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Functioning of Patients with Myasthenia Gravis after Plasmapheresis

Funkcjonowanie pacjentów z miastenią po zabiegu plazmaferezy

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Abstract

Introduction. Myasthenia gravis is an autoimmune disease of the postsynaptic part of the neuromuscular junction. A characteristic symptom of myasthenia gravis is the so-called fatigability or muscle fatigue, which also affects the respiratory muscles. Plasmapheresis is one of the treatments that improve breathing and reduce the symptoms that cause muscle weakness.

Aim. The aim of the study was to assess the functioning of patients with myasthenia gravis before and after plasmapheresis.

Material and Methods. The research was conducted in 2019 in the Anaesthesiology and Intensive Care department on a group of 30 patients with clinically confirmed myasthenia gravis. The author's questionnaire was used to assess the functioning of patients. The research was approved by the Bioethics Committee.

Results. The patient's condition improved after the procedure in each of the analysed areas of functioning (physical activity, movement, breathing, swallowing, mood, social functioning). The plasmapheresis most preferably affected the area of physical activity of the subjects. The conducted statistical analysis proved a significant difference ($p=0.000$) for all results, in every aspect (before and after the procedure). At the same time, the highest average increase was observed in relation to physical activity — an increase of 3.17 points.

Conclusions. Plasmapheresis had a positive effect on patients' health in both physical and mental aspects. (JNPN 2019;8(4):143–147)

Key Words: myasthenia gravis, plasmapheresis, functional assessment

Streszczenie

Wstęp. Miastenia jest chorobą autoimmunizacyjną, postsynaptycznej części złącza nerwowo-mięśniowego. Charakterystycznym objawem miastenii jest tak zwana męczliwość albo nużliwość mięśni, która dotyczy również mięśni oddechowych. Jedną z metod leczenia poprawiającą oddychanie i zmniejszającą objawy, które powodują osłabienie mięśni jest zabieg plazmaferezy.

Cel. Celem pracy była ocena funkcjonowania pacjentów z miastenią przed i po zabiegu plazmaferezy.

Materiał i metody. Badania prowadzono w 2019 r. na oddziale Anestezjologii i Intensywnej Terapii na grupie 30 pacjentów z klinicznie potwierdzoną miastenią. W ocenie funkcjonowania chorych posłużono się autorskim kwestionariuszem ankiety. Na prowadzenie badań uzyskano zgodę Komisji Bioetycznej.

Wyniki. Stan pacjenta polepszył się po zabiegu w każdym analizowanym obszarze funkcjonowania (aktywność fizyczna, poruszanie się, oddychanie, przełykanie, nastroj, funkcjonowanie w towarzystwie). Najkorzystniej zabieg plazmaferezy wpłynął na obszar aktywności fizycznej badanych. Przeprowadzona analiza statystyczna, dowiodła występowania istotnej różnicy ($p=0,000$) dla wszystkich wyników, w każdym jej aspekcie (przed i po wykonaniu zabiegu). Jednocześnie najwyższy wzrost średniej zaobserwowano w odniesieniu do aktywności fizycznej — wzrost o 3,17 punktu.

Wnioski. Zabieg plazmaferezy wpłynął pozytywnie na stan zdrowia pacjentów zarówno w aspekcie fizycznym jak i psychicznym. (PNN 2019;8(4):143–147)

Słowa kluczowe: miastenia, plazmafereza, ocena funkcjonalna

Introduction

Myasthenia gravis is an autoimmune disease of the postsynaptic part of the neuromuscular junction, which is caused by the presence of antibodies against the acetylcholine receptor, a muscle-specific tyrosine kinase protein or MuSK-MG, LRP4 protein and probably other synapse proteins [1].

The clinical symptoms of myasthenia gravis are very variable and may be very concealed in the first period. The improvement of well-being after rest is characteristic: often in the morning patients feel better, and during the day the symptoms get worse. Fatiguability may affect ocular muscles, bulb muscles (tongue, throat, larynx), facial muscles and limb muscles. The most common symptoms are drooping eyelids or double vision. Weakening of facial expression muscles causes so-called transverse smile, and involvement of the bulb muscles is manifested by a speech decrease, chewing and swallowing disorders. Weakening of the neck muscles causes the head to drop. Patients may also have difficulty climbing stairs, raising their hands up. Symptoms of the disease may be variable, may be exacerbated by physical exertion, emotional fluctuations, hormonal changes, infections or overheating [2]. Significant worsening of myasthenia gravis symptoms, affecting limb muscles, respiratory muscles and bulb muscles, resulting in respiratory failure may lead to life-threatening myasthenic crisis. Currently, symptomatic and/or surgical treatment of myasthenia gravis is offered to patients. In the case of very severe symptoms, and above all in myasthenic crisis, plasmapheresis is used, i.e. plasma exchange to flush out circulating serum antibodies. Patients should be in the Intensive Care unit at this time. Plasmapheresis is an expensive and labour-intensive method of treatment requiring expensive and specialized equipment, trained and experienced nursing and medical staff. Despite this, this method allows for the rapid elimination of specific pathogenic agents, such as antibodies [3].

When analysing the subject of functioning of patients with myasthenia gravis (including the quality of life of patients with myasthenia gravis), it should be stated that the number of reports on this subject is negligible. However, this subject is interesting and very important for both specialists in the field of neurology and the patients themselves.

The aim of the study was to assess the functioning of patients with myasthenia gravis before and after plasmapheresis. Six important areas of patient functioning were assessed: physical activity, movement, breathing, swallowing, mood and social functioning.

Material and Methods

The studies were conducted in 2019 at the Department of Anaesthesiology and Intensive Care on a group of 30 patients qualified for plasmapheresis treatment. The criteria for inclusion in the study were: 18 years of age, clinical diagnosis of myasthenia gravis, good clinical condition enabling the study to be conducted and written consent of the patient to participate in the study. Taking into account the sociodemographic characteristics of the study group, it was found that the majority of patients are women in the age range of 41–50 (the average age was 46). Most of the respondents came from the city, were married and had higher education (Table 1).

Table 1. Characteristics of the study group

Variable	N	%
Gender		
Woman	29	3.3
Man	1	96.7
Age		
Up to 40 years	10	33.3
41–50 years	12	40.0
50 years and more	8	26.7
Education		
Primary	1	3.3
Vocational	1	3.3
Secondary	12	40.0
Higher	16	53.3
Place of residence		
City	24	80.0
Village	6	20.0
Marital status		
Single/divorced	4	13.3
Married	26	86.7

The studies were carried out using the diagnostic survey method using a proprietary survey. The survey consisted of two parts. The first part contained questions about sociodemographic variables, the second part included questions about important areas of functioning of a patient with myasthenia gravis. Respondents answered the questions contained in the survey before and after the plasmapheresis treatment assessing their functioning on a scale from 1 to 6 (from bad to very good).

The Microsoft Excel spreadsheets and Statistica 12.0 program were used for the statistical analysis. The descriptive analysis calculated the mean (\bar{x}) along with the standard deviation (SD). The non-parametric Wilcoxon test (*Z*) was used to check the significance of differences

between two dependent measurements. The significance level $p \leq 0.05$ was considered statistically significant. The research was approved by the Bioethics Committee.

Results

When assessing the patient’s areas of functioning (Table 2, Figures 1–6), it was found that in terms of the subject’s physical activity — the average physical

Table 2. Areas of patient functioning before and after plasmapheresis

Term study	N	$\bar{x} \pm SD$	Min-Max	Z	p
Physical activity before and after plasmapheresis					
Before plasmapheresis	30	1.33±0.711	1.0–4.0	4.741	0.000
After plasmapheresis	30	4.50±1.253	2.0–6.0	4.741	0.000
Movement					
Before plasmapheresis	30	1.50±0.820	1.0–4.0	4.669	0.000
After plasmapheresis	30	4.57±1.165	2.0–6.0	4.669	0.000
Breathing					
Before plasmapheresis	30	2.10±1.094	1.0–6.0	4.623	0.000
After plasmapheresis	30	5.17±1.053	3.0–6.0	4.623	0.000
Swallowing					
Before plasmapheresis	30	2.06±1.030	1.0–5.0	4.622	0.000
After plasmapheresis	30	5.06±0.960	3.0–6.0	4.622	0.000
Mood					
Before plasmapheresis	30	2.03±1.189	1.0–5.0	4.782	0.000
After plasmapheresis	30	4.97±1.245	2.0–6.0	4.782	0.000
Social functioning					
Before plasmapheresis	30	2.20±1.324	1.0–6.0	4.623	0.000
After plasmapheresis	30	4.93±1.143	2.0–6.0	4.623	0.000

N — number of subjects, \bar{x} — mean, SD — standard deviation, Z — test value for groups $n > 25$, p — the value of the significance level for the test

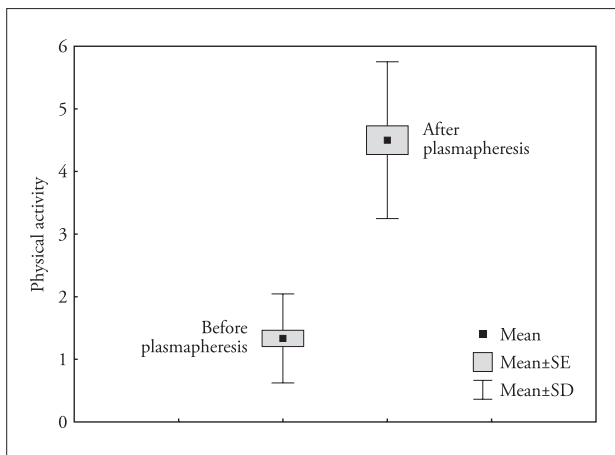


Figure 1. Distribution of average values in relation to physical activity

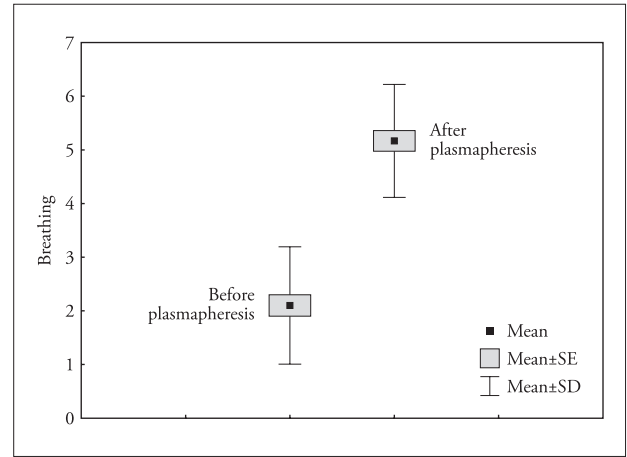


Figure 2. Distribution of average values in relation to the function of breathing

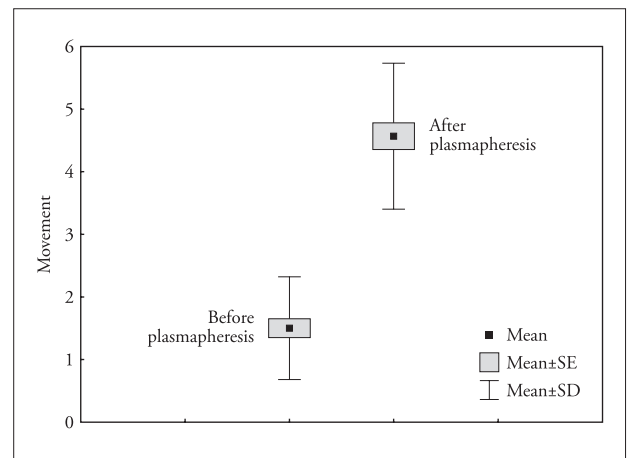


Figure 3. Distribution of average values in relation to the function of movement

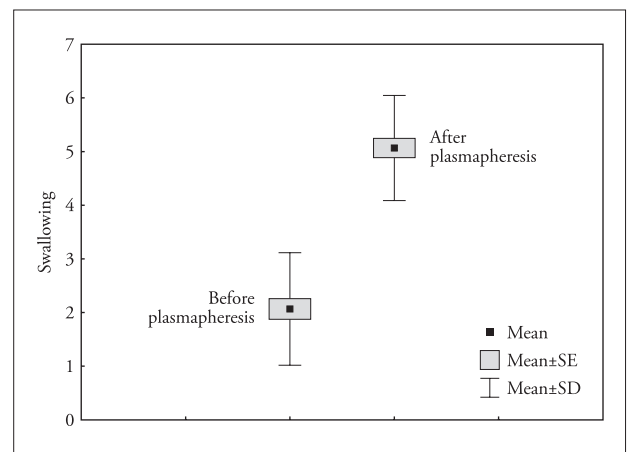


Figure 4. Distribution of average values in relation to swallowing

activity before the treatment was 1.33 points. After the plasmapheresis procedure, the average physical activity increased to 4.5 points (increase by 3.17 points) (Figure 1), at the same time indicating its significant improvement. When analysing the patient’s movement aspect, it was found that the average movement before the procedure was 1.5 points. After plasmapheresis, it increased to 4.57 points (increase by 3.07 points) (Figure 2). In the case

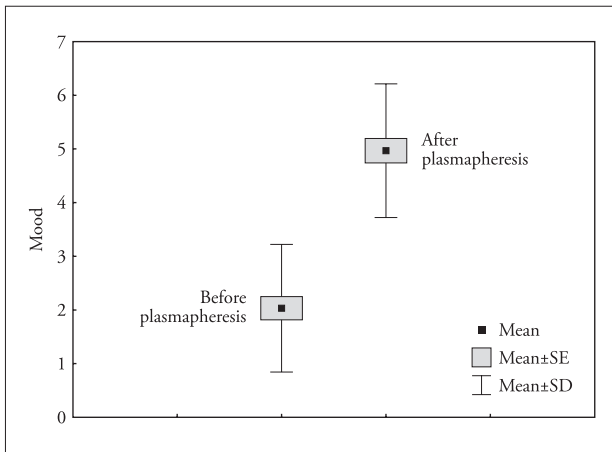


Figure 5. Distribution of average values in relation to mood

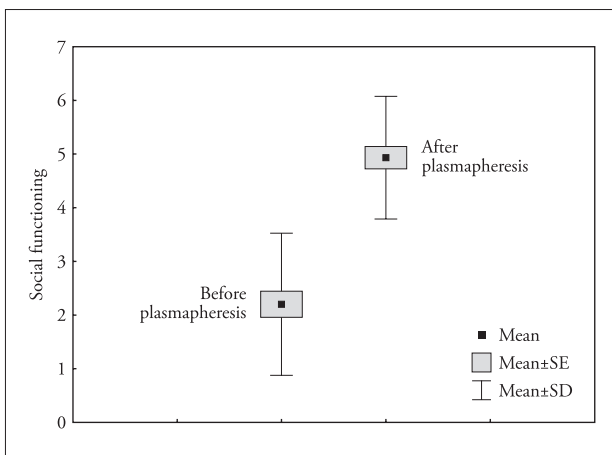


Figure 6. Distribution of average values in relation to functioning in the society

of breathing, the average before the procedure was 2.1 points. After the procedure, it increased to the value of 5.17 points (increase by 3.07 points) (Figure 3). Analysing the swallowing aspect, it was found that the average before the procedure was 2.06 point. After the plasmapheresis procedure, it increased to 5.06 points (increase by 3.00 points) (Figure 4). The average mood also increased significantly. Before the procedure, it was 2.03 points. After the procedure, it increased to 4.97 points (increase by 2.94 points) (Figure 5). In the aspect of social functioning, the average before the procedure was 2.2 points. After the procedure, it increased to the value of 4.93 points (increase by 2.73 points) (Figure 6).

The conducted statistical analysis proved a significant statistical difference ($p=0.000$) for all results in every aspect (before and after the procedure). At the same time, the highest increase in the average was observed in relation to physical activity — an increase of 3.17 points.

Discussion

The Polish study that referred to the topic of plasmapheresis was conducted by Błaszczuk et al. [4].

The authors conducted the study using the method of analysing medical records of patients hospitalized in the Neurology Ward of the Provincial Specialist Neuropsychiatric Care Complex in Kielce. The effectiveness of plasmapheresis has been assessed in relation to three neurological diseases. A group of 29 patients was studied, 11 of whom were patients with myasthenia gravis. The study results indicated the safety and small side effects of plasmapheresis and encourage its use as one of the therapy methods.

Subsequent studies conducted by Rozmilowska et al. [5,6] referred to the assessment of the quality of life of patients with myasthenia gravis who underwent the plasmapheresis treatment. The results of the study showed an improvement in the quality of life of patients with myasthenia gravis after the therapeutic plasma exchange. The quality of life makes it possible to assess the impact of the disease and the treatment undertaken on the patient's functioning in terms of physical, mental and social aspect.

Bilińska and Sitek from the Gdańsk centre [3] conducted a study on a group of 32 patients with myasthenia gravis and 32 healthy people as a control group assessing, among others, their quality of life and acceptance of the disease. In assessing the quality of life, they used the QLQ-30 v. 3 Quality of Life Rating Scale. The obtained results showed a reduction in the quality of life of patients with myasthenia gravis and were confirmed statistically. Both deterioration in overall quality of life and poorer physical/fitness, emotional, social and cognitive functioning were found among the patients, as well as worsening of somatic symptoms of the disease. Unfortunately, this study did not manage to compare patient subgroups with respect to the type of therapy conducted despite many clinical data, and in the group of people with myasthenia gravis no one was subjected to the plasmapheresis procedure. The main assumption of the study was primarily to assess the impact of the disease on the quality of life.

Italian researchers Padua et al. [7] came to similar conclusions; they assessed the quality of life of 46 patients of the Neurology department in Rome with diagnosed myasthenia gravis and compared them with the group of healthy people using the SF-36 questionnaire. The obtained results indicated that patients with myasthenia gravis were characterized by limitations in all areas of functioning.

Conclusions

Plasmapheresis had a positive effect on patients' health in both physical and mental aspects. The patient's condition improved in each of the analysed areas of functioning. The plasmapheresis treatment best affected the area of physical activity of the subjects.

Implications for Nursing Practice

The nurse's role is to efficiently carry out the plasmapheresis procedure, which is also safe for the patient [8]. For this purpose, care should be taken to properly prepare the apparatus and the position in which the procedure is performed, so that the patient does not feel uncomfortable and proper supervision can be exercised. Equipment and substitution fluid are assembled before the planned procedure. After preparing the patient and connecting the monitoring equipment, the anaesthesiologist connects the vascular input. It is required to insert a dialysis catheter into the upper vein through the internal jugular, subclavian and femoral vein. Temporary plasmapheresis catheters should be inserted under stringent sterility conditions to minimize the risk of catheter blood infections. If possible, the catheter should be inserted under ultrasound control. This may increase the effectiveness of the procedure and reduce the percentage of complications. If the vascular insert is already in, care must be taken to avoid complications within it. It should be remembered to change the dressing daily according to the current procedure and to provide documentation of the performed procedure.

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