

CONQUER

News for our community

Complement inhibition in MG: a novel treatment approach



By Rabia Malik, MD
Rush University Medical Center

October 2017 was an exciting month for the myasthenia gravis (MG) community. Patients and physicians alike were heartened to hear that after more than 60 years the FDA had approved a new treatment for MG called eculizumab (brand name Soliris®). This medication offers a novel approach to treating MG by a process called “complement inhibition”. To understand how this medication works, it is important to understand what we know about the disease process in MG.

Normally, when a wave of electricity reaches the nerve terminal, packets of chemicals called acetylcholine (ACh) are released into the space between the nerve ending and the muscle fiber.

(continued on p. 4)

Join Us!

TRIVIA NIGHT

Calling all Einsteins! Join us for competitive trivia and a chicken/pasta dinner on Saturday, January 26, 2019, at the Elmhurst American Legion Post, 310 W. Butterfield Road, Elmhurst, Illinois. Sign up your team of six for \$210 per table. Space is limited, so call 800-888-6208 soon to register your team.

Details inside!



Happy Holidays!

About MG

Myasthenia gravis (MG) can strike anyone at any age. MG is a highly misdiagnosed and undiagnosed autoimmune disease in which communication between nerve and muscle is impaired, causing weakness. Its primary symptoms are erratic, vary in severity and occur in any combination such as: droopy eye lid(s); double or blurred vision; weak arms, hands, neck, face, or legs; difficulty chewing, smiling, swallowing, talking; undue fatigue, difficult breathing or shallow respiration; or sense of balance difficulty. MG can masquerade as overwork, under-rest, anemia, vitamin deficiency, disease of the involved organ(s), or even emotional disturbances. Its cause is unknown; there is no cure.

Conquer Myasthenia Gravis was formed October 29, 1972, by a local group of caring individuals who wanted to help patients achieve the best possible quality of life, while living with and managing their MG. We are a volunteer-led organization.

OUR MISSION: To facilitate the timely diagnosis and optimal care of individuals affected by myasthenia gravis and to improve their lives through programs of patient services, public awareness, medical research, professional education, advocacy, and patient care.

Conquer Myasthenia Gravis

275 N. York Street, Suite 401
Elmhurst, IL 60126-2752
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www.myastheniagravis.org

FROM YOUR EXECUTIVE DIRECTOR

Countdown to Cold



What's on your getting-ready-for-winter list? Mine includes a flu shot and unearthing the car window scraper. It's also time to practice fall prevention (salt for the sidewalk, keeping the driveway clear). Recovering from a broken bone can be tough when you're weak with MG.

American Bone Health adds this: Wear shoes with good traction. Dress warm so your muscles are relaxed. Take care when getting out of a car. And (most challenging for me): Slow down. Allow yourself enough time to get somewhere without rushing.

We wish you a happy – and unrushed – holiday season!

Joan Wincentzen, Executive Director

Conquer MG Patient Assistance Program

This Conquer MG program will help cover the cost of medical bills, prescription drugs and durable medical equipment up to \$1,000 per person per year. We've exceeded budget this year, paying over \$17,000. You may be eligible if you are an MG patient, reside in Illinois, Indiana, or Wisconsin, and have some kind of financial hardship. Medical costs don't have to be MG-related. Contact the MG office (800-888-6208 or info@myastheniagravis.org) for the application, or find it on our website.



Contact Update Form (PLEASE PRINT)

Mail to: Conquer MG, 275 N. York Street, Suite 401, Elmhurst, IL 60126

Name _____

Address _____

City _____ State _____ Zip _____

Email _____

- ☐ Please note the above change in my contact information.
- ☐ Please remove my name from your mailing list.
- ☐ Please send **Conquer** via e-mail instead.

Events & Activities

► Better Breathing through Technology



On November 4, 45 myasthenia gravis patients and friends attended Conquer MG's Fall Patient Seminar at Advocate Lutheran General Hospital in Park Ridge, Illinois. Sally O'Meara, RN, carefully explained the features and mask options for assistive breathing devices such as Bi-PAPs and home ventilators, and which work best for MG. Some attendees were surprised to learn C-PAP machines (for sleep apnea) are not designed to help with the breathing weakness experienced by myasthenia gravis patients.

Thank you to our speakers, Sally O'Meara and Sara Hasemeyer. See their presentations on our YouTube channel at <https://tinyurl.com/ycp58uwb>.

Calendar Club Winners

September – Betty Anderson, Carla Burnett, Dorothy Mykisen, Gail Olley

October – Deanne Scanlan, Joel Schohn, Anonymous (2)

November – George Dulzo, Miriam Moroney, Julia Rogers, Ted Wienski

► New MG Meet-Up in Oak Park

A new MG support group is starting this February. Led by Victor Yipp, the group will meet once a quarter at the West Suburban Medical Center in Oak Park, Illinois. See page 9 for details.

► YOU are Needed!

Chicago-area folks, we sincerely need thoughtful and/or energetic individuals who can do a two-year stint on the Conquer MG board. Board meets in Elmhurst 4-6 times a year. Call 800-888-6208 for details.



► Year 7: 5K Fun Run/Walk



Get ready to Conquer MG like Vikings! In 2019, our 7th annual fundraiser is set for Sunday, June 2. The new location is Danada Forest Preserve in Wheaton, Illinois. Run or stroll the 5K through the woods, with or without your Nordic beards and braids. Watch your inbox for details! Planners needed!

► MG Awareness at the Zoo



Thank you, Apex Management & Special Events, for making Myasthenia Gravis part of Brookfield Zoo's Holiday Magic 2018. Check out tree #162; it's the one with MG snowflakes!



► Try Your Luck in 2019!

Join the fun and help us get vital MG information in the hands of over 1,700 people in the coming year. Donations to our annual Calendar Club fundraiser are \$12 per ticket or \$50 for five tickets. Each month we pick four lucky \$25 winners. And at year end, there's a Grand Prize (\$500!) and Runner-Up (\$100!). Call the office and we'll fill out your tickets and add them to the drawing box. This could be your year!



Complement inhibition in MG: a novel treatment approach

By Rabia Malik, MD

Department of Neurological Sciences, Rush University Medical Center

(Continued from p. 1)

ACh binds to a receptor on the surface of the muscle fiber and allows activation of a chain of events that leads to muscle contraction (Figure 1.).

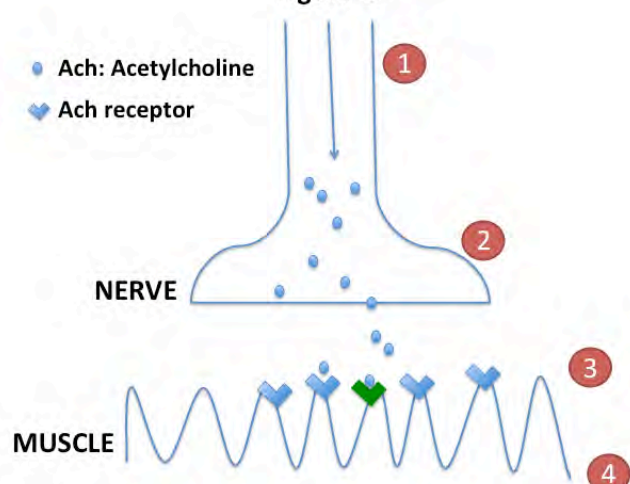
In a large majority of patients with MG, proteins called antibodies against the acetylcholine receptors cause dysfunction in this pathway and can lead to failed transmission at the nerve-muscle junction (Figure 2.). This breakdown of communication between the nerve and muscle leads to the painless muscle weakness of MG.

Eye muscles are commonly involved, leading to droopy eyelids or double vision. However, the disease can affect other areas of the body and may cause speech and swallowing difficulty, as well as weakness of the arms and legs. When the weakness is limited to the eyes, the disease is referred to as “ocular MG” but if it involves other muscles, then it is referred to as “generalized MG”.

There are several different ways that these “bad” antibodies may cause problems. Aside from blocking the receptors one of the presumed mechanisms is that it leads to the **long-standing activation of the complement system**. The complement system is part of the immune system that amongst other things plays a role in defending our bodies against certain bacterial infections. The uncontrolled activation of this system by the disease-causing antibodies **damages the surface of the muscle** and can further prevent normal binding of ACh.

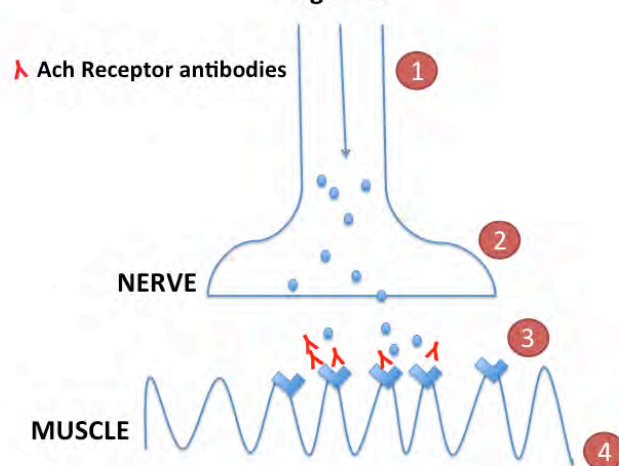
Soliris® has been approved for patients with acetylcholine receptor antibody positive generalized MG. This treatment, which **acts by inhibiting the complement system**, showed benefit for those with

Figure 1.



1. Electrical impulse arrives at the nerve ending
2. ACh moves out of the nerve toward the muscle
3. ACh binds to the receptor on the muscle
4. Muscle contraction occurs

Figure 2.



1. Electrical impulse arrives at the nerve ending
2. ACh moves out of the nerve toward the muscle
3. ACh cannot bind well to the receptor
4. Muscle contraction does not occur



poorly controlled MG. Although most MG patients respond well to existing medications that suppress or modulate the immune system, called immunosuppressants, 10-15% remain quite symptomatic and are felt to be treatment-resistant.

There are a number of things to consider when discussing Soliris with your neurologist. It is a medication that is given through the vein. The treatment is fairly intensive and requires an infusion weekly for the first four doses followed by treatments every 2 weeks. Each infusion, on average, is 35 minutes long.

As frequent and long-term infusions are needed, **catheters like portacaths (port) for access** are preferred. These are placed, under local anesthesia, just under the skin in the right upper chest area. They are easily felt as a disc-shaped structure roughly the size of a quarter. The port is attached to a catheter that is placed into a large vein near the heart. This option reduces the pain of multiple needle sticks to the veins in the arms and allows a quicker access. These ports can stay in use for years. The risks of ports include potential for infection or blockage. In rare cases it may dislodge.

Ports are preferred to **peripherally-inserted central catheters (PICC line)**. PICC lines are placed in a vein of the arm and the catheter is guided to a large vein near the heart. The PICC line hangs from your arm and is held in place by a dressing. Using PICC lines reduces the discomfort and damage to the veins from multiple needle sticks, but unlike ports, they are uncomfortable given that a part of the catheter is outside the skin. This limits arm movements and greater care is required to reduce the potential for infections. The PICC lines cannot stay in more than a few months.

The cost for Soliris is substantial. Because it needs to be administered intravenously, there also are associated costs for the infusions such as nursing care.

Currently, **a self-injected or subcutaneous form** of complement inhibitor for generalized MG **is being tested**. Rush University Medical Center in Chicago is a participating site for this research study. Enrollment for this study was recently completed and results for the first part of the study are expected in early next year. If effective, this would bring down significantly the cost of treatment.

There is **risk for serious or life-threatening meningitis infection** with use of Soliris. Patients **need to get immunized** at least two weeks prior to starting treatments. Vaccinations reduce but do not eliminate the risk of infection.

There is **no data** to support its use in **MG patients who do not have AChR antibodies**, and it is **not known how long patients need to be on treatment** or if there is an end point. Since only one of the pathways for disease propagation is being combated by this medication, it is felt that this treatment on its own is insufficient and should be given along with another immunosuppressant.

In spite of the limitations, **the approval of Soliris brings a sense of optimism for both MG patients and physicians who treat this disease**. This treatment adds to the arsenal of medications that are being used to combat this illness and also reflects an increasing interest in research and drug development that we hope will lead to other therapeutic options in the near future.

ANOTHER COMPLIMENT INHIBITOR MEDICATION FOR MG

On December 10, Ra Pharmaceuticals, Inc. announced positive top--line results from the Company's Phase 2 clinical trial evaluating zilucoplan for the treatment of generalized MG. Patients showed significant improvement after 12 weeks with the subcutaneous therapy.

Aye-Tee Monaco

Rituximab and MuSK MG A Personal Narrative

Aye-Tee's myasthenia is caused by antibodies to a protein called muscle specific tyrosine kinase (MuSK).

The views expressed here are the author's and not necessarily those of Conquer MG



My daughter got married in October last year, and I could see, smile, walk her down the aisle with my husband, dance the hora, enjoy the good food, and laugh with our family and friends...I felt absolutely fantastic! Anti-MuSK myasthenia gravis (MuSK MG) was a distant memory that day and it has become a distant memory since my last rituximab treatment. Rituximab infusions have given back my health/life!

I was diagnosed in April 2011 with MuSK MG. From the start, my kind and brilliant neurologist, Dr. Jeffrey Allen at Northwestern Memorial, knew that the protocol treatment like azathioprine or CellCept® with corticosteroids would not be effective in managing MuSK MG. He appealed relentlessly for the rituximab treatment. When Dr. Allen moved I found an equally caring and brilliant doctor closer to my permanent home in

Dallas, Dr. Sharon Nations at UT Southwestern. She too appealed to the insurance company on my behalf, but was similarly turned down despite her success with rituximab and MuSK MG. My prednisone and azathioprine doses varied with my symptoms, but were never fully effective. I suffered many prednisone side effects: severe osteoporosis, cataracts, glaucoma, a swollen moon face, and becoming pre-diabetic.

In 2015 my liver showed stress from prolonged use of azathioprine. Despite an increased dose of prednisone, I could not hold up my head, bend, or go up the stairs; I experienced double vision, droopy eyelids, slurred speech, and difficulty in swallowing and breathing. We appealed once again to the insurance company with medical studies from my neurologist, and supporting letters

this time from my endocrinologist, ophthalmologist, and primary care physician testifying to the damage that prednisone had done to my body. This appeal failed, too.

Because my husband's company at that time was privately insured, we appealed to the company to make an exception for the treatment. I had my first round of rituximab in September of 2015. After two weeks, I experienced remarkable improvement. By my second infusion, I was symptom free.

Rituximab is resoundingly effective in battling MuSK MG, not just for me but for many (as evidenced from the many studies). I am happily in remission, taking no medication for MuSK MG. I urge insurance companies to recognize its value, not only for the good of MuSK MG sufferers, but also for the cost savings they could realize by avoiding treatments related to chronic use of steroids.



Saturday, January 26, 2018
Elmhurst American Legion
310 W. Butterfield Road, Elmhurst, Illinois
Doors open at 6 p.m. Trivia starts at 7 p.m.
\$210 per table/team of 6

Includes delicious chicken/pasta/soda dinner
Cash bar for beer/wine * 50/50 Raffle
Cash prize for winning team!
Space is limited, so call 800-888-6208
to register without delay!

Support Groups



Our groups offer:

- Information about myasthenia gravis and ways to cope with its symptoms
- Good listeners who care about your concerns
- Assurance, comfort, and friendship

AREA	2019 DATES	TIME	LOCATION
Chicago - North Suburban	Saturday, Jan 5 Tuesday, Apr 9 Tuesday, Aug 13 Friday, Oct 25 Tuesday, Dec 10	9:30 AM - 11:00 AM	Glenbrook Hospital, 2100 Pfingsten Road, Glenview, IL. 2nd floor conference rooms A1-A2 Use main entrance and South (Blue) parking. December meeting is a holiday party.
	Saturday, June 8	11:30 AM - 1:00 PM	Potluck indoor picnic
Chicago - South Suburban	Sunday, Feb 10 Other dates TBD	1:30 PM - 3:30 PM	Metro South Hospital, private dining room in cafeteria, 12935 S. Gregory, Blue Island, IL. Use ER entrance parking garage, off Union St.
NEW! Chicago - Oak Park	Sundays February 10 May 12 August 11 November 10	1:00 PM - 2:30 PM	West Suburban Medical Center, 3 Erie Court, Oak Park IL, lower level conference room A-B. Use main entrance or enter via parking garage lower level. Near CTA Green Line, Austin Station
Chicago - West Suburban	Sundays February 10 May 19	1:00 PM - 2:30 PM	Central DuPage Hospital, Conference room 3, Women & Children's Pavilion, 25 N. Winfield Road, Winfield, IL. Use NW entrance, park in Visitor Lot 3 or use valet parking.
Rockford	Saturdays January 12 April 13 July 13 October 12	1:00 PM - 3:00 PM	St. Anthony Medical Center, St. Francis Room, 5666 E. State St., Rockford, IL. Use main entrance and adjacent parking.
Springfield	Sundays Jan 20, Mar 17, Apr 21, May 19, July 21, Aug 18, Sept 15, Oct 20, Nov 17	3:00 PM - 5:00 PM	Parkway Christian Church, 2700 Lindbergh Blvd. in Springfield, just east of the Parkway Point Shopping Mall. Use office entrance and adjacent parking.
Northwest Indiana	Saturdays Mar 23, May 25 July 27, Oct 26	10:00 AM - 12:00 PM	Schererville Public Library, ask at desk for meeting room, 1001 W. Lincoln Highway, Schererville, IN. Adjacent parking.
Quad Cities Iowa / Illinois	Saturday, Feb 9 Other dates TBD	10:00 AM - 12:00 PM	Davenport Public Library - Eastern Avenue Branch, Room A, 6000 Eastern Avenue, Davenport, IA. <i>Note: Location of future meetings may change.</i>

Immune Globulin (IG) Therapy

IG therapy can be used to treat rapidly worsening MG. It also is used with some myasthenia patients as a maintenance therapy. Immune globulin is a human blood product pooled from multiple donors who are carefully screened. By providing the body with normal antibodies from donated blood, IVIg treatments appear to temporarily modify the immune system. Source: Conquer MG website

In Appreciation

SPECIAL THANKS to everyone listed who supported our mission from August 1 through November 30, 2018. Your continuing financial support keeps our organization going! We also thank those who asked not to be listed but support Conquer MG work.

Monica and Edwin Aitken
Laura Austin
Vicky Benyr
Thomas Cawley
Charles River Laboratories
Claudia Cox
Richard A Culhane
Lisa Dahl
John Davenport
James Dee
Angelo Tasoulis
Anne Flanz
Matthew Gerald
Anna Goldstein
Fred and Sharon Grabenhofer
George and Jackie Green
Carolyn Hildreth
Mary Kay Hoffmann
Jane Horan
Mary Ippolito
Rachna Jain
Greg Jillson
Marci Einstein and John Treece
Dianne Kaufman
John Keating
David Kelly
Holly Kost
Michael and Linda Kyser
Russ and Julie Laney
John and Diane Leistman
Steve and Dorothy Litwitz
Linda Loland
Ross Major

John Manion
Toni Martinez
Gary Nelsen
Harry O'Leary
Beverly O'Sullivan
Carol Oswald
Michael Quane
Srinath Rao
Bob Ratzel
Edward Ravens
Darlene Renko
Ed Rickert
Connie Rowe
Sharon Ruddy
Lois and Arnold Samuels
Deanne Scanlan
Myrna Schiattareggia
Lonnie Smith
Rosemary Sprague
Alberta Steinhoff
Nancy Van Laten
Geraldine Volpe
Vinnie Volpe
Larry and Mary Jean Weller
Joey Whitt
Ted Wienski
Victor Yipp

In Honor of Laura Austin
Mary Reynolds

In Honor of Dr. Barend P. Lotz
Susana Guzman-Holland

In Honor of Connie Rowe
Elizabeth Matzinger

In Honor of Nancy Van Laten
Arlene Sangmeister

In Honor of Joan Wincentsen
Jim and Nancy Bulger

In honor of the men and women who serve our country, past and present
Carolyn Grebner

In Memory of Nicholas Albers
Edward and Carolyn Albers

In Memory of Fred Arnold
Loretta Fleming

In Memory of Rod Brown
Charlene and Andy Rismann

In Memory of Debra Clyden
David Clyden

In Memory of Luther Isaac Dickens
Andrew Arbury

In Memory of Michael J. Jamen
Ida Jamen and Family

In Memory of MaryAnn Konopka
John and Carol Myers

In Memory of Alexander Niemczura
Darlene Niemczura
In Memory of Lorraine Schohn
Joel Schohn

In Memory of Duane and Dorothy Seggerman
Virginia Seggerman
Donald Seggerman

In Memory of Rita Spector
Patti and Alan Friedman
David Paulson

In Memory of Joseph M. Vellano
Robert Bryla

Infusion Q & A

By Samantha Masmar, RN
Home infusion nurse, Kabafusion



Q: What is the difference between intravenous (IVIG) and subcutaneous (SCIG) immunoglobulin?




A: With intravenous immunoglobulin (IVIG), a nurse or other health care technician places a needle in the vein of a patient, and the patient receives an infusion of immunoglobulin straight into the blood stream. The infusion typically lasts 2-4 hours, and can take place in a hospital, outpatient facility, or (with medical supervision) in the patient's home. With this approach, large doses of immunoglobulin can be administered, and the patient usually only needs to be infused every 3-4 weeks. Side effects with IVIG usually are due to the rate of the infusion, or dehydration.

With subcutaneous immunoglobulin (SCIG), a patient typically is trained to give IG medication to himself at home. He does this by injecting a needle into the fatty tissues under the skin (for instance, in the abdomen, using the same technique as an insulin injection), where it enters the circulation slowly over a few days. Due to the slower absorption, side effects are less common. SCIG is usually given every week, lasting up to 2 hours and in smaller doses.

Conquer Myasthenia Gravis
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Upcoming Events

- JANUARY 26
Trivia Night
- JANUARY - DECEMBER
Support Group Meetings
- JUNE 2
Conquer MG 5K Fun Run/Walk
Save the date!

