

Understanding Your Child's Myasthenia Gravis



- What is MG?
- How can I know that my child has MG?
- Is there a cure for MG?
- What kinds of treatments are there for my child's MG?
- What can my child and I expect with MG?
- What are the prospects for my child?

What is MG [My-as-theen-ee-a Gra-vus]?

Myasthenia gravis (MG) is an autoimmune disorder in which antibodies attack and destroy nerve muscle connections resulting in muscle weakness. MG is a rare disease affecting about 20 per 100,000 people, or about 60,000 people in the U.S. But, Juvenile MG is even rarer making up about 10 to 15% of all MG cases in North America.¹ There are 3 major types of MG found in children and teens.

1. **Transient neonatal MG** appears temporarily in infants born to women with MG. This happens when antibodies common in MG cross from the mother through the placenta into the fetus. Typically neonatal MG lasts only a few weeks and these infants are *not* at greater risk of developing MG later in life.
2. **Congenital MG (CMG)** is an extremely rare non-immune form of MG which is inherited. Those with congenital MG begin experiencing symptoms at birth. Some infants have their diagnosis made during infancy; however, others are not recognized as having CMG until later in life, even during adult years. For more about Congenital MG, please see the MGFA brochure:

Terms & Definitions

- Acetylcholine** – molecule that acts as a nerve transmitter
- Antibody** – a protein created by the body's cells to fight infections.
- Autoimmune** – when a body mistakenly attacks its own cells and tissues.
- Congenital** – existing since birth
- Corticosteroids** – are hormones that are involved in a wide range of bodily responses such as stress and regulate inflammation and metabolism
- Electromyography** – test of the electrical activity produced by skeletal muscle.
- Intravenous** – in vein
- Juvenile** – childlike, young
- Neonatal** – newborn
- Placenta** – the organ that nourished the unborn baby in the mother's womb
- Transient** – temporary

¹ Vanderpluym, MD, Juliana; Vajsar, Jiri; Dominique Jacob, MD, Francois; Mah, MD, Jean K.; Grenier, MD, Danielle; Kolski, MD, Hanna; Clinical Characteristics of Pediatric Myasthenia: A Surveillance Study, July 2013, Pediatrics, ISSN: print, 0031-4005; online: 1098-4275, American Academy of Pediatrics

Congenital Myasthenia, listed on the MGFA website at:
<http://myasthenia.org/LivingwithMG/InformationalMaterials.aspx>.

3. Juvenile MG (JMG) is an autoimmune disorder in which antibodies interfere with nerve to muscle communication causing muscle weakness. Symptoms of weakness develop after exertion so tend to become more pronounced as the day progresses. The variability in symptoms between early and late in the day and the usual slow buildup of the weakness can lead to the diagnosis of JMG only being made after weeks or months of symptoms. On occasion, the symptoms can develop rapidly. Drooping eyelids and/or incomplete movement of the eyes (causing crossing of the eyes and double vision) are frequently the earliest symptoms. However, any skeletal muscle can be involved so other symptoms include problems chewing, swallowing, and speaking clearly, shortness of breath or limb muscle weakness.

How can I know that my child has MG?

MG can be diagnosed in several ways. When the clinical history and neurologic examination suggest the possibility of MG, the following tests can be used to confirm a diagnosis:

- **Blood test**—this test looks for antibodies which interfere with the nerve/muscle communication process to receptors on the muscle membrane: acetylcholine receptors (AChR) and muscle specific kinase receptors (MuSK).
- **Drug testing:** Anticholinesterase inhibitors (edrophonium chloride given via IV use, neostigmine for injection or pyridostigmine for orally) can be used to prolong the activity of the body's natural neuromuscular transmitter, acetylcholine. An individual is then observed to see if the weakness temporarily reverses. These tests are done under careful medical supervision.
- **Electromyography/nerve conduction studies (EMG/NCS) test** the electrical connection between motor nerves and the muscles they supply.
- **Genetic tests** if CMG is suspected.

Multiple tests may be needed to confirm the diagnosis.

Is there a cure for MG?

Unfortunately, there's no cure for MG. However, treatment and care has improved markedly over the past 50 years. Your child has good prospects for a full life with proper treatment and care.

What kinds of treatments are there for my child's MG?

A treatment plan for your child will depend on his or her health, medical history, severity of the MG symptoms, and response to medications and therapies. In addition, the anticipated course of the condition and your involvement as a parent should be part of the process.

Medications & Therapies

Your child's doctor will work with you and your child to find the right medication or combination of medications. This therapeutic regimen will lessen the symptoms of MG and allow your child to be themselves. It may take some time to find that optimal approach. And, the best approach may change over time. Monitor how well medications are working with your child and your child's doctor. You may wish to keep a diary or chart noting times and effects. For instance, not only when medication was given but also when it seemed to take effect; when its help began to wane; side effects and any other significant details. In talking with your child's doctor, your input will be important.

- **Symptomatic Treatment:** Assuring adequate sleep and planning each day to allow for rest periods between more vigorous levels of activity is an important factor in minimizing symptoms of MG. Optimum nutrition and level of aerobic conditioning (being active enough to be in "shape") also help individuals with myasthenia function at their best.
- **Pyridostigmine** is a first line therapy for juvenile MG. It provides clinical improvement by prolonging the activity of the body's natural neuromuscular transmitter, acetylcholine. This oral medication tends to be well tolerated. However, it can have side effects such as abdominal cramps, diarrhea and, in very rare instances, worsening weakness. Typical dosing is 0.5 to 1 mg per kilogram of weight per dose for every 4 to 6 hours while awake with a maximum of 60 mg per dose. The maximum total dose is 7 mg per kilogram of weight per day. The absolute maximum of 300 mg per day is for older children and adults.² [One kilogram = 2.2 pounds.]
- **Immune-modulating Treatment: Corticosteroids** modulate the immune system and may reduce symptoms for some patients. There are concerns about use of high dose, daily corticosteroids in children. They can lead to side-effects such as weight gain, slowing growth, bone loss, facial changes, mood changes, high blood sugar, high blood pressure, and susceptibility to infection.³
- **Other medications that suppress or moderate the immune system** can be used to avoid long-term use of corticosteroids. The medication which has been used most extensively is azathioprine. Other agents include tacrolimus, cyclosporine, cyclophosphamide and mycophenolate. There are no clinical trials published regarding the use of these medications for treatment of children with MG. Careful monitoring of children on these medications is required as side effects include liver enzyme elevation, nausea, abdominal pain and bone marrow suppression. These medications can take several months to produce clinical improvement.
- **Intravenous Immunoglobulin (IVIg)** IV infusions of pooled human antibodies (IVIg) are effective at decreasing the clinical symptoms in many individuals with MG. The clinical improvement is frequently noted within several days to a week and lasts for variable intervals (generally not longer than 4-6 weeks). There is no consensus on the best dosing schedule for using IVIg to treat children with MG. Possible side effects are headache and nausea or less likely aseptic meningitis or rarely renal failure. IVIg therapy is expensive and usually requires preauthorization with third party payers.

² Ibid.

³ Liew, Wendy KM; Kang, Peter B. [Update on Juvenile Myasthenia Gravis](#), Current Opinion in Pediatrics, December 2013.

- **A Thymectomy** is surgical removal of the thymus gland. A thymectomy with or without an existing thymoma (tumor, usually benign) has been felt to improve clinical status in adults with MG. Early removal of the thymus in a child with generalized, antibody positive juvenile MG (i.e. within two years from diagnosis) has been associated with better long-term outcomes. The clinical benefit of thymectomy appears over time (long term). Therefore, thymectomy surgery should not be done on an urgent basis in a clinically ill or weak child as the risks of anesthesia and surgery outweigh the long term benefit.

For more information on these medications and therapeutics, see the MGFA's brochures on each at <http://myasthenia.org/LivingwithMG/InformationalMaterials.aspx>.

What can my child and I expect with MG?

Because MG is an uncommon disease many doctors have never seen a case before. Even those who have some experience with MG may only treat a few MG patients. Ideally, you may find it helpful to find a physician, usually a neurologist, who has significant experience with MG. This specialist is most likely found at a major medical center. But if that isn't possible for you, you may want to work with your physician to learn more about MG. MGFA can offer important resources to the physician open to learning more about MG. Visit www.myasthenia.org/HealthProfessionals/EducationalMaterials where you and your doctor can find the MGFA book, *Myasthenia Gravis: A Manual for the Health Care Provider* as a PDF. This manual is also available as an iBook - *Myasthenia Gravis: A Manual for the Health Care Provider* - #iBooks, <https://itun.es/ca/04G68.l>.

It can only be viewed on an Apple device. You can also reach out to the MGFA at 1-800-541-5454 to locate a neurologist who is part of our Medical/Scientific Advisory Board and who has strong experience with MG. If we do not have a physician in your locality, you can also seek a physician through the American Academy of Neurology's website at <http://patients.aan.com/findaneurologist/>.

- As you might imagine or have already experienced, MG is frequently undiagnosed. That means you may have already taken a long journey to get to this point. Once you have your child's diagnosis, you will still have many challenges. But you can get through them. Every parent and child has his or her own way of coping. Find those things that help you and your child manage stress. Focus on understanding MG and its treatment, along with finding ways to manage its impact and maintain as much normalcy in your child's and family's life as possible.
- Talk with your child about their illness in an age appropriate way. Don't deny or hide the problem (which may cause more worry for your child). Stress that he or she can cope with this challenge and that there is help from family, friends, health professionals and others.
- Share with others. Let other people in your life and your child's life know about his MG and its impact. But share this information in a matter of fact manner. Help others understand that although they need to be aware of your child's MG, you, your child and your family are maintaining as normal a lifestyle as possible. Ease any fears or misunderstandings others may have about your child's MG. For instance, can your child join a sleepover, a party, game, outing, etc.? How can another adult address any challenges?

What are the prospects for my child?

With proper treatment and care, the child with MG can look forward to a long and fulfilling life. MGFA includes volunteers who have MG who are nurses, support group leaders, communications executives, engineers and educational event planners. Some have grown up with MG.

Keys are:

- Work with your child’s doctors to find a therapeutic regimen that works for your child. That includes finding a medication schedule that works with your child’s life.
- Build strong relationships with your child’s teachers, coaches, school nurse and other key adults in his or her life. Help them understand your child’s health challenges and how they can assist in managing his or her challenges. See MGFA’s Fact Sheet, *A Parent’s Guide: Steering your Child with MG to Success*.
<http://www.myasthenia.org/LinkClick.aspx?fileticket=kdeE9EQOKMc%3d&tabid=339>
- Maintain a healthy lifestyle that includes exercise, nutritious food, positive thinking patterns, problem solving and relaxation, rewarding activities, love and support.
- Visit the MGFA website to learn more about MG and to find other resources for parents of children with MG. Here are a few:
 - Your Child and Exercise
<http://www.myasthenia.org/LinkClick.aspx?fileticket=-VVS9MCpDuQ%3d&tabid=339>
 - A Parent’s Guide—Steering your Child with MG to Success --
<http://www.myasthenia.org/LinkClick.aspx?fileticket=kdeE9EQOKMc%3d&tabid=339>
 - MGFA’s Literature on MG, medications, emergencies and related issues.
<http://myasthenia.org/LivingwithMG/InformationalMaterials.aspx>
 - See Patient Stories at <http://myasthenia.org/CommunitySupport/PatientStories.aspx>
 - Drugs to Avoid at <http://myasthenia.org/LivingwithMG/DrugstoAvoid.aspx>
- Get more information and ideas from other expert sources such as these:
 - **MG Parents** Facebook page--to reach this group of parents of MG children go to Facebook and type in MG Parents in the “Find Friends” search box – if you are a parent ask to join the group.
 - American Psychological Association www.apa.org/helpcenter/chronic-illness-child.aspx
 - The American Academy of Pediatrics at HealthyChildren.org:
<https://www.healthychildren.org/English/health-issues/conditions/chronic/Pages/Coping-With-Chronic-Illness.aspx>
 - Many health organizations are likely to have guidance on coping with a child’s chronic illness. The University of Michigan’s site has a very helpful article with links to many other resources. Go to: <http://www.med.umich.edu/yourchild/topics/chronic.htm>
 - Other sites with helpful resources include: www.kidshealth.org; Bandaides & Blackboards: <http://www.lehman.cuny.edu/faculty/jfleitas/bandaides/> a site for and from youngsters with chronic illness; Women’s and Children’s Health Network/Parenting and Child Health: <http://www.cyh.com/healthTopics/HealthTopicDetails.aspx?p=114&np=304&id=2578>.

Managing Myasthenia Gravis

Maintain a Positive Outlook
Track Patterns
Work around MG
Safety



At this time there's no cure for MG. Although each of us finds our own path in coping with MG – here are some thoughts about how to manage this chronic illness.

Maintain a Positive Outlook

First, work to maintain a positive outlook despite the stress MG is causing for you, your child and your family. Although that may be hard, it will help you cope and maintain a loving and encouraging environment for your child and family. By remaining upbeat, you'll also reduce stress, thereby helping to manage MG which can be reactive to stress like many illnesses are.

Track Patterns

Look carefully at your child's energy and strength/weakness patterns. MG is known as a snowflake disease because each person experiences it somewhat differently. You may find it helpful to track his or her patterns.

- You can use **myMG**, an app that allows a patient to record energy levels, quality of life and symptoms. The app can be used on a phone or on the computer. You can generate summary reports over time to share with your child's doctor. Go to <http://mymg.myasthenia.org/home> to learn more.
- By tracking your child's patterns, you may find ways to better manage their MG. For instance, make a plan for rest times throughout the day, or avoid significant exertion at certain times of day. Or, know the kinds of stress that may propel your child toward an exacerbation of their symptoms.
- Armed with this knowledge you can better work with other adults in your child's life such as teachers, school nurses, coaches, family, friends and neighbors. To learn more read MGFA's brochure & webpage, *A Parent's Guide -- Steering Your Child with MG to School Success*.
<http://www.myasthenia.org/LinkClick.aspx?fileticket=kdeE9EQOKMc%3d&tabid=339>

Work around MG

Clearly this is much easier said than done. MG is a chronic disease, which currently has no cure – that means you and your child will have to find peace with this challenge and learn to manage any limitations it brings.

- Find the images and ideas that help you cope.
- Create a place of calm where you can retreat when needed to recharge. This place can simply be in your mind – a beach where the surf roars, the sun shines, birds call or any other imaginary image that helps you relax.
- Consider Yoga, meditation, running or watching funny movies.
- Help your child find ways to beat stress too.

Safety

An important element in the care of those with MG is to know what Drugs to Avoid and how to handle Emergencies. Everyone with MG and their caregivers should be aware of this important information.

Many common medications can have serious adverse effects on people with MG. Please see Drugs to Avoid at <http://myasthenia.org/LivingwithMG/DrugstoAvoid.aspx> print it out for your future reference and share it with all medical personnel and other adults caring for your child such as a school nurse.

Order the following free literature from MGFA: Emergency Management 1 and Emergency Management II – You'll find these important brochures at <http://myasthenia.org/LivingwithMG/InformationalMaterials.aspx>.

- **Emergency Management 1** is directed to medical personnel including Emergency Management Services (EMS) or Emergency Medical Technician (EMT). You may want to share this brochure with these professionals in your community. Find them at your local hospital(s) and through the local EMT services that many communities have affiliated with the fire department.
- **Emergency Management 2** is directed to the MG patient or MG parent.

In addition, you can print out or order by mail Emergency Alert Card 1 and Card 2.

<http://myasthenia.org/LivingwithMG/InformationalMaterials.aspx>

- **Emergency Alert Card 1** includes key facts for First Responder Management.
- **Emergency Alert Card 2** lists Drugs to be Avoided or Used with Caution.

These cards should be kept together and placed in an easy to find place. A child's backpack and wallet may be appropriate places to have a set. Another set at home in an obvious place such as the refrigerator door or a bulletin board is also a good idea.

In Closing

You, your child and your family are facing a significant challenge with MG, but with work you can manage it. We urge you to become educated, to take a positive approach and to seek out help from credible sources.