



MG Communicator

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BiPAP Myths "Blown Away"

Submitted by: Sally O'Meara, R.N.
Nurse Educator at Oakland University

Myth: CPAP and BiPAP machines are the same

Actually: CPAP stands for Continuous Positive Airway Pressure. It is commonly used to treat obstructive sleep apnea. A CPAP machine uses air pressure blown through a tight-fitting face mask to prevent the airway from collapsing during sleep. BiPAP stands for Bi-level Positive Airway Pressure. BiPAP has two levels of pressure— high pressure to help inhale air into the lungs and low pressure to allow air to be exhaled from the lungs. BiPAP is used by patients with neuromuscular diseases because the weak respiratory muscles cannot exhale against the high CPAP air pressure.

Myth: BiPAP is only used in the hospital during MG crisis

Actually: BiPAP is a common intervention to help avoid intubation for MG patients who are in myasthenic crisis. BiPAP functions like a ventilator, but uses a tight-fitting mask on the face rather than a tube down into the trachea. BiPAP can be used by patients at home during the night to correct shallow breathing during sleep, and during the day to relieve shortness-of-breath. Home BiPAP machines are small, quiet, and easy to use.

Myth: A sleep study is required in order to get a BiPAP

Actually: A sleep study is required to diagnose obstructive sleep apnea and qualify the patient for a CPAP machine. Different criteria are used to determine that BiPAP machines are needed for patients with neuromuscular disease. Test results used to qualify MG patients for BiPAP therapy include:

- Forced Vital Capacity < 50% of expected value
- Maximum Inspiratory Pressure (MIP) < 60 cm H₂O.
- Arterial Blood Gas (ABG) shows carbon dioxide level > 45 mm Hg

Myth: MG patients must fail CPAP before getting a BiPAP

Actually: Patients with sleep apnea are required to start with a CPAP machine before insurance will pay for a BiPAP machine. Medical equipment companies are often unfamiliar with the qualifying criteria used for patients with neuromuscular diseases, and may sometimes tell patients that CPAP must be tried first. Patients should always check with their insurance companies about their individual policies and coverage requirements, but the test results shown above are generally-accepted guidelines. Additional information about qualifying guidelines can also be found at:

https://www.resmed.com/us/dam/documents/articles/1010293_RAD_Guidelines.pdf

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"Blessed" with MG

By Susan Southerton



Fred Hoonhorst

The Hoon Open is my favorite event! It is such a good time. I just had to talk with the man who started it all. I wanted to hear his story, so last month Fred Hoonhorst took time out of his morning to share.

Fred never set out to move mountains. Twenty-seven years ago, a friend offered the use of his golf course to hold the first golf outing for our MG foundation. With no experience, Fred and his family

dove in. You see, Fred's MG knocked him down for a time. Being a hard working father of six in his prime, working two jobs to provide for his young family. Not being able to talk or walk became a life changer. His struggle with physical limitations caused 'a great pause' in his life and he came to accept that he could not do it alone. Fred shared that his hope came from his faith. When he set his sights on his eternal home, things turned around! He accepted his limitations and had a new motto, "I've been blessed (with MG) and I want to be a blessing and be of value to others."

It took some time to get the medications in place which brought about remission of his MG symptoms. Once under control he had many years with a fulfilling career before retirement as owner of Hoonhorst Concrete. The blessing he brings is not

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Check This Out

Holiday Party—Monday, December 4, 2017 at the Plainfield Senior and Community Center 6:00 p.m.
5255 Grand River Dr. NE, Grand Rapids, MI 49525 This is not a support meeting. It is just a fun night with great food, good people, volunteers, friends and family, medical advisors and trustees. As always—door prizes and caroling. More information at www.mg-mi.org

MG-MI Annual Meeting and Conference is being planned for 2018 — watch for more information

28th Annual Hoon Open Outing—The 1st Saturday in August
August 4, 2018 in Rockford —8:00 a.m.

Eenhoorn Run/Walk for MG

The date is set— Saturday, June 23, 2018 — 10:00 a.m. start

Plan on a fun filled morning for all. The MG cars from the “Old Speckled Hen” MG Car Club will be joining us again. Little cars for the little know MG condition. Save the date and join us during June Awareness!

Place MG Snowflakes @ a local business in your area — it’s easy; email or call the office to find out how.

Consider Volunteering. Here are some needs:

Join the Walk Committee and help us plan the Eenhoorn Run/Walk for 2018.

Chair a local support meeting, of course we would assist you with this. It is a challenge for the office staff to host as many meetings across the state as we would like to. We would like to be in your areas more often :)

Assist in spreading the word — contact your local newspaper when a support meeting is in your area.



Design the 2018 Eenhoorn Run/Walk t-shirt.

You design it — best design will win a prize. Submit your design soon to myasthenia.info@gmail.com/ The winner will be selected by the Run/Walk Committee.
Want to also join the Committee and help in planning? Email or call the office.

Thank you!

Partners from the 2017 Hoon Open

LUNCHEON SPONSORS \$1000:



D.L. NEU & ASSOCIATES
INTEGRATED SYSTEMS



HOSPITALITY SPONSORS \$500:



TEE SPONSORS \$300:

Concrete Central
Grand Rapids Glass & Door, Inc
Grand Rapids Gravel
Hammersmith Equipment Co
Hoonhorst, Fred and Gloria
Macatawa Bank
The Shop Bodyshop
Van Laan Construction Supply

GREENS SPONSORS \$100:

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Shear Salon
State Farm —Ken Jipping
Twohey Maggini, PLC
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Weller Truck Parts
Yff & Scholma P.C.

Meet those who make it happen. The Hoon Open has given the foundation a leg to stand on year in and out. Each member of the Hoonhorst family pitches in—from Fred and Gloria, their children, grandchildren and friends. Darl brings a team each year (27 and counting), the outstanding BBQ Karl serves at the turn is made by Denise, and Dawn organizes the raffle. Kathy Hart, office manager of Hoonhorst Concrete, commits a great deal of time securing sponsors,

teams and prizes each year. This is a very fun and dedicated group that makes the golf outing run smoothly every year and did we mention already IT IS A GREAT DEAL OF FUN? You will not find many opportunities on that morning to volunteer but you can bring a team and help get sponsors or prizes. In 2018, Fred and Gloria and their family along with Kathy Hart will be found on the first Saturday in August at the 28th Annual Hoon Open. Plan on joining us!



Concrete Central with Darl Hoonhorst



Kent Mudie and Fred



Dr. Amy and crew!



13th Hole Volunteers



Fred and Doug Vandenberg



The Hubers Tim and Mark with Fred



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Office Hours

11 a.m. to 3:00 p.m.

Monday through Friday
also by appointment

From Your Director

As we look back on the past year, I am very excited about the progress we have made raising awareness for myasthenia gravis and our continuing efforts to increase educational opportunities for patients, family members, and doctors. These programs and projects would not be possible without the help of many people. It has been a joy to meet and hear your stories; thank you for sharing.

First, I would like to thank our loyal donors, volunteers, Medical Advisory Board, trustees and corporate partners for their support and generosity. We could not do what we do raising awareness, providing educational materials, events and underwriting research grants without your donations of time, talent and treasure.

Our trustees do great work and have taken on additional duties recently. Their hard work and dedication allow us to further our mission by providing hope to the MG community statewide. Next time you call, e-mail or write please thank them for what they do for MG-MI and the MG community. I want to thank our Board of Trustees and Medical Advisory Board who are committed to the mission of MG-MI. Recently a long time board member, Susan Richards, retired from the board. I want to thank her for her service to the foundation and helping us grow into the organization we are today.

If you have not already returned a Patient Survey this year, please do so NOW. You can find it online at our website. Please consider supporting the foundation's Annual Appeal with a donation. Another way to support our programs is through your holiday buying at AmazonSmile or your local Spartan Nash 'Direct Your Dollars' register receipts.

MG-MI continues to improve the way we deliver our messages. This year we hosted a conference in Lansing on three topics with almost 100 people in attendance, making this the largest MG gathering in the region. If you missed the 2017 conference we hope you will be able to attend the 2018 conference that is being planned right now.

Another project we are working on will increase awareness for MG through videos for awareness and education. We hope to use these on our website and social media channels.

So, as we look toward the new year, we continue our commitment to work for you by raising awareness, providing the most up to date information through the 'MG Communicator', MG-MI emails, facebook, our website, other social media channels and fund research.

Thank you again for your support of MG-MI. Feel free to contact our office if you need any materials. You are our best advocates and we hope you continue to spread the word about our MG Foundation of MI.

The foundation relies on your donations to provide programs— PLEASE DON'T FORGET MG-MI WITH YOUR ANNUAL DONATION!

Sincerely, Sue Southerton

("Blessed"...continued)



Fred and Gloria

just to MG-MI, but to other ministries in his church and to those in Haiti as he gives back what he is able to. In 2002 Fred received the 'Volunteer of the Year' award from the National MGFA. Fred gives all the credit to his wife, Gloria, for her unending support and continued care. They celebrated 50 years of marriage this past year.

Side by side they walk with the unwelcomed but accepted MG condition.

Thank you Fred for sharing your time and talents. **Fred, you are a blessing to others who have also encountered MG.**



Facebook Reach

2,000+ likes
1,465 fans

Support Meetings

10 Locations
24 Meetings
262 Attending
6 Guest Speakers

MG Research and Clinical Trials

New Data Proves Long Term Use of Eculizumab Data from an open-label extension study involving the use of eculizumab was on full display at the American Association of Neuromuscular & Electrodiagnostic Medicine (AANEM) Annual Meeting in Phoenix, Arizona. The new data shows the drug to be safe and effective to treat patients with myasthenia gravis. Eculizumab is an antibody directed toward the complement system that has already been approved for paroxysmal nocturnal hemoglobinuria (PNH) and atypical hemolytic uremic syndrome (aHUS). The open label study (ECU-MG-302) was for patients who completed the 26-week, double-blind, placebo-controlled REGAIN study which tested the use of eculizumab in patients with anti-acetylcholine receptor refractory generalized myasthenia gravis. In the open label extension, patients received eculizumab (maintenance dose: 1200 mg every 2 weeks). The study found that patients who took eculizumab for 26 weeks in the REGAIN study and then continued to take the drug in the extension study (n=56), had stable scores on their myasthenia gravis activities of daily living (MG-ADL) questionnaire. Further, those switching from placebo to eculizumab (n=60) showed a statistically significant improvement in their scores within 1 week of treatment.

Argenx Announces Orphan Drug Designation for ARGX-113 for the Treatment of MG Argenx a clinical-stage biotechnology company developing a deep pipeline of differentiated antibody-based therapies for the treatment of severe autoimmune diseases and cancer, announced that the Office of Orphan Products Development of the U.S. Food and Drug Administration (FDA) has granted orphan status for the use of ARGX-113 for the treatment of myasthenia gravis. ARGX-113 has the potential to eliminate patient symptoms while minimizing common side effects seen with current treatments by reducing the pathogenic IgG levels. Orphan drug status is granted by FDA to a drug or biological product to treat a rare disease or condition. Orphan drug designation qualifies for various development incentives, including tax credits for qualified clinical testing, a waiver from FDA's application User Fee for marketing application, and a 7-year period of marketing exclusivity in the US. The granting of an orphan designation request does not alter the standard regulatory requirements and process for obtaining marketing approval. Safety and effectiveness of a drug must be established through adequate and well-controlled studies. To read the press release, go to MGFA's Website.

MGFA Webinar Answers Patient Questions

The MGFA hosted a patient webinar on Tuesday, Oct. 3, featuring MG doctors Jonathan Goldstein and Sami Khella. Dr. Goldstein and Dr. Khella responded to general questions submitted in advance by MG patients. The webinar was open to all patients and caregivers. A recording will be available on the MGFA web site.

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(BiPAP Myths ...continued)

Myth: BiPAP masks are huge and uncomfortable

Actually: BiPAP masks used by hospitals to treat inpatients are designed for economy rather than comfort. They are intended to be used by one patient for a short period of time and then discarded. In contrast, a wide variety of BiPAP masks are available for home use to accommodate patient preferences and sleep positions. For patients who dislike the full face masks, nasal masks cover only the patient's nose while nasal pillows fit snugly underneath the nostrils. BiPAP masks are designed to allow for unobstructed vision, facial hair, and eyeglasses while minimizing the risk of skin breakdown on the face.

Myth: BiPAP is not needed until breathing gets really bad

Actually: It's never too early to think about breathing needs. Subtle signs that indicate breathing may be impaired include:

- Morning or daytime headaches
- Difficulty waking up
- Shortness of breath when lying down
- Morning grogginess
- Unexplained sleepiness

MG patients who have weak respiratory muscles should speak with their neurologist about home BiPAP before breathing symptoms become severe. It is better to start BiPAP when pulmonary function tests are borderline, rather than to wait until signs of respiratory failure finally appear. ■



Help MG Just by Dining Out

Like to eat out? Then sign up, and a percentage of your purchase will be given back to MG. What is eScrip? It is a system that rewards customers loyalty by contributing a percentage of purchases made by participants. Your family is encouraged to register your existing credit/debit cards as supporters.

How does it work? Participating merchants contribute each time you make a purchase using your registered card(s). There are no receipts to collect, no vouchers or certificates to buy... and every purchase counts. How can I contribute?

- 1) sign-up online and register the credit card(s) you use when dining out
- 2) dine out and use your card—we'll get 2.5%
- 3) you will get an online survey from eScrip about your dining experience, fill it out and we'll get another 2.5% (That is 5% total).

Thousands of restaurants give back when you dine with them. A list is on the eScrip website. Cards accepted: VISA, MasterCard, American Express and Discover.

Check it out at <http://escrip.rewardsnetwork.com/>. Then sign up and invite your family and friends to sign up too.

Ask the Doctor

By Christopher Glisson, DO

"Potential difficulties with cold weather and its effects on MG: some tips for dealing with a frozen or non-functioning diaphragm due to cold weather exposure."

Over the past winter, we heard from two MG patients who reported difficulty with breathing when outside in cold weather. One patient described the sensation as, "my diaphragm froze" making her feel unable to exhale. Another reported that it was "a struggle to take a breath in." Interestingly, these patients both expressed that this had not occurred in the setting of a crisis or otherwise-severe weakness; it seemed instead to be an acute attack that was isolated to exposure to cold weather.

Based on our collective experience with many patients with MG, this phenomenon of worsening symptoms with exposure to extreme cold (or extreme heat, for that matter) is not uncommon. The process of "neuromuscular transmission" (by which nerves communicate with muscles to perform a specific task) has long been known to be negatively affected by extremes of temperature. In patients with MG, neuromuscular transmission is already impaired by virtue of the disease. Therefore, when temperatures are cold the communication between the nerves and muscles may be further impaired, resulting in transient worsening of symptoms.

Although it may seem based on the similarity of the questions asked above that this phenomenon is restricted to the breathing muscles, this transient weakness with cold can actually affect any of the muscles in the body. Many patients have noted leg, arm and hand weakness with exposure to extremes of temperature. Like so many other aspects of MG, symptoms (and the pattern and severity thereof) can vary to a great degree between patients, and even for the same patient at different times.

The National Institute on Aging (NIA) has published the following recommendations to assist those who are especially susceptible to the effects of cold weather. They are applicable to patients with MG, regardless of age, if it is noticed that symptoms worsen with exposure to the cold.

- Wear several layers of loose clothing when it is cold. The layers will trap warm air between them. Tight clothing can keep blood from flowing freely and lead to loss of body heat.
- To keep warm at home, wear long underwear under your clothes along with socks and slippers. Use a blanket or afghan to keep legs and shoulders warm and wear a hat or cap indoors.
- Make sure your home is warm enough. Set your thermostat to at least 68 to 70 degrees. Even mildly cool homes with temperatures from 60 to 65 degrees can trigger hypothermia in older people.
- Check with your doctor to see if any medications (prescription or over the counter) you are taking may increase your risk for hypothermia.

While these tips may be valuable for the winter ahead, it is important to recognize that heat can have the same effect on patients with MG. If you notice that your symptoms vary based on temperature, it is important to notify your neurologist and take steps to mitigate your exposure accordingly.

QUESTIONS?

If you have a question about MG, community resources or a treatment therapy, please mail or email it to us and we'll do our best to get you an answer.



MuSK-MG STUDY

Catalyst Pharmaceuticals Inc. hosts a webinar on a new clinical study for patients with-Myasthenia Gravis (MG). Catalyst Pharmaceuticals is presenting an informational webinar in which Dr. Gary Ingenito, Catalyst's Chief Medical Officer, reviews the clinical study requirements and answers questions about potential participation in it's upcoming clinical study evaluating amifampridine phosphate in patients with Myasthenia Gravis. Registration is open for interested persons. To learn more go to their website: <http://www.catalystpharma.com/>. The webinar will be recorded for archived listening.

Ways you can support the programs we bring to you:



**Lasting Good—
include MG-MI, in your
estate planning.**



Snowflakes Fundraiser
at a local business in
your area — it's easy
call the office to
find out how.

Please note that the views expressed in this newsletter are those of the individual author and do not reflect any official position of the Myasthenia Gravis Foundation of Michigan. Each person's situation is unique. If you have any medical questions please discuss them with your doctor, as they know your situation best.