

## INSIDE THIS ISSUE

Doctor on Call ..... 1

President's Corner ..... 6

Danny Weil,  
The Wall of Death ..... 9

Medical News  
Roundup ..... 10

Personal Financial  
Planning ..... 15

IWMF Doc Star:  
Shirley D'Sa ..... 17

IWMF Connect ..... 18

Support Group  
News ..... 21

Crossing Continents... 24

International Scene.... 25

Cooks' Happy Hour ... 29

## DOCTOR ON CALL

BY STEVEN P. TREON MD, PHD, FRCP



Dr. Steven Treon (DFCI) and Dr. Lia Palomba (Memorial Sloan Kettering Cancer Center) with colleagues from Pharmacyclics, Inc., and Janssen Oncology Inc., at the U.S. Food and Drug Administration, Washington D.C. in 2015. Dr. Treon, Dr. Palomba, and Dr. Ranjana Advani (Stanford University) oversaw the trial that led to the first ever approval of a drug (ibrutinib) for WM. Dr. Treon stands third from the left in the front row. Dr. Palomba is to his right.

*Dr. Steven Treon is a Professor of Medicine at Harvard Medical School, Director of the Bing Center for Waldenstrom's Macroglobulinemia at the Dana-Farber Cancer Institute, Chair of the WM Clinical Trials Group, and Principal Organizer of the International Workshops on WM. He is the recipient of the Robert A. Kyle Award for Outstanding Contributions to WM and the Jan Gosta Waldenström Lifetime Achievement Award, and was elected a Fellow to the Royal College of Physicians in London for his work in WM. Dr. Treon's laboratory was the first to discover highly recurring mutations in MYD88 and CXCR4 in WM. He was the Principal Investigator for the pivotal trial that supported the approval of ibrutinib by the US Food & Drug Administration, the European Medicines Agency, and Health Canada.*

*Doctor on Call, cont. on page 2*

# OFFICERS & TRUSTEES

## FOUNDER

Arnold Smokler

## PRESIDENT EMERITA

Judith May

## PRESIDENT

Carl Harrington

## SECRETARY-TREASURER

Beverly Docteur

## VICE PRESIDENTS

Elena Malunis

Michael Sesnowitz

Guy Sherwood, MD

## BOARD OF TRUSTEES

Stephen Ansell, MD

Peter DeNardis

Marlyn Friedlander

Sue Herms

Marcia Klepac

Robert A. Kyle, MD

Barry Nelson

Linda Nelson

Lisa Wise

## BUSINESS OFFICE

Jennifer Silva, Operations Manager

## IWMF SCIENTIFIC

### ADVISORY COMMITTEE

Stephen Ansell, MD, Chairman

Mayo Clinic

Bart Barlogie, MD

Mount Sinai Hospital

James R. Berenson, MD

Institute for Myeloma & Bone Cancer  
Research

Morton Coleman, MD

Weill Cornell Medical College

Meletios A. Dimopoulos, MD

School of Medicine,

University of Athens, Greece

Stanley Frankel, MD

Columbia University

Morie Gertz, MD

Mayo Clinic

Irene Ghobrial, MD

Dana-Farber Cancer Institute

Eva Kimby, MD

Karolinska Institute, Sweden

Robert A. Kyle, MD

Mayo Clinic

Véronique Leblond, MD

Hôpital Pitié Salpêtrière, France

Gwen Nichols, MD

Leukemia & Lymphoma Society

Roger Owen, MD

St. James's Institute of Oncology,

United Kingdom

Lia Palomba

Memorial Sloan Kettering Cancer Center

Surinder Sahota, PhD

University of Southampton, United Kingdom

Sheeba Thomas, MD

MD Anderson Cancer Center

Steven Treon, MD

Dana-Farber Cancer Institute

Brian Van Ness, PhD

University of Minnesota

Mary Varterasian, MD

## Doctor on Call, cont. from page 1

The approval of ibrutinib as the first ever approved agent for use in symptomatic Waldenstrom's macroglobulinemia (WM) by the US Food & Drug Administration and many other international regulatory agencies ushered in a new era in the treatment of WM. A regulatory pathway was also chartered with the approval of ibrutinib for treatment of WM, attracting interest by pharmaceutical companies and providing a roadmap for the investigation and approval of other agents for WM.

With the approval of ibrutinib, debate has emerged on how WM is best managed and what priorities should be given for the development of medications like ibrutinib that require indefinite daily administration versus those that have a defined length of administration. While there is precedent for the use of daily medications to treat a multitude of disorders in everyday life, such as high blood pressure, cholesterol, or diabetes, the debate of "daily" versus "defined" has taken on more urgency as the high cost of drugs like ibrutinib is pitted against the efficacy and safety of other drugs traditionally used in the management of WM. However, such comparisons are not easy to undertake or justify because of many factors that need to be weighed beside actual drug costs. These other factors include the costs associated with treatment administration, supportive medications (such as Neulasta or Neupogen), blood transfusions, and treatment of short- and long-term side effects.

Treatment options for WM have traditionally relied on rituximab alone, or with alkylators (cyclophosphamide or bendamustine), nucleoside analogues (fludarabine or cladribine), immunomodulatory agents (thalidomide), or proteasome inhibitors (bortezomib or carfilzomib). While many lessons were learned from trials examining single agent and

*Doctor on Call, cont. on page 3*



The IWMF *Torch* is a publication of:

International Waldenstrom's

Macroglobulinemia Foundation

6144 Clark Center Avenue • Sarasota, FL 34238

Telephone 941-927-4963 • Fax 941-927-4467

E-mail: [info@iwmf.com](mailto:info@iwmf.com) • Website: [iwmf.com](http://iwmf.com)

This publication is designed to provide information about the disease Waldenstrom's macroglobulinemia. It is distributed as a member service by the International Waldenstrom's Macroglobulinemia Foundation, Inc., to those who seek information on Waldenstrom's macroglobulinemia with the understanding that the Foundation is not engaged in rendering medical advice or other professional medical services.

### EDITORS

Alice Riginos, Shirley Ganse

### ASSOCIATE EDITOR

Sue Herms

### SUPPORT GROUP NEWS

Penni Wisner

### IWMF DOC STAR EDITOR

Ron Ternoway

### IWMF CONNECT

Jacob Weintraub, MD

### INTERNATIONAL CORRESPONDENT

Annette Aburdene

### CULINARY EDITOR

Penni Wisner

### CARTOONIST

Linda Pochmerski

### FORMATTING & PRODUCTION

Sara McKinnie

### TORCH ADVISORY PANEL

Carl Harrington, Robert Kyle, Judith May, Guy Sherwood

IWMF is a 501(c)(3) tax exempt non-profit organization Fed ID #54-1784426.

Waldenstrom's macroglobulinemia is coded 273.3 in the International Classification of Diseases (ICD) of the World Health Organization.

combination rituximab therapy, the most important ones have centered on how much disease eradication can occur (i.e. depth of response), how long such therapies can keep disease from progressing or coming back (i.e. durability of response), and the short- and long-term toxicities they can produce.

Rituximab was the first modern biological therapy used to treat indolent lymphomas such as WM. Rituximab binds to CD20 and attracts immune cells and immune system proteins such as complement that destroy CD20-expressing cells. The malignant clone in WM patients is made up of B-cells that exist in three morphologically distinct stages, including CD20-expressing mature B-cells, CD20-expressing lymphoplasmacytic cells, and non-CD20-expressing plasma cells. Most of the IgM made in WM patients comes from plasma cells, and for many patients the production of IgM produces symptoms such as hyperviscosity syndrome, IgM-related neuropathy, cryoglobulinemia, and cold agglutininemia that necessitate treatment.

Two schedules for single agent rituximab monotherapy were investigated in WM: a “standard” one with 4 weekly rituximab infusions and an “extended” one with 4 additional weekly infusions at weeks 12-16 following standard administration. With “standard” rituximab administration, the overall response rate (ORR) that includes minor responses (i.e. responses that reduce IgM by at least 25%) is 40%, while the major response rates (i.e. responses that reduce IgM by at least 50%) are 20-30%. With “extended” rituximab, the ORR is higher (50-60%), with major response rates of 40%. Deeper responses, such as very good partial responses (VGPR), where IgM is reduced by 90%, and complete responses (CR), where the IgM levels return to normal and no WM-related IgM is detected, are rare with rituximab, and the average time that these responses last is 13-29 months.

Responses to rituximab are slow, with time-to-best-response requiring up to 18 months. Since the malignant WM clone is also made up of plasma cells that do not have CD20, sparing of these cells usually occurs after rituximab alone. This, in turn, results in persistence of IgM-producing plasma cells that can trigger symptoms brought on by the production of IgM, as well as its accompanying light chain proteins (kidney failure, amyloidosis). A flare in serum IgM commonly occurs with rituximab and can induce hyperviscosity symptoms and/or aggravate symptoms attributed to IgM. With prolonged rituximab use, allergic reactions can occur in 10% of WM patients. Moreover, prolonged rituximab use can deplete good CD20-expressing B-cells and result in deficiencies of IgA and IgG that can lead to recurring sinus and bronchial infections necessitating antibiotics and, in

more severe cases, requiring the use of intravenous gamma globulin (IVIG).

To extend rituximab activity, rational combinations have been sought. Laboratory studies have provided logical justification for some, but not all, rituximab combinations. Many agents used with rituximab target non-CD20-expressing plasma cells and provide an important overlap to eradicate the entire WM clone. With most combination rituximab therapies, improvements in ORR and deeper responses have occurred. The ORR with rituximab and alkylators, nucleoside analogues, and proteasome inhibitors is 80-90%, with deeper VGPR/CR responses seen in 30-40% of WM patients. The use of maintenance rituximab has also contributed to deeper responses in WM. With deeper responses, improvements in the durations of response have been recognized. The attainment of VGPR or better has been observed to predict for longer response durations with many rituximab combinations. These findings were also recognized in a retrospective study that examined the outcome of 159 WM patients receiving rituximab-based therapy. CR or VGPR attainment was associated with average response durations that exceeded 90 and 75 months, respectively. For those that attained less deep responses, the average response durations were 43 and 31 months, respectively, and were 11 months in those without response or stable disease.

While depth and durability of response have increased with combination rituximab regimens, so has toxicity. Treatment-related adverse events following rituximab combinations have included myelodysplasia (damaged bone marrow stem cells), secondary malignancies such as acute leukemia, prolonged suppression of blood counts, immune system suppression, and neuropathy. Avoidance of nucleoside analogue drugs like fludarabine, limitations on alkylator drug exposure, adoption of weekly bortezomib regimens, and use of neuropathy-sparing proteasome inhibitors have impacted short- and long-term toxicity with rituximab combinations. Efforts to maintain and induce deeper responses with rituximab alone or in combination have shown promise and continue to be evaluated. Conversely, the use of stem cell transplant therapy following initial therapy has been avoided due to toxicity.

The discovery of highly recurrent MYD88 (95-97%) and CXCR4 (30-40%) mutations in WM patients has provided important new insights into the biology of WM and in the development of targeted therapies. Mutated MYD88 triggers the activation of Bruton's tyrosine kinase (BTK) in WM cells. Activated BTK turns on NF- $\kappa$ B, a protein in WM cells that



enhances the growth and survival machinery of WM cells. In addition, mutated MYD88 stimulates the production and activation of hematopoietic cell kinase (HCK), a protein that acts as a major switch for the activation of other WM cell growth-promoting pathways including AKT and ERK. The activity of BTK and HCK are both blocked by ibrutinib.

These basic science findings enabled a pivotal clinical trial of ibrutinib in symptomatic, previously treated WM patients and allowed the first ever “breakthrough” designation for an oncology drug, thereby enabling accelerated review. Sixty-three patients at the Dana-Farber Cancer Institute, Memorial Sloan Kettering Cancer Center, and Stanford University Medical Center took part in the Phase II trial wherein patients received 420 mg a day of ibrutinib. Dose reduction was permitted for toxicities. The results of this study were originally published in the *New England Journal of Medicine* in 2015 and were just updated at the 2017 Annual Meeting of the American Society for Hematology (ASH). The findings showed an ORR of 90%, with 78% of patients attaining a major response, while 27% had a VGPR. Those patients with MYD88 mutations but no CXCR4 mutation showed the highest levels of ORR, major response, and even VGPR. Most of these patients show continued response beyond 5 years. Patients with both MYD88 and CXCR4 mutations had slower responses and lower ORR and major responses. Their responses lasted on average 4 years, which still favorably compares to other treatments used in previously treated patients. In contrast, those patients without a MYD88 mutation showed poor response activity and duration of response.

The high rates of response and durability of response with single agent ibrutinib were also observed in a study published in *Lancet Oncology* in 2017 wherein more heavily pre-treated, rituximab refractory patients received treatment. The impact of CXCR4 mutations was also observed in this study, in which WM patients with mutated CXCR4 had slower responses and slower improvements in their hemoglobin levels. A clinical trial of single agent ibrutinib in symptomatic patients with untreated WM was also reported at the 2017 ASH meeting. High levels of overall response (97%) and durable responses were also observed in this study, along with the impact of CXCR4 mutations on time-to-response and response rates. A randomized trial (iNNOVATE) comparing ibrutinib and rituximab versus rituximab in patients with symptomatic untreated and previously treated WM has been fully enrolled, and results are expected to be available in early 2018.

In comparison with other therapies used to treat WM, use of ibrutinib results in rapid responses, with a time to at least a minor response of 4 weeks. In 10% of patients, atrial

fibrillation can occur but does not limit ibrutinib continuance in most patients. Risk of bleeding with procedures and concurrent use of blood thinning medications remain a concern, as do decreased blood counts in heavily pre-treated patients. Unlike rituximab-based therapies, IgA and IgG levels remain unchanged with ibrutinib, and infectious complications are uncommon. Persistent low-grade musculoskeletal, skin, and gastrointestinal toxicities can occur with ibrutinib and result in dose reduction and treatment cessation in some WM patients. Withholding ibrutinib for procedures or adverse events can lead to rapid increases in serum IgM, constitutional (fevers, chills, sweats) complaints, and decreased hemoglobin, signifying that residual tumor cells have the potential to rapidly propagate disease. For these reasons, ibrutinib therapy should not be stopped unless medically indicated. These findings contrast with what is typically observed following rituximab-based therapy, wherein the typical post-treatment course is disease latency, followed by slow disease recurrence over time.

The lack of CR observed in WM patients on ibrutinib, regardless of MYD88 or CXCR4 mutation status, also indicates an “intrinsic” resistance. Signaling studies of surviving WM cells in patients on prolonged ibrutinib therapy (>6 months) showed that while BTK activity was suppressed, an alternative pathway by the IRAK1/IRAK4 proteins was not effected by ibrutinib and contributed to WM cell survival. “Acquired” ibrutinib resistance (i.e. resistance after the patient responds) is also an emerging problem in WM patients. Mutations that prevent ibrutinib from binding to BTK were identified in half of a handful of WM patients who progressed after responding to ibrutinib. Nearly all these patients had mutated CXCR4. MYD88 mutated WM cells engineered to express one of these BTK mutations showed ibrutinib resistance, reflecting the importance of these mutations as contributors of “acquired” resistance.

While in most WM patients deep responses and long-term disease control can be attained with prolonged ibrutinib therapy, those without MYD88 mutations and those with mutated MYD88 and CXCR4 may be at higher risk of either non-responsive disease, suboptimal responses, or “acquired” resistance in the latter. “Intrinsic” resistance in patients with mutated MYD88 and no CXCR4 mutations can also lead to rapid disease progression if ibrutinib is stopped. For these reasons, a strategy dependent on disease control with ibrutinib alone should not be viewed as optimal for WM. Many insights into WM cancer biology, as well as mutated MYD88 and CXCR4 signaling, have provided important clues for rational drug development aimed at eradicating the malignant clone in WM.

As previously mentioned, one of the important limitations of rituximab is sparing of IgM-producing plasma cells that do not express CD20. These cells make up 10-15% of the WM clone. Daratumumab targets CD38, highly expressed on WM plasma cells. Strategies using daratumumab and rituximab, as either dual therapy or with other therapeutics, are of interest and offer a means to target the entire WM malignant clone. A Phase II study of daratumumab in previously treated WM has been initiated ([www.clinicaltrials.gov](http://www.clinicaltrials.gov) NCT03187262) and will offer critical insights into targeting the plasma cell compartment, along with the potential to combine rituximab with other agents aimed at expunging the entire WM clone.

As mentioned before, a randomized study (iNNOVATE) is also examining the combination of ibrutinib with rituximab ([www.clinicaltrials.gov](http://www.clinicaltrials.gov) NCT02165397). Since activating CXCR4 mutations promote ibrutinib resistance, a clinical trial combining the CXCR4-blocking antibody ulocuplumab with ibrutinib has also been initiated at our institution in WM patients with CXCR4 mutations ([www.clinicaltrials.gov](http://www.clinicaltrials.gov) NCT03225716).

Compounds that inhibit IRAK1/IRAK4 are also under intense pre-clinical investigation and are aimed at overcoming intrinsic ibrutinib resistance in MYD88 mutated diseases. A research project to develop IRAK inhibitors is being supported by the IWFM and has produced highly selective and potent IRAK inhibitors that show synergistic WM cell killing when combined with ibrutinib.

BCL-2 is a protein that is overexpressed in WM cells and blocks the killing effects of ibrutinib. The BCL-2 inhibitor, venetoclax, showed major response activity in 4 WM patients treated in a Phase I study. A clinical trial examining venetoclax in previously treated WM patients is near complete enrollment ([www.clinicaltrials.gov](http://www.clinicaltrials.gov) NCT02677324) and will inform a planned successor study of venetoclax with ibrutinib.

Lastly, other BTK inhibitors are currently under investigation (e.g. BGB-3111, acalabrutinib, GS-4059) in WM, and a study comparing the efficacy and safety of BGB-3111 directly to ibrutinib is currently enrolling subjects ([www.clinicaltrials.gov](http://www.clinicaltrials.gov) NCT03053440). The activity and safety of these other BTK inhibitors will provide important insights on the use of this class of agents and help to better determine their positioning relative to other therapeutics used to treat WM.

## Welcome To New Board Member Lisa Wise

Elected as a new trustee in 2017, Lisa Wise brings a career of invaluable experience to her role in the IWFM. With a graduate degree in education and human development from Harvard, Lisa went on to a professional career of 19 years as a family and patient-centered healthcare specialist. She is co-leader of the Eastern Pennsylvania support group, and has been a dedicated student at the Penn Program for Mindfulness as well. Her IWFM duties include chair of the LIFELINE Committee and a member of the Support Group Committee.

Lisa lives in the suburbs of Philadelphia with her husband of 27 years and is the proud mom of 20 year-old fraternal twin sons, one of whom carries diagnoses of Crouzon syndrome and hydrocephalous. As a cancer patient, and born into a cancer-cluster family, Lisa navigates through an extensive palliative care journey, and these pivotal experiences inform, shape, and guide her work today.



*Board Member Lisa Wise, who leads a busy and committed life, catches her breath relaxing in a natural environment.*

---

## PRESIDENT'S CORNER

---



Waldenstrom's macroglobulinemia (WM) is a rare disease. A disease or disorder is defined as rare in Europe when it affects fewer than 1 in 2,000. A disease or disorder is defined as rare in the United States when it affects fewer than 200,000 at any given time. Although exact numbers are not available, in the US about 1,500 new cases of WM a year are diagnosed, and, perhaps 20,000 WMers live with the disease. WM is a rare disease indeed.

As I travel around the US and the world to represent the IWMF, I am struck by how rare a community we are. I constantly hear from other patient organizations, clinicians, researchers, and pharmaceutical companies that the IWMF is very special. The IWMF stands out among rare diseases as a rare *community* that cares and shares in a unique and special way.

Since this is the first issue of the *Torch* for 2018, let me give some examples of how we made progress in 2017 in caring and sharing. This progress has enabled us to grow our rare community to serve you better, and better fulfill our vision to *support everyone affected by Waldenstrom's macroglobulinemia while advancing the search for a cure*.

### Financial support

We received over 2.5 million dollars in support in 2017, our second highest total ever, behind 2016, which included a one-time \$500,000 estate gift through the Ben Rude Heritage Society. Thank you for your support!

If you're still in the mood to make New Year's resolutions, how about joining the Ben Rude Heritage Society by naming the IWMF in your will? There is no minimum value for a legacy gift, so why not add the IWMF now? See <http://iwmflegacy.com/?pageID=1004> to join.

### Research support

In 2017 we committed an astounding 2.2 million dollars to fund six new research projects over the next two years as part of the IWMF-LLS Strategic Research Roadmap. We now have 13 active WM research projects totaling over \$4.7 million dollars funded by WMers just like you and me. Importantly, we have the best minds in the world working on our orphan disease with projects in Germany, Italy, the Netherlands, Spain, the UK, California, Connecticut, Massachusetts, Minnesota, and Missouri. Add in the Australians working on the WhiMSICAL database and the sun *never* sets on WM

research. When you go to bed, someone is working on finding a cure. When you get up, someone is working on finding a cure. When you have lunch, someone is working on finding a cure. Isn't that incredible? And we did that working together!

We also laid the groundwork for future progress in 2017 with the third IWMF-LLS Strategic Research Roadmap meeting in New York in October. Fifteen of the brightest minds in WM met to hone our research needs. In November we issued the Third Strategic Research Roadmap RFP (Request for Proposals). We hope to be able to fund at least two new pieces of research in 2018. The number of projects we are able to fund depends upon the generosity of our members and their friends and family. Can you ask them to support you and the IWMF in 2018?

### Educational support

To help you and other members understand WM, here are a few highlights of what we accomplished in 2017:

- Held very successful patient education meetings in the US, Canada, the UK, France, Scandinavia, Australia, and Finland that reached nearly 900 WMers. The US Educational Forum in Phoenix, AZ, set a new attendance record and received a rating of 4.85 out of 5! In 2018 our Educational Forum will be at the Westin Hotel in Rosemont, IL, May 18-20, 2018. Registration information is available on page 8.
- Conducted two very successful joint webinars with CancerCare. The second one, entitled "What's New in the Treatment of WM" featuring Dr. Jorge Castillo and Dr. Jeffery Matous, had attendance of over 1,000 WMers from 17 countries. If you missed it, you can still listen to it at <https://www.iwmf.com/news-and-events/news/cancercare-webcast-whats-new-wm-treatment-now-online>
- Created a new Frequently Asked Questions booklet that is written in easy-to-understand language. If you haven't downloaded your free copy, go to <https://www.iwmf.com/media-library/iwmf-publications> and give yourself a New Year's present. I'm proud to say this booklet is available in English, Traditional Chinese, Simple Chinese, German, Spanish, French, Italian, and Finnish.

### Patient support

We didn't stint on patient support in 2017 either. Highlights include:

- Implementing a new Online Forum, *IWMF Connect*, to replace *IWMF-Talk*. If you're not a member, join the 2,100 WMers who participate. <https://www.iwmf.com/get-support/iwmf-connect-and-online-discussion-forums>

*President's Corner, cont. on page 7*



- Expanding LIFELINE, our one-on-one peer telephone service.
- Adding new international affiliates in Mexico and India. We now have 16 affiliates worldwide.
- Increasing the quality and quantity of our support groups. We now have over 60 support groups worldwide.
- Adding six new partners to supplement and expand the services that we can offer. See what our partners can do for you at:

<https://www.iwmf.com/about-us/partners>

Just as with research, the sun *never* sets on IWMF Education and Support. No matter when you need help, you can find it at the IWMF website or our affiliates' or partners' websites. If you need to talk to a person, call LIFELINE or call the IWMF office (1)-941-927-4963 or come to a support group meeting. We also expanded the IWMF office in 2017 to better serve WMers everywhere.

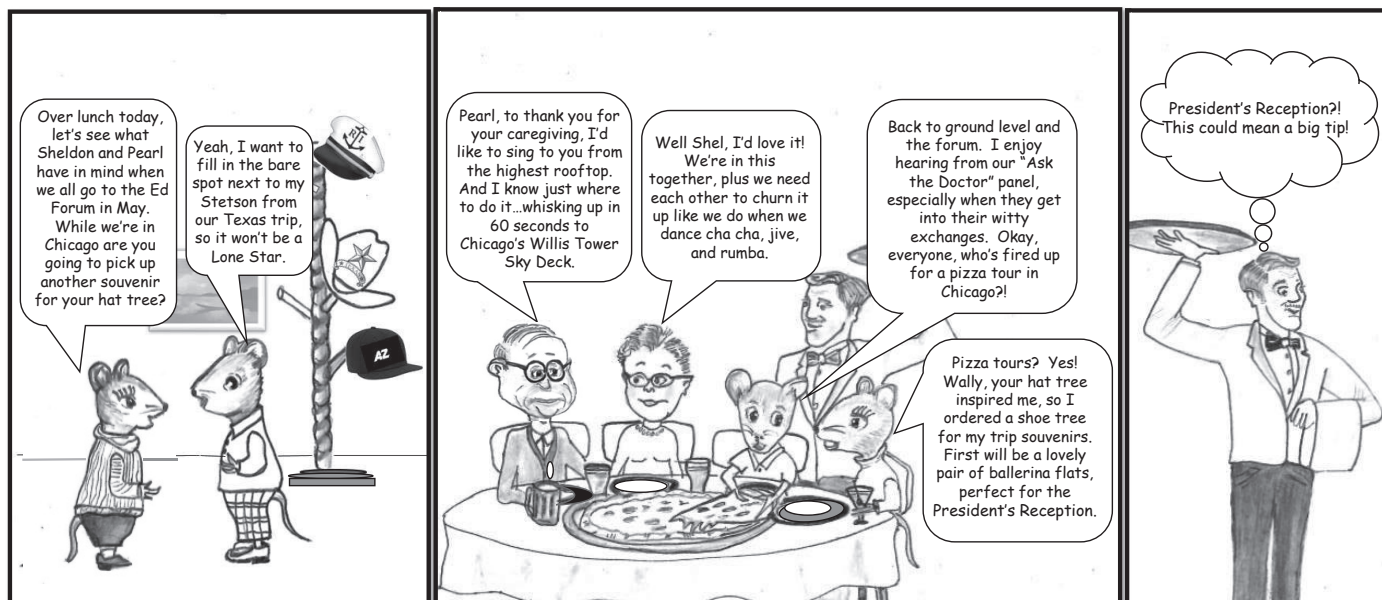
Happy New Year and best wishes for 2018! Please keep sharing and caring for one another. We want people newly diagnosed with WM to find even more knowledge and support than we did when we were diagnosed. And we want them to find better treatments with milder side effects. To do that, we all need to continue sharing and caring for each other.

As you know, the IWMF is patient-led and patient-funded. That means all of the Trustees, all of the affiliate heads, all of the support group leaders, all of the LIFELINE folks, the *Torch* staff, the Research Committee, and many more are volunteers. If you have skills you'd like to share to benefit your fellow WMers, contact Jennifer Silva, the IWMF Operations Manager at [jsilva@IWMF.com](mailto:jsilva@IWMF.com) or the head of the WM organization in your country. If you want to provide financial support to the IWMF, contact Dave Benson at [dave@dbenson.com](mailto:dave@dbenson.com) or Brian Miller at [bmiller@iwmf.com](mailto:bmiller@iwmf.com)

Let's join together in 2018 to further build our rare community. We are all in this together!

Have a happy and healthy 2018!  
Carl

*WALLY and WINNIE, WM Model Mice by Linda Pochmerski*



After Winnie and Wally sit down to lunch with their friends Shel and Pearl to discuss each other's trip desires, there are pleasant signs of gratitude, caring, the joy of learning, and a start-up shoe tree to look forward to. Now, as we might imagine, Shel's singing will be music to Pearl's ears, Wally will grab a front seat to listen to the doctor's panel, and Winnie's shoe tree will be adorned with style and fashion. And yes, there really are pizza tours in Chicago. Enjoy!

*Don't  
miss this one!*

## **23rd Annual IWMF Educational Forum**

**May 18-20, 2018**

**The Westin O'Hare Hotel**

**6100 North River Road, Rosemont, IL 60018**

**[www.westinohare.com](http://www.westinohare.com)**

## **Imagine a Cure: Closer Than Ever!**

**So much exciting progress has been made in research and treatment options.  
Come join the celebration. We have a space just for you. Register today!**

### WHAT IS THE ED FORUM?

Imagine several hundred WMers and caregivers gathering for an information-packed weekend of learning, connecting, and networking! The IWMF Ed Forum offers one-stop shopping for WM patients and caregivers from around the world. Be there in person to experience the magic of landing on Planet Waldenstrom and discovering that "You are NOT ALONE!"

### WHO WILL BE THERE?

The IWMF Ed Forum features the world's leading medical experts in WM research and clinical care. Enjoy being surrounded by friendly "Waldenfriends" and caregivers who know how it feels to carry the same diagnosis, speak the same disease language, share strategies for symptom management, and swap treatment experiences and lab numbers. We are all in this together!

### NEWLY DIAGNOSED OR A FIRST-TIMER?

The exciting Early Bird Session on "The ABCs of WM" is a great introduction for newbies and first-timers, but it's open to ALL attendees. Enjoy a continental breakfast before it begins.

### ED FORUM FAVORITES...

Highlights each year include: asking your questions for the lively and ever-popular "Ask the Doctor" panel discussion, mingling at the President's Reception, enjoying the festive Welcome Dinner alongside IWMF Trustees and world-class physicians, and exploring a plethora of different and diverse Breakout Sessions addressing multiple issues and hot topics. There is truly something for everyone at the IWMF Ed Forum!

### CALLING ALL SUPPORT GROUP LEADERS!

Are you a Support Group Leader? Be sure to attend the informative workshop on Thursday, May 17, led by Marcia Klepac, IWMF Support Group Coordinator.

**Don't miss the event of the year!**

**Get all the details and register online at**

**[www.iwmf.com/news-and-events/iwmf-educational-forum](http://www.iwmf.com/news-and-events/iwmf-educational-forum)**

**OR fill out and send the registration form enclosed with this *Torch* mailing**



---

# DANNY WEIL, THE WALL OF DEATH . . . AND WM!

AS TOLD TO SHIRLEY GANSE

---



*Danny Weil on his 70 year-old Indian Scout motorcycle on rollers in front of his Thrill Arena.*

We all have certain fond memories of childhood. Not all of us, however, get the opportunity to act upon them in later life. When Danny Weil was young, he loved going to carnivals and their sideshows. The people, the excitement, and the fun were a natural for Weil, who says he's "a 'people person' who loves to meet and greet." It all came together for him when he opened his own Wall of Death motorcycle sideshow in 2016.

Native to Texas, Weil knew he really liked motorcycles by the time he was 25, and as a machinist and toolmaker, he enjoyed fixing and restoring antique motorcycles. Working for himself since 1992, he went to various motorcycle shows and rallies, where he always went to see the Wall of Death shows. But in 2001, at the Sturgis Motorcycle Rally in South Dakota, his talents were called upon. An antique motorcycle that was part of a Wall of Death show needed fixing; he helped them out, and he was on his way to becoming passionate about a disappearing bit of American popular entertainment.

Many of us are probably of the age that we remember this Wall of Death sideshow at our local fair or carnival. Daring motorcyclists run their cycles in a tight circle up a vertical wall and then do various tricks while contending with almost 3 Gs created by centrifugal force. It takes a fair amount of strength to perform this show, and Weil was almost 53 years old when he first "rode the wall" in 2012.

For almost ten years, Weil continued to help the Wall of Death show crew with their motorcycles, and then he went on the road with them from 2012 to 2015. With this experience, he decided he'd rather travel a show of his own. It took him ten months to build the tented arena in his backyard and another several months to get the insurance and DOT papers in order. But he couldn't figure out why he was getting so tired. Not only that, Weil says, "I never went to a doctor or had an annual physical; I felt weird on occasion, and my fingers and toes were always cold...but I didn't think anything of it."

But Weil finally did go for a checkup, and a blood test resulted in his being sent to see Dr. Nuruddin Jooma at Florida Cancer Specialists. "I was really freaked out by what he said, so I decided I'd just go to the Mayo Clinic in Jacksonville for a second opinion." So in January of 2013 (Weil's second year of riding the wall), the Mayo visit not only confirmed Waldenström's with a bone marrow biopsy, but also found he had thrombocytosis, a very high platelet count. Since his IgM was only 1200 and he could live with the symptoms, Weil declined treatment and went back to riding the wall.

Weil's Wall of Death show is strenuous...not only in its performance, but in the set up and take down. With his three-man crew, and maybe a couple of hired hands, it takes eight hours to load their 40' flatbed trailer (which doubles as a stage) and semi, and ten hours to unload and set it up. The twenty wall panels at 225 lbs each take a lot of muscle to wrestle around. It takes its toll.

By early 2014, Weil went back to the Mayo Clinic and found his IgM was over 5000; rituximab only was suggested, so he decided to go back to Dr. Jooma for treatment since Florida Cancer Specialists was closer to home. He had an infusion every three months until September of 2017, and he has high praise for Dr. Jooma, a "cool guy who brought his kids to see my performance."

Weil's first year of operation of his own show was 2016. Set up on a carnival midway, it includes a 70 year-old Indian Scout motorcycle on rollers (so it can run – like an exercise bicycle) in front of the tent, complete with a siren to attract an audience. Weil says "as a talker who likes people, I try to pull them in." Both modern as well as sturdy cycles from the '20s and '30s are used in the show.

The "well" is 26 feet in diameter and 12 feet high, and while riders can experience 3 Gs at the highest speed – it takes 25 mph to stay on the wall – riders don't go any faster than needed to stay up. Each show is 15 minutes long plus a stage show out front for ten minutes. Two cyclists can be on the wall at the same time, and do a variety of tricks while going around. Do they get dizzy? Weil indicates "you can get used to it; some can never overcome it. Maybe if it's hot and you're hungry, you just ride the low line and breathe through it."

So how does he do it during chemotherapy? "Sometimes I sit it out if I'm too tired, but usually I go for it," Weil admits. Injuries can happen: sometimes by going too slow, or a chain breaks, or two riders clip each other. Some riders can stand up, do sidesaddle, or put their legs on top of the handlebars. It can be very noisy, but no one uses earplugs. No one was hurt at all last season. "I never fell once, and that's a record for me," he says.

*Danny Weil, The Wall, cont. on page 10*

Weil says his rituximab treatments were virtually side effect-free. Little slowed him down the past year even though he was still tired a lot, toes and fingers still cold, and feeling weird every once in a while.

Weil's passion clearly includes not only the physical thrill of the event, but also the culture and the history of it. He recounts some history: "the first Wall of Death sideshow was on Coney Island in 1915, and its popularity at local fairs increased until just before World War II when about fifteen shows were circulating around the United States." Rural America enjoyed carnival shows that traveled from town to town, but with the advent of huge amusement parks like Disneyland (1955) and the Six Flags organization (1957), local carnivals started to disappear. "By the early 1980s only one show remained, a little piece of lost magic from America," Weil lamented.

Now four Wall of Death shows circulate, but Weil is the only operator still interested in doing local fairs and carnivals in rural areas. "It's much more fun with families and kids at fairs," he says, comparing his to the other shows which concentrate on entertaining at motorcycle events such as the large yearly gathering at the Sturgis Motorcycle Rally.

Weil and his wife Julie winter over in Florida in their house, where he spends his time restoring engines for antique

motorcycles. But they live in a 5th wheel house-trailer when touring, generally in the East and Midwest. During the season, mid-March until the end of November, a carnival or fair might have a ten-day run, and the Wall of Death show goes on every hour from noon until 10pm or so. That's a grueling schedule for someone who contends with Waldenström's, but for Weil, like for so many people, passion means doing what you love, if at all possible.

Weil knew he was a cancer patient when he started to set up his business in 2015. He says, "Anybody can go out and buy a brand new Corvette for their mid-life crisis – but it takes a real nut to do what I did. Don't let WM keep you from doing something you want to do." He feels really lucky that he has a unique thing to do: helping people have fun at a fair, and he sees it as a privilege to do that.

What does he see for the future? Weil feels it's ironic that he never went to doctors and now he has to rely on them. He thinks his brain is a bit foggy from the WM or the rituximab, or both, and says he doesn't have a lot of confidence to do stuff in his shop now. "If I can sit in the ticket box and talk to people and interact with them on the midway; provide a venue for my crew; and have kids come to see the show, I'm happy just keeping a piece of American nostalgia alive."

---

## MEDICAL NEWS ROUNDUP

BY SUE HERMS, IWMF TRUSTEE AND RESEARCH COMMITTEE MEMBER

---

**Ibrutinib Receives Approval in England for WM and in Australia for CLL/SLL** – Ibrutinib (Imbruvica) has received approval in England through the Cancer Drugs Fund for the treatment of relapsed WM patients who are not suitable for chemoimmunotherapy such as bendamustine and rituximab. The Cancer Drugs Fund was introduced in England in 2011 in order to provide a means by which National Health Service patients in England can get cancer drugs rejected by the National Institute for Health and Care Excellence because they are not cost effective. In Australia, the drug has been approved for treatment of chronic lymphocytic leukemia and small lymphocytic leukemia patients only, but approval is still being pursued for WM patients.

**New Shingles Vaccine Approved in US and Canada for Seniors** – GlaxoSmithKline announced that the US Food and Drug Administration (FDA) and Health Canada have approved its shingles vaccine Shingrix for the prevention of shingles in adults aged 50 and over. Shingrix is not a live-virus vaccine like the current Zostavax vaccine. It contains an adjuvant to help boost the body's immune response and in clinical trials has shown greater protection against shingles among older recipients. Four years after injection, the new vaccine remained about 90% effective in people over 70

who were in the trial study. It also reduced the incidence of nerve pain, called postherpetic neuralgia, which can follow a shingles outbreak. Shingrix is given intramuscularly in two doses 2-6 months apart. Based on FDA approval, the Advisory Committee on Immunization Practices for the US Centers for Disease Prevention and Control (CDC) has recommended that Shingrix be preferred over Zostavax for immunocompetent adults (those with normal immune systems). At this point the CDC recommendation does not address immunocompromised adults. Canada has not made this distinction. A final decision from the CDC is expected in early 2018. Shingrix is awaiting approval in Europe, Australia, and Japan.

**United Kingdom Advisory Issued for Ventricular Arrhythmia, Hepatitis B Reactivation, and Opportunistic Infections in Patients Using Ibrutinib** – An update from the Medicines and Healthcare products Regulatory Agency of the United Kingdom has advised healthcare professionals that cases of ventricular arrhythmia, hepatitis B virus reactivation, and opportunistic infections have been reported in patients using ibrutinib (Imbruvica). The update advises physicians to 1) temporarily discontinue ibrutinib in patients

*Medical News Roundup, cont. on page 11*

---

who develop symptoms suggestive of ventricular arrhythmia (palpitations, chest pain, shortness of breath, dizziness, fainting) and assess benefit-risk before restarting therapy; 2) establish hepatitis B virus status before initiating therapy; 3) for patients with positive hepatitis B serology, consult with a liver disease expert before starting treatment to minimize the risk of reactivation; and 4) consider standard-of-care prophylaxis for patients who are at an increased risk of opportunistic infections.

**Mayo Clinic Looks at Efficacy of DRC Therapy Based on MYD88 Mutation Status** – Mayo Clinic researchers published an article in the *British Journal of Haematology* on the efficacy of dexamethasone, rituximab, and cyclophosphamide (DRC) for relapsed/refractory and treatment-naïve WM patients, especially with regard to their MYD88 status. One hundred patients were evaluated between January 2007 and December 2014. In the 50 relapsed/refractory patients, the overall response rate was 87%, and median progression-free survival and time-to-next treatment were 32 months and 50 months, respectively. In the previously untreated group, overall response rate was 96%, and the median progression-free survival and time-to-next treatment were 34 months and not reached, respectively. Twenty-five of 29 patients who were genotyped harbored the MYD88 L265P mutation. The response rates and outcomes in these patients were independent of MYD88 mutation status. Moderate to severe adverse effects included neutropenia (low neutrophils), thrombocytopenia (low platelets), and infections.

**Mayo Clinic Study Compares Mortality Risk in WM Patients with Wild-Type MYD88 to Those with the MYD88 L265P Mutation** – Due to the low prevalence of the wild-type (unmutated) MYD88 genotype in WM, clinically relevant data are sparse in these patients, with one previous study showing a nearly 10-fold increased risk of mortality in the group. For this study, published in the *American Journal of Hematology*, a large group of patients evaluated at Mayo Clinic from 1995-2016 was assessed for the impact of these genotypes on their clinical course. Of 557 patients, MYD88 status was known for 219 patients, and 174 (79%) exhibited the L265P mutation. The estimated median follow-up was 7.0 years. Median overall survival was 10.2 years for MYD88 L265P versus 13.9 years for wild-type MYD88. The time-to-next therapy from frontline treatment and the presenting features were similar in the two populations. The authors suggest that these findings warrant further validation.

**FDA Grants Accelerated Approval to Acalabrutinib for Mantle Cell Lymphoma** – The US Food and Drug Administration has granted accelerated approval to acalabrutinib (ACP-196 or Calquence) for the treatment of patients with mantle cell lymphoma who have received at least one prior therapy. Acalabrutinib is in the same drug class as ibrutinib but purports to be more potent with fewer off-target

side effects. Approval was based on the results of a Phase II trial of 124 mantle cell patients. The overall response rate was 81%, with a complete response rate of 40%. The median duration of response has not been reached with 15.2 months of follow-up. The median time-to-best-response was 1.9 months. The most common adverse reactions were anemia, thrombocytopenia (low platelets), headache, neutropenia (low neutrophils), diarrhea, fatigue, muscle aches, and bruising. The dosing was 100 mg/twice daily. A Phase II study of acalabrutinib in WM is ongoing.

**Phase III Results Reported for PI3K Inhibitor in CLL/SLL** – Verastem, Inc. reported results from a Phase III clinical trial evaluating the efficacy and safety of duvelisib, an oral dual inhibitor of PI3K-delta and PI3K-gamma, in patients with relapsed or refractory chronic lymphocytic leukemia (CLL)/small lymphocytic lymphoma (SLL). Duvelisib was superior to ofatumumab, which is an approved standard of care treatment for CLL/SLL, with an improvement in median progression-free survival of 13.3 months, compared to 9.9 months for ofatumumab. Duvelisib was also reported in this trial to have a manageable safety profile. Duvelisib is in the same drug class as idelalisib. Verastem plans to file a New Drug Application with the FDA in the first half of 2018 and will explore its use in other cancers.

**New Monoclonal Antibody to Be Tested in Combination with Rituximab for Relapsed/Refractory NHL** – BioInvent International AB is testing its monoclonal antibody BI-1206 in combination with rituximab in a Phase I/IIa clinical trial of patients with relapsed or refractory non-Hodgkin's lymphoma. The clinical trial identifier number on [www.clinicaltrials.gov](http://www.clinicaltrials.gov) is NCT02933320. BI-1206 targets CD32b, an Fc gamma receptor highly expressed on B-cell malignancies and implicated in cancer cell resistance. Preclinical studies have demonstrated its efficacy by preventing rituximab internalization into tumor cells, thereby leaving more rituximab available on the cell surface to exert an anti-tumor effect.

**New Combination Therapy with Ibrutinib to Be Explored in Clinical Trial of CLL and NHL** – The University of California at San Diego will be testing a two-drug therapy combination of ibrutinib (Imbruvica) and cirmtuzumab in patients with chronic lymphocytic leukemia and several forms of non-Hodgkin's lymphoma. This Phase Ib/IIa study will look at both safety and effectiveness. Cirmtuzumab is a humanized monoclonal antibody that targets a protein called ROR1, which is normally expressed by embryonic cells for growth but is abnormally expressed by cancer cells to aid in their growth. The researchers believe that combining the two drugs may provide better potential for eradicating cancer cells, possibly allowing patients to achieve a complete response and stop therapy.



### **FDA Approves First CAR T-Cell Therapy for Lymphoma**

– The US Food and Drug Administration has approved a second CAR T-cell therapy and the first one for lymphoma, in this case, diffuse large B-cell lymphoma (DLBCL). Gilead Sciences is marketing the therapy, called axicabtagene ciloleucel (Yescarta), and has priced it at \$373,000. The treatment carries serious risks, and the FDA is requiring that hospitals and their associated clinics dispensing Yescarta be certified. As part of that certification, staff involved in prescribing, dispensing, and administering Yescarta are required to be trained to recognize and manage the side effects of cytokine release syndrome and nervous system toxicities.

*The following are summaries of selected abstracts about treatments and survival trends in WM patients presented at the 2017 ASH Annual Meeting on December 9-12, 2017, in Atlanta, GA. These and other abstracts are online at <https://ash.confex.com/ash/2017/webprogram/start.html>. For a specific abstract search, type its title in the Search box; for a general search of all abstracts pertaining to or including WM, type “waldenstrom” in the Search box.*

### **Bendamustine and Bortezomib-Containing Regimens Produce Higher Response Rates and More Durable Responses Versus Cyclophosphamide-Based Therapy in Frontline Waldenstrom Macroglobulinemia, Abstract #1488**

– A retrospective study from the Bing Center at Dana-Farber Cancer Institute looked at data from 182 WM patients who received frontline therapy with bendamustine-rituximab (Benda-R), bortezomib-dexamethasone-rituximab (BDR), or cyclophosphamide-dexamethasone-rituximab (CDR) between 2005 and 2016. Of this number, 116 patients also received maintenance rituximab following frontline therapy. Patients treated with BDR were more likely to be less than 65 years old, have serum IgM greater than 4,000 mg/dL, and receive treatment for symptomatic hyperviscosity, while patients with extramedullary (outside the bone marrow) disease and peripheral neuropathy were more likely to be treated with Benda-R and CDR, respectively. For patients treated with Benda-R, BDR, and CDR, the median progression-free survival rates were 5.5 years, 5.8 years, and 4.9 years, respectively. The estimated 5-year overall survival rates among patients treated with Benda-R, BDR, and CDR were 95%, 96%, and 81%, respectively. Maintenance rituximab was associated with a decreased risk of progression, and the median progression-free survival for patients who did and did not receive maintenance was 6.8 years versus 2.8 years, respectively. The authors concluded that all three regimens produce high response rates and durable progression-free survival, but the risk of progression is lower in patients treated with Benda-R and BDR when compared to CDR. Maintenance rituximab was associated with both major and deep responses to therapy, as well as superior progression-free and overall survival.

### **Carfilzomib-Based Combination Regimens Are Highly Effective Frontline Therapies for Multiple Myeloma and Waldenstrom’s Macroglobulinemia, Abstract #1880**

– Researchers from MD Anderson Cancer Center retrospectively looked at both multiple myeloma and WM patients who received carfilzomib as initial therapy at their institution from April 2014 to November 2016. Six WM patients treated with carfilzomib, rituximab, and dexamethasone, followed by maintenance dosing of the same combination, were identified. After a median of 6 cycles, the overall response rate was 66%. Only one patient had severe toxicity, a debilitating neuropathy attributed to rituximab, prompting therapy discontinuation after 3 cycles. At a median follow-up of 33.5 months, overall survival was 100%. MYD88 and CXCR4 mutational status did not seem to impact response to therapy.

### **Long-Term Follow-up of a Prospective Clinical Trial of Carfilzomib, Rituximab and Dexamethasone (CaRD) in Waldenstrom’s Macroglobulinemia, Abstract #2772**

– This update to a clinical trial of 31 symptomatic WM patients treated with carfilzomib, a neuropathy-sparing proteasome inhibitor, along with rituximab and dexamethasone, was provided by the Bing Center at Dana-Farber Cancer Institute. These patients had not been previously treated with either a proteasome inhibitor or rituximab. Thirty patients completed induction therapy, with reduction in bone marrow tumor burden from a median of 60% to 7.5%. Twenty patients also completed maintenance therapy with the same drug combination and had a further bone marrow tumor burden reduction to 2.5%. The best overall response was 80.64%, and the major response rate was 71%. All patients are alive, and 14 patients remain in follow-up without progression. Only one peripheral neuropathy event occurred. This treatment was associated with a significant decrease in IgG and recurring sinus and bronchial infections, necessitating IVIG therapy in 19.4% of patients.

### **Prospective Phase II Study of Ixazomib, Dexamethasone and Rituximab in Previously Untreated Patients with Waldenstrom Macroglobulinemia, Abstract #1487**

– Ixazomib is an orally administered proteasome inhibitor with limited toxicity to the peripheral nerves and is active in multiple myeloma. This study, conducted by the Bing Center at Dana-Farber Cancer Institute, is the first to evaluate ixazomib in WM patients. The drug combination was administered to 26 symptomatic, previously untreated patients, first as induction therapy and then as maintenance. At the end of the study, median serum IgM decreased from 4,528 to 871 mg/dL, median hemoglobin increased from 10.2 to 13.6 g/dL, and median bone marrow involvement decreased from 55% to 5%. Major responses were observed in 69% of patients with CXCR4 mutations versus 82% of those with wild-type CXCR4. The most common adverse events included infusion reactions, nausea, insomnia, rash, neuropathy, edema, diarrhea, and high blood sugar.

**Indications for Hematopoietic Stem Cell Transplantation in Patients with Waldenstrom's Macroglobulinemia: A Consensus Project of the EBMT Lymphoma Working Party (LWP)/European Consortium for Waldenstrom's Macroglobulinemia (ECWM)/International Waldenstrom's Macroglobulinemia Foundation (IWMF), Abstract #2026** – Defining the role and timing of high-dose therapy with autologous stem cell transplantation (ASCT) and of allogeneic stem cell transplantation (alloSCT) in the treatment of WM are major challenges. A panel of international experts was formed to reach a consensus on several of these issues, which the panel did with the following statements: 1. ASCT is not an appropriate treatment option as part of the first line therapy in patients responding to induction therapy; 2. ASCT is an appropriate treatment option following second and subsequent relapses in high risk patients (according to ISSWM prognostic guidelines) with chemosensitive disease; 3. Choice of first line therapy should avoid the use of stem cell toxic agents in transplant eligible patients; 4. ASCT is not an appropriate treatment option in patients responding to and tolerating B-cell receptor inhibitors, provided they are available; 5. AlloSCT could be considered for patients who relapse post-ASCT; 6. AlloSCT should be considered in high risk patients in third or subsequent relapses provided they have received immuno-chemotherapy and B-cell receptor inhibitors; 7. AlloSCT should be considered in patients who are relapsed/refractory to immuno-chemotherapy and resistant to B-cell receptor inhibitors; 8. If B-cell receptors inhibitors are available, alloSCT should not be considered in patients who have not been treated with these drugs; and 9. Treatment decisions for SCT should be based on the clinical course (response and duration of response) rather than on ISSWM prognostic guidelines or on biological factors (such as CXCR4 and other molecular abnormalities).

**Ibrutinib Is Highly Active as First Line Therapy in Symptomatic Waldenstrom's Macroglobulinemia, Abstract #2767** – The activity of ibrutinib in untreated, symptomatic WM patients is not known. Researchers at the Bing Center at Dana-Farber Cancer Institute investigated the activity and safety of ibrutinib in a clinical trial of 30 such patients. All patients expressed the MYD88 L265P mutation, and 14 (47%) had a CXCR4 mutation. The overall response rate was 96.7%, and the major response rate was 80%; 5 patients (17%) achieved a very good partial response, but no complete responses were observed. With a median follow-up of 8.1 months, 2 patients who were CXCR4 mutated met the criteria of progressing disease while on therapy. Overall, treatment was well tolerated, and moderate to severe adverse events included joint pain, bruising, procedure-related bleeding, high blood pressure, muscle cramps, neutropenia (low neutrophils), rash, urinary tract infection, elevation in AST and ALT liver enzymes, foot pain, and thrombocytopenia (low platelets). Three patients (10%) had treatment-related

atrial arrhythmia and continued ibrutinib with medical management. Delays in ibrutinib response were associated with expression of mutated CXCR4.

**Long-Term Follow-up of Previously Treated Patients Who Received Ibrutinib for Symptomatic Waldenstrom's Macroglobulinemia: Update of Pivotal Clinical Trial, Abstract #2766** – This multicenter report presented data from long-term follow-up of patients involved in the groundbreaking Phase II clinical trial that led to FDA, EMA, and Health Canada approval of ibrutinib in WM patients. In the original trial, 63 symptomatic patients who had received at least one prior therapy were enrolled. The median time on ibrutinib at the time of this follow-up study was 46.6 months. Improvements in responses occurred with prolonged treatment, with overall and major response rates of 90.4% and 77.7%, respectively. No complete responses were observed, but 27% achieved a very good partial response. At best response, median serum IgM decreased from 3,520 to 821 mg/dL, median bone marrow involvement decreased from 60% to 20%, and median hemoglobin level increased from 10.5 to 14.2 g/dL. The median progression-free survival for patients with MYD88 mutated/CXCR4 wild-type has not been reached; by comparison, the median progression-free survival was 45 months for patients with MYD88 mutated/CXCR4 mutated and 21 months for those with MYD88 wild-type/CXCR4 wild-type. Three patients died due to disease progression. Adverse events included anemia, atrial fibrillation, GERD (acid reflux), high blood pressure, neutropenia (low neutrophils), pneumonia, skin infection, and thrombocytopenia (low platelets). Four patients came off therapy for toxicity.

**Impact of Ibrutinib Dose Intensity on Patient Outcomes in Previously Treated Waldenstrom Macroglobulinemia, Abstract #623** – This retrospective analysis from the Bing Center at Dana-Farber Cancer Institute examined the impact of interrupted ibrutinib therapy in the 63 patients enrolled in the Phase II trial that led to FDA, EMA, and Health Canada approval of ibrutinib in WM. Fifty patients (79%) held ibrutinib at least once, and there were 102 drug hold events in total. An increase in serum IgM level was observed on 63 occasions at the next assessment after a drug hold. The median increase in serum IgM level was 50%. Following the restart of ibrutinib therapy, the median time to a response of stable disease or better was 125 days. Serum IgM increases persisted longer for patients who were MYD88 mutated/CXCR4 mutated. Holding ibrutinib for more than one week during the entire treatment duration was associated with a shorter progression-free survival. These findings suggest that ibrutinib holds should be minimized and ibrutinib restarted as soon as clinically indicated to achieve best patient outcomes.

**Ibrutinib Discontinuation in Waldenstrom Macroglobulinemia: Etiologies, Outcomes, and IgM Rebound, Abstract #802** – Discontinuation of ibrutinib has been associated with an adverse prognosis in patients with chronic lymphocytic leukemia and mantle cell lymphoma. However, the outcomes following discontinuation in WM patients has not been previously evaluated. The Bing Center at Dana-Farber Cancer Institute identified 189 patients seen at its institution from May 2012 to April 2017 who received ibrutinib therapy, 51 (27%) of whom subsequently discontinued it. Ibrutinib was discontinued due to progressive disease in 27 of these patients, followed by toxicity (15 patients), non-response (5 patients), and miscellaneous reasons (4 patients). A baseline platelet count of less than 100,000 and CXCR4 mutation were associated with higher odds of ibrutinib discontinuation. An IgM rebound was observed in 37 patients (73%) following discontinuation; 6 patients developed symptomatic hyperviscosity requiring emergency plasmapheresis. Patients without IgM rebound were more likely to respond to salvage therapy than patients with rebound. The median overall survival was 32 months following discontinuation, and response to salvage therapy was associated with a decreased risk of death following discontinuation. The authors concluded that patients who must discontinue ibrutinib require close monitoring and that continuation of ibrutinib until initiation of next therapy should be considered.

**Patient-Reported Symptoms during Ibrutinib Holds: A Withdrawal Syndrome, Abstract #4058** – Researchers from the Bing Center at Dana-Farber Cancer Institute identified 189 WM patients seen at its institution between May 2012 and April 2017 who received ibrutinib therapy; of this number 89 (47%) held ibrutinib at some point during therapy. Twenty of the patients who held ibrutinib experienced “withdrawal” symptoms, including fever, body aches, chills, night sweats, headache, fatigue, joint pain, and weakness. The rate of withdrawal symptoms was lower in patients who started ibrutinib with IgM levels greater than 4,000 mg/dL and higher in patients who had achieved a very good partial response on ibrutinib. The symptoms began within 2 days of drug hold and resolved rapidly following re-initiation of ibrutinib therapy.

**MYD88 Mutation Status Impacts Overall Survival and Risk of Histological Transformation in Waldenstrom’s Macroglobulinemia, Abstract #4006** – Mutations in MYD88 and CXCR4 are present in 93-95% and 30-40% of WM patients, respectively. Mutations in MYD88 trigger NF-kappa B dependent growth and survival of WM cells through the BTK/IRAK and AKT/ERK pathways. The mutational landscape of MYD88 wild-type (unmutated) is under investigation and is likely to involve alternate signaling pathways. Given the uncommon incidence of MYD88 wild-type disease, researchers from the Bing Center at Dana-Farber

Cancer Institute investigated the impact of MYD88 wild-type status in a population of 46 WM patients and compared their findings and outcome to 262 patients with MYD88 mutations who were diagnosed over the same time period. The median follow-up for all patients was 74.7 months. During the follow-up period, there were 23.9% and 5.7% deaths, respectively, in the MYD88 wild-type and MYD88 mutated patients. The estimated 10-year survival was 73% and 90% for MYD88 wild-type and MYD88 mutated patients, respectively, and did not differ by CXCR4 mutation status. Transformation to diffuse large B-cell lymphoma occurred in 15.2% and 0.76% of MYD88 wild-type and MYD88 mutated patients, respectively.

**WhiMSICAL (Waldenstrom’s Macroglobulinemia Study Involving CART-wheel): Empowering Patients Internationally to Contribute Patient-Derived Data for Observational Research, Abstract #1502** – Patient-derived data are an attractive option to increase the breadth of knowledge about rare diseases like WM because large trials are difficult to undertake. WM patients are well connected through the IWmf and its affiliates, using the Internet and social media. As a patient group, they are relatively well informed about their disease, its parameters (IgM, hemoglobin), and available treatments. This multi-center international study utilized CART-WHEEL, an ethically approved online rare cancer database for patient-derived data, plus the digital connectedness of WM patients, to develop a patient-derived dataset. At the time of this abstract, 279 participants had completed an online consent form and provided data on symptoms, pathology results, treatments, their tolerance of treatments, and how treatments were accessed. The participants were predominantly from the US, Australia, Canada, UK, New Zealand, and the Netherlands. Fatigue was the most common symptom reported at diagnosis (44% of participants), followed by B-symptoms of weight loss, fever, and night sweats (29%), peripheral neuropathy (21%), shortness of breath (12%), leg cramps (11%), and nosebleeds (10%). Forty different first line treatment combinations were documented, with median time-to-first-treatment for US patients of 48 days, and the rest of the world 122 days. For US patients, 31% of therapies were government funded, whereas in the rest of the world 61% were government funded. Further recruitment of patients and additions to the database, including quality-of-life metrics, are planned.

*The author gratefully acknowledges the efforts of Peter DeNardis, Wanda Huskins, Pavel Illner, John Paasch, Colin Perrott, Howard Prestwich, Charles Schafer, Ron Ternoway, and others in disseminating news of interest to the IWmf Connect community. The author can be contacted at [suenchas@bellsouth.net](mailto:suenchas@bellsouth.net) for questions or additional information.*



## **– Personal Financial Planning –**

### **Finding the Perfect Blend**

**How do you like your coffee? If you are a coffee drinker, you probably like your coffee prepared a certain way. Do you like it scalding hot or iced? Do you have a favorite blend? Maybe you like it sweet and add sugar, milk or cream. However you like your java, hopefully you have found the right mix for what appeals to you.**

In the same way, there is a right blend for you to balance the important priorities in your life. As with the perfect coffee, you can set goals for your personal philanthropy that will make life just a little sweeter.

For example, if you own highly appreciated stock and are concerned about paying capital gains tax, you can benefit by donating some of your stock to support our cause. You will receive an income tax deduction for the value of the stock without paying any gains on the amount transferred. It is important to transfer, not sell, the stock for you to receive the full benefits. By making a charitable gift of stock, rather than a cash gift, you avoid capital gains but still receive an income tax deduction. And you can even use the cash you would have otherwise given to buy more of the same stock at a new, higher basis.

But to make the gift even sweeter, blend your gift of stock today with a gift of additional stock (or other assets) in your will. By giving stock now, you receive income tax and capital gains tax benefits immediately. By adding to your gift later, your giving will become even more powerful (and this additional gift won't change your lifestyle). You can add as much or as little as you want so that your total gift has just the impact you desire. Blend your giving to maintain maximum control with minimal taxes and effort. There are just as many ways of blending gifts as there are ways of making coffee. Call or email us today to learn how you can combine your gifts for maximum impact, benefit today and continue to support our cause in the future.

**By blending your gifts, you can create just the right recipe to provide the tax or income benefits you want today while leaving a greater impact tomorrow.**

### **Sweeten Your IRA**

**Have you ever tried to pour coffee before it is done percolating? The result is usually coffee everywhere! Even if you avoid spills, your coffee just won't taste right if it hasn't finished brewing. There are other times in life when we may be forced to take something sooner than desired. An example of this is the required minimum distribution (RMD) from your IRA. Did you know that once you reach 70½, the government will require you to take distribution from your IRA, even if you do not need the money or might think it better to preserve your IRA for something important, such as a rainy day? The RMD could also substantially increase the taxes you have to pay on your income.**

If you are faced with an RMD this year, consider a better use for the funds. Make a gift of your RMD (up to \$100,000 this year) directly to charity. Contact your IRA administrator and ask for the forms to make a charitable transfer to support our mission. The transfer counts against your RMD, but because you never received the IRA distribution, you will not be taxed on this amount.

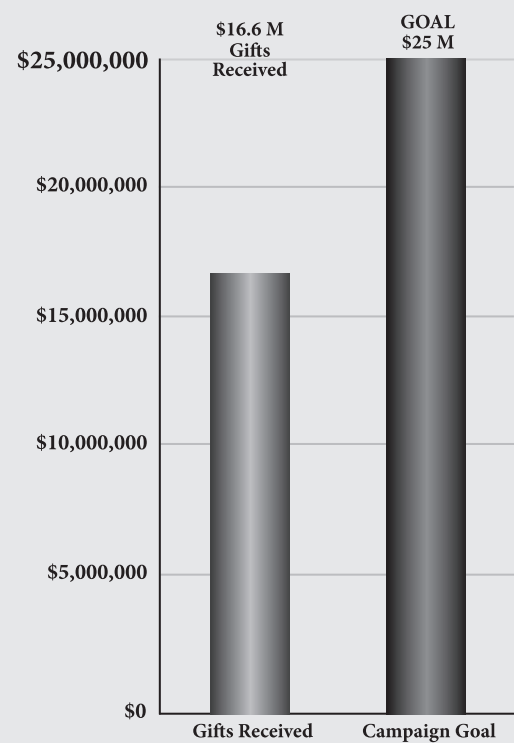
**Rather than pay income tax on your required minimum distribution, transfer the funds to charity instead.**

While this helps with your immediate RMD concerns, consider amplifying your annual gift with a legacy gift. Your IRA rollover gift can be combined with a bequest made in your will or additional IRA beneficiary designation gift to make an even greater difference. By giving directly from your IRA today and supplementing it with a bequest, you can make your giving go further by giving when the timing is right for you.

**If you want your IRA to continue your giving legacy, ask your plan administrator to add the IWMF as a remainder beneficiary on your account.**



**Imagine a Cure Campaign Progress Report  
as of October 31, 2017**



*Many WMers have found blending their gifts to be a recipe for providing the tax or income benefits they want today while assuring a greater impact tomorrow.*



## IWMF DOC STAR: SHIRLEY D'SA

EDITED BY RON TERNOWAY

*"Don't be bamboozled by the fact that Waldenstrom's macroglobulinemia is a rare disease. Many, many, people around the world are working to improve patient outcomes – gaining a better understanding through scientific study and working hard to translate this into clinical benefits."*

A newly diagnosed WM patient at the University College London Hospitals (UCLH) who hears such a positive message has every reason to be encouraged by the dulcet tones of the doctor sitting across the desk. The speaker, Dr. Shirley D'Sa, is one of the United Kingdom's leading WM specialists.

Roger Brown, chairman of the IWMF's affiliate WMUK, has this to say of the role Shirley D'Sa plays in the WM community of the UK today: "Dr. D'Sa is a key member of our WM community. UCLH has grown to be one of the largest WM centres in Europe with 400 patients, and, under Shirley's chairmanship, the UK Doctor Forum has raised the profile of the disease amongst her fellow clinicians from an obscure corner of the rare cancer world to national prominence."

In 2017 WMUK initiated the Rory Morrison Clinical Award in order to honor a physician who has, in the opinion of the UK WM community, gone above and beyond the call of duty for patients. The award was named after Rory Morrison, a hugely popular BBC Radio announcer and founding member of WMUK. Rory, who died in 2013, was treated at UCLH. His widow, Nikki Morrison, was a member of the award selection committee.

Dr. D'Sa was named the first recipient of the award at a WMUK meeting last July. "A hugely popular and fitting choice, and warmly welcomed by the audience," said Brown. "The citation was for 'her tireless work in improving the treatment of Waldenstrom's patients,' and nobody could fit that role better." Two months later, in September, Dr. D'Sa was to play a key part in obtaining funding for ibrutinib for relapsed UK WM patients through the National Health System's Cancer Drugs Fund.

A final observation from Roger Brown: "In a world where physicians, sadly, are often judged by the number of academic papers they produce, Shirley has resolutely focused on improving treatment for patients rather than research-driven projects. At the same time, she has transformed UCLH into a key UK center for WM clinical trials."

Of Indian descent, Shirley D'Sa was born in London but grew up in Uganda and Kenya, where her parents were teachers. "My parents were born in Uganda due to the work of *their* parents during the British Empire. I completed my primary school education in Kenya. Our family repatriated to the UK, and I continued my high school education in Wimbledon, London SW19."



*The Rory Morrison Clinical Award, created in 2017 to honor a physician who has, in the opinion of the WMUK community, gone "above and beyond the call of duty." Dr. Shirley D'Sa is the first recipient of the award. (Photo courtesy of Roger Brown)*

Shirley was interested in science and in people, and, with the positive role models of an aunt and uncle who were doctors, as she puts it: "I liked what I saw – it was no more calculated than that."

After training in Medicine at St. George's Hospital Medical School (University of London), Shirley qualified in 1990, and next came three years of internal medicine and a Membership of the Royal College of Physicians (MRCP) diploma. She then worked at the Royal Marsden Hospital in London in hemato-oncology and decided hematology was to be her specialty. Seven more years of research and training culminated in 2002 when she became a certified specialist in hematology and took up a consultant post at University College London Hospitals National Health Service Trust in central London.

"Finding an exciting clinical and academic niche is hugely rewarding," she said. "Although I started out in myeloma, I have branched out into the management of an interesting and challenging disease, Waldenstrom's, which is a truly multi-faceted disease where, as a physician, there is never a dull moment! Thinking laterally and outside the box is needed all the time. And as biological developments have led to a greater understanding of the disease, I genuinely feel that the future for treating the disease is brighter than ever."

*IWMF Doc Star, cont. on page 18*



“The ongoing developments in the biological sciences which are helping to shape medicine of the future have created a transformative time in hematology like never before. The trick will be to make these advances affordable in terms of application in the real world.

“I am in awe of patients and their carers as they grapple with the disease, often with temerity and bravery, despite the challenges. Working with patients as colleagues in WMUK has been a huge privilege. I am really proud of our notable achievements, most recently working to get ibrutinib on the National Health Service’s Cancer Drugs Fund for WM patients – at last!”

So how does a WM Wonder Woman achieve a modicum of work-life balance? “Even work-work balance is a struggle sometimes, such are the competing priorities!” she ruefully chuckles. “I have three lovely children of whom I am so proud, aged 19, 15, and 11, and wonderful family in Stockholm, Sweden, where we spend a lot of time. My husband’s contribution to family life has enabled me to follow my work and achieve as much as I can.

“We love to travel and also enjoy sports – I joined a tennis club this year. Now that the children are more self-sufficient, I also



Dr. Shirley D'Sa and family.

enjoy fitness activities such as yoga and exercise training. I love listening to amazing BBC radio programs about a huge range of topics which appeal to my philosophical side.”

Final words from Dr. Shirley D’Sa to the *Torch* readership: “In general WM is an obedient disease – it dutifully responds to treatment, and this can restore well-being. Even if the disease behaves out of character or affects unusual areas, strategies exist to overcome these challenges. *Do not be afraid to be hopeful. There is every reason to be so!*”

---

## IWMF CONNECT: WINTER 2018

BY JACOB WEINTRAUB, MD

---

The holidays are past, and the New Year is upon us. Discussion continues unabated, with new and old subjects discussed. Speaking of new subjects, there was even a discussion about whether or not to dye one’s hair during or after treatment! New members joined, and we said good-bye to some old friends.

### HUMAN INTEREST/ARTICLES

IWMF Connect Manager and IWMF Trustee **Peter DeNardis** posted several links of general interest.

One was a link to an interview with **Ali Handal**, a musician who has WM but faces her WM head on and continues to pursue her passions. More about Ali Handal in the October issue of the *Torch*, page 8.

<http://www.thelcn.com/lcn05/ali-handal-is-fearless-sassy-singer-songwriter-takes-on-cultural-taboos-cancer-20171102&template=mlcn>

Another link from Peter was to “Life after cancer: the real battle happened after treatment.” Many of us are quite familiar with much of the content of this article. It presents examples and comments by people affected by cancer and is further proof that we’re not alone in this adventure.

<http://www.latimes.com/science/healthy-living/la-he-hl-life-after-cancer-20171012-story.html>

One more link from Peter related to using humor to deal with a cancer diagnosis. Some examples from the article Pete found funny and others cut “a bit too close to the bone,” but humor is still a valuable coping mechanism. He cited the last paragraph that stated, “Devoid of self-pity, cancer humor proves that raging fear passes, when transmuted through ironic camaraderie – with friends or prospective readers or animals – into emotional clarity. The gift of these creative works: they foster a sense of community with the living and also with the dead. We are not alone in what we go through.” <https://www.nytimes.com/2017/10/12/well/live/cancer-humor.html>

**Wanda H** also posted links to several articles of general interest.

The first was an article entitled “Tragedy, Perseverance and Chance – The Story of CAR-T Therapy.” The article discusses this new breakthrough treatment, which has been in the news recently and discussed on IWMF Connect. While this is a *New England Journal of Medicine* article, it addresses many of the human issues associated with the new treatment. <http://www.nejm.org/doi/full/10.1056/NEJMp1711886>

*IWMF Connect, cont. on page 19*

Another link posted by Wanda was about the psycho-social impact of chemo and how people undergoing treatment may have more psychological issues related to the treatment than to actual physical side effects. This article comes from the European Society for Medical Oncology (ESMO), and the findings were presented at the ESMO 2017 Congress.

<http://www.esmo.org/Conferences/ESMO-2017-Congress/Press-Media/Press-Releases/Patients-Feel-Psycho-social-Impact-of-Chemo-More-Acutely-Than-Physical-Side-Effects>

Wanda also posted a link to an article from the organization CancerCare about the financial impact of a cancer diagnosis. This is based on a survey of patients and family members and discusses medical and nonmedical expenses including gasoline, food, non-prescription medicines, and special clothing and wigs. There are some striking findings, and the survey presents a side of having cancer that isn't often reported.

[https://media.cancercare.org/publications/original/349-financial\\_hardship.pdf?1498075790](https://media.cancercare.org/publications/original/349-financial_hardship.pdf?1498075790)

**Meg M** posted a link to an article entitled "Who Actually Is Reviewing All Those Preauthorization Requests?" She said it was a "real eye opener" for her. *[IWMF Connect Editor's note: Having been through this process myself, I also found this article to be eye opening and guaranteed to raise your blood pressure a few points just from reading it.]*

<https://www.medpagetoday.com/blogs/revolutionandrevelation/69125>

## MEDICARE

This is a subject that comes up periodically as we age and people ask about Medicare and which Medicare supplement plans work best. Although discussion on this subject was presented in the last issue of the *Torch*, there was yet more conversation about different aspects of enrolling and finding the best plan, especially relating to coverage of Imbruvica. This extended discussion also included information about patient assistance programs offered by the Leukemia & Lymphoma Society and by Johnson & Johnson.

**Marcia** commented that Medicare Part A and B do not cover prescription drugs outside a hospital setting. She said that Medicare Part D appears to be the same throughout the country, no matter the insurer. Even with a Medicare Advantage Plan, the cost is essentially the same, and there may be some limits on coverage, so "buyer beware."

**Anita L** posted that the Medicare Part D plans she is aware of all treat Imbruvica as a Tier 5 drug. Because of its cost, the deductible and the donut hole are both satisfied in the first month, so that a patient is out of pocket \$4,000-\$5,000. After that, he or she pays the 5% catastrophic portion, which is about \$550 to \$600 monthly. This is where the Leukemia & Lymphoma Society co-pay assistance and other such programs can help. Anita has Humana Part D, but the four top-rated plans are all similar.

**Linda C** added that she agreed with Anita's post. Her experience is that Imbruvica cost assistance from the manufacturer or from non-profit foundations is based on income. It is worthwhile to investigate. She and her husband were referred to investigate their qualifications for assistance by their oncologist through the specialty pharmacy that provides Imbruvica.

**Steven D** offered a recap and a website to help select Medicare plans. As noted above, Medicare A and B won't cover Imbruvica since it is a prescription medicine. Coverage for such medications is through Medicare Part D, but some Medicare Advantage Plans (also known as Medicare Part C) might also cover these drugs. The following website shows the Part D plans available in a patient's home area and which plans cover Imbruvica at what basic cost. Steven uses this site every year during the open enrollment period.

<https://www.medicare.gov/find-a-plan/questions/home.aspx>

## HAIR

**Ann** asked what she thought might be a frivolous question – does anyone continue to color or bleach hair while on treatment despite being advised not to? She is currently on bendamustine, waiting for the addition of Rituxan and not anticipating a loss of hair.

**Anita L** offered that she was never advised not to dye her hair and continued to have her hair "foiled" for 30+ years. She did this through fludarabine, R-CVP, Velcade/dexamethasone, solo Rituxan, bendamustine/Rituxan, and now nearly four years of Imbruvica. She has had no issues and felt this is not a frivolous topic.

**Mary S** posted that she also has asked this question about hair and has been given different answers by different experts. She has continued to dye her hair throughout the time she has had WM, while on treatment and when she was on watch and wait. She has used products that are supposed to be less toxic. She said there is some research on the association of hair dyes and lymphoma. Many of the newer products are possibly less toxic, and she suggested that Ann check with her own oncologist about this.

**Pavel I** said that his hair doesn't need help to change color. However, since he has been on Imbruvica, his (remaining) hair is getting curly, like it was 60 years ago.

**Amy D** also reported that when her hair grew back after chemo for breast cancer, she had "chemo curls" which unfortunately didn't last long.

## IBRUTINIB/IMBRUVICA

As usual, Imbruvica continues to be discussed. Its scientific aspects have been covered at IWMF Educational Forums and in more formal articles posted on IWMF Connect and in the *Torch*. Here is an aspect that has not appeared in this forum in the past.

**Ron T** asked if anyone using Imbruvica for an extended period has experienced sudden onset of persistent high fevers when the medication has been stopped. Ron was in a clinical trial and had to stop Imbruvica due to a precipitous drop in absolute neutrophil count to below 500. Three days later, he experienced sudden onset of fevers as high as 104°F, which led to hospitalization and antibiotic treatment. No infection was found and the antibiotics were stopped. Tylenol helped to temporarily resolve the fevers. While his fevers persisted after he was released from the hospital, his neutrophil count increased; Imbruvica was restarted and the fevers resolved.

**Karen W** posted that her husband has had to stop Imbruvica twice due to minor surgeries. Each time on day 5 after stopping, he developed fevers of 104°F, loss of appetite, and weakness. He was started on antibiotics the first time, but no infection was found. The second time the fevers occurred, they knew it was due to stopping the medication. His doctor at Stanford is perplexed by this reaction.

**Julie T** had been on Imbruvica for only two months before stopping the drug but experienced “withdrawal” symptoms, including nausea, aches and pains, and headaches, but no fever. She added that the prescribing pharmacy was very interested in the side effects she was reporting.

A similar scenario was reported by a number of others. **Debbie B** said that her husband developed a sudden and scary spike in his liver enzymes. After three days off the medication, he felt horrible with aches and pains, no appetite, and the return of night sweats. When his liver function returned to normal, Imbruvica was restarted, although at a reduced dose, and symptoms resolved.

**Jane P** asked if unusual symptoms like these (perhaps not quite so unusual now) are reported somewhere other than in this forum. She is hoping that as patients share these problems with their doctors, they are recorded somewhere so that other doctors have access.

**Steve D** then posted a link to a Food and Drug Administration site that discusses how to report serious side effects, which also include those from medications.

<https://www.fda.gov/Safety/MedWatch/HowToReport/ucm053074.htm>

Finally, **Ron T** added that Dr. Jorge Castillo at Dana-Farber Cancer Institute has reported that the ibrutinib withdrawal syndrome occurs in about 20% of people who stop the drug. Symptoms include those that have been discussed here, especially fever and other “unpleasant things.”

Just recently we were informed of the passing of four long-time members of the IWMF Connect family: Renee Paley-Bain, Jack Whelan, Richard Orr, and Dudley Killiam.

### FRIENDS WE WILL REMEMBER

**Renee Paley-Bain** was a frequent contributor to IWMF Connect, a LIFELINE volunteer for the IWMF, and a friend to many IWMF members. She was an author in her own right and married to Donald Bain, a noted author and ghost writer. *[IWMF Connect Editor's note: I met Renee in Boston in October 2001 at the First International Patient and Physician Summit on WM, a conference organized by the Bing Center at Dana-Farber Cancer Institute. I had just been diagnosed with WM. Renee was very encouraging and supportive, and she helped me learn about going on with life despite having WM.]* She will be missed by all whose lives she touched.

**Jack Whelan** also was a long-time member of the WM community, volunteered as a Support Group Leader for many years, and served as an IWMF Educational Forum photographer. He also contributed frequently to this group. His final battle actually was with prostate cancer, which had become quite aggressive. He, too, will be missed by many people in this community.

**Richard Orr** was represented in the online community by his wife and caregiver, Barb. He contributed occasionally, but Barb frequently contributed with insightful and compassionate notes, and she worked diligently to try to get the best of care and advice for her husband. Contributions from both will be missed.

**Dudley Killiam** was the caregiver for his wife who has WM. Dudley was very interested in understanding the science of the disease and was known and appreciated for offering his encouraging and supportive posts to others in our group.

\*\*\*\*\*

There is always a much wider range of topics and discussions on IWMF Connect than can be presented in this limited space. You are all invited to join and just “listen” or participate. If you have any discussion topics that you are particularly interested in, please let me know and I will try to include those discussions in a future column. I wish you all good health in 2018!



---

# SUPPORT GROUP NEWS

EDITED BY PENNI WISNER

---

## Please note!

Contact information for all support groups is found on [www.iwmf.com](http://www.iwmf.com) under GET SUPPORT.

Details of support group meetings and other upcoming events are posted under EVENTS. Please check there to confirm details of future events.

## CALIFORNIA

### *Sacramento and Bay Area*

The most recent meeting was held at Kaiser Hospital in Roseville on Saturday, November 4. The group of twelve ranged from a newly-diagnosed person to one who has had WM for 24 years. The group watched a DVD of the “Ask the Doctor” session from the Phoenix Forum, broke for yummy refreshments, and had a fruitful discussion about each other’s progress in dealing with WM.

### *Monterey*

The new group in this area, facilitated by **Suzie Shook**, has met twice – in July and October. Suzie plans the next meeting for January. She began her WM journey in 2008 at age 52 with various symptoms. After a definitive diagnosis of WM was made in March 2009, she started treatment which allowed her to enjoy a long remission until recently. A change in lab results got her active again in the IWMF community so she organized the support group in the Monterey Bay area. Suzie is an operating room nurse who loves her job, and she is optimistic about her long-term prognosis, even as she is getting close to retreatment.

## COLORADO AND WYOMING

On a beautiful crisp fall day, October 14, the group felt honored to have Dr. Jorge Castillo, of the Bing Center for Waldenström’s Macroglobulinemia at Dana-Farber Cancer Institute, visit Denver to speak about WM. Sixty-two patients and caregivers came to hear him, share a breakfast snack, and integrate the newly diagnosed people into the group. About



*The group listened intently as Dr. Castillo spoke about diagnostic criteria and symptoms indicating a need for treatment.*



*Dr. Castillo was available to answer questions at the conclusion of his talk in Denver.*

eight to ten new patients join each year, and all of them love the camaraderie, the knowledge, and the sharing of ideas and hope that the group provides. One of the new patients drove all the way from Santa Fe, NM. Dr. Castillo spoke about diagnostic criteria and what has changed over the last few years. He talked about WM symptoms, whether or not to treat, and emphasized that the decision of when to treat and which treatment to use is unique to every patient. Dr. Castillo reviewed all the current treatments and their potential issues and success rates, as well as new drugs in the pipeline. He also encouraged patients to take advantage of the WM clinical trials available here in Denver, through the Colorado Blood Cancer Institute. Dr. Castillo spoke and answered questions for about 90 minutes. WMers are indeed lucky to have such giving doctors who share their personal time with us so we can become more empowered patients.

## CONNECTICUT

The biannual meeting was held in October in the West Farms Mall community room in Farmington. The meeting began with a 30-minute “meet and greet” session, after which

*Support Group News, cont. on page 22*



*Dr. Castillo speaking to the Colorado and Wyoming support group.*

participants shared individual stories. Fifteen WMers and caregivers attended, including the oldest WM member, age 100, whose story was featured in October in the *Torch*. The next meeting is scheduled for the spring of 2018 with details to be listed on the IWFM event calendar.



Over the past few years, Dr. Sherine Elsawa has taken a special interest in the Chicago support group. We wish her well in her new position at the University of New Hampshire. We will certainly miss her!

## ILLINOIS

### Chicago Area/SE Wisconsin

The group welcomed Dr. Sherine Elsawa back as the speaker for the final 2017 meeting at Advocate Lutheran General Hospital in October. After doing WM research at Northern Illinois University, she has recently moved to the University of New Hampshire to continue her research and teaching. She is a special friend of WMers and answered questions for the attendees about her recent research, as well as our immune system issues. It is always a privilege to see doctors who obviously care for the patients of this rare disease. During the break, there was time for more personal discussions. Stay tuned for the next meeting in the spring. Since the IWFM Educational Forum is in Chicago in May, the group's program committee is discussing whether to meet in April or just focus on attending the Forum at the Westin Hotel in Rosemont, IL.



The Chicago support group met at Advocate Lutheran Hospital for the talk by Dr. Elsawa.



Back stage setting for the Chicago support group at Advocate Lutheran General Hospital: a tempting buffet, plenty of hand sanitizer, and a concern for recycling.

## INDIANA

Dr. Rafat Abonour sure knows how to draw a crowd. Twenty-eight people – the largest meeting so far, including seven first-time attendees – found their way to the Leukemia & Lymphoma Society (LLS) conference room to hear about Waldenström's and WM treatment. Dr. Abonour is an oncologist at the Melvin and Bren Simon Cancer Center at Indiana University Medical Center, Indianapolis. He discussed WM's various presenting symptoms, available treatments, and how the different treatments work. He stressed, and patients have heard it before, that one treats the symptoms, not the numbers. Despite this advice, he reminded the group to be aware of three numbers and why: IgM, hemoglobin, and, at least once a year, LDH, a test that measures damage in tissue, muscle, and blood (an indicator of potential problems). Dr. Abonour also talked about WM as an indolent disease and the fact that many WMers are long-term patients. The support group does not meet in the winter months because of anticipated inclement weather but plans to meet again in early spring.

## EASTERN OHIO, WESTERN PENNSYLVANIA & WEST VIRGINIA

A small group of WM patients and family members met on a lovely September afternoon at the home of **Marcia and Glenn Klepac** in Pittsburgh, PA, to catch up with each other and warmly welcome a new member. Members shared personal stories and basic WM information, all valuable pieces of the WM puzzle to help the new member along the path of understanding and support. Delicious potluck contributions of appetizers, salads, desserts, and drinks sparked a spirited conversation of the challenges in living with this rare disease. A satisfying afternoon for all!

## PENNSYLVANIA

### Philadelphia

On September 17 Dr. Mary McMaster gave an excellent presentation to the group about Familial Risks of WM. Dr.

*Support Group News, cont. on page 23*





*Dr. Mary McMaster's talk in September about familial risks of WM drew a good turnout of very interested listeners.*



*In November, Philadelphia support group members enjoyed warmth, cheer, information, and friendly support appropriate to the season at the home of Lisa Wise. The turnout was impressive, and so, too, were the refreshments!*

McMaster is a leading expert on this topic and a senior clinical specialist at the National Cancer Institute. Dr. McMaster was instructive, answered many questions, and gave a thorough explanation of the genetics of cancer and WM.

In November the group assembled for the third time at the home of **Lisa Wise** for warmth, cheer, information, and friendly support. This annual meeting at Lisa's home is the highlight of the year. The group congratulated Lisa on her recent election to the IWFMF Board and thanked her for her work as Chair of the IWFMF LIFELINE committee. Then Dr. Stephen J. Schuster, director of the Lymphoma Program at The Abramson Cancer Center at the University of Pennsylvania, gave an outstanding talk on the history of cancer treatment. He described in detail Kymriah, the first FDA-approved CAR-T cell immunotherapy for acute lymphoblastic leukemia in children. He also described his current trials for CAR-T cell therapy in lymphoma patients who have no alternative therapy options. The trials are showing positive results and are well received. Dr. Schuster's excitement was infectious: "Every day is a discovery and has me jumping up and down; this stuff [that we are learning] is mind blowing. Every day is an epiphany; I don't know why every kid isn't going into science..."



*The Northern Virginia/Washington DC/Western Maryland support group heard Dr. Mary McMaster (center) of the National Cancer Institute speak in October about predisposition to Waldenstrom's macroglobulinemia.*

### **NORTHERN VIRGINIA/WASHINGTON DC/ WESTERN MARYLAND**

In a meeting co-sponsored with the Leukemia & Lymphoma Society (LLS) in October, 40 WMers and caregivers gathered to hear Dr. Mary L. McMaster, senior clinical specialist at the National Cancer Institute, speak to the group about predisposition to Waldenstrom's macroglobulinemia. This was Dr. McMaster's second visit to the group; she spoke at the inaugural meeting three years ago, also in October. Attendees were encouraged with the hope of a better tomorrow with WM, and over the years, several group members have participated in her studies. The group is so appreciative of her interest in WM as well as her long years of dedicated work. **Bonnie Beckett** and **Chuck Ross**, members of the IWFMF Advocacy Committee, also attended the meeting. Bonnie offered an inspiring report about how all IWFMF members can make a difference with cancer care and funding of research for NIH and other organizations. She provided information on, and links to, bills now before Congress that could affect the availability and affordability of drugs. She encouraged the attendees to visit the offices of their senators and representatives in Congress and also to make phone calls on behalf of cancer care and research. Bonnie has also started posting this information on IWFMF Connect so that other US members can make their views on these bills known.

### **WASHINGTON**

Fifteen members of the support group attended the Lymphoma Research Foundation workshop on November 11 in Seattle, many mainly for the WM session. As the session speaker was unable to attend at the last minute, we had a very enjoyable and useful meeting among ourselves and welcomed some new members to the group. We are now gearing up for Dr. Steven Treon's visit on Saturday, January 13, with a symposium which will also include University of Washington Medical Center Neurologist Dr. Barbara Jane Distad speaking on peripheral neuropathy and local experts discussing healthy living with WM. See the IWFMF website for event details.



---

# CROSSING CONTINENTS FOR WM EDUCATION

BY CHRISTOPHER PATTERSON, ADMINISTRATIVE DIRECTOR  
OF THE BING CENTER AT DANA-FARBER CANCER INSTITUTE

---

In these exciting times of new genomic findings and drugs to treat Waldenstrom's macroglobulinemia (WM), many clinicians outside of North America are eager to learn about new ways to manage their WM patients. WM has become a focal point for many national and international hematology and oncology congresses, and many institutions abroad have incorporated WM into their teaching programs.

Dr. Steven Treon, Professor of Medicine at Harvard Medical School and Director of the Bing Center for WM at Dana-Farber Cancer Institute, was invited to speak at many of these meetings in Europe and Asia this past fall. Starting in September in Germany, he chaired a congress on new drug development for WM in Berlin. He then presented "grand rounds" at the University of Würzburg, Germany, where innovative drugs and immune therapies are being developed for the treatment of plasma cell malignancies under the leadership of Professor Hermann Einsele. "Grand rounds" are an important teaching tool for doctors, residents, and medical students and consist of a discussion of the medical issues and treatment of specific patients, in this case patients with WM.

From there Dr. Treon went to Vienna, Austria, where an international congress focusing on new drug development for WM and other B-cell diseases was being held. A week later, he was back in Europe, this time in Prague, the Czech Republic, to speak to physicians from Baltic and Eastern European countries on genomic and treatment breakthroughs on WM. The conference was sponsored by the International Myeloma Foundation.

The next week, Dr. Treon went to Ireland for a three-day visit to University Hospitals in Dublin, Galway, Cork, and then Belfast, where he gave a state-of-the-art keynote lecture on October 13 entitled "Genomic and Treatment Advances in Waldenstrom's Macroglobulinemia" at the Annual Meeting of the Hematology Association of Ireland (HAI). Hematologists-in-training at the HAI joined Dr. Treon the next day for a master class devoted to the management of WM patients and challenging WM cases. Case discussions focused on use of the MYD88 and CXCR4 molecular markers for treatment

decision-making, choice of therapeutic regimens, treatment-related complications, and management of patients with peripheral neuropathy and Bing Neel Syndrome.

Tokyo was the destination of Dr.

Treon's travel the following week, where he was invited to speak at the 79th Annual Meeting of the Japanese Society of Hematology (JSH). At a workshop on WM, Dr. Treon provided an update on findings made possible by whole genome sequencing of WM, including mutations in MYD88 and CXCR4 and their impact on treatment decision-making for WM. He also discussed current rituximab-based therapies, including the use of bendamustine and proteasome inhibitors, and the role of ibrutinib in the treatment of WM. The session was chaired by Dr. Michihide Tokuhira of the Department of Hematology at Saitama Medical Center, Saitama Medical University. That afternoon, Dr. Treon spoke in the General Session of the JSH, chaired by Professor Kiyohiko Hatake, chief of the Clinical Chemotherapy Center at the Japanese Foundation for Cancer Research, and provided an overview of treatment advances being made possible from whole genome sequencing of WM patients.

From this glimpse into a portion of Dr. Treon's 2017 itinerary, it is easy to see that physicians and their institutions on a global level are showing a strong and an increasing interest in WM learning and then sharing this information with their colleagues through lab research, clinical trials, and patient advocacy. These meetings have always been (and will always be) the keystone to improving physician education and knowledge and producing better outcomes for patients, all of which are important goals for Dr. Treon and other experts in the field.



*Professor Treon speaking to delegates of the 79th Annual Meeting of the Japanese Society of Hematology.*

## Have Your Say

The *Torch* welcomes letters, articles, or suggestions for articles. If you have something you'd like to share with your fellow WMers, please contact *Torch* editor Alice Riginos at [ariginos@me.com](mailto:ariginos@me.com)

---

# INTERNATIONAL SCENE

EDITED BY ANNETTE ABURDENE

---

## AUSTRALIA

### **WhiMSICAL database support by IWMF worldwide.**

The IWMF and its global affiliates have driven a fourfold growth in membership of WhiMSICAL database. Participation has grown from 69 to 280 members in the six months to November 2017. Multiple social media platforms have been used including online discussion forums, Facebook, email mailing lists, and community newsletters. The international expansion has confirmed WhiMSICAL CART-WHEEL database as a robust global data-collection platform for WM patient-derived data.

The WM patient questionnaire completion has continued to be good in providing data on symptoms, pathology results, treatments, patient tolerance, and how participants were accessed. The continuously expanding patient-derived dataset is utilized by the principal investigators of the study to provide a foundation for hypothesis generation around WM. Further recruitment and continuous data entry will give an increasingly robust body of data, helping to increase knowledge of the range of presentations and treatment experiences of WM patients. WhiMSICAL has the potential to map real-world therapeutic efficacy, along with international patterns of treatment access.

Initial analysis of real-world clinical data on presentations and treatments from participants in the WhiMSICAL project was submitted to the American Society of Hematology (ASH) annual meeting held in December in Atlanta. None of the research would have been possible without the WM patients who have contributed their demographic data, tumor type details and gene testing, family cancer gene testing, reports from clinicians, treatments, side effects, and other psychosocial and family information via the CART-WHEEL database. Increased participation in WhiMSICAL will provide the “big data” required to make a major impact to demonstrate how truly informative this WM research can be.

Additional WM clinicians who have joined the WhiMSICAL database team as authors of the ASH abstract are: Dr. Marie Jose Kersten of the Netherlands; Dr. Ruth Spearing, New Zealand; Dr. Lia Palomba and Dr. Sheeba Thomas, USA; Dr. Shirley D’Sa, UK; and Dr. Loic Ysaebert, France.

### **Leukaemia Foundation provides WMozzies new member recruitment assistance Australia-wide.**

Leukaemia Foundation blood services coordinators are assisting in recruiting new members for WMozzies in 20 locations around Australia. WMozzies membership is only 10% of the estimated number of people in Australia with WM. The Leukaemia Foundation professional staff is assisting by distributing the new WMozzies brochure to WM patients with whom they are in contact. Leukaemia Foundation blood



*Light the Night 2017 in Sydney: Light the Night is a unique annual fundraising event sponsored by the Leukaemia Foundation of Australia.*

services coordinators are continually visiting all major centres throughout Australia where WM patients are treated.

### **WMozzies support for Leukaemia Foundation’s Light the Night annual fundraising.**

WMozzies in Sydney had the eighth largest turnout in the Light the Night fundraiser in Australia. The 2017 annual event raised nearly \$1.7 million Australian dollars. Light the Night is a unique fundraising event bringing Australia’s blood cancer community together in more than 140 locations to remember and reflect during a moving ceremony and short lantern walk. Raising money at Light the Night helps provide blood cancer patients with life-changing practical and emotional support in local communities right across the country, from diagnosis through treatment and beyond. Services offered include free transport to medical appointments, counseling, information, education, and practical support to people when they need it most. Families from regional Australia may have free accommodation in capital cities so they can access life-saving treatment at major hospitals.

Details of how the money raised is used are provided in the Leukaemia Foundation’s annual financial reports at: <http://www.leukaemia.org.au/about-us/annual-reports>

**Andrew Warden, WMozzies, reporting.**

## CANADA

WMFC participated in September’s Blood Cancer Month for the first time to raise awareness of WM and thank all those who sent gifts in support of WM research.

The WMFC welcomes **Lisa Skoric** as a Board Member. She joins **David Johnston**, Treasurer, **Betty McPhee**, Vice President and new Support Group Liaison, **Raffaella Mercurio**, who now monitors and seeks new content for our website, **Ian Roberts**, and **J.A.Clancy**.

We also want to thank **Rebecca Hinchcliffe** who has been for the past year responsible for our digital marketing and is

*International Scene, cont. on page 26*

now expanding her role to also include some administration, website maintenance, and event planning. She has been a great asset to the foundation established in memory of her grandfather, Nick Carrick.

As we embark on a new year, we reflect on the past years and how far we have come in understanding this rare cancer known as Waldenström's macroglobulinemia and encourage you to continue to support our efforts in the future.

Registering with the WMFC is an important step for those living with WM in Canada as it greatly improves the visibility of WM within the Canadian community for funding of treatments and research. As a rare cancer, this is vitally important. Some benefits of registering and becoming a member of the WMFC include a 25% discount to Canadian educational forums, first-to-receive access to new abstracts, updates, and research, as well as connecting with others in your community living with WM.

A quote from Milton Olson: "As each bird flaps its wings, it creates 'uplift' for the bird following. By flying in a 'V' formation, the whole flock adds 71% greater flying range than if the bird flew alone."

Lesson: People who share a common direction and sense of community can get where they are going more quickly and easily because they are travelling on the thrust of one another.

Take care.

**Arlene Hinchcliffe, President, WMFC, reporting.**



*In October the Nova Scotia/New Brunswick Support Group held a luncheon meeting hosted by Jim and Jill Mason in Halifax, Nova Scotia. Twenty patients and caregivers attended. We were very pleased to welcome from Toronto Betty McPhee, WMFC Board Member and Support Group Liaison for Canada.*

### **From the Halifax Support Group**

On Saturday October 21 the Nova Scotia/New Brunswick Support Group held a luncheon meeting hosted by Jim and Jill Mason in Halifax, Nova Scotia. Twenty patients and caregivers were in attendance for this semi-annual meeting, including four first-timers from as far away as Saint John, New Brunswick (approximately a four-hour drive, one way). We

were very pleased to welcome from Toronto **Betty McPhee**, WMFC Board Member and Support Group Liaison for Canada. The agenda consisted of a summary of the October 4 Cancer Care WM webinar from one of our members, WMFC update and information-sharing from Betty, and the ever-popular sharing and discussion of our own individual stories.

During the meeting we also discussed the possibility of hosting an educational forum in Halifax for the fall of 2018. There was enthusiastic support to pursue this endeavour. A committee will be formed, with more information to follow. The meeting adjourned after three hours. Some members left to participate in the Leukemia & Lymphoma Society of Canada Light the Night event.

**Betty McPhee, WMFC, reporting.**



*At the meeting of the Finnish Waldenström's macroglobulinemia patients, Dr. Pekka Anttila spoke to a full room. The Waldenström patients in Finland are a small but well-informed group.*

## **FINLAND**

This year the Finnish Waldenström's macroglobulinemia patients' group had an opportunity to get together at a two-day event organized with the support of the Association of Cancer Patients in Finland. The event was held at the Scandic Grand Marina hotel and Marina Congress Center in Helsinki, a fine location for the weekend's activities.

The venue is one that caters to large happenings of various kinds, and the arrangements were taken care of accordingly. Present were not only the Waldenström's group of patients, but patients from many hematologic cancer patient groups: myelomas and myeloproliferative diseases, chronic lymphocytic leukemia, and lymphomas. The family members of attending patients were also invited to join in the two-day program of lectures and discussions for the separate patient groups.

The myeloma patients had a very special guest lecturer: Dr. Rafat Abonour, Professor of Medicine, Indiana University School of Medicine, where Dr. Abonour is medical director of the bone marrow transplant program and medical director

*International Scene, cont. on page 27*





*The venue for the meeting of the Waldenström's macroglobulinemia patients' group was excellent, but the weather in Helsinki was cold and the sea breeze felt more like a hurricane. We were happy to stay indoors in the hotel and the congress center, enjoying the view of the passenger terminal from the restaurant windows.*

of the stem cell laboratory. In addition, there were six Finnish hematologists presenting on the new findings in their fields of study. All lectures had time set aside for patients' questions and discussion.

For Waldenström patients it was a special treat to meet with Dr. Pekka Anttila. He is currently one of the foremost Finnish researchers on the subject of Waldenström's macroglobulinemia, working at the University Hospital of Turku. Dr. Anttila spoke about recent developments regarding treatments and then had time for our questions. He showed genuine interest in the patients' points of view about the various treatments that we have had.

During the two days there were times set aside for patient group discussions on various topics; we got to pick and choose the groups that were of particular interest to us. The feeling that we came away with was that we are survivors all – and not simply surviving but living well while facing our various challenges.

For me personally, this was an event I attended as a WM patient in remission, living a full and busy life. In addition, the occasion was very special because I could attend this event with my son, who has survived Precursor T-cell Lymphoblast Lymphoma after a life-saving allogeneic stem cell transplant from his brother.

**Taina Lukkaroinen, Waldenström Finland, reporting.**

### FRANCE

Seventy-four participants from all over France gathered on September 23 in Limoges at the initiative of Waldenström France. This patient-doctor day was organised by Hematolim, the hematology network of Limousin. Hematolim provided us with a very well organised program, and the meeting also took place in a beautiful conference room.

The morning was reserved for participants to gather and meet up with both old and new acquaintances – and there



*The patient-doctor day in Limoges sponsored by Waldenström France drew 74 participants from all over France*

were numerous new faces this year. A convivial meal followed. The afternoon program was quite busy: Professor Arnaud Jaccard, head of the hematology and cell therapy department and specialist in amyloidosis, gave us a detailed presentation of Waldenström's disease and the advances in research. Dr. Mohamed Touati, hematologist and president of the Hematolim network, and Professor Jean Feuillard, MD, PhD, of the hematology laboratory, Centre Hospitalier Universitaire de Limoges, enlightened us on the role of analysis and the "mystery" of electrophoresis in particular. Throughout the session patients had the opportunity to ask their questions of each of the speakers.

A novelty this year was a practical workshop based on a new tool for entering test results and monitoring the changes in the state of one's health. This was recently created by a member of the association and tested by a whole group of enthusiastic volunteers.

The day was a great success thanks to the quality of the program, as well as the warmth of the meetings between patients.

The various presentations were recorded and are available to members of the association on the Waldenström France website: <http://portail.waldenstromfrance.org>

**François Soulié, Waldenström France, reporting.**

### INDIA

To borrow a phrase from cricket that is popular in India: we've been off to a measured and solid start. In August this year, I helped organize a support group meeting in the city of Bangalore. Two patients attended this meeting: Ms. Jaya Mani and my mother, Mrs. Rajini Seroo. We also had another supporter attend, whose mother, like mine, is a WM patient. At present we have been able to establish contact with over 50% of the patients on our list. After writing to two doctors (Dr. Amit Rauthan, an oncologist at Manipal Hospital in Bangalore, and Dr. Sunil Parekh, a hematologist at Bombay Hospital) and meeting a third (Dr. Sharat Damodar at the Mazumdar Shaw

*International Scene, cont. on page 28*

Cancer Centre in Bangalore) we have tentatively decided to meet as a group at the Sixteenth International Myeloma Workshop in New Delhi. Dr. Damodar is working with his colleagues to conduct a session on WM at the Workshop. We are also planning a session where WM patients, supporters and caregivers can meet for a one-day event during the same time frame, most probably in Bangalore. Dr. Damodar has also connected me with a few more doctors in the city, and I plan to meet them over the next few weeks.

#### **Saurabh Seroo, W-M India, Reporting.**

#### **UNITED KINGDOM**

“It was the best of times, it was the worst of times,” to quote Charles Dickens in *A Tale of Two Cities*, words which might well sum up this report. Good news was final ibrutinib approval via the Cancer Drugs Fund, two and a half years since EU licensing, and the first ten relapsed patients have been treated. This triggered the treatment data collection scheme funded by Janssen UK into the Rory Morrison Clinical Registry, via major WM hospital centres. I cannot thank enough all those who have contributed to this victory in the face of austerity – fellow UK Blood Cancer Alliance members, Lymphoma Canada, the IWMP, and Janssen UK who have bent over backwards to make ibrutinib available to the WM community. I must also thank many in our community who filled in our questionnaire for submission to our health service to demonstrate unmet need for better treatment.

We aim to expand the Registry project to more sites than the six or more already volunteered and to collect complete retrospective data, but this needs substantial fundraising to pay for data entry. Patient power is also being used to “encourage” consultants not enrolled to join; UK patients are very vocal at having their data collected for research!

With tacit Registry endorsement by the health regulator (no mean feat), we have a pathway to assist future approvals for other WM targeted drugs in the UK. On that front, we await acalabrutinib trial results, where UK involvement was significant, plus more information on venetoclax and ixazomib from the upcoming meeting of the American

Society of Hematology where several of our specialists will attend. Meanwhile, we are encouraged by recruitment to the BGB-3111 versus ibrutinib clinical trial at eight UK sites.

The bad news has been a spate of deaths of longstanding WMUK supporters, particularly of Gill Bennet and Phil Manning, for whom these new treatments came too late. Phil was a founder trustee of WMUK and did great things here and in Europe to raise the profile of WM. He had his transplant at the same time as me, making mortality more vivid! WMUK treads a fine line – trying to be optimistic to the newly diagnosed whilst at the same time knowing that this is a nasty disease, with the UK having some of the lowest survival rates in Western Europe.

Back to the positive, WMUK led the way at a major event celebrating Blood Cancer Awareness Month in September. Eight volunteers had giant names and their WM wishes displayed in Paternoster Square, next to St Paul’s Cathedral. The display was viewed by thousands and attracted much media attention. Our Patron, Charlotte Green, launched the event. There were 104 names, representing the number of UK people diagnosed daily with blood cancer.

Our fundraisers are ever more creative – recently including relay-swimming the English Channel, trekking to Everest Base Camp, rallying to Mongolia in a beaten-up car, and cycling the height of Everest up the hills of Bath. We welcome bizarre challenges! Our highlight event is the annual Bake4 Rory Day at the BBC before Christmas, where Rory’s friends consume large amounts of cake and last year raised £4,000. This year, we hope to draw in more of the WM community to support the day on social media and are busy selling unique Rory baking kits of apron, spoon, and tea towel.

By the time you read this we should have announced the venue for our big 2018 meeting, probably in London. Details and booking will be at <http://www.wmuk.org.uk/>

**Roger Brown, WMUK, reporting from: The Larder at Butlers Retreat, Epping Forest. (It’s a real place: check it out!)**

### **IWMP Publications**

*Torch* readers who receive their copy by mail will have found two very useful printed inserts upon opening this first issue of 2018.

The handout IWMP Publications shows a complete listing of the publications currently available, together with a convenient display of the languages into which each publication can be downloaded, including English, Chinese (traditional and simplified), German, Spanish, French, Italian, and Norwegian.

The Publications include Booklets, Treatment Options Guides, and Fact Sheets.

Don’t let this important and informative insert slip out and fall to the floor when you open your issue 19.1 of the *Torch*! Hold on to it for future reference!

**Let’s face it: the time comes for each of us when we need to reach for an IWMP publication.**

---

# COOKS' HAPPY HOUR

BY PENNI WISNER

---

It's once again beta-carotene season and I am thinking about pumpkins and squash. Maybe you bought some to use as home décor. Don't throw them out! One year I rescued a couple of 10-pounders from the sidewalk, brought them home, and cooked them. That supply lasted a year!

But perhaps you have noticed that delicata squash is having a "moment." Sometime in October, I start keeping an eye out for recipes that might be fun to cook for the various fall and winter holidays. Some of these, if I get around to trying them, might enter a more permanent rotation. This year several featuring delicata squash caught my eye. They are pale yellow with thin green lengthwise stripes, skin delicate enough to eat (no peeling needed!), and a mild, sweet flavor.

As you read this in January, we might all be ready to give up the rich and sweet foods that larded our holiday feasts. And maybe our waistlines. But we want – at least I do – winter foods with lots of flavor that continue to satisfy not just bodily hunger, but also that deeper hunger for comfort and care. So I will only briefly mention the rich custard, blue cheese tart studded with roasted delicata.

Instead, I encourage you to try this: look for smallish delicata and slice off the tops and bottoms. Use a melon baller to scoop out the seeds. Mix together a stuffing of cooked grain such as rice or quinoa or barley, steamed and roughly chopped kale, currants or dried cranberries, a little cooked onion or shallot, and chopped almonds. Season well with salt, pepper, and plenty of herbs such as Herbes de Provence, za'atar, or even some curry powder. Add a little chile flake or Spanish smoked hot paprika if you like. Stuff the squash with the filling and stand them up in a baking dish. Drizzle with olive oil and a little more salt and pepper and bake in a preheated 375-degree F oven until tender, about 30-45 minutes.

I have also noticed delicata showing up on restaurant menus, particularly in salads. First the squash is roasted, which is easily done. Cut the squash in half lengthwise, scoop out the seeds, slice the vegetable into half-moons about 1/3-inch

thick, drizzle with olive oil, and season with salt and pepper and any dried herb blend you have around (this last is optional but tasty). Roast the squash at 375 degrees F until tender and browned. Flip the pieces once during roasting so they brown on both sides. Add the squash to salads made with winter greens such as spinach, kale, or dandelion greens.

I have been known to stand at the stove and eat a whole batch of the roasted squash. They make a terrific "cook's snack." However, if I can get them to last long enough, my goal is to mix the squash with roasted mushrooms, poblano chiles, and onions and use the mix as a stuffing for tacos with a dried chili salsa and a crunchy cabbage garnish. And maybe I would add a little crema made from plain kefir seasoned with salt, pepper, and pressed garlic.

One more pumpkin idea: A few years ago my neighbor, who cooks several times a week for her vegan sons, gave me a recipe for a pumpkin-coconut curry that I have continued to make and evolve. My partner, who likes neither squash nor coconut, loves this. Heat a few tablespoons olive or canola oil in a large Dutch oven over medium heat. Add 1 large finely chopped onion, 1 teaspoon or more finely chopped fresh ginger, 3 large minced garlic cloves, 1½ teaspoons (more or less to taste) ground cumin, 1 teaspoon turmeric, and 1 tablespoon tomato paste and saute until onions are soft and translucent. Add 4 cups water, 10 ounces red lentils, 1 medium butternut squash (peeled and cut into large cubes), 1 small can peeled tomatoes, and 1 can coconut milk. Stir well. Then add 2 teaspoons curry powder, a pinch cayenne, and several dried hot red chiles or chile flakes to taste. Bring to a boil and cook until the lentils have cooked into a mush and the squash is very tender. You may need to add more liquid as the soup cooks. Before serving, remove and discard the chiles, add salt to taste, and puree with an immersion blender. A nice garnish is a sprinkling of the middle-eastern spice mix dukkah (or slivered, toasted almonds) and julienned fresh mint.

*Our motto: Eat Well to Stay Well*

## IWMF Ed Forum 2018

**The wait is over! Your 2018 IWMF Educational Forum brochure with registration form is also enclosed!**

Planning is well underway, and it's not too early to register for the popular annual event, to be held this year on May 18-20 in Rosemont, IL.

Information is included on some special options to take place during this exciting and educational weekend, along with instructions for making your room reservations at the Westin O'Hare Hotel, which is very convenient to Chicago O'Hare Airport.

The Educational Forum is a unique opportunity for WMers to come together in a very special way and learn from the medical experts and from each other. There's nothing else quite like it!



**BETWEEN SEPTEMBER 1 AND NOVEMBER 30, 2017, THE FOLLOWING CONTRIBUTIONS TO THE INTERNATIONAL WALDENSTROM'S MACROGLOBULINEMIA FOUNDATION WERE MADE IN MEMORY OF:**

**Mrs. Elaine Berkowitz**  
Noah and Phyllis Fields

**Charles S. Bryan**  
Dorothy Bryan  
Mildred Cox  
Winston M. and  
Marion O. Ditmars  
Morris and Marilyn Fabian  
Alice Fisher  
Andrea K. Higginbottom  
Marty and Steve Janny  
Donald and Patricia Matthews  
Edwin and Doris McGwire  
Erin Meyer  
John and Patricia Millar  
William Millar  
Jay and Jayne Neary  
Paul and Elsie Nicolette  
Thomas Paris  
Wendy Pfeffer  
Richard and Barbara Pratt  
Joan Raymond  
Edwin and Shirley Sandford  
Paula Schwalje  
David and Ann Van Wagoner  
John W. Van Wagoner

**Rocco Canaza**  
William and Joy Howe

**Gloria Dillan**  
Patricia and Raymond Bologna

**Ron Draftz**  
Sue Herms

**John Eldridge**  
Anonymous

**Theodore Evanish**  
Patricia Franks Evanish

**David Fellows**  
Bobbie Krueger  
Norma Fienberg  
Beth Black Barnette  
Arlene and Keith Bronstein  
Jerry and Renee Cohen  
Linda and Wayne Goldstein

**David Fellows (cont.)**  
Bradley and Diane Mautner  
Ginger and David Russ

**Richard Freeman**  
Ms. Merle Weston

**Patrick Fulton**  
Joan Fulton  
Volney and Paula Julianel

**Elizabeth Gangale**  
Judy and Bob Wagner

**Tom Hanson**  
Virginia Hanson

**David Hill**  
Ken and Nadine Dale

**Eunice M. Hoyle**  
Carol Hoyle Ballard

**K. Edward Jacobi**  
Ellen Peifer

**Kevin Kennedy**  
Walter and Diane Kennedy

**Dudley Killam**  
Vinh Bui  
Sue Herms  
Alan Kirschbaum  
Ann Pert  
Richard and Mary Wendt

**Terry Kilsby**  
Kevin Kilsby

**Bonnie Koch**  
Charles Koch

**Bryan Lauch**  
Susan and Carl Rentschler

**Don Lindemann**  
Ellen Smith

**Ann Lindsay**  
Margaret P. Stevenson  
Foundation

**Charles Lyons**  
Jim Robinson

**Phil Manning**  
Carl Harrington

**Alfred Mathurin**  
Diane Mathurin

**William S. McIlrath**  
Inge and Geoff Brieger  
Robert B. Clement  
Lucia and Henry Heingold  
Pauline McIlrath

**Mary Ellen Mohr**  
Steve Mohr

**Raymond Nadolny**  
Patti and Dave Oilar

**Peggy O'Donnell**  
Kathryn Gurland

**Donna Olson**  
Kim Cain  
Michael and Lisabeth  
Hock Coakley  
Katelina Galiotti  
Hemant Keskar  
Michele Rambasek  
David A. and Susan M. Roback

**A. Edward Ooghe**  
Donna Ooghe

**Roy C. Parker**  
Eileen Parker

**Jo Ann Parrish**  
Richard and Karen Clark  
Meredith and Matt Connely  
Jamie McDonald  
Candace Nicole Salmon

**Don Pennington**  
Mick and Elaine Denning  
Michael and Kathryn Donohue  
Linda Phillips  
Dave Stokey  
Tobin and Pam Taylor

**Carl Petersen**  
Mary Petersen

**Edwyn Dale Peterson**  
Gary Alexander  
Kathleen Lewis Beard

**Karen Pindzola**  
John and Gloria Albert  
Tony and Mary Blanco

**Robert Reichart**  
Carol Rup

**Robert Richardson**  
Sally Stephens

**Henry C. Rimmer Jr.**  
Christina Rimmer

**Karla Roy**  
Robin Tucker

**Ronald Russell**  
Donna Roumagoux

**Kathleen "Kay" Savio**  
Lee Savio

**Richard (Dick) Shelly**  
Margaret Turnbow

**Raymond Simpkins**  
Charles and Grace Wagner

**Richard Swain**  
Barbara Swain

**Noel Dwain Wells**  
Harlon Wells

**Jack Whelan**  
Marie and Fred Carter  
Carl Harrington

**Marcia Wierda**  
Richard and Bonnie Blauwkamp  
Mr. and Mrs. Robert VanderVeen  
Andy and Tracie Wierda

**Aren Jennifer Wish**  
Carol and Joel Wish

**Ron Yee**  
Sue Herms

**BETWEEN SEPTEMBER 1 AND NOVEMBER 30, 2017, THE FOLLOWING CONTRIBUTIONS TO THE INTERNATIONAL WALDENSTROM'S MACROGLOBULINEMIA FOUNDATION WERE MADE IN HONOR OF:**

**Anwar Al-Kunani, MD**  
Edward Goldberg

**Dr. Orlando and Sue Arana**  
Edward Goldberg

**Gail Arcari**  
John Reisch  
Gregory and Dee Ann Slover

**Shanthi Aribindi, MD**  
Edward Goldberg

**Laura Baber, MD**  
Edward Goldberg

**Gayle Backmeyer**  
Joseph Backmeyer

**Drs. Jeffrey and Debra Bakal**  
Edward Goldberg

**Deanna Ball**  
Mary Lee Ball

**Bruce Bedingfield, DO**  
Edward Goldberg

**Stan and Susan Rakoff Bedows**  
Edward Goldberg

**Percival A. Bigol, MD**  
Edward Goldberg

**Michelle Blazek**  
Scott Blazek

**Bernhard L. Brijbag, MD**  
Bernard F. Graham  
Charitable Foundation

**Mary Brown**  
Don Brown

**Chris and Bob Budzinsky**  
Edward Goldberg

**Phil Carver**  
Nate Carver

**Dr. Jorge Castillo**  
Cindy Furst  
Ellen Hartline  
Margaret Link

**Cheri Chadima**  
Gloria Chadima  
Rachel DeNicola  
Carol Neveu  
Heather and Jason Printz

**Micki Chambers**  
Kelly and Josh Horeni

**Mary Chapman, RN**  
Edward Goldberg

**Steven and Robin Chess**  
Edward Goldberg

**Mary Clausen, R.Ph**  
Edward Goldberg

**Grete Cooper**  
Edward Goldberg

**Kathy and George Coutrakon**  
Edward Goldberg

**BETWEEN SEPTEMBER 1 AND NOVEMBER 30, 2017, THE FOLLOWING CONTRIBUTIONS TO THE INTERNATIONAL WALDENSTROM'S MACROGLOBULINEMIA FOUNDATION WERE MADE IN HONOR OF:**

**Pete DeNardis**

Edward Goldberg  
Sue Herms

**Robert Dick, MD**

Edward Goldberg

**Jane Dobies**

Karen Dobies

**Dr. Mark Glazer and  
The 10 Point Faith Glazer**

Edward Goldberg

**Glen Durmas**

Ms. Charlotte Doss

**Marty Edelman**

Phyllis and Stephen Lishnoff  
Ms. Blossom Miller  
Larry and Barbara Shapiro

**Geoffery Engel, MD**

Edward Goldberg

**Dr. Gregg and Candy Fenske**

Edward Goldberg

**Kitty Filippone**

Barbara Barnowitz

**Dr. Cal and Kristie Fischer**

Edward Goldberg

**Stanley Fisher**

Jodi and Brian Lynch

**Julia Flora-Tostado**

Edward Goldberg

**Enrique Garcia**

**Valenzuela, MD**

Edward Goldberg

**Dr. Morie Gertz**

Teresa and Wayne Holimon  
Elena Malunis

**Dr. Gene and Mrs. Gail Gitin**

Edward Goldberg

**Dr. Mady Gomez and  
Mr. Javier Gomez**

Edward Goldberg

**Evaristo Gomez, MD**

Edward Goldberg

**Louis Gurkin**

Edward Goldberg

**Ali Handal**

Jennifer and Michael Poole

**Dennis Harleman, DMD**

Dennis and Lyn Harleman

**Carl Harrington**

Eleanor Levie

**Marci Heller**

Ms. Laura Simmons

**Katherine Hens**

Peter and Katherine Hens

**Jan Hergesheimer**

Mr. Robert Hiday

**Dr. Tom Hoffman**

Edward Goldberg  
David and Penny Kirby

**Wanda Huskins**

Edward Goldberg

**Pavel Illner**

Edward Goldberg

**James Ishida**

Connie and Robert Mutel

**IWMF Board of Trustees**

Jennifer Silva

**Jamie Hopping and  
Steve Bernstein**

Edward Goldberg

**Joe Janda**

Robert and Sheila Benecke

**Chris Johns**

Edward Goldberg

**Drs. Lynwood and  
Sandra Jones**

Edward Goldberg

**Russ Jones**

Edward Goldberg

**Michelle M. Kadlec**

Edward Goldberg

**Ramesh Kancherla, MD**

Edward Goldberg

**Penny Kirby**

David and Penny Kirby

**Peter Kloepfer**

Edward Goldberg

**Lauren Knisley, MSN, NP-C**

Edward Goldberg

**Dr. Edward and  
Debbie Kogan**

Edward Goldberg

**Marcella Koslowske**

Marcella Koslowske

**Dr. Robert Kyle**

Marlyn Friedlander

**Ricardo Leigh**

Sarah and Dale Gidcumb

**Eleanor Levie**

Carl Harrington

**Ruth Lizotte**

Gretchen Lizotte  
Peter Lizotte

**Sandy Loebmann**

Edward Goldberg

**Dr. Arnaldo and  
Emma Lopez**

Edward Goldberg

**Shuo Ma, MD, PhD**

Edward Goldberg

**Glen Machesney**

Suzanne Machesney

**Doug and Pat Magilvy**

Joan K. Magilvy

**Elena Malunis**

Andrew Warden

**Gerri McDonald**

Patricia Sirls

**Larry and Gina Melby**

Edward Goldberg

**Phil Mellon's 80th Birthday**

Joseph and Nancy Naylor

**Doris Nixon**

Edward Goldberg

**Lillian O'Donnell**

Brooklyn Center American  
Legion Auxiliary Post 630

**Mildred Olivier, MD**

Edward Goldberg

**Susan and Mehmet Orhan**

Edward Goldberg

**John Paasch**

Edward Goldberg

**Al Pemberton**

Dale and Jeri-Lynn Cross

**Tess Perez, MD**

Edward Goldberg

**Colin Perrott**

Edward Goldberg

**Stephanie and  
Dr. Sandy Rakofsky**

Edward Goldberg

**Vince and Raquel Sanudo**

Melvin Goldberg

**Allen and Vicki Saxon, MD**

Edward Goldberg

**Liz Schupp, MD**

Edward Goldberg

**David Schwartz, MD**

Edward Goldberg

**Samuel Schwartz, MD**

Edward Goldberg

**Governor and  
Mrs. Rick Scott**

Edward Goldberg

**Ahmed and Linda Shaaban**

Edward Goldberg

**Sunny Sharma, MD**

Edward Goldberg

**Zach and Lindsay Shubert**

Carol D. Shubert

**Rose and Don Shustack**

Ellen and Herb Hersh

**Jay Singer**

Anita and Carey Heber

**Cinda Spavins**

Ms. Sharon DePerry

**Carl Stoel**

Kathryn and Doug Irving

**Estelle Sugarman**

Jay Sugarman

**Orawan Sukabachana, MD**

Edward Goldberg

**Dr. John and Mrs. Sue  
Sullivan**

Edward Goldberg

**Maureen Sullivan**

Joseph Hauswirth

**Ron Ternoway**

Edward Goldberg

**Julia Tobias Bearss**

John Bearss

**Marcy Traxler**

Edward Goldberg

**Dr. Steven Treon**

Michael and Betsy Boxer  
Ellen Hartline

Stan Rottell

Laura Winchell

**Ed Goldberg and  
Linda Tritek**

Dorothy Zimbrakos

**Josh Tunca, MD**

Edward Goldberg

**Beth Tze**

Edward Goldberg

**Carol Van Fossen**

Elizabeth Van Fossen  
Emily Van Fossen

**Tomas Villafior, MD**

Edward Goldberg

**Lisa Weldy**

Bob Darvill

Julie Kates

**Dr. Jeff and Rhonda Wener**

Edward Goldberg

**Walter West**

Noel Crowe

**Scott Whitman**

Edward Goldberg

**Dr. Erol and  
Mrs. Gulay Yorulmazoglu**

Edward Goldberg

**Dr. Clive Zent**

Daniel Marder



International Waldenstrom's  
Macroglobulinemia Foundation

6144 Clark Center Avenue  
Sarasota, FL 34238

Telephone 941-927-4963 • Fax 941-927-4467

E-mail: [info@iwmf.com](mailto:info@iwmf.com) • [www.iwmf.com](http://www.iwmf.com)

IWMF is a 501(c)(3) tax exempt non-profit organization

Fed ID #54-1784426

**FOR THE WHOLE STORY, SEE PAGE 9!**

**DANNY WEIL, THE WALL OF DEATH . . . AND WM!**



*Danny Weil on his 70 year-old Indian Scout motorcycle on rollers in front of his Thrill Arena.*



*Wall of Death Thrill Arena overview with Danny Weil on the left promoting the show.*



*Funding provided by a grant from Pharmacyclics LLC, An AbbVie Company and Janssen Biotech, Inc.*