.3	
Median (Q_1÷Q_3)	99 (71÷115)	130 (111÷147)	0.001

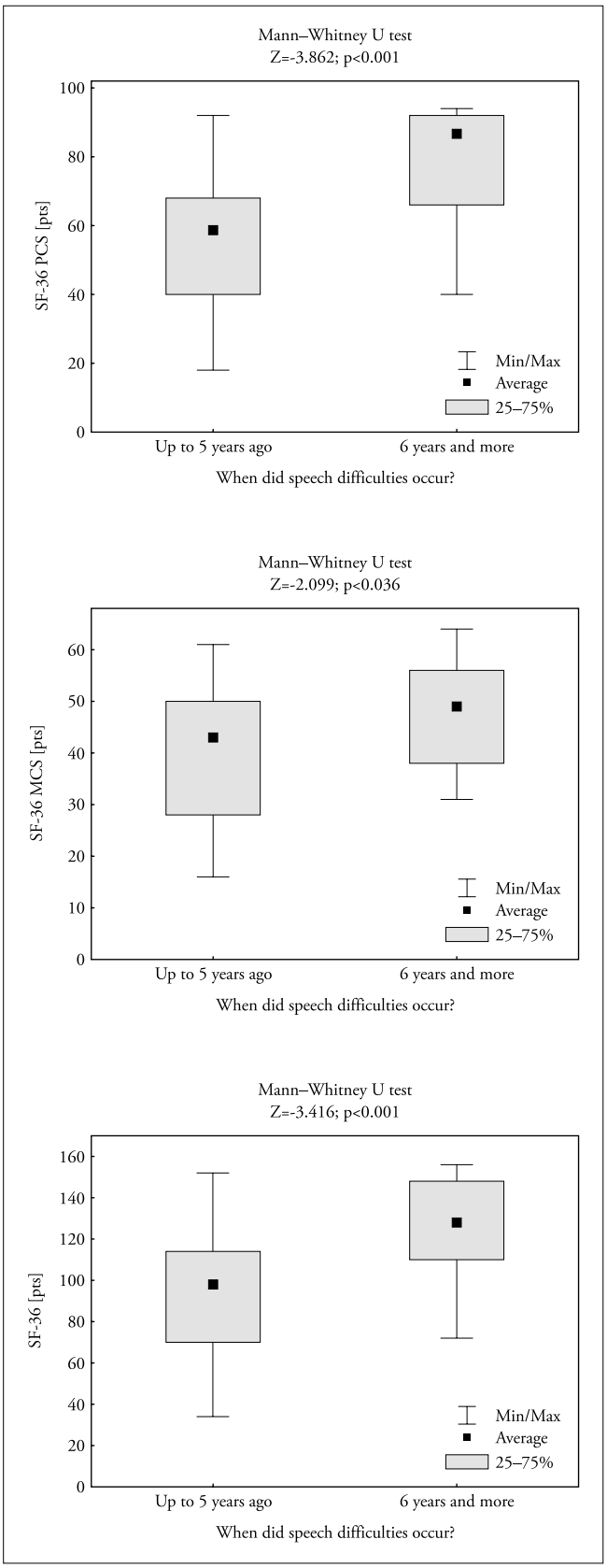


Figure 5. Comparison of the average number of points for the quality of life assessment (SF-36 questionnaire) in groups of patients who had first speech difficulties at different times and Mann–Whitney comparison test results

Discussion

In the study group, the majority of patients had confirmed SCA1 (52 persons), with SCA2 — 13 persons and with SCA8 — 1 person. Although SCA3 predominates in western Europe and in the world, SCA1 and SCA2 as the most frequently occurring types in Poland. The lack of diagnosis of 4 persons was the consequence of the fact that in the Institute of Psychiatry and Neurology in Warsaw the examination was carried out only on 10 types of SCA, as noted by Sułek-Piątkowska [7] as a result, 25% of patients had negative results in molecular tests. Despite benefits which physical rehabilitation may offer, mentioned among others by Martins et al. [12], 31.4% of the respondents did not attend any such activities. Patients who did not attend physical rehabilitation complained about the lack of access to such rehabilitation. Access to specialists is certainly limited for those who come from towns or villages. People with disorders often live alone and their health condition does not allow them to attend rehabilitation in health centers. The fact that 3 women considered such rehabilitation as not needed arises, most probably, out of their insufficient knowledge on the disease or the fact that symptoms were not severe yet. Despite their well-functioning, these women cannot forget that the disease is progressive.

In the context of Acceptance Illness Scale, it was noticed that women accepted their illness worse than men, and more often felt useless (p=0.312), which may be the result of biologically determined differences in the psyche of the two genders. Although psychologists do not consent to gender determination, in Polish society there is an adopted female pattern which assumes females are mothers, caregivers, family life organizers [13]. Following this assumption, disability to take care of family may make women, in their subjective opinion, less valued or even useless, because they cannot meet social expectations imposed on them. Self-esteem is also an important factor in the quality of life.

Because no statistically significant difference was observed in the degree of disease acceptance in groups of patients of different age, and what is interesting, in groups of patients whose first symptoms of the disease occurred at different times, or groups of patients whose difficulties with speech occurred at different times, it can be concluded that patients who have been suffering from the disease for not a long period, and those who have been suffering for years exhibit similar degree of disease acceptance or the lack of it. The causes of such results may arise from a situation when patients with less visible symptoms look at their relatives whose symptoms are more advanced: they may automatically project their own functioning among family or friends in the next years, and so they start to adopt negative

attitudes towards their health condition and therefore do not accept their disease.

Patients with higher education exhibited higher degree of disease acceptance than those with vocational education (p=0.031), which was the outcome of their greater knowledge on how to deal with the condition with opportunities offered by modern science. Those patients were more wealthy, which allowed them to attend private logopedic therapies or physical rehabilitation.

The respondents were afraid not only of the progressive process of the disease and its influence on physical functioning, but also of the reaction of the society; they could feel how other people felt in their company. This was connected with gait abnormality and speech impairment typical for ataxia. Patients were often perceived as intoxicated persons. Such stereotypes make patients withdraw and isolate from social life, which leads to discrimination and therefore plays an important role in social stigmatizing.

Some of the respondents had much higher degree of disease acceptance than others despite long duration of disease or progressive symptoms. In order to explain this phenomenon, the following point needs attention: the degree of disease acceptance may depend on psychological benefits which this disease may bring. This disease may be used as a very powerful excuse to all misfortunes or passive attitudes. Patients may also use it for their own benefits (through evoking sympathy).

Another aspect, though sometimes omitted, is that the disease and its symptoms may be perceived as blessing (for instance when it makes patients sensitive, or helps them learn new things, changes the way of their perception and attitudes towards life). Ailments help such patients understand and give meaning to their lives. The way such stigma-disease is perceived is to a large measure conditional on the system of values and worldview of the stigmatized person. There were some cases of patients who assessed their lives much better than those who did not suffer from the disease. It can be concluded that patients, due to their psychological defense mechanisms, were able to adjust to a new situation, and therefore, exhibit higher level of life satisfaction than healthy persons.

Comparing the results of different groups examinations, it was noticeable that the resulting normal value was 20.15±7.3 in AIS scale for a group of patients suffering from SCA. This result was lower than the majority of results for patients, who suffered from chronic somatic illnesses [14]. Patients with respiratory diseases had normal value at 29.09±8.46, those with urinary diseases at 28.62±10.34, patients with circulatory system diseases at 27.78±9.86, those with locomotor diseases at 26.63±8.55, with nervous system diseases at 27.02±8.92 and diabetics at 25.76±10.34. Respondents with SCA, therefore, showed lower degree of disease acceptance than those aforementioned. Normal value of AIS scale was also lower (23.5±6.7) in groups of women treated for lesions of the cervix and it was 28.76 [15].

The studies in the context of SF-36 questionnaire showed that women better than men rated their life quality in the sphere of physical functioning (p=0.040) and worse than men in the sphere of bodily pain (p=0.017). It was observed that men, in the sphere of mental health, were more peaceful and calm than women in the last 4 weeks. Women more often than men claimed that their health condition would get worse (p=0.039). Those results were compatible with results of AIS. In the remaining scales, differences were not statistically significant (p>0.05). Patients whose speech difficulties occurred 6 or more years before, assessed their life quality much worse (p<0.05). Such results are understandable because all symptoms progress with time and become more severe to patients of this group than to patients who had first speech difficulties less later than 6 years before.

Both examinations, the one in the context AIS questionnaire, and in the context of SF-36 questionnaire, showed statistically significant difference in the assessment of life quality in groups of patients with different education. Patients with higher education showed higher quality of life in physical and mental spheres than those with vocational education (p<0.05).

In groups of patients who differed in terms of time since the first symptoms occurred, lower level of physical functioning (p=0.009) and worse general level of life quality (p=0.021) was observed in patients who had first symptoms more than 15 years before. Such results are related to progressive nature of the disease, progressive age of the respondents and all natural limitations it imposes. Similar situation was with patients who had first speech difficulties 6 or more years before. They considered their physical, mental and general activity levels and life quality as worse (p<0.05). Speech difficulties also progress with time. Patients, apart from physical limitations, encountered limitations in social life, even in relations with those people who were the closest [16]. Unfortunately, there is a lack of good quality studies that have evaluated speech and language therapy [17].

So far, no other examination in the context of AIS and SF-36 questionnaires has been carried out on patients with SCA, therefore there is no possibility to compare patients from different countries. It is therefore necessary to continue examinations.

Conclusions

Patients with SCA rate their life quality very low. Such life assessment depends on gender, time since speech difficulties occurred, education, and time since

first symptoms occurred. This disease has a negative influence on many spheres of functioning in patient's life. Patients exhibit very low degree of disease acceptance. This dependents on gender and education. Women worse than men accept their disorder, more often feel useless. Patients with higher education show higher level of disease acceptance than those with vocational education. Patients feel stigmatized, they show reveal psychosocial difficulties, and their personal relationships get worse. This phenomenon particularly affects men. It is necessary to educate patients, their caregivers as well as medical staff in order to develop desirable health behaviors. Understanding this disease will help improve the quality of life and therefore will improve level of disease acceptance.

Implications for Nursing Practice

1. SCA3 predominates in western Europe and in the world, however, SCA1 and SCA2 are the types most frequently occurring in Poland.
2. Interpersonal relationships get seriously worse in SCA, especially among men, however, women often feel useless.
3. It is necessary to educate patients, their caregivers as well as medical staff in order to develop desirable health behaviors.

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