

# Ocular Myasthenia

Much has been written about Myasthenia Gravis (MG) in recent years, because there now seems to be a plausible, scientific explanation for the cause of this disease. The word "gravis" seems no longer appropriate, as current forms of treatment have allowed patients to live fully functional and independent lives. This review will summarise the therapeutic advances that have been made in the ocular form of MG, stressing the most effective and practical ways to manage double vision (diplopia) and droopiness of one or both lids (ptosis). The term Ocular MG as used here implies and describes symptoms that are confined to eye muscles.

## **Anatomy & Pathophysiology**

MG is an autoimmune disorder, i.e. it is caused by an anti- body which attacks and diminishes the integrity of one of the body's own components. Making antibodies is a normal daily process from the moment we are born and exposed to such foreign proteins as viruses and bacteria. For reasons, which are still not fully understood, some patients make antibodies against their own body proteins, and in MG, these antibodies are directed against the region where the nerves have their junctions on their muscles.

All the muscles in our body are activated by nerve impulses, which travel along nerve trunks emanating from the brain and spinal cord. When their nerve impulses reach the neuromuscular junction (the point at which a nerve fibre terminates on a muscle fibre) a chemical is released called acetylcholine (ACh), which attaches to the receptor on the muscle, resulting in a muscular contraction.

Patients with MG produce a blocking antibody that deposits on the muscle receptor and prevents the entry of all molecules. This leads to muscular fatigue and sometimes- frank paralysis. Characteristic of MG is the randomness of neuromuscular blockade. It may be confined only to a small muscle that moves one eye upward, outward, downward or laterally; or to one of the larger muscles which moves the face, arm or leg or breathing muscles. Regardless of the muscles involved, the goal of effective treatment is to either reduce the concentration of the blocking antibody or to increase the concentration of ACh at the neuromuscular junction.

## **Clinical Characteristics**

Ocular MG is characterised by the abrupt or insidious onset of weakness or fatigueability of one or both lids or the eye muscles. If the lid is involved it droops, otherwise known as ptosis; with extra ocular muscle involvement the patient sees double looking in the direction of the weak muscle.

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For instance, they may see well in all directions except upward, which tells the examiner that one of the elevator muscles is weak.

To compensate for the weakness, the patient can tilt their head or turn their face to allow the relatively stronger eye muscles to work. In the above example, the patient would tilt their head back, thus throwing their eyes relatively downward out of the field of action of the relatively weak elevator muscle.

Because of the frequency with which eye muscles are involved in MG, droopiness of one or both upper eyelids (ptosis) and double vision (diplopia) are common impediments. Diplopia or ptosis eventually occur in 90% of patients with MG and account for the initial complaint in 75%.

About 80% of patients with ocular onset MG will progress to involvement of other muscle groups within the first two years; thus only 20% of patients have pure ocular MG. Patients in whom the disease has been confined to the ocular muscles for three or more years seem unlikely to progress to the generalised disease.

Eye muscle strength in MG, just like swallowing, speech and leg strength may be normal or mildly affected in well-rested patients, but weakness can usually be elicited with exercise. For instance, if a patient is asked to look forward for sixty seconds, thereby testing the endurance of the vertical eye muscles and the upper lid that concomitantly rises, the patient may evolve from a normal state to extreme weakness with flagrant diplopia or ptosis.

Although upward eye movements are said to be involved earliest, extra ocular muscle involvement along the horizontal plane is equally as common. Essentially any pattern of disordered eye movement may develop so that isolated weakness or total immobility of the eyes can occur, sometimes mimicking other medical conditions such as strokes, tumours, thyroid eye disease, infections and multiple sclerosis.

## Clinical Course

The natural history of Ocular Myasthenia has been well studied. Of one hundred and sixty-eight patients with generalised MG, in whom the average follow-up was twelve years, and the last examination was compared with the initial examination, 68% were unchanged, 14% were improved, 14% complete remission and 5% were worse. Most of the deterioration seemed to occur during the first three - five years of illness during which time the

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incidence of a severe exacerbation is highest. More specifically, of those patients who present pure ocular MG, 58% will show symptoms of generalised muscular weakness during the first seven months of their illness, another 29% between seven months and thirteen months, 7% during the first two and three years and only 6% after the third year. Therefore, if a patient with ocular MG continues to have only ocular symptoms after three years, there is a 94% chance that his symptoms will not increase.

The salient symptoms of Ocular MG, double vision and droopiness of one or both lids, are frequently influenced by environmental, emotional and physical factors.

Bright sunlight, emotional stress, viral illness, surgery, menstruation, pregnancy, immunisations and other physical factors may all precipitate a change in the expression of Ocular MG, although not in a predictable direction.

Spontaneous remissions can occur in any patient and sometimes persist for years. Most patients with MG show a fluctuating course, often with a particular muscle group (e.g. ocular, speech, swallowing, arm or leg strength) being maximally involved. Fortunately MG only rarely follows an acutely or chronically progressive course. Age, sex, and pattern of onset do not predict the eventual course of the disease.

## Diagnosis

The diagnosis depends on the patient's story and physical findings, on a blood test for the myasthenic antibody (positive in about 60% of patients in the Ocular MG) and on a very sensitive electrical test (single fibre EMG). In a few cases it is necessary to test the response to an injection of Tensilon (edrophonium). This drug temporarily increases the amount of ACh at the neuromuscular junction, and so, when it is carefully injected into a vein, it may produce instant improvement in muscle weakness, be it of eye muscles or others, but this lasts for only a minute or so. The test carries some dangers, and should only be done by someone who is experienced in using it, preferably in hospital.

## Anticholinesterase Medication

An anticholinesterase medication, usually Mestinon (pyridostigmine), is the first line of treatment. Used judiciously, it can often relieve the ocular complaints. However, it is often unsuccessful for the following reasons: (1) The diplopia may be only partially relieved and still be bothersome. (2) The

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dosage schedule may be inappropriate. (3) By improving the ptosis it may unmask the diplopia. (4) The side effects may be intolerable, especially in the older patients. If Mestinon is ineffective, then other medication, such as corticosteroids (prednisolone) or drugs like azathioprine that suppress immunity may succeed. However, if the disease is limited to the eyes, it is important to balance the potential benefit to the patient against the risks imposed by the short and long-term side effects of the medication.

## **Prednisolone Treatment**

Low dose alternate day treatment often leads to complete remission of symptoms in patients who have not responded to anticholinesterase therapy. The dose is usually increased slowly (e.g.: by 5mg a week) from a low starting value (e.g.: 5mg), either until symptoms recover or an agreed 'ceiling' dose is reached (e.g.: 50mg alternate days).

This dose is then usually held until the patient has been free of symptoms for 2-3 months, before reducing the dose slowly (e.g.: 5mg month), aiming to define the 'effective minimal dose', i.e. the lowest dose at which symptoms are fully controlled. If an attempt is made to withdraw prednisolone completely, the ocular symptoms will often come back. Before starting prednisolone treatment, it is important that patients should be made aware of the potential side effects.

## **Other Treatment**

If medications are ineffective or not tolerated, other methods can be used to relieve the patient's ptosis and double vision. Fresnell lenses, for instance, are flat, plastic prisms that can be attached to a diplopic patient's eyeglasses to relieve them of diplopia. By optically "bending light", the patient can comfortably look straight ahead and downward to read with both eyes open.

Ptosis crutches have been used in some patients to mechanically elevate a droopy lid. These are manufactured and fitted by only a handful of opticians and should be prescribed only in extreme circumstances. Sometimes just taping a portion of an eyeglass can avert diplopia, or minimise the action of weakened eye muscle(s). Most specialists would not recommend Thymectomy for patients purely with Ocular Myasthenia.

## **More Information**

If you wish to have more information about M.G. or the Myasthenia Gravis Association and its activities please contact:

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