Summer 2015

Myasthenia Gravis

Abbe R. Feaver
Otterbein University, abbe.feaver@otterbein.edu

Myasthenia Gravis

Abbe R. Feaver, RN, BSN

Otterbein University, Westerville, Ohio

Introduction

Myasthenia gravis (MG) is a chronic autoimmune neuromuscular disease characterized by ocular, bulbar, facial, and skeletal muscle weakness due to a defect in the transmission of nerve to muscle impulses at the neuromuscular junction (Mestecky, 2013, p. 110).

Approximately 14-20 in 100,000 people are diagnosed with myasthenia gravis in the United States (Leis, Moore, Kofler, Beric, De Visser, 2014, p. 112). Typically, diagnosis occurs in middle-aged adults with women generally acquiring the disease at a younger age than men (Mestecky, 2013, p. 110). MG has the potential to greatly affect the quality of life of a patient.

Although the disease may go into remission for any given amount of time, there is currently no known cure for myasthenia gravis. Patients experience varying degrees of symptoms, however, there are several treatment options available to help those with MG decrease symptoms and lead full lives.

Significance

MG is an uncommon disease that can greatly affect the lives of the patient and their family members. Patients may experience myasthenic crisis, which is life threatening, however, it is important to remember that they continue living with the disease and its effects every day. Family Nurse Practitioners must design an individualized care plan to treat and manage symptoms in a patient's every day life. It is also imperative to note that official diagnosis of MG may be delayed in many patients because the symptoms vary in each case. It is important to have a solid knowledge base regarding this disease in order to adequately diagnose patients in a timely manner. This is a manageable disease but can become life threatening, therefore, earlier recognition and treatment is essential.

Signs & Symptoms

Bulbar Symptoms:

- Eye muscle weakness
- Ptosis, double vision, blurring
- Slurred speech
- Stuttering
- Low endurance of mastication
- Droop of corners of mouth
- Facial weakness/droop
- Dysphagia

Trunk/Limb Symptoms:

- Proximal symptoms
- Weak neck muscles: head drooping
- Arms, shoulder weakness
- Wrist, finger weakness
- Ankle weakness

The hallmark sign of MG is fluctuating muscle weakness and fatigue that is exacerbated with activity and improves with rest. Weakness is typically least bothersome in the morning, when acetylcholine stores are highest, and symptoms worsen as the day progresses (Abbott, 2010, p. 471).

Underlying Pathophysiology

In a normally functioning neuromuscular system, transmission of nerve impulses to the muscle occurs at the neuromuscular junction (NMJ). The neurotransmitter acetylcholine is released from the motor neurons, crosses the synaptic cleft, and attaches to the acetylcholine receptor on the muscle fibers, consequently generating a muscle contraction. This mechanism is altered in patients with myasthenia gravis.

Myasthenia gravis is a relatively rare, autoimmune, neuromuscular disorder. "MG is a B-cell driven, T-cell dependent, complement and antibody-mediated disease, due to autoantibodies directed against molecules at the neuromuscular junction" (Avidan, Panse, Berrih-Aknin, Miller, 2014, p. 146). These antibodies are directed against proteins concentrated at the NMJ, which include nicotinic acetylcholine receptor (AChR), the muscle-specific kinase (MuSK), or the lipoprotein-related protein 4 (LRP4). The most common antibody found in MG patients is against AChR (Kusner, Ciesielski, Marx, Kaminski, Fenstermaker, 2014, p. 1). "The binding of anti-AChR antibodies to their target impairs neuromuscular transmission by complement-mediated destruction of the postsynaptic membrane" (Avidan et al., 2014, p. 146).

The thymus gland is the chief organ for immunological self-tolerance and usually decreases in size and is replaced with fat as one ages (Weeks, 2012, p. 31). Many patients with MG have thymus abnormalities including hyperplasia or thymoma, which may play a role in disease initiation. These mechanisms include the expression of self-antigens by thymoma cells and impairment of autoreactive T lymphocytes (Ha & Richman, 2014, p. 652). The exact role of thymus in MG is not fully understood, however, researchers suspect that viruses or bacteria may trigger this autoimmune response.

Patients with MG experience cycles of remission and exacerbation. There are many factors thought to exacerbate the symptoms associated with MG. These include pregnancy, emotional stress, thyroid disease, menstruation cycle, and viral or bacterial illness particularly upper respiratory infection (Abbott, 2010, p. 470).

Significance of Pathophysiology

The estimated amount of MG patients has more than doubled in the last twenty years (Avidan et al., 2014, p. 146). This could be because of many factors including better diagnosis, improved treatment, and an increasing aging population. The average age of onset for females is 20-30 years; the average age of onset for males is 60-80 years. This is significant information to consider because of the increasing aging population. It is probable that because people are living longer, there is an increase in the amount of people experiencing the onset of MG.

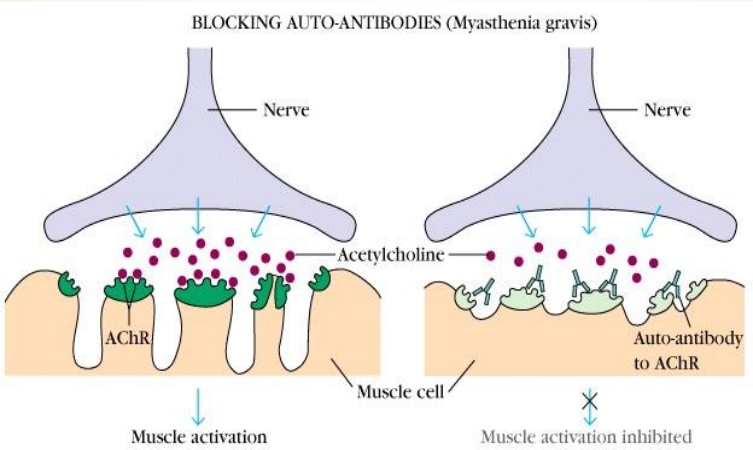
Although there is plenty still unknown, MG is considered to be one of the best understood of all autoimmune diseases. Before improved corticosteroid therapies were developed, one third of patients diagnosed with myasthenia gravis died (Abbott, 2010, p. 470). With continued improvements in research, diagnosis, and treatment the prognosis is good in terms of quality of life, daily functions, and survival (Sieb, 2013, p. 408).

Understanding the pathophysiology is of extreme importance so diagnosis is completed quicker and more accurately, and treatment options are performed. As mentioned previously, it is now known that MG can be divided into subcategories of antibody specificity. Understanding exactly where and how the antibodies are impeding function at the NMJ leads to more accurate treatment options and prolonged periods of remission.

Knowing the significance of pathophysiology of MG is crucial because these patients are susceptible to myasthenic crisis, a life threatening condition. During myasthenic crisis, the respiratory and accessory muscles are significantly weakened as well as the diaphragm (Weeks, 2012, p. 33). These patients often require a mechanical ventilator until the exacerbation subsides. Although a critical condition, with improvements in knowledge about the disease and proper treatments, mortality from myasthenic crisis alone is less than 5% (Weeks, 2012, p. 32).

Qualitative research also assists in understanding the significance of the disease process. "Incorporating the patient's perspective provides invaluable information regarding which impairments are most relevant to patients as well as patterns of impairment" (Barnett, Bril, Kapral, Kulkarni, Davis, 2014, p. 1). A patient may verbalize difficulty with daily activities due to weakness but may have a one time clinical presentation displaying the opposite. Concerning MG, this is the hallmark sign of fatigability. A solid knowledge base of the pathological processes taking place will only enhance the understanding of the patient's perspective of symptoms, thus, leading to better treatment and care plans.

OTTERBEIN UNIVERSITY



Implications for Nursing Care

The highly variable symptom presentation of myasthenia gravis can pose considerable challenges to accurate diagnosis. Because of the similarities of symptoms with many other neurological diseases and disorders, providers may unknowingly give a wrong diagnosis. Physical examination, thorough neurological assessment, and detailed medical history are of utmost important. Providers, such as nurse practitioners, must have a substantial understanding of MG and its presentation in order to differentiate other diagnoses. It is now becoming possible to identify the antigenic target in almost all cases of MG, consequently leading to more specific care plans and focused therapies (Lewis, 2013, p. 97). The earlier MG is recognized and treatment started, the better the outcomes and quality of life for the patient.

Education plays a huge role in nursing care for those affected by MG. Knowing the disease process, how it affects ones body, and ways to improve or avoid symptoms is of utmost importance. Nursing care plays a primary role in educating patients on how to enhance their daily lives while living with a chronic condition. Because of the increase in diagnosis and aging population as previously stated, providers are highly likely to come in contact with a patient diagnosed with MG during their career. Understanding the pathophysiology as well as the patient's perception of the disease and its symptoms will only serve to improve overall care and clinical decision-making.

Conclusion

Myasthenia gravis was once an unknown neurological disease with poor outcomes. However, with continuous advanced research, it is now one of the most understood autoimmune diseases today. Although it is still a serious condition, patients with this diagnosis are expected to have longer survival rates and increased quality of daily life activities. Nurse practitioners are providers that will have contact with these patients very frequently and keen assessment skills are needed for accurate diagnosis. Continuous research and treatment plans may lead to not only longer periods of remission, but possibly a cure.

References

Abbott, S. (2010). Diagnostic challenge: Myasthenia gravis in the emergency department. *American Academy of Nurse Practitioners, 22*, 468-473. doi:doi 101111/j.1745-7599.2010.00541.x

Avidan, N., Panse, R., Berrih-Aknin, S., & Miller, A. (2014). Genetic basis of myasthenia gravis a comprehensive review. *Journal of Autoimmunity,* 146-153. doi:doi:10.1016/j.jaut.2013.12.001

Carolina, B., Bril, V., Kapral, M., Kulkarni, A., & Davis, A. (2014). A conceptual framework for evaluating impairments in myasthenia gravis. *PLOSONE, 9*(5), 1-9. doi:doi:10.1371/journal.pone 0098089

Ha, J., & Richman, D. (2014). Myasthenia gravis and related disorders: Pathology and molecular pathogenesis. *Biochimica Et Biophysica Acta (BBA) – Molecular Basis of Disease,* 651-657. Doi:

Kusner, L. L., Ciesislski, M. J., Marx, A., Kaminski, H. J., & Fenstermaker, R. A. (2014). Survivin as a potential mediator to support autoreactive cell survival in myasthenia gravis: a human and animal model study. *PLOS ONE, 9*(7), 1-10. doi 10.1371/journal.pone.0102231

Leis, A. A., Moore, A. R., Kofler, M., Beric, A., & De Visser, M. (2014). Keep an eye out for myasthenia gravis patients with an eye out. *Frontiers In Neurology, 5*1-3. doi:10.3389/fneur.2014.00112

Lewis, R. (2013). Myasthenia gravis: New therapeutic approaches based on pathophysiology. *Journal of the Neurological Sciences, 333*, 93-98. doi:doi:10.1016/j.jns.2013.06.018

Mestecky, A. (2013). Myasthenia gravis. *British Journal Of Neuroscience Nursing*, *9*(3), 110-112.

Myasthenia Gravis: Causes, Signs, Symptoms, Investigations, Treatment-Medications, Surgery. (2015). Retrieved from http://www.epainassist.com autoimmune/myasthenia-gravis

Myasthenia Gravis. (2014). Retrieved from http://www.beverlydoc.com/our-services myasthenia-gravis/

Sieb, J. P. (2014). Myasthenia gravis: an update for the clinician. *Clinical & Experimental Immunology, 175*(3), 408-418. doi:10.1111/cei.12217

Weeks, B. (2012). Myasthenia gravis: Helping patients have better outcomes. *The Nurse Practitioner, 37*, 31-36.