

The End.

Everything that has a beginning has an end. Let's close the book on ALS. The End.

April 26, 2021

Edition 2

Always believe that something wonderful is about to happen.



The Jim Heller Fund Grants \$150,000 to ALS TDI for New Drug Lead

The C9orf72 gene mutation was previously discovered and is the most common **known** cause of ALS (and frontotemporal dementia). Recently, scientists at ALS TDI discovered that inhibition of a family of enzymes known as protein arginine methyltransferases (PRMTs) are key contributors to C9orf72-mutation ALS. Specifically, ALS TDI scientists discovered that by inhibiting certain PRMTs, cells that typically die from exposure to C9orf72 mutation-associated proteins can be completely protected. They all survive! This is an important finding because it may reveal a therapeutic option for people with C9orf72 that may be more effective than the still unproven treatments currently in development which require repeated lumbar punctures.

However, for ALS TDI to advance this finding, the team must assess the treatment approach in animal models. Work in mouse models that are well-suited to test this specific hypothesis will validate the approach scientifically.



ALS TDI's biotech lab in Cambridge, MA

These specific mice were initially developed at the University of Massachusetts Medical School and express the same toxic proteins that challenge the cells in ALS TDI's initial experiment. This further study will show that neurons can be protected from C9orf72 mediated cell death in brain and spinal cords, slow disease progression, all while not causing negative side effects or toxicities. These types of findings are also essential to attract potential partnerships from well-capitalized biotech and pharmaceutical entities who could fund necessary clinical trials. This could be even more important for all people with ALS based on the fact that we have reason to believe that PRMTs may be involved in other subtypes of ALS as well.

www.als.net

What is ALS?

Amyotrophic lateral sclerosis (ALS) is a progressive nervous system disease that affects motor neurons in the brain and spinal cord, causing loss of muscle control. ALS is often called Lou Gehrig's disease, after the baseball player who died from it. It often begins with muscle twitching and weakness in a limb, or slurred speech. Eventually, ALS affects control of the muscles needed to move, speak, eat and breathe. There is no cure for this fatal disease and currently no effective treatments.



"The support of the Jim Heller Fund has allowed us to bring in a new animal

model to test potential therapeutics aimed at the target. Our expertise in animal model development is held in the highest regard in the research community, and through this effort we plan to both identify potential treatments and also help the research community to understand best practices for its use."

- Fernando Vieira, M.D., CEO & Chief Scientific Officer at ALS TDI

The Opus Group Makes \$225,000 Contribution to The Jim Heller Fund



Yes, you read that correctly. This incredibly generous donation nearly doubled the size of our Fund overnight! Because of the generosity of The Opus Group, we were able to fund the drug development lead at ALS TDI (see pg. 1) and more. Many, many thanks to Opus and its leadership team of Tim Murnane, Dave Menke, Tom Becker and Ed Gschneidner. Jim worked for Opus from 1993-2008 and was a leader and a friend to many in those 15+ years. When asked, Jim's Opus colleagues mention his humor, integrity and genuine interest in others as some of his best traits. **THANK YOU, OPUS!** You have just become part of a passionate group of ALS advocates who are determined to change the ALS landscape. It's not an incurable disease ... it's an underfunded one.

“Jim was an incredible guy, and all of us who were lucky enough to work with him learned so much from him. He was a great leader and mentor, but most of all he was a dear friend. We hope that our gift in his memory can help make a difference as the Fund and Lori continue to fight this battle.”

- Tim Murnane, President & CEO, Opus Holding, LLC

Fund Update

We remain **so very grateful** for each and every contribution we receive. Without your donations, we would be unable to fund very important research and therapeutic developments needed to end ALS. As a reminder, all donations are tax deductible, as they are going straight to a 501c3 charitable foundation.

\$243,355 raised via hundreds of individual donations-8/20-4/21

(\$50,000) granted to ALS TDI for PMP Program-12/20

(\$10,000) granted to Team Gleason for ALS Patient Equip.-12/20

\$225,000 donation from The Opus Group-3/21

(\$150,000) granted to ALS TDI for Therapeutic Development-4/21

(\$10,000) granted to Team Gleason for ALS Patient Equip. -4/21

\$248,355 current Fund* balance

**To date, we have raised a total of \$468,355 in Jim's memory with your help. THANK YOU!!*

www.givemn.org/jimheller/story

What Have I Been Doing, You Ask?

- I am filling a (volunteer) role as the Clinical Outreach Rep for **ALS TDI's Precision Medicine Program (PMP)**. I am working to increase program enrollment of people living with ALS. The PMP allows those living with ALS to become a partner in research, as the data tracked by ALS TDI through the program becomes part of the largest and most comprehensive database needed to end ALS. At the same time, this data also empowers people living with ALS to make decisions about their own treatments, supplements, etc. by providing them with information that is less subjective and *specific to their* disease progression.
- I also participate in **I Am ALS's Legislative Affairs Committee**, made up of people living with ALS, and caregivers, family and friends of people who are, or have been, afflicted by this disease. We have a weekly Zoom call each Tuesday, and then we head off to the task of contacting all US Senators and House of Representative members to attempt to educate them on issues facing ALS patients. It can be a full-time job, trust me.



“Keep Your Fork”

Remember when you were a kid and your mom was clearing the table after dinner but told you to “keep your fork” because dessert was coming? Well, Jim and I decided during our battle with ALS that the fork would be our symbol. It was a reminder to us that the best is yet to come. Only God knows what and when that will be for each of us. I would encourage those living with ALS and their families/caregivers to keep the faith. For some of you, the fork may symbolize the eventual cure to this disease, and maybe to others you will feel the same assurance of eternity and sense of peace from the fork that we did. In any event, please remember that **the best is yet to come.**

“Alone we can do so little; together we can do so much.”

-Helen Keller

“So true.” 😊

-Lori Larson Heller

Reflections on One Year

Our Jim lost his battle with ALS on May 16th, 2020. I was always pretty good at math, so I think this means we are only a couple of weeks away from the one-year anniversary ... a day we both *do* want to remember and *do not* want to remember. There are far too many lessons learned this year to share here, so I'll focus on the big one, **gratitude**.

Did you know that gratitude should live in your heart not only when good things happen and we are grateful, but also when terrible things happen to good people? Yep, that's right. So, I'm supposed to be filled with gratitude even though I lost my person, I am alone, the future I dreamed of is gone, and there are very few minutes of each day that I don't think about my loss? Yep, that's right.

I have taught myself to be grateful over the past year. I am grateful that although I had to lose Jim, I had time left with him after the diagnosis. I am grateful that we were in a position to have me stop working to spend time with him. I am grateful that we happened to live five minutes away from an ALS Center of Excellence at HCMC. I am grateful that we fell into the hands of such compassionate doctors as Dr. Maiser and Dr. Tonkin, along with the most fabulous infusion nursing crew at HCMC anyone could ask for. I am grateful that I had the strength and courage to be Jim's caregiver until the day he died. I am grateful that we met Pastor Debbie just two weeks before Jim died, that she was willing to come into our home during the pandemic when she otherwise was not making house visits, that we instantly felt like we had known her forever, that she turned around while leaving from a visit just two days before Jim died to say "but if you need me, Jim, or take a turn for the worse, I will be here" (after we had decided her visit would be in another two weeks and had no inclination his death was near), and that she came immediately when called two days later to pray with us when we learned Jim wouldn't make it through the night.

(cont'd.)

I am grateful that both of Jim's boys and their families not only made trips from Atlanta to visit Jim in the week leading up to Jim's passing, but they also chose that visit to have very meaningful conversations with their dad on this specific trip. I am grateful that although Jim was in bed with "a touch of pneumonia", something in him made him rally to get out of bed and have me call the kids to come that Thursday night to visit rather than waiting until Friday morning as planned (and then Jim was never lucid again after going to bed on Thursday night). I am grateful that an angel in the form of a hospice nurse showed up on Friday night, happened to be a palliative care nurse, and had randomly decided to take a weekend shift which was not her norm. She was a gift from God in her knowledge and the way she knew to deliver the unexpected news to us. I am grateful that I was not alone and that most of the kids were with us and gave me comfort when we found out the end was here. I am grateful that I had the strength to administer the last dose of meds to Jim once the nurse had left, and I am grateful that I had the wisdom to know that I should crawl into bed and hold him, and *then* he took his last breath.

Keep in mind that we thought we had at least months, if not year(s), of time left with Jim. The kids did not plan those trips nor have those words with him because we thought it was their last visit. On May 14th, 2020, Jim had simply come down with a rough cough and, again, we were told he had a touch of pneumonia. Not much more than a day later, he went home to Heaven. ***So, you see, we have a lot to be grateful for. We could choose to be disillusioned that he was taken from us, or we can choose to be grateful we ever had this wonderful man in our lives. We choose the later, and we are grateful.***

New News

- March 25, 2021 was a special day as I officially changed my name to Lori Larson Heller!
- Reminder: All 32 MLB teams have declared June 2nd as Lou Gehrig Day. This is a great way to honor a great man while bringing more awareness to ALS. Watch for announcements from your home team on ways you may be able to participate.
- I applied for, and was awarded, 1 of only 4 scholarships given out by NYU Compassionate Use and Preapproval Access (CUPA) to take a course called "Expanded Access, Clinical Trials, and Right To Try". The course is a full-time, six-week class that is put on by the University of Arizona Law School in conjunction with NYU. The \$2,000 cost of this course is covered via the scholarship, and I will be taking the course online. The education I receive from this course will be very beneficial in my ALS advocacy work.

May is ALS Awareness Month

If you are so inclined, please think about doing a random act of kindness for someone on or around May 16th as a way to honor Jim and give a nod to ALS awareness month. Have you always wanted to pay for the next person's coffee in line at the coffee shop? Now's your chance. It's something we can all do without having to open our wallets. Just open your heart.

www.givemn.org/jimheller/story