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The clinical absolute and relative scoring system—A quantitative scale measuring myasthenia gravis severity and outcome used in the traditional Chinese medicine

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Summary Myasthenia gravis (MG) is a chronic autoimmune disease caused by autoantigen against the nicotine acetylcholine receptor at the neuromuscular junction. With modern treatment facilities, the treatment effect and outcome for MG has been greatly improved with MG and non-MG patients enjoying the same life expectancy. Many classifications of disease distribution and severity have been set up and tested all over the world, mainly in the western world. However, the absolute and relative scoring system for evaluating the severity and treatment effect of MG in China where traditional Chinese medicine (TCM) has been practiced for thousands of years has not been introduced worldwide. The TCM has achieved a great success in the treatment of MG in the country with a huge population. This article serves to introduce this scoring system to the world.

Introduction

Being a chronic autoimmune disease of neuromuscular transimission, Myasthenia Gravis (MG) manifests as fluctuating, fatigable weakness involving different muscular groups.[1] The annual incidence of MG is reported to be 0.25—4 patients per 100,000 residents,[2] with the first peak of onset around the second and third decades of life and the second peak around the fifth and sixth decades. In most cases, it is caused by pathogenic autoantibodies directed toward the skeletal muscle acetylcholine receptor (AChR), but in others, non-AChR components of the postsynaptic muscle endplate, such as the muscle-specific receptor tyrosine kinase (MUSK), might serve as targets of autoimmune attack.[3] The exact origin of the autoimmune response in MG is unknown, however, abnormalities of the thymus gland (hyperplasia and neoplasia) almost certainly play a part in patients with anti-AChR

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antibodies. In most patients, a hyperplasia of the thymus (70—85%) and, in some cases, a thymoma (10—15%) can be found. Genetic predisposition is also likely to influence which patients develop this disorder. MG affects striated muscles, with the hallmark being painless, fluctuating or fatiguing weakness of involved muscles. Patients complain early about diplopia with uni- or bilateral ptosis. Additional manifestations are bulbar symptoms such as speech and chewing disorders and dysphagia. Weakness of mimetic musculature, proximal limbs, and trunk musculature may occur. In severe cases, muscle weakness results in respiratory failure and even death of the patient.

Advances in medical therapy have continuously increased the life expectancy of MG patients without definitively curing the disease. MG patients often are not able to participate fully in daily life, primarily due to their muscle weakness. The persistent experience of weakness may negatively influence patients' perceived quality of life, especially among individuals for whom demands of work, family and other responsibilities require significant physical involvement. According to the pathogenesis of MG, several therapeutic strategies are applied, ranging from acetylcholine esterase inhibitors such as pyridostigmine, imunosuppressors and modulators (e.g., azathioprine, corticosteroids, methotrexate), to plasmapheresis, immunoadsorption, intravenous immunoglobulins or removal of the thymus.[2] Anticholinesterase drugs are the first line of therapy for MG and enhance neuromuscular transmission by inhibiting cholinesterase activity at the neuromuscular junction. However, they do not alter the natural history of MG because they have no impact on the underlying autoimmune process, and the relief of symptoms is often incomplete. The application of the second-line medications (immunosuppressive agents) or thymectomy is eventually required in most patients.[4–6] Plasmapheresis, immunoadsorption and intravenous immunoglobulins are just short-term measures effective for severe exacerbations and are mandatory for MG crisis or threatening crisis. Even with these therapeutic approaches, complete remission of the disease cannot be achieved and relapse of symptoms is frequent besides some serious side effects of cardiac arrhythmia, osteoporosis and hypotension.[7] Thus, alternative approaches with better efficacy and fewer adverse effects are required.

Traditional Chinese Medicine (TCM) is the primary available health care method in China and other Asian countries and has been practiced for thousands of years in these regions to treat many diseases including cancer, inflammation, cardiovascular disease, and Parkinson's disease because of its herbal medicines' long-term immune-regulatory function and few side effects.[8] As a comprehensive system of medicine, TCM is characterized by its own unique theoretical basis and practical experience and has been attracting increasing interest worldwide because of its unique paradigm and purported remarkable efficacy with fewer side effects. The TCM and western biomedical sciences have different viewpoints about the etiology and pathology of diseases and different diagnostic methodologies.[9] In western medicine, a disease is the result of one or more pathogenic factors while in TCM a disease is the combined outcome of both pathogenic factors and maladjustment of the body. Hence, TCM does not focus solely on the disease defined by specific pathological changes (e.g., the level of blood pressure or sugar) rather on the overall health and functional state of the patient. In other words, western medicine focuses on the person's disease while TCM focuses on the diseased person, thus resulting in different thoughts and therapies on the same disease. The immune-regulatory activity of Chinese herbal medicine has been elucidated in several studies with vital roles in immune effector cells, cytokine production and antibody production.[10–13] Traditional Chinese herbal medicines have also been used to treat MG by regulating the immunity of the patients against diseases, with good treatment outcomes reported in the English literature[13–15] as well as in Chinese literature.[16–18] The health care practitioners in China usually use a clinical absolute and relative scoring system to evaluate the severity of MG and treatment effect after appropriate medical management.[19] This clinical absolute and relative scoring system (CARSS)[19] was initially proposed by Xian-Hao Xu in early 1990s and modified in late 1990s in China, incorporating both the initial disease-severity assessment and the improvement extent after treatment. It has been widely practiced across China. However, in the English medical literature, the assessment of the clinical status of MG patients is mainly through scales constructed in the western world.[20–34] The CARRSS has its own advantages in assessing the clinical status of patients with MG and thus is worth publicizing worldwide. Its absolute score reflects the disease severity while the relative score reflects the quantity of improvement after treatment. This article serves to introduce the CARSS used in TCM for the treatment of MG.

The CARSS

Clinical absolute scores

Evaluation aspects

The Clinical absolute scoring system was used to evaluate the patients in the following eight aspects: ptosis, upper eyelid fatigue, eyeball horizontal movement, upper limb fatigue, lower limb fatigue, facial muscles, chewing and swallowing function, and respiratory muscle function[19] (Table 1). These aspects deal with myasthenic severity of seven groups of frequently involved muscles including muscles of eye up-gazing, horizontal movement of eyeballs, upper and lower limbs, facial expression, chewing and swallowing, and respiratory function. At evaluation of the clinical absolute score, all patients should have stopped taking the acetylcholinesterase inhibitor for 8 h.

For ptosis, ask the patient to look straight ahead, and record the level of cornea being covered by the upper eyelid (Fig. 1). The score is given based on the position of the upper eyelid covering the cornea in the clock: 0 (normal): 11—1 O'clock, 1: 10—2 O'clock, 2: 9—3 O'clock, 3: 8—4 O'clock, and 4: 7—5 O'clock. Both eyes should be checked with a total score of 8.

For upper eyelid fatigue test, ask the patient to gaze upwards and record the duration (s) when ptosis occurs. Ptosis is established if the eyelid drops down and touches the pupil at the position of 9—3 O'clock in the clock. Both eyes should be checked with a total score of 8. The score is given