Care for Children with Myasthenia Gravis

Myasthenia gravis is an autoimmune disease that affects the neuromuscular junction, causing people to feel weak and get tired quickly. It is an uncommon disease, with a prevalence of about 14 cases per 100,000, and juvenile cases account for about 10% to 15% of the diagnosed cases in North America. We asked Nancy Kuntz, MD, Director of the MDA Care Center at Ann & Robert H. Lurie Children's Hospital of Chicago, about caring for kids with this disease.

Recognizing MG

MG may begin with drooping eyelids or weakness in the face and throat muscles. For some, the symptoms are limited to weakness around the eyes, called ocular MG. For others, weakness can progress to muscles throughout the face, neck, and limbs, called generalized MG.

"In children, there is a higher frequency of the disorder beginning with weakness around the eye muscles, causing double vision or drooping eyelids," Dr. Kuntz says.

MG is a condition that pediatricians may suspect from a wellness visit by observing muscle weakness and asking the right questions. "When a given amount of exercise makes certain muscle groups weak, it's a sign of MG," she says. "A common example may be a child reading out loud, and by the end of the session, they go from having a normal voice to slurring words. If they don't talk for an hour, their voice goes back to normal. Another example may be if they eat a piece of sticky candy or tough meat and their jaw gets so tired that they have trouble continuing to chew."

Dr. Kuntz emphasizes the importance of getting an accurate diagnosis before treating MG. "Other disorders can be confused for myasthenia gravis," she says. "There are also children who have congenital forms of MG. You want to know whether a gene mutation is creating the abnormal transmission from muscle to nerve because that is treated differently than an autoimmune problem causing MG."

Managing MG in kids

For kids with MG, paying attention to how much activity they have in a day and reeling it in if it's too much is an important part of managing their symptoms.

"Children often can respond to systematic treatment like rest and change in activity and a cholinesterase inhibitor," Dr. Kuntz says.

Cholinesterase inhibitors improve the transmission from the nerve to the muscle across the neuromuscular junction and can relieve symptoms within minutes. The one most commonly used is pyridostigmine bromide (Mestinon).

In some mild cases, restricted only to eye muscles, Mestinon is the only treatment needed. "But most of the time, in particular in the involvement of muscles beyond the eye, they need some kind of immunosuppressive therapy," Dr. Kuntz says.

Corticosteroids, such as prednisone, are immunosuppressive and effective, but they can come with negative side effects, including stunted growth, osteoporosis, and mood disturbances. Corticosteroids were the mainstay of MG treatment until non-steroid immunosuppressants, such as azathioprine (Imuran), were developed.

Children with generalized MG can have critical weakness affecting swallowing and breathing. In these cases, they begin treatment with immunomodulating therapy.

Those at risk of respiratory failure or aspiration due to weak breathing and swallowing muscles may undergo plasmapheresis to remove antibodies from the blood. Patients may also

receive intravenous immunoglobulin (IVIG) infusions to help decrease the immune system's attack on the nervous system. Sometimes, chronic intermittent IVIG transfusions are given as maintenance therapy. According to Dr. Kuntz, chronic IVIG infusions are used more frequently in children than in adults because children are more susceptible to side effects from long-term immunosuppression with oral agents such as corticosteroids and mycophenolate.

There are also clinical trials pending in children and adolescents using neonatal Fc receptor (FcRN) inhibitors and complement inhibitors. The US Food and Drug Administration recently approved these agents for use in adults and are promising for use in children if the safety profile looks positive.

A watchful attitude

It's vital that children are under good surveillance by a care team to understand what can cause an exacerbation of MG symptoms. A myasthenia crisis, the most serious complication of MG, is an episode of extreme muscle weakness, particularly of the diaphragm and chest muscles that support breathing.

"Education is critical for everyone. One of the first things to do is to educate the family and primary care doctors," Dr. Kuntz says. "For example, certain kinds of anesthesia and medications for unrelated disorders can impact the neuromuscular transmission and can make a person go into a myasthenia crisis." Dr. Kuntz points out that Milk of Magnesia and antibiotics like azithromycin are among the common medicines that can adversely affect MG.

The care team can also help children navigate the challenges of living with MG. In her practice, Dr. Kuntz has seen that helping these young patients learn to manage the disease can make a big difference.

"Children with MG just want to be like everyone else," she says. "Often, with medical monitoring and a little adjustment in their schedule, they can do the things they want to do."

Myasthenia gravis resources:

- Share MDA's Myasthenia Gravis Fact Sheet with patients and families.
- Visit the Myasthenia Gravis Foundation of America Professionals webpage for recommended protocols, the latest research grants, and other useful information.