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Original research article

Improvement of quality of life after therapeutic plasma exchange in patients with myasthenic crisis

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ABSTRACT

Introduction: We sought to evaluate quality of life patients with myasthenic crisis before and after therapeutic plasma exchange.

Materials and methods: In our study we conducted an assessment of the quality of life with the use of the questionnaire SF-36, when executed eleven therapeutic plasma exchange. The assessment was made on baseline and after 4 weeks. We also did neurological clinical evaluation before and after TPE.

Results: Patients in the study showed significant improvement in quality of life after performed therapeutic plasma exchange. The changes were observed in physical functioning, which confirmed the results of the statistical significance of $p < 0.05$. In the analysis, the assessment of mental functioning not obtained the results of statistical significance, but the results also showed improvement in self-assessment. We observed high correlation between general health and physical mental functioning, between the role limitations due to physical health problems and role limitations due to emotional problems, and general health perception and bodily pain.

Conclusions: Therapeutic plasma exchange significantly improves the quality of life of patients with myasthenia gravis during the crisis.

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1. Introduction

Myasthenia gravis (MG) is an acquired autoimmune disease with pathomechanism based on production of antibodies directed against acetylocholine receptors (anti-AchR) of the postsynaptic membrane of the neuromuscular junction. The epidemiological examinations showed that the disease occurs with frequency of 10–15 persons per 100,000 population. The main symptom of myasthenia is progressive muscle fatigability, the typical of which are: apokamnosis symptoms of limbs muscles and mimetic muscles, diplopy, eyelids drooping, ''snarling'' smile the so-called Gioconda's syndrome, difficulty chewing, dysphagia, dysarthria [1]. In conformity with

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Osserman's classification two basic types of the disease can be distinguished: ocular (type 1) and generalized (type 2). The criteria for classification of patients into particular types of myasthenia are exclusively clinical symptoms [2,3].

The diagnostics of myasthenia is based on marking the level of antibodies anti-AChR, which occur at 85% patients. The remaining 15% described as seronegative may have other types of antibodies, such as against muscle-specific tyrosine kinase (anty-MuSK) or antititin antibodies [4–6]

In case of rapid increase of the symptoms and acute course of the disease leading to respiratory inefficiency talk about the myasthenic crisis. It is estimated that every fifth patient has at least one myasthenic crisis in a lifetime and, if the disease is accompanied by thymoma, the risk increases to 50% [7]. The myasthenic crisis usually occurs at patients with generalized type of disease, the symptoms of respiratory inefficiency are accompanied by significant fatiguing of muscles, swallowing disorders, bulbar palsy [8].

The standard procedure in case of the occurrence of the myasthenic crisis is treatment of the patient in an intensive care ward. It is important to implement the therapy (plasmapheresis or intravenous supply of immunoglobulins) as soon as possible, which will allow the shortening of application of mechanical ventilation and limitation of the risk of complications [9].

The patients with diagnosed myasthenia are characterized by a significant variability of the course of their disease. Some of them complain on numerous clinical symptoms, which in a small or a bigger extent make everyday life difficult. The patients understand the concept of health as a functional condition and a quality of life with the disease. The evaluation of the quality of life has in recent years become an important element of everyday medical practice, since the purposes of modern medicine are both prolonging patients' lives as well as improvement and approximating the quality of life to the condition before the disease. In our research we mainly use the definition of the quality of life condition by the health condition (health related quality of life – HRQOL). This value is estimated subjectively by a patient and involves the comparison of the present health condition with the expected condition using respectively general questionnaires, specific and mixed questionnaires [10].

The aim of our study was to evaluate for the first time in our Polish population patients with myasthenic crisis the quality of life before and after therapeutic plasma exchange.

2. Materials and methods

2.1. Patient population

Into the prospective study eleven cases diagnosed with myasthenia during the myasthenic crisis who had been qualified for TPE were enrolled. Only conscious, cooperating, able to fulfilling the questionnaires subjects were included into the study. Demographic patients' characteristics were presented in Table 1. TPE were conducted in patients submitted to therapeutic plasma exchange during years 2012–2015 in the Ward of Anaesthesiology and Intensive Therapy. The work described in this article has been carried out in accordance with the Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans Nr KNW/0022/KB1/68/15; uniform requirements for manuscripts submitted to biomedical journals.

Table 1 – Demographic and clinical characteristics of patients enrolled into the study.

TPE (*n*)	11
Average age (years)	59 (51–71)
Gender (%)	75 (female) 25 (male)
The mean duration of illness (years)	14 (3–32)
Tymectomia (%)	80 (%)
Main neurological symptoms before TPE (%)	Apokamnosis symptoms of limbs muscles and mimetic muscles (100%) Diplopy (81%) Eyelids drooping (75%) Difficulty chewing (63%) Dysphagia (75%) Dysarthria (81%) Dyspnoea (81%)
Electrical activity in repetitive nerve stimulation (%)	Positive (100%)
Antibodies against AchR before TPE (nmol/l) [range/mean]	15.8–1300 (253.17)
Antibodies against AchR after TPE (nmol/l) [range/mean]	39.4–320 nmo/l/179.7
Treatment (%)	Anticholinesterase drugs (100%) Prednisone (27%) Azathioprine (18%)
CRP (mg/l) [range/mean]	0.48–20.92/4.24

TPE – therapeutic plasma exchange, CRP – C-reactive protein.

2.2. Therapeutic plasma exchange

TPE (plasmapheresis) is a method of blood purification allowing to remove inflammatory mediators and antibodies by using extracorporeal circuit. The procedure is used in a variety of conditions, including autoimmune diseases like myasthenia (indication IA during American Society for Apheresis) [11]. TPE is an invasive procedure, but when performed by properly trained and qualified staff, it is relatively safe [12–14]. Life-threatening episodes like shock (anaphylactic or septic), hypotension requiring vasopressor drugs or bleeding are rare [12–14]. The most frequent, but not very severe complications are urticaria, pruritus, hypocalcaemia and mild hypovolemia. Adverse effects are associated more commonly with the administration of fresh-frozen plasma (FFP) used as a replacement solution compared to human albumin solutions.

Patients with diagnosed myasthenia were qualified to TPE by neurologist, based on clinical state and the level of acetylcholine receptor binding antibodies. In Poland TPE is performed in Intensive Care Units, regarding patient's clinical state, often requiring respiratory or circulatory support. Because of many different indications (plasmaphereses are used in treatment of about 150 diseases) and various equipment available, one standard TPE protocol does not exist. We developed our own TPE protocol based on previous

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