The lived experience of adults with myasthenia gravis: A phenomenological study

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Abstract
Myasthenia gravis (MG) is a disorder of the neuromuscular junction (NMJ) that causes fatigue and fluctuating muscle weakness (Hickey, 2009). The physiology of this disease is well understood and there are numerous medically focused articles that outline historical data, randomized controlled trials of treatment options and unusual case studies. The nursing literature about MG is limited and dated.

The aim of this study was to examine and understand the lived experiences of adults with MG. An interpretive phenomenological approach has been used that applies the research methodology of van Manen (1990). Seven people living with MG were interviewed and their experiences of the disease recorded. Questions were broadly worded about various topics related to MG and were guided by individual experiences. Thematic analysis revealed that MG affects every aspect of a person’s ‘lifeworld’: their sense of time, body, space and their relationships with others.

The findings of this study highlight three main themes embedded in the data that a person with MG experiences: living with uncertainty, living with weakness and living with change. These experiences have been interpreted and discussed to gain a deep understanding of the meaning of the disease. This study raises awareness of MG for neuroscience nurses and provides a unique view of this disease.

Key words: myasthenia gravis, lived experience, phenomenology.
strategy was effective because it yielded a vast amount of information that addresses all aspects of MG and has provided a sound literary background for this study.

Definition and pathophysiology
In MG the main muscles affected are skeletal (Alshekhlee, Miles, Katirji, Preston, & Kaminski, 2009), in particular the voluntary muscles innervated by motor nuclei of the brainstem (Ropper & Samuels, 2009). Ineffective neuromuscular transmission occurs because of the destruction of nicotinic post synaptic acetylcholine receptors (AChRs) by acetylcholine receptor antibodies (Lindsay, et al., 2010). This results in decreased strength of muscle contractions and, with repetition, there is less acetylcholine (ACh) available which causes the muscle to become fatigued (Abbott, 2010). Acetylcholine receptor antibodies are found in up to 90% of people with MG (Cavel-Greant, 2008).

Classification and presenting symptoms
Ocular symptoms can occur in up to 90% of people with MG (Ropper & Samuels, 2009). Ptosis can be unilateral or bilateral (Barker, 2008) and occurs because the extra ocular muscles are weak (Hickey, 2009). Ptosis can consequentially cause blurred or double vision (Barker, 2008). Lindsay, Bone and Fuller (2010) suggested up to 40% of people with ocular myasthenia will develop generalised myasthenia. Bulbar weakness can result in facial weakness, dysarthria and dysphagia (Kaminski, 2009). Facial weakness can produce an expressionless mask-like appearance that makes it difficult for a person to smile, causing a characteristic ‘myasthenic snarl’ (Lindsay, et al., 2010). Dysphagia associated with MG can account for weight loss of 5-10kg for a person in the months prior to diagnosis (Kaminski, 2009). Progression to generalised weakness involves the diaphragm, intercostal and neck muscles which can result in breathlessness and dyspnoea (Hickey, 2009). Limb weakness is usually bilateral in the muscles closer to the torso, especially the hips, shoulders, upper arms and legs (Cavel-Greant, 2008). Fatigue is also a symptom of MG (Hickey, 2009) which may increase during the day and improve with rest.

Thymus involvement
A thymoma is a rare tumour of the thymus gland caused by epithelial neoplasms (Kaminski, 2009). Thymoma is more common in men between the ages of 50 and 60 years (Ropper & Samuels, 2009). Matsui et al. (2009) found that MG is unrelated to ethnicity, however Kaminski (2009) suggested there is a higher incidence of the disease among African-Americans. MG with thymoma occurs more frequently among Māori or Pacific Island people and presents at a younger age in these populations (Fink, Wallis, & Haydock, 2001).

Older adult considerations
There is a higher incidence of MG in adults over 65 years of age (Caserta, et al., 2010; Matsui, et al., 2009; Vincent, Clover, Buckley, Evans, & Rothwell, 2003) and may be under-diagnosed in older adults. Vincent et al. (2003) believed this may be due to co-existing morbidities older people have that may mask the symptoms of MG, while Matsui et al. (2009) suggested increasing incidence could be the result of the ageing population in general, improvements in diagnostic tests or an increased awareness of MG. Thymoma is more common with late onset patients, as is an increase incidence of bulbar symptoms and progression to more severe disease (Kaminski, 2009).

Pregnancy considerations
Pregnancy can unpredictably alter the course of MG (Gurjar & Jagia, 2005; Kaminski, 2009). Complete remission, clinical improvement, acute exacerbation and change in symptoms of MG have been documented with
pregnancy (Gurjar & Jagia, 2005). While most women can have a successful pregnancy and delivery (Cavel-Greant, 2008), it is important to note that the treatments for maternal MG can affect the foetus (Kaminski, 2009).

**Myasthenic crisis**

Myasthenic crisis involves a sudden onset of severe muscle weakness that can cause respiratory failure leading to the patient requiring intubation and ventilation (Alshekhlee, et al., 2009). It can be caused by a lack of anticholinesterase medication or precipitating factors such as infection, surgery or stress (Hickey, 2009). Myasthenic crisis should be diagnosed and treated promptly and it is important that nurses are able to identify this medical emergency (Chaudhuri & Behan, 2009; Kutzin, 2011). A rare cholinergic crisis can also occur due to the toxic effects, or overmedication, of anticholinesterase drugs (Ropper & Samuels, 2009).

**Diagnosis**

The Tensilon test involves administering intravenous edrophonium, a short-acting anticholinesterase inhibitor (Moore & Shepard, 2014). Edrophonium works by inhibiting enzyme action, which prevents the breakdown of ACh molecules and improves muscle weakness (Cavel-Greant, 2008). The diagnosis is considered positive if there is an improvement in muscle strength lasting approximately five minutes (Moore & Shepard, 2014). Nerve conduction studies (Hickey, 2009), Immunological tests to determine elevated AChR antibodies MG (Heldal, Owe, Gilhus, & Romi, 2009) and a CT of the mediastinum to detect thymoma (Barker, 2008) may also be used to diagnose MG.

**Treatment options**

Because there is no cure, treatment options are aimed at managing the symptoms of MG and can include medication, plasmapheresis, intravenous immunoglobulin (IVIG) and thymectomy (Fowler, 2013). Anticholinesterase drugs are the first line approach for managing the symptoms of MG (Barker, 2008). They work by inhibiting cholinesterase, the enzyme that breaks down ACh (Lindsay, et al., 2010). Anticholinesterase drugs allow larger amounts of ACh in the synapse for a longer time resulting in more effective nerve transmission (Woodward & Waterhouse, 2009). Pyridostigmine bromide (Mestinon) is the most common anticholinesterase drug used to treat the symptoms of MG (Kumar & Kaminski, 2011). Doses are individualised and improvement in muscle weakness can occur within 15-30 minutes after oral administration (Hickey, 2009). Some people may require additional immunosuppressive drugs, such as the corticosteroid prednisone, which is most commonly used and can provide significant improvement in patients (Kumar & Kaminski, 2011). Other immunosuppressant drugs that can be used include Cyclosporine, Azathioprine (Imuran), Mycophenolate mofetil, Tacrolimus, Cyclophosphamide, Methotrexate and Rituximab (Kumar & Kaminski, 2011).

Plasmapheresis is used as an option for patients in myasthenic crisis and prior to thymectomy (Kaminski, 2009; Lindsay, et al., 2010). The benefits of plasmapheresis can last up to six weeks (Barker, 2008), but there can be complications and side effects (Kumar & Kaminski, 2011). IVIG can also be used as a short-term treatment for worsening symptoms (Ropper & Samuels, 2009). Thymectomy can result in an improvement in patients with generalised MG by inducing remission (Costello, 2006).

**Lived Experience**

Van Manen (1990) built on the work of early 20th Century European philosophers by developing a modern extension of traditional phenomenological methods. Van Manen (1990) advocated a ‘human science’ approach to researching the lived experience and used this term interchangeably with the terms ‘phenomenology’ and ‘hermeneutics’. Van Manen (1990) believed phenomenology is the study of essences and descriptions of meanings as they are lived; lived experience is the explication of phenomena as they present themselves to consciousness.

There is limited literature on the lived experiences of people with neurological conditions, especially neuromuscular disorders (LaDonna, 2011). Chen, Shih, Hayter, Hou & Yeh (2013), found patients with MG experienced physical and emotional difficulties that impacted on quality of life. For this study, the experiences of people with other neurological conditions with similar symptoms to MG such as Guillain-Barré Syndrome, stroke, multiple sclerosis, motor neuron disease, muscular dystrophy and cerebral palsy were explored. Some people have published books about their individual experiences with MG (Atkins, 2010; Byars, 2007; Gray, 2011; Hill-Putnam, 2010; Smart, 2006). The focus of these publications are stories, journeys and reflections on life events, and they all include specific examples of how they have experienced MG.
Method
An interpretive phenomenological approach developed by van Manen (1990) was used for this study. This method aims to understand thorough interpretation and identify common themes of the lived experience. The purpose of this qualitative study was to develop detailed, insightful perspectives to understand the lived experiences of adults with MG. This study was approved by the Canterbury District Health Board (CDHB), the Upper South B Regional Ethics Committee and the Research Manager of Māori Health. Seven adults with generalised MG aged between 31-86 years volunteered to participate in this study. Each person was interviewed once on a broad range of topics related to MG. The transcribed interviews were sent to the participants to review before they were returned to the researcher for data analysis.

Relevant text was analysed using van Manen’s (1990) ‘highlighting’ approach. Identified themes were clustered into four ‘existentials’ that represent a person’s ‘lifeworld’: lived time, lived body, lived space and lived other. Lived time (temporality) is subjective time and involves the “temporal way of being in the world” (van Manen, 1990, p.104). Lived body (corporeality) refers to the way “we are always bodily in the world” (van Manen, 1990, p.103) and means our physical or bodily presence reveals something about us. Lived space (spatiality) is ‘felt’ space and “refers us to the world or landscape in which human beings move and find themselves at home” (van Manen, 1990, p.102). The concept of lived other (relationality) involves the lived relations maintained with others in shared interpersonal space (van Manen, 1990).

Results
Thematic analysis revealed that MG affects every aspect of a person’s ‘lifeworld’: their sense of time, body, space and their relationships with others. The ‘existentials’ were used as major headings and examples from the transcripts have been included to illuminate the essence of each dimension. It is essential that these dimensions are considered, as a whole to truly reflect the lived experience.

Lived time: past, present and future time
Each person interviewed in this study recalled where they were in relation to time when they first noticed symptoms “What is happening to me?” and when they were diagnosed “It was he who picked it, he put all those symptoms together”. This was significant and had meaning for them. Present time was dominated by medication “I counted twenty pills a day!” and fatigue “I have to have my sleep in the afternoon”. There was also a fear of what may happen by some of the participants in this study “I just don’t want to go backwards”. MG is intrusive and alters a person’s sense of time regularly along the disease continuum.

Lived body: physical and psychological effects
The physical symptoms of MG can subtly or severely invade a person’s body. The people in this study described difficulty with swallowing “I just couldn’t swallow” and communicating “I couldn’t speak, I was talking all funny”. The effects of visual disturbances “I thought I was going blind” and fatigue “I would spend most of my time sleeping” were also significant. MG is unpredictable and a person may have little or no control during acute exacerbations. Experiences also included an array of emotions including fear “I was scared about the breathing” and anxiety “I just wanted to hide away, actually” in addition to grief due to loss of body function.

Lived space: restricted, improved and home space
The experience of MG restricts a person’s dimensions of space, both mentally “I wasn’t with it really, I was in just another kind of world” and physically “I wasn’t allowed out of bed”. A person’s landscape can become cluttered with equipment and have limits set by health professionals in the acute phase of illness. Some of the people in this study reported feeling more positive “I’ve turned the corner” when they improved and developed a sense of determination “It’s not going to defy me”. Within home space, MG can intrude on everyday activities “It’s a shame how you give things up” resulting in isolation “I can’t drive anymore” and financial concerns “I could not work”.

Lived other: relationships with family, friends, health professionals and faith
Relationships with others was significant in this study at every stage of illness. Family concern “My son, he seems a bit worried” and support “I could not have done without them” were acknowledged, in addition to strained relationships “We do have our words of course” the disease caused. Friendships changed and were either altered “I can’t relate to people in the same way that I could” or strengthened as a direct result of the disease...
“It was a wonderful experience with the patients”. Despite often getting conflicting information from health professionals “But even doctors will all say different things”, having doctors and nurses that were trustworthy “To have someone I was able to trust” was a significant finding. Medical and nursing students were spoken about often positively throughout this study “Students have got to learn too”. Whether it was the support of family and friends, the importance of faith “I had a lot of people praying for me” or the trust they placed in health professionals, it was essential people with MG have relationships with others. This appeared to be a method of coping with the disease.

The findings of this study highlighted the many different ways a person experiences MG. In viewing the results through the dimensions of lived time, lived body, lived space and lived other, clear embedded themes related to the phenomenon of MG emerged. The themes of uncertainty, weakness and change are illustrated in the following diagram (Figure 1).

![Figure 1](image.png)

**Figure 1 (Above): The interrelated experiences in adults with myasthenia gravis.**

Living with uncertainty, weakness and change are all meaningful experiences in people with MG and, when interlinked represent the lived experience of this disease. At different stages of the disease, these intertwined circles expand and contract with a sense of fluidity, but are always present to some degree in a person living with MG.

**Discussion**

It is important to note that these findings are the researcher’s interpretation of the experience of MG from the data obtained. More participants may have represented the MG population in a different way and collection of structured quantitative data may have enhanced the results of this small study.

**Living with uncertainty: altered life equilibrium**

People with MG experienced symptoms that are constantly changing, which can result in varying degrees of uncertainty that can occur at any time along the disease continuum. Throughout the duration of chronic illness, Hickey (2009) suggested a sense of uncertainty can occur as the level of disability waxes and wanes. Uncertainty is a characteristic of chronic illness (Kaminski, 2009), and this dynamic experience can have a major impact on a person (Hansen, Rortveit, Leiknes, Morken, Testad & Joa 2012). A sense of uncertainty is always present in some people with MG.

**Living with weakness: altered physical strength and energy**

The experience of weakness can occur at any stage in people with MG and was often the first sign that something was wrong. MG is essentially an invisible disease unless a person exhibits signs of muscle weakness or fatigue. The weakness associated with MG has been well described in the autobiographical literature (Atkins, 2010; Cavel-Greant, 2008; Gray, 2011; Hill-Putnam, 2010; Smart, 2006) but the meaning of weakness has been elusive. Muscle weakness was found to be subtle or severe, acute or chronic and had a profound effect on each individual that was interviewed in this study and therefore a significant experience of MG.

**Living with change: altered control, outlook and daily life**

The people in this study made significant changes to their daily lives as a result of their experiences of MG, although, it was not perceived or viewed in a negative manner. There were three main areas of change which illuminated the lived experience. Having control or lack of control and autonomy was significant and similar to people with cerebral palsy (Sandström, 2007) and amyotrophic lateral sclerosis (King, Duke & O’Connor, 2009). A positive outlook and determination were all experienced by people in this study which was also found in people with Guillain Barré Syndrome (Forsberg, Ahlström & Holmqvist, 2008) and muscular dystrophy (Boström & Ahlström, 2004). Daily life change highlighted the alteration and adaptation people with MG have made in relation to self-care, hobbies, employment and driving.
Conclusion

The pathophysiology of MG is well established in the literature and the results of this literature review provide a broad understanding of the aspects of this disease. The nursing literature available is limited, dated and highlights the need for quality nursing research within this specialist area. The literature review draws attention to the lack of information on the lived experiences of people with MG, a lack of nursing research and a lack of New Zealand led research. However, there is emerging literature which examines MG from a cultural viewpoint (Chen, et al., 2013) which supports some of the findings in this study.

What is the meaning of this disease?
The experiences in people with MG tell us that it is an individual experience with similar aspects that are shared by others with the disease. MG is an intrusive disease that causes physical and psychological challenges differently for each individual affected. The meaning of MG is multifaceted. The experiences of the people interviewed in this study are unique to their 'lifeworld'. This study has identified that people who have the same diagnosis of MG experience the disease in different ways, but do have common elements that are similar to other types of neurological disease. A person with MG lives in a dynamic equilibrium in their world where the experiences of uncertainty, weakness and change are interlinked and always present in some shape or form. These experiences serve as a constant reminder that disease is present, even if a person is in remission.

Relevance to Clinical Practice

MG, as a chronic health condition, can be severe and debilitating. The disruptions it causes to the physicality of one's body, to lifestyle, and to one's self-image make it imperative that gaining understanding into patient experience is essential. Nurses are in a unique position to understand the disease process and its meaning for the patient (Benner & Wrubel, 1989) and have a significant role when helping patients with MG adapt and cope with their illness. This study raises awareness of MG for nurses and other health professionals. It provides a unique view of the disease by exploring the meaning of MG and fills a gap in the nursing literature. The findings support existing studies in the area of neurological illness and add to the existing body of neuroscience knowledge. Despite its small sample size, this study may enhance clinical practice for nurses caring for people with MG, in particular ensuring accurate assessment, care planning, and collaboration with allied health, patient education and future research.

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