



SÉZARY SYNDROME PART TWO: TREATMENT OPTIONS

In the treatment of Sézary syndrome, we have yet to establish a unified algorithm to map out which drugs to use and when to use them. This lack of uniformity is due in large part to the equivocal efficacy of most treatments, usually in the 35% response rate range, meaning that no specific treatment is clearly superior. Furthermore, treatment strategy is very nuanced and individualized based on many patient factors including patient comorbidities, prior treatment failures and access to specific treatments.

Research is underway exploring genomics in CTCL which aims to allow clinicians to match drug choice to patients’ disease molecular profiles.

Physicians are increasingly utilizing combination therapies in treating Sézary syndrome. What drugs or therapies are selected is driven by the overall burden of malignant lymphocytes in the blood, lymph nodes and skin. As we select various treatments and combine them, it is with the goal of reducing disease in various body compartments in a customized fashion.

For those people with relatively few cancerous cells circulating in their bloodstream, treatment is usually a pairing of a systemic category A medication with a skin-directed therapy such as phototherapy or topical creams.

However, for individuals with many malignant cells circulating in their blood, systemic agents with good evidence of targeting the bloodstream are initiated. This group commonly includes targeted agents such as mogamulizumab, brentuximab, and alemtuzumab, HDAC inhibitors such as romidepsin as well as immunotherapies such as pembrolizumab and more classical chemotherapy agents like doxorubicin.

Category A Medications

In a review of category A medications, *Bexarotene* is a retinoid (Vitamin A analog), which helps to impose a more normal cellular differentiation cycle on abnormal lymphocytes which are proliferating unchecked in Sézary. It is available in capsules, and can be given alone or with other medications. The main side effects reported with bexarotene are suppressed thyroid function and an increase in blood cholesterol. Due to these predictable drug side effects, patients also are started on thyroid replacement and cholesterol lowering medications when starting bexarotene.

Sézary Syndrome...continued on page 10



2021 Issue 3

From the President and the Chief Executive Officer	3	Living With Lymphomatoid Papulosis.....	6
USCLC Workshop Overview - CBCL.....	4	Frequently Asked Questions: COVID-19.....	8
Help Ensure the Future of the Foundation	5	AADA Legislative Conference	11
		Your Community Center.....	Backpage

What Is Cutaneous Lymphoma?

Cutaneous lymphomas are cancers of lymphocytes (white blood cells) that primarily involve the skin. Classification is based on lymphocyte type: B-lymphocytes (B-cell) or T-lymphocytes (T-cell). Cutaneous T-cell lymphoma (CTCL) is the most common type of cutaneous lymphoma that typically presents with red, scaly patches or thickened plaques of skin that often mimic eczema or chronic dermatitis. Progression from limited skin involvement is variable and may be accompanied by tumor formation, ulceration and exfoliation, complicated by itching and infections. Advanced stages are defined by involvement of lymph nodes, peripheral blood, and internal organs. ☺

FORUM

The newsletter of the Cutaneous Lymphoma Foundation
ALL RIGHTS RESERVED

Cutaneous Lymphoma Foundation
PO Box 374
Birmingham, MI 48012-0374
telephone: (248) 644-9014
fax: (248) 644-9014
email: info@clfoundation.org
www.clfoundation.org

Forum is published by the
Cutaneous Lymphoma Foundation.
Editor: Autumn Lintz
Layout & Design: Deb Van Zegeren

Disclaimer

The Cutaneous Lymphoma Foundation does not endorse any drugs, treatments or products reported in this newsletter. Information is provided for informational purposes only. Because the symptoms and severity of cutaneous lymphoma vary among individuals, the Cutaneous Lymphoma Foundation recommends that all drugs and treatments be discussed with the reader's physician(s) for proper evaluation, treatment and medical care.

The Cutaneous Lymphoma Foundation's patient educational newsletter, Forum, has been made possible in part thanks to the following generous supporters:



CUTANEOUS LYMPHOMA FOUNDATION

Cutaneous Lymphoma Foundation is a 501(c)(3) non-profit organization.
Donations are tax deductible to the extent allowed by law.

BOARD OF DIRECTORS

OFFICERS

Laurel Carlson	President
David Elefant	Vice President
Jeff Ward	Treasurer
Marianne Tawa, RN, MSN, ANP	Secretary

DIRECTORS

Steven Horwitz, MD	Director
Youn Kim, MD	Director
Michael W. Young	Director
Susan Thornton	Chief Executive Officer

RESEARCH ADVISORY COUNCIL

Christine Eischen, PhD (Co-Chair)	Thomas Jefferson University
Pierluigi Porcu, MD (Co-Chair)	Thomas Jefferson University
Jaehyuk Choi, MD, PhD	Northwestern University
Alejandro Gru, MD	University of Virginia
Bradley Haverkos, MD, MPH	University of Colorado
Michael Khodadoust, MD, PhD	Stanford University
Anjali Mishra, PhD	Sidney Kimmel Cancer Center
Alison Moskowitz, MD	Memorial Sloan Kettering Cancer Center

MEDICAL ADVISORY COUNCIL

Michi Shinohara, MD (Chair)	University of Washington
Wei Ai, MD	University of California San Francisco
Kim Bohjanen, MD	University of Minnesota
Francine Foss, MD	Yale University
Richard Hoppe, MD	Stanford Cancer Center
Erick Lansigan, MD	Dartmouth-Hitchcock Medical Center
Cecilia Larocca, MD	Dana-Farber/Brigham and Women's Cancer Center
Leora Lowenthal, LICSW, OSW-C, MPA	Beth Israel Deaconess Medical Center
Sue McCann, MSN, RN, DNC	University of Pittsburgh
Amy Musiek, MD	Washington University School of Medicine
Steve Rosen, MD, FACP	City of Hope
Debjani Sahn, MD	Boston University
Jasmine Zain, MD	City of Hope

MEDICAL ADVISORY COUNCIL EMERITUS ADVISORS

Madeleine Duvic, MD	University of Texas MD Anderson
Lars French, MD	University Hospital Zurich
Ellen Kim, MD	University of Pennsylvania
Robert Knobler, MD	Medical University of Vienna
Liliane LaRoche, MD	Hospital Avicenna
David Ramsay, MD	New York University School of Medicine
Alain Rook, MD	University of Pennsylvania
Martin Weinstock, MD, PhD	Brown University
Sean Whittaker, MRCP	Guy's and St. Thomas' Hospital

STAFF

Holly Priebe	Chief Operating and Financial Officer
Autumn Lintz	Education Manager
Debra Van Zegeren	Information Technology Manager
Susette Arnot	Administrative Assistant

FOUNDATION NEWS

Thank you

We would like to thank Hilary Romkey for her seven years of service with the Cutaneous Lymphoma Foundation, as well as her dedication to people affected by cutaneous lymphomas. We wish her nothing but the best for what comes next.

Welcome Autumn Lintz

Please join us in welcoming Autumn Lintz to the Cutaneous Lymphoma Foundation team. Autumn is serving as the Foundation's Education Manager. She brings with her a wealth of knowledge and experience in program management, as well as a deep passion for empowering people through education.



FROM THE BOARD PRESIDENT

Laurel Carlson

Happy Holidays!

Here we are at the end of our second year impacted by the COVID-19 virus. The year 2021 has seen more coronavirus variants, multiple vaccination opportunities, continued mask-wearing, and even more questions about what we, as patients, should do to try and keep ourselves safe and healthy. The challenges continue and our organization is working tirelessly to meet the changing needs of our patient community.



"As a member of our community, you are never alone."

Laurel Carlson

The end of 2021 also marks my 10th anniversary as a member of the Board of Directors. When I recall my first board meeting in the fall of 2011, I remember feeling honored to be part of such a distinguished group of people, but I also remember feeling very intimidated and unsure about how I would be able to contribute to help make a difference in the lives of people affected by my disease. I had no medical background and, to be honest, the doctors who I had seen for the previous thirty years were competent, but none were very familiar with my disease and I always felt they were learning about it along with me. It was a bit of a shock to find myself surrounded by people who were not only more familiar with my disease than I was, but who were also very dedicated to specifically helping those of us who were affected by it. I will admit that I was almost afraid to speak at our meet-

From the President...continued on page 9

FROM THE CHIEF EXECUTIVE OFFICER

Susan Thornton



"...Each of you who has engaged in some way has made a difference."

Susan Thornton

"The smallest act of kindness is worth more than the grandest intention." – Oscar Wilde

As we wrap up this calendar year and reflect on the last 12 months, I'm struck by how fast the time has gone and how much has happened in a short time. One of my personal aha's of the last two years is how deeply grateful I am for the people in my life and the connections that make every day rich and rewarding.

Thinking about those connections, I'm excited about the opportunity we have to expand our cutaneous lymphoma community around the globe. This year, we established new relationships with our sister foundation in the Netherlands, launched in 2018 and led by a passionate, dedicated

voluntary group of patients, caregivers, and clinicians. You may have met Mieke and her husband, Ton. They were our guests on Friday evening at our first International Patient Conference in September. We also co-hosted our first patient education seminar in Marseille, France, delivered entirely in French, with the clinical team lead by Professor Martine Bagot and our colleague

From the CEO...continued on page 9

UNITED STATES CUTANEOUS LYMPHOMA CONSORTIUM - 2021 WORKSHOP OVERVIEW

The United States Cutaneous Lymphoma Consortium, or USCLC, hosted their Annual Workshop on Saturday, June 26th on the biology and treatment of cutaneous B-cell lymphomas. Dr. John A. Zic, from Vanderbilt University Medical Center, was one of the presenters at the workshop and was kind enough to give some of the takeaways from the meeting to the Cutaneous Lymphoma Foundation.

Primary Cutaneous Lymphoma: International Collaborative Recommendations for Clinical Trial Design, Assessment, Response Criteria, Endpoints, and Update to Staging

Elise Olsen, MD, Duke University

About seven years ago, Dr. Elise Olsen brought together a group of cutaneous lymphoma experts from around the world to standardize how medications are studied in clinical trials for patients with cutaneous lymphoma. The group has worked towards developing criteria to determine the effectiveness of the medications being tested in clinical trials. Dr. Olsen presented the results of their collaboration which are soon to be published in the medical journal *Blood*.

The publication of their report is significant to both physicians and pharmaceutical companies. For physicians, it can provide guidelines both in treating their patients and running trials at their facilities. For pharmaceutical companies, it can be used for designing trials, assessing patient response, and determining the endpoint of the trial in order to determine if it is successful. The outcomes of the group's collaborative efforts have the potential to help individuals diagnosed with cutaneous lymphoma, whether it be B or T-cell.

Low-Grade B-cell Lymphomas - Primary Cutaneous Marginal Zone Lymphoma and Follicle Center Lymphoma

Dr. Michi Shinohara presented on low-grade, or slow growing, cutaneous B-cell lymphomas. The two subtypes considered low-grade are the cutaneous follicle center lymphoma (pcFCL) and the cutaneous marginal zone B-cell lymphoma (pcMZL). Both have an excellent prognosis for patients.

Dr. Shinohara focused on how a new patient should be "worked up" or diagnosed. Most patients typically have a nodule on the skin that is biopsied. If the pathology report comes back with a diagnosis of B-cell lymphoma, it is important that imaging tests such as a CT scan or PET/CT scan are performed. While most B-cell lymphomas start in the lymph cells, these types of lymphomas start in the skin, so it is important to include imaging tests to look for an increase in the size of lymph nodes. Additional tests may also include flow cytometry (either of a skin biopsy or the peripheral blood) to test for abnormal B-cells in the blood. Dr. Shinohara also concluded that in most cases bone marrow biopsies were not necessary.

Dr. Zic mentioned that one of his takeaways from Dr. Shinohara's presentation is that both pcMZL and pcFCL are typically slow growing and respond very well to treatment, *except* when pcFCL is found on the leg. For some reason, when the follicle center lymphomas are on the leg, they behave more aggressively and need to be watched more closely and may need more aggressive treatment than if they occur elsewhere on the body.

High-grade B-cell Lymphomas - Diffuse Large B-cell Lymphoma, Leg Type and Intravascular Large B-cell Lymphoma

Alejandro Gru, MD, University of Virginia School of Medicine

The next speaker was Dr. Alejandro Gru on the high-grade, more aggressive cutaneous B-cell lymphomas which include diffuse large B-cell lymphoma (DLBCL), leg type, and intravascular large B-cell lymphoma.

It is unusual to have the name of the specific body part in the description of a cancerous lymphoma, however about 80% of DLBCL do occur in the leg - there is something about lymphoma in the leg that causes it to behave more aggressively. This type of cutaneous B-cell lymphoma (CBCL) occurs most frequently in elderly individuals and slightly more frequently in women than men. It is a more rare form, occurring in only 10-15% of CBCLs. Like other cutaneous lymphomas, its cause is unknown. DLBCL, leg type, can be an aggressive lymphoma and it is recommended to be treated systemically, typically with rituximab and CHOP chemotherapy.

Dr. Gru also spoke about other rare CBCL lymphomas, demonstrating, as noted by Dr. Zic, the complexity of high-grade B-cell lymphomas. Because of this complexity, Dr. Zic emphasized how important it is for individuals diagnosed with a high-grade B-cell lymphoma to have their biopsy slides read by a pathologist with B-cell lymphoma experience.

Included in Dr. Gru's presentation was intravascular large B-cell lymphoma which is a very rare type affecting the B-cells found within the blood vessels as opposed to the 2nd and 3rd layer of the skin where the majority of CBCL cells are typically found. This makes this type of lymphoma easy to miss. This variant may manifest in a variety of ways, but most patients may mention tender areas, indicating a biopsy is in order. The median age of those diagnosed is around 67 years old.

Pseudolymphomas: Clinical and Histologic Spectrum and Controversies

Joan Guitart, MD, Northwestern University School of Medicine

Dr. Joan Guitart spoke briefly on pseudolymphomas, or "false" lymphoma. Under the microscope, the pathologist may be concerned that they are looking at a lymphoma. However, when more testing is done and the patient is followed over time, it's found the patient does very well, causing a dilemma for the pathologist - was the cancer diagnosis correct or was it a case of pseudolymphoma? Dr. Guitart refers to this as a "grey zone" - an instance where a pathologist has difficulty

diagnosing a lymphoma despite what is seen under the microscope. Dr. Zic's take-away was how important it is for the clinician directly examining the patient and the pathologist to work together to determine whether what the patient has is truly a cancer or, despite what the pathologist sees on the slide, is actually a pseudolymphoma.

Dr. Guitart also discussed cutaneous lymphoid hyperplasia. Hyperplasia does not mean cancer; it means that there are many, many cells in a particular tissue. By studying the existing literature, Dr. Guitart developed a list of known causes of cutaneous lymphoid hyperplasia. A common cause was bug bites. Sometimes a bug bite will trigger the immune system and a nodule will form that sits in the skin for months and months afterwards and mimics a cutaneous lymphoma, but it is not. It was noted, however, that in some instances a cutaneous lymphoid hyperplasia papule may develop into a marginal zone lymphoma over time.

In closing, Dr. Zic reassured patients that it is very unusual for an individual with cutaneous B-cell lymphoma to develop cutaneous T-cell lymphoma and vice versa - so patients don't need to worry about developing one if they have the other. He concluded with the observation that there are many people doing extensive research to make sure the right care is provided to the right patient at the right time.

Dr. Zic's full presentation, including brief summaries of multiple research projects presented at the conference, is available on the Foundation's YouTube channel at <https://bit.ly/304rwFs>



Cutaneous B-Cell Lymphoma

Saturday, June 26, 2021

Agenda

Cutaneous B Cell Lymphomas - Part I	
10:03-10:18	Primary Cutaneous Lymphoma: International Collaborative Trial Design, Assessment, Response Criteria, Endpoints
10:18-10:32	Low-grade B-cell lymphomas - Primary Cutaneous Marginal Zone Lymphoma (M. Shinohara)
10:32-10:49	High-grade B-cell lymphomas - Diffuse Large B-cell Lymphoma (A. Gru)
10:49-11:08	Pseudolymphomas: Clinical and Histologic Spectrum and CTCL and CBCL Research-Based Abstract Presentations

HELP ENSURE THE FUTURE OF THE CUTANEOUS LYMPHOMA FOUNDATION

Did you know you can support the Cutaneous Lymphoma Foundation in achieving its vision of a life free of cutaneous lymphoma and lower your tax bill with an IRA charitable rollover? If you have an IRA, or certain other types of retirement plans, and are at least 70 ½ years old, you can reduce your income taxes (even if you don't itemize) with a qualified charitable distribution (QCD) to the CL Foundation from your IRA. The donation can count as part of your required minimum distribution for 2021, thereby reducing your taxable income. So a charitable-minded account owner who must take out, say, \$25,000, can divert part or all of it to his or her favorite charity, such as the CL Foundation. While the gift can't be claimed as a charitable deduction, the payout doesn't count as taxable income either. So a QCD doesn't raise adjusted gross income in a way that could trigger higher income taxes, Medicare premiums, or taxes on Social Security benefits. Both Medicare Part B and Part D payments rise with income, so minimizing adjusted gross income can be important. A QCD is simple to make, however, there are some technical rules in that it cannot exceed \$100,000, must be made directly to the Cutaneous Lymphoma Foundation by your IRA custodian, and must be completed by December 31.

Please check our website at <https://bit.ly/3HoAQoR> for additional information about ways everyone can support the CL Foundation. If you have any questions or would like to discuss this further, please contact Holly Priebe, COFO, at 248-644-9014 Ext. 200 and your personal estate planning attorney. ☺

The above summary is provided for informational purposes only and is not intended to be legal, tax or investment advice. Please contact your attorney or financial advisor for advice related to your personal circumstances.

LIVING WITH LYMPHOMATOID PAPULOSIS: A STORY OF HOPE

Shared by Victoria (Torie) M.

Most people think that the year 2020 was the year of COVID-19, but for us 2020 was the year that changed our lives forever. My name is Torie, my daughter's name is Mia, and this is our story of hope.

Not Looking For a Diagnosis

In April 2019, Mia had a growth on her leg that just wouldn't heal. It was going on six weeks and the lesion was getting necrotic. I have been working in dermatology for 20 years, so after two weeks of the growth not going away and getting better, I started applying cortisone. I even injected it with cortisone. The growth kept getting bigger and bigger. I asked a doctor who I had worked with to biopsy the growth to get rid of it, not looking for a diagnosis. We just thought it was an unusual bite from a weird arthropod. The diagnosis came back as "arthropod assault," so it confirmed our thoughts because, until that day, we had not had a child with a form of CTCL come into our office. Fast forward to the end of November when a couple of lesions presented again. We kept thinking it was another arthropod assault and that Mia's immune system was hypersensitive to bites now because of her skin's experience with the bug bite from April.

Weeks went by again with no improvement, after trying topical and injectable cortisone. A physician assistant I work with thought it was time to do a biopsy again, this time ruling out lymphomatoid papulosis. I had a weird feeling for weeks before the biopsy, that these lesions were a form of cutaneous lymphoma. I only had experience with adults having mycosis fungoides, but still my intuition told me differently. The biopsy report was sent to a different laboratory with an excellent dermatopathologist, who diagnosed it immediately after two different types of skin biopsies. My daughter Mia now had a diagnosis of lymphomatoid papulosis type A.

I Can Figure This Out

More growths kept appearing, so immediately my life went into searching for a "cure" for Mia. Working in dermatology for 20 years is a blessing and a curse at the same time. It's an amazing blessing because I have access to doctors at any time, and even have the lightbox and Xtrac devices in my office that I use to treat Mia. Where the curse comes in is that I can't figure out the "cure" for my daughter. I kept saying to myself that I could figure this out because this is a dermatology issue and that is what my career is in. I spent every day looking for medications, treatments and specialists that would make everything better. It was my mission to figure it all out. Even

having access to so many physicians, lymphomatoid papulosis is one in a million in children so I couldn't find the answer we were looking for.

We had started topical and injectable cortisone therapies again which made the lesions worse. We started lightbox treatments, oral antibiotics and antiviral medications which weren't making things better. We kept trying every topical treatment from rose hip oil, Manuka honey, even CBD cream. We live in Florida which has sunlight year round, but natural sunlight was not making it go away. Knowing what we know now, COVID-19 brought many stressors and stress doesn't help matters. In the end, we decided to put Mia on methotrexate because other therapies were not working. She has been taking MTX for six months now.

On a Mission

I was on a mission to solve the mystery. I had been told by some doctors that "I should stop looking for the reason why this is happening because doctors can't find the reason." That was not acceptable to me. I said to myself, "How dare someone say those words to me, because I am A MOTHER WHO WILL FIND AN ANSWER FOR MY MIA."

I subscribed to the Cutaneous Lymphoma Foundation newsletter, I reached out to patients in support groups, and searched daily to find the doctors that were well-versed with LyP. Dermatologists know about it, but not many know enough about the treatment options that are not typical. I found two doctors in Texas who know about LyP well. We were on the next plane to Houston, even in the middle of a pandemic. Many blood tests were done to try to figure out why this was happening. I had asked about a particular virus that I had read if exposed to it, it can cause this diagnosis. The blood



Torie (left) and Mia (right)

test wasn't run because the oncologist said he was running so many other levels to start. When I received the call about the blood test results, the oncologist stated that he was confused with Mia's results because her T-cell levels showed a significant viral infection that potentially could have landed her in the hospital. I thought to myself, if he is confused then I am really confused as to how to proceed. Mia's growths did not stop coming, so our quest for answers did not stop either.

Finding an Angel

I found the Cutaneous Lymphoma Foundation website when we were waiting for the final biopsy reports in January. I had watched every video three times and read every bit of information that was available on the site. I was looking for someone or something to answer my questions. I kept looking for any outlet that would give me the chance to connect with other parents who had children with lymphomatoid papulosis. Again, my career is in dermatology, but I didn't have anyone to share this with or anyone who could help me with this process. I had found a couple of support groups for LyP, but it was mostly patients in the groups.

I had FINALLY found an angel on the LyP Facebook group. The angel is named Mindy. Her daughter was 14 and looked just like Mia. Mindy was literally an angel sent to us. I finally had another parent to talk to. Our daughters were so similar and were both experiencing the same struggles. It was the first time I had someone who understood what we were going through. Her daughter was diagnosed with LyP D a couple of years before Mia, so she really helped to guide me. There were only two other parents I connected with during these months because they were all I could find with children that have LyP. I would call the Foundation and countless look for any parent groups I could find to meet parents going through the same thing.

Never Stop Trying

Fast forward to July 2021, we traveled to Dallas to see a doctor we found who helped this skin condition with more holistic approaches, mostly lifestyle changes and specific antibiotic regimens. This doctor ran the blood level that I had inquired about the year prior. Guess what? The virus that I had read so much about, that could potentially be a cause for LyP, Mia was positive for it. It's ok to question doctors, pathologists, treatments and medications. I have learned so much through all of this, that I actually educate my colleagues on the condition. Stand up for yourselves, your kids, and NEVER stop trying

or give up. No one knows you or your children better than you do. Hope is everything!!

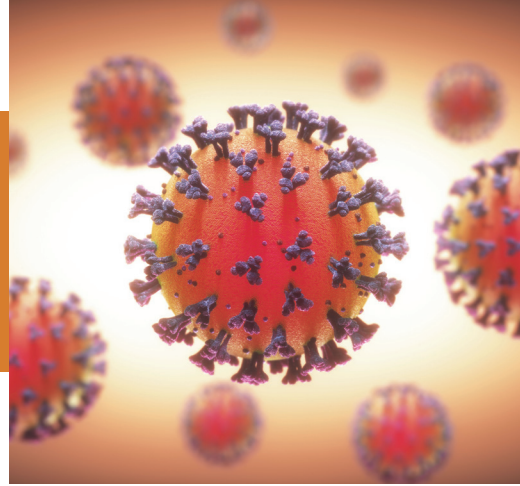
It doesn't change treatments or outcomes, but for me it has helped me change my focus. Instead of focusing on the "why or the how," it has opened my eyes to only focus on "Mia." As parents, our job is to protect our kids, make them feel safe and to be advocates when they need help. I think some days my focus shifted from Mia to the how and why. That is where I went wrong. Mia didn't need me to be searching endlessly for the answers. Mia needed me to be her mom and make her feel safe. Mia is not defined by LyP. Mia is the most authentic, caring, giving, empathetic, selfless and hilarious human I know. We have grown as a family and individuals. We have had discussions as to what we think our silver lining will be through all of this. My amazing Mia said that maybe she can help other kids with the same thing. Of course she would say that, that is who Mia is. Mia has shown up for every blood test, skin treatment and doctor's appointment with strength and courage. I have learned so much from my daughter. She has taught me what strength and courage are.

Never Give Up on Hope

One day we woke up and decided that we were going to put our energy into making a difference and to not focus on the challenges. That we were going to spend our time bringing awareness and raising money for research. Our hope is that one day there will be a cure for cutaneous lymphoma. My hope would be to have a parent support group. To be able to share experiences or ask questions of other parents who are dealing with the same thing is so important, so that we can be the strongest force for our kids. Our hope is for every person dealing with cutaneous lymphoma to know that they are not alone. As you all know, having cutaneous lymphoma can bring different challenges on different days. Through all of this, we have really learned to appreciate the good days and most of all to never give up on hope. 🌈

Disclaimer - Stories of Hope are an amazing opportunity to share, grow and learn from a peer's experiences. Stories of hope are not intended to serve as medical advice; given the individual nature of a diagnosis, all diagnosis and treatment decisions should be made alongside your medical team.

Frequently Asked Questions COVID-19



What is the current recommendation regarding booster shots for people with cutaneous lymphoma and would you consider a patient with cutaneous lymphoma immunocompromised?

The recommendation at this time regarding boosters is that if you received two shots, you should receive a booster six months after your second shot if you are immunocompromised or over the age of 65. On whether you would be considered immunocompromised, it will depend on what stage of disease you are at. For early stage disease where you are not receiving systemic treatments, you could forgo the booster shot. Later stage disease and patients receiving systemic treatments should discuss getting the booster with their doctor.

If I am a double vaxxed patient, is it safe to host overnight guests and family over the holidays and, if so, what precautions should be taken?

As vaccinations in patients who are being treated for lymphoma may not be as effective, depending on their stage and current treatments, precautions should be taken. Discuss with your doctor, make sure your guests are vaccinated, and if they are not, ensure they have a negative COVID-19 test within 72 hours of their visit.

Should hematology patients receive a booster shot?

It is being recommended to receive the booster as hematology patients may not have as strong an immune response as a non-hematologic patient. Just like when someone receives a bone marrow transplant and they are revaccinated with all childhood vaccinations, receiving the booster will help level the playing field.

Please note: These FAQs were derived from a webinar hosted on 10/12/21. Please be sure to check the CDC website for the most current and up-to-date recommendations.

I have developed a cutaneous patch around the vaccination site. Is this something I should be concerned about, particularly if I am getting a booster?

Some patients are reporting a rash around the vaccination site and you may get another rash with the booster, but it is still recommended to get the booster if you qualify for it.

Do you recommend that patients who were vaccinated prior to being six months off rituximab vaccinate again after being nine months off of rituximab?

Most people who have received rituximab did not mount an immune response from the COVID-19 vaccine. It is best to discuss this with your doctor.

Have patients with cutaneous lymphoma who have tested positive for COVID-19 shown increased cutaneous lymphoma disease activity afterwards?

There have only been a couple of case reports, which is a detailed report of the symptoms, signs, diagnosis, treatment, and follow-up of an individual patient, regarding this. At this time, there isn't enough data to answer the question.

Does the lack of reaction to the vaccine correlate with patients not building antibodies?

If you don't mount an antibody response, it is possible that the immune system response is not optimal. However, there is a whole other component of the response that is the T-cell response, which is longer lasting, but this is hard to measure. At this time, it is not known if the antibody response correlates to the actual immune response. 📧

From the President...continued from pg 3

ings, and it took some time for my confidence to develop. But I was determined to try to be the voice for all patients and take advantage of the incredible opportunity that I had been given.

I had never forgotten the frustration and sense of loneliness I felt when I was diagnosed in 1983. I knew of no other person with cutaneous lymphoma and I had no way to even try to meet one. Looking for any kind of connection, I once attended a cancer support group meeting. I felt so out of place there that I never returned. To say that finding the Cutaneous Lymphoma Foundation was a life changer for me would be an understatement. And now every month (on its first Tuesday), I continue to meet others during our online networking group meetings who are receiving the same life-changing support that I once discovered. But I also know that for every person who attends one of these meetings and has benefitted from speaking with its other participants, there are several others who have been personally helped by a resource on our website, one of our live events, or by a phone conversation with a member of our staff.

I will admit that when I joined the board, I also felt a slight apprehension. I knew that many non-profits failed to last beyond their first few years, and I was now part of an orga-

From the CEO...continued from pg 3

Guy Bouguet who leads Ellye, the French lymphoma organization. Discussions are underway with others throughout Europe to explore ways we can work together to connect all patients and family members, and share valuable resources. Yes, there is a lot of work to do, but it's a big step in the right direction.

It's challenging to summarize a year's worth of events and projects in a short newsletter column. However, know that each of you who has engaged in some way has made a difference. Each individual act of kindness and care sends ripples out into the world, touching people in ways you may never know. Connecting, supporting, and serving each other along the way is genuinely what this community is all about.

In the following pages, you will learn about cutaneous B-cell lymphoma from the United States Cutaneous Lymphoma Consortium annual workshop shared by Dr. John Zic, the follow-up article about Sézary syndrome by Dr. Jennifer DeSimone, and read a heartwarming story authored by Victoria M, the mom of a teen living with lymphomatoid papulosis (LyP).

In addition to the programs and new initiatives to connect our global community, we've been active in the policy and advocacy arena. CL Foundation representatives participated in

nization that could easily become one of those. I can't begin to list the many successes and recouped failures that have occurred over the past ten years, but what I proudly see today is not only a healthy organization, but a thriving community that has exceeded my expectations.

"One of the most vital ways we sustain ourselves is by building communities of resistance, places where we know we are not alone." - bell hooks

As a member of our community, you are never alone. Once small and with limited resources, our community is now not only much larger, but it continues to identify and develop more ways to help it grow. Each of you is a valued member of our community and your voices and financial gifts are important and necessary to help us continue to grow and help everyone who is affected by cutaneous lymphomas.

Thank you and best wishes for a Happy and Healthy New Year,

Laurel

several events to bring your voice to legislators on the impact of living with cutaneous lymphoma and how policy impacts individuals. We have engaged in collaborative efforts to bring your voices to government agencies in Canada and Europe too. Thank you to everyone who participated in any of the surveys to capture your experience to share with these government review panels. Your participation makes a difference.

As this year comes to a close and 2022 begins to take shape, I invite you to become an active member of our global community. Your engagement, interaction, and willingness to support each other will help make life better for everyone impacted by this disease. This quote by Helen Keller says so much:

"Alone, we can do so little; together, we can do so much."

– Helen Keller

Come join us - become a part of your community. Let's go do more together.

See you in 2022!

Jessie

Sézary Syndrome...continued from pg 1

Interferons are substances which our bodies produce naturally and may be given as a form of immunotherapy. Interferons stimulate anti-cancer immune function. However, interferons bring about a wide range of responses. While there is no clear protocol when using interferons in treatment, most patients use this option for the long run, with good general effectiveness. Interferons are given by self-injection three times a week. Early in the treatment schedule, many patients report flu-like symptom reactions, which tend to improve over time. Frequent lab monitoring of blood count and liver enzymes is required on interferon. Some patients experience depressed mood and mental fog which lead to termination of treatment.

There is a class of drugs termed “targeted agents” which means that the drug identifies and impairs a specific protein rather than impacting all cells broadly, as is the case with traditional chemotherapy. In general, these drugs tend to be better tolerated than older generations of chemotherapy with less nausea, vomiting, hair loss and neutropenia/immunosuppression.

Included in this group is **Romidepsin**, an enzyme inhibitor, which shows effectiveness in about one third of patients, although it can take over three months for progress to be noted. Romidepsin has a long infusion time, between four and five hours, and can cause some strong side effects such as fatigue and nausea for certain patients.

Protein-Targeting Drugs

Alemtuzumab (also known as Campath) targets the CD52 protein expressed in malignant cells. Trials show a very good overall response rate when this is given as an injection, in the office, three days a week. However, this treatment may eradicate some healthy cells as well, and patients must also take prophylactic antibiotics and antiviral medication.

Another choice, this one targeting the CD30 protein, is **Brentuximab**, which is usually given as an infusion every three weeks. While the response rate of 60% is good, it has some possible severe side effects such as neuropathy which must be closely monitored.

Mogamulizumab, given by infusion weekly for 4 weeks, then every other week thereafter, targets the CCR4 protein molecule. The response rate is around 28%, but patients must be careful to complete a wash-out period from this medication, if they plan to schedule a stem cell treatment in the near future.

Other Treatment Options

Outside of these protein-targeting drugs, other treatment options include **Total Skin Electron Beam** radiation, which attempts to uniformly deliver electrons (radiation) to the entire

surface of the skin. As part of a combination therapy, long-term remission has been recorded using low-dose Electron Beam Therapy along with immunotherapy.

A promising treatment option is **Immunotherapy**, which is designed to enhance the native anti-cancer response. Using Extracorporeal Photopheresis (ECP), blood is extracted from the patient, then lymphocytes are separated out, irradiated with ultraviolet type A light (UVA) which exposes the genetic material of the cancer cells, allowing the body to better identify the cancer cell target. The blood is then re-infused into the patient to help the patient’s immune system recognize and attack the cancer cells. ECP is given in two consecutive treatments over four or five hours every two to four weeks. Responsiveness to this treatment option varies, with highest reports around 60%. Four to five hours of continuous high flow large bore IV access is required and for some patients a port must be installed for this reason.

Immune Checkpoint Inhibitors are molecules that boost anti-cancer immune function by preventing cancer cells from evading routine immune surveillance by healthy T cells. Pembrolizumab is a PD-1 inhibitor in this category. A study completed in 2016 showed a 38% response rate in Sézary, however, some patients reported flares during the treatment. This is explained by the fact that these drugs turn on T cells in efforts to fight disease, but in CTCL when the T cells are abnormal, the drug may also stimulate the malignant T cells leading to disease worsening. As a class, these drugs augment immune activity and the main side effects are autoimmune inflammation of the thyroid, lungs, gut and rarely the heart and nervous system.

Currently, the only treatment aimed at cure rather than remission is **Stem Cell Transplant** (SCT) therapy. Candidates for stem cell transplant should have excellent disease control prior to undergoing the transplant. Preparation for SCT involves using drugs and radiation to eliminate the patient’s current immune system prior to engrafting a new donor immune system. The hope is that the new immune system is better able to fight Sézary. 🏡

Jennifer DeSimone, MD, FAAD
Inova Schar Cancer Institute

The Sézary Syndrome Part 1 and 2 articles were based on Dr. DeSimone’s presentation at the 2021 Virtual Patient Conference. You can watch a recording of the presentation at: <https://bit.ly/3kRo6gS>

To see the referenced NCCN guideline for Category A Medications, please visit: <https://bit.ly/3kReKS8>

AADA ANNUAL LEGISLATIVE CONFERENCE

The Cutaneous Lymphoma Foundation was pleased to participate in the American Academy of Dermatology Association’s (AADA) Virtual Legislative Conference and Day on the Hill. As a member of the Coalition of Skin Diseases, the Foundation was invited to participate in this annual event, providing an invaluable opportunity to add the patient voice. We joined over 200 dermatologists and 40 patient advocates from 30 states to speak to members of Congress - making this one of the most well-attended AADA legislative events yet.

The AADA’s legislative “asks” were especially pertinent to the cutaneous lymphoma community:

- Ensure Medicare Stability for Patients & Physicians - members of Congress were asked to take action to prevent Medicare physician payment cuts that impact patients’ access to care and will help ensure financial stability for physician practices that are struggling from the effects of the pandemic. The relevance to patients is the potential 10% cut to Medicare payments could cause dermatologists to lay-off staff (causing long delays in getting appointments), not invest in equipment (for example phototherapy units), or limit the number of Medicare patients they will see.
- Preserve Patients Access to Treatments - the focus of this “ask” is the Safe Step Act (H.R. 2163/S. 464). Step therapy protocols or “fail first” strategies currently used by health plans prevent physicians from prescribing treatments that will provide the best treatment results in the most effective manner. Step therapy protocols require patients to try one or more prescription drugs before coverage is provided for a treatment selected by the patient’s physician. The Safe Step Act establishes an exemption process to the protocols used by health plans, preserving the physician’s right to make treatment decisions in the best interest of the patient.

(Summaries from AADA one-pagers prepared for members of Congress)

A highlight of each conference is recognizing the cutaneous lymphoma clinicians participating in the event. It is gratifying to know that our clinicians use their own time to advocate on behalf of the patients they serve.

If you would like to know more about how you can participate in advocacy, we recommend watching Your Voice Matters: How (and Why) to Take Action on Public Policy - the patient advocacy presentation from our recent International Patient Conference. You can find the video on our YouTube channel, CutaneousLymphomaFndn, or on our Advocacy page on the website (www.clfoundation.org/Advocacy). 🏡

PROGRAMS & SERVICES NEEDS ASSESSMENT SURVEY

Thank you for your participation!

We appreciate you taking time to provide your feedback. Your input helps us to both analyze the programs offered as well as shape future efforts. For more specifics about what we learned please watch our 2021 Annual Update where we highlight a look back at 2021 and forward to 2022.

<https://bit.ly/3Ct3MrP>

UPCOMING EVENTS & OPPORTUNITIES

Join Community Connections

Make sure to check out the Cutaneous Lymphoma Community Connections, a place where you can interact with others facing the same or similar experiences as you. In order to provide privacy and encourage open communication with each other, Community Connections is open exclusively to patients and their loved ones.

To learn more, visit www.clfoundation.org/connections

Upcoming Events

To learn about upcoming events, visit www.clfoundation.org/upcoming-events or scan the Q-code.



Don't miss out, make sure to subscribe to the Cutaneous Lymphoma Foundation's YouTube channel (CutaneousLymphomaFnd) and follow us on Facebook and LinkedIn.



CUTANEOUS LYMPHOMA FOUNDATION
PO BOX 374
BIRMINGHAM, MI 48012

TIME SENSITIVE MATERIALS ENCLOSED

The Cutaneous Lymphoma Foundation is Your Cutaneous Lymphoma Community Center

YOUR place to belong...

- a library of educational information
- a resource center for finding specialists, clinical trials, financial aid and more
- an activity center with programs and events focused on education and connection
- a common area providing a safe space to share your experiences with peers
- a town hall to bring voices together to make change
- a research lab with a visitor gallery -- a place to invest in finding a cure, including a birds eye view into how the work is getting done

Ensure the doors stay open and the lights stay on, for you, and the next person affected by cutaneous lymphomas.

DONATE TODAY -- ONLINE, BY PHONE OR BY MAIL

Online: <https://www.clfoundation.org/giving-online>

By Phone: (248) 644-9014, Ext 100

By Mail: PO BOX 374, Birmingham, MI 48012

(a remittance envelope has been enclosed for your convenience)

