

Our banner for the March newsletter honors National Doctors Day on March 30th



Research Round Up

by Dean Frolich, PhD

The first study that I am highlighting this week, I am happy to say, was partially funded by SFA. In the paper entitled "[A randomized, non-comparative phase 2 study of neoadjuvant immune-checkpoint blockade in retroperitoneal dedifferentiated liposarcoma and extremity/truncal undifferentiated pleomorphic sarcoma](#)," the investigators conducted a randomized, non-comparative phase 2 trial (NCT03307616) of neoadjuvant, or pre-surgical, immune checkpoint blockade, or ICB, (nivolumab or nivolumab/ipilimumab), which removes the "brakes" that tumors put on the immune system, in patients with resectable retroperitoneal dedifferentiated liposarcoma (DDLPS) and extremity/truncal undifferentiated pleomorphic sarcoma (UPS). The patients with UPS received radiation therapy along with the immune checkpoint blockade. The primary end point of pathologic response was a median of 8.8% in DDLPS and 89% in UPS. Lower densities of immune cells called regulatory T cells in the tumor before treatment were associated with a major pathologic response. Additionally, increased numbers of a different kind of immune cells called B cells in tumors following neoadjuvant treatment was associated with overall survival in DDLPS. This research shows that neoadjuvant ICB is associated with how the immune system interacts with the tumors in DDLPS and UPS and that neoadjuvant ICB along with radiotherapy has significant efficacy in UPS, however additional studies need to be done before this treatment is routinely used for patients with these sarcoma subtypes.

Next, is a study entitled, "[Proteomic characterization identifies clinically relevant subgroups of](#)

[soft tissue sarcoma](#).” This is another study that looks at the functional classification of soft tissue sarcoma; this time by the proteins expressed in the tumors of 272 patients from 12 different subtypes in China. In their analysis, they find a similarity between angiosarcoma (AS) and epithelial sarcoma (ES), and elevated expression of one protein in particular (SHC1) is correlated with poor prognosis in these subtypes. Additionally, they find that when they group the subtypes by the proteins expressed that there are three different groups each driven by different cell pathways and with different patient outcomes. Their analysis also identified the three immune subtypes with different tumor microenvironments. Additional research needs to be done, but this study continues the insights that have developed by characterization of multiple subtypes of sarcoma using a variety of testing methods.

Lastly, in a paper titled, “[KDM3B inhibitors disrupt the oncogenic activity of PAX3-FOXO1 in fusion-positive rhabdomyosarcoma](#),” researchers screened thousands of drug compounds in cell culture and a mouse model of fusion-positive rhabdomyosarcoma (FP-RMS). FP-RMS is driven primarily by the fusion of two genes into one called PAX3-FOXO1. Through their screening, they identified a previously unstudied compound called P3FI-63 and with further investigation determined that it works through proteins called histone lysine demethylases (KDMs) with the greatest affinity for KDM3B. Additionally, they found that a structurally similar drug, P3FI-90, had improved solubility and potency. They were then able to demonstrate that P3FI-90 decreases the growth of FP-RMS cells in vitro and in mice through downregulating PAX3-FOXO1 activity. Although this is a study very early in the drug development process, the results indicate that this could be a therapeutic approach for FP-RMS.

In conclusion, P3FI-90 inhibits multiple KDMs with highest selectivity for KDM3B that can be further developed for the therapy of highly malignant FP-RMS and possibly other “transcriptionally addicted” cancers.

Clinical Trials Corner

by Kristi Oristian, PhD

This month SFA is highlighting a study called [Brightline-4: A Study to Test How Well Brigimadlin is Tolerated by People With a Type of Cancer Called Dedifferentiated Liposarcoma](#). This is a multi-center, open-label, phase 3 study for patients with advanced dedifferentiated liposarcoma. This study, sponsored by [Boehringer Ingelheim](#), is open to people ages 18 and older in the United States. All patients enrolled in the study will receive the study drug, brigimadlin.

Brigimadlin is an experimental new medicine being developed to treat sarcoma. Brigimadlin is a type of drug called an MDM2 inhibitor, which works inside the body to disrupt the interaction between the proteins MDM2 and p53. In some tumors, excess MDM2 protein binds with p53 and allows cancer cells to survive. Brigimadlin is designed to break the bond between MDM2 and p53 so that tumor cell death can occur.

Patients participating in the study will take brigimadlin as a tablet once every 3 weeks and will be allowed to continue taking the medicine for as long as they continue to see benefit from the drug and are able to tolerate it. Patients will be asked to visit the study site at regular intervals where doctors will monitor tumor size and collect information about any side effects they are experiencing from the drug. This study will help doctors determine if brigimadlin is safe and effective for patients with dedifferentiated liposarcoma.

There are additional eligibility and exclusion criteria, including minimum organ function requirements and positive MDM2 immunohistochemistry or amplification. Patients interested in this study should review these criteria with their doctor. To learn more about this study, patients can talk to their doctor, contact the investigator at the [site](#) nearest you or your primary treatment center, or contact the [study sponsor](#).



One Woman's Winning Battle with Leiomyosarcoma

by Christine Summers, daughter and caregiver of Sarcoma Survivor, Kathy

It's exactly one year and ten days since the basketball-sized tumor was removed from my mother's uterus. Only three weeks prior to that day, the tumor had been the size of a softball. The tumor was so big when the doctors went in – they expected it to still be softball-sized – they couldn't see everything they were doing and could only hope to get it all out without hurting Mom in other ways. The expansiveness of the tumor was stunning and unexpected. Let me back up to another year prior to this date to add some necessary context.

Mom was recovering from the first big illness in her life – breast cancer. It was a stage 1 cancer that had not spread and was not high risk. No chemo was necessary, only weekly radiation treatments where Mom had found friends at each visit who she would cling to over time. A double mastectomy and some reconstructive surgeries had occurred, but she was recovering well at that point. Regular scans were being done, and life was moving along smoothly while she watched her grandkids' sporting events and theatre productions.

[Read Kathy's Full Story Here.](#)

Webinar Announcement

LIVE VIRTUAL SESSION WITH THE EXPERTS

LIPOSARCOMA

MARCH 27TH, 2024
3-4 PM ET

RAPHAEL E. POLLOCK, MD, PhD
Director, The Ohio State University
Comprehensive Cancer Center
Klotz Chair in Cancer Research

GARY SCHWARTZ, MD
Director, Case Comprehensive Cancer Center
Vice Dean for Oncology, School of Medicine
Gertrude Donnelly Hess MD Professor in Oncology Research

 **SFA**
Sarcoma Foundation of America
finding the cure in our time

SFA is proud to launch a new series of subtype-specific education sessions where patients will learn from and have the opportunity to interact with experts with experience caring for patients with their specific subtype of sarcoma. This series will empower patients with knowledge and resources that are important strategic priorities for SFA to promote joint care decision making as part of the sarcoma journey.

This first session will focus on liposarcoma. Expert panelists will discuss diagnosis, treatment, clinical trials, and current research specific to liposarcoma. Patients will have the opportunity to ask questions, learn about what makes liposarcoma unique among other sarcoma subtypes, and get information about current research and resources specific to their disease.

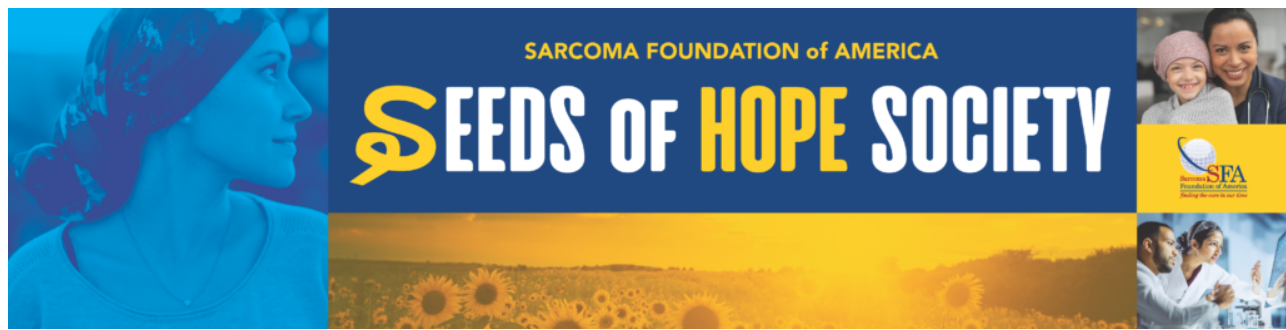
This session will empower patients to participate in shared decision making with their care team and find resources to support them on their sarcoma journey. Additionally, this session will be a resource for clinicians, caregivers, and industry partners to learn about liposarcoma from experts in the field.

Panelists

Dr. Gary Schwartz, MD, Director, Case Comprehensive Cancer Center

Dr. Raphael Pollock, MD, PhD, Director, The Ohio State University Comprehensive Cancer Center

[REGISTER NOW](#)



This month, SFA is launching the Seeds of Hope Society – a growing, passionate community of sustaining givers who have committed to making a recurring donation to SFA. When you stand with SFA with your recurring gift, you make a commitment to sarcoma patients and their families that they are not alone—that you are a champion they can count on through all the seasons of their lives.

As an important partner to SFA, Seeds of Hope Society members receive exclusive information and updates throughout the year, including:

- Quarterly exclusive updates of your impact on sarcoma community, including stories and experiences of those affected by sarcoma and a roundup of articles of interest to the sarcoma community.
- Special recognition in SFA's annual report and in SFA's program at the annual Stand Up to Sarcoma Gala.

Your active recurring gift to SFA qualifies you as part of the Seeds of Hope Society and the special member benefits listed above. Every dollar makes a difference!

Here's how your monthly gift matters to those living with a sarcoma diagnosis:

- **\$10 a month** supports increased awareness about the need for more research funding and treatments.
- **\$25 a month** can provide education and resources for sarcoma patients to ensure they have access to the best treatment options.
- **\$100 a month** can help advance research and clinical trials to realize lifesaving treatments for people diagnosed with sarcoma.

Turn your support into a flexible, convenient, and sustainable impact all year long by joining the **Seeds of Hope Society** community with a recurring gift!

JOIN SEEDS OF HOPE TODAY

In the Community

On what would have been her brother Ben's 41st birthday, Jessy Tolkan used Instagram to host a fundraiser for Sarcoma Foundation of America (SFA) in his memory. Unfortunately, Ben lost his battle with sarcoma in October 2020. Jessy has been a committed supporter of SFA, contributing significantly to our RTCS Milwaukee event by



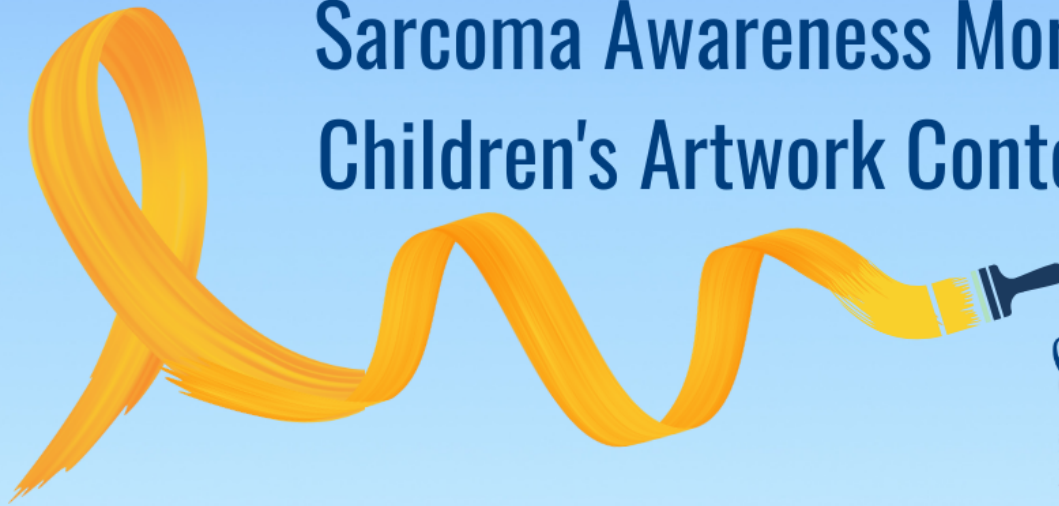
bringing together a large team and actively participating in our Gala committee. Moreover, she makes it a point to gather with friends and family on her brother's birthday, ensuring that his memory remains cherished and never forgotten. [Read Ben's story here.](#)

We are honored to share the story of a courageous young man named AJ who lost his father to sarcoma at the tender age of six. Inspired by his mother's determination to raise money for a cure, he has embarked on a remarkable mission. In June-July 2024, he will walk 200 miles from Porto, Portugal, to Santiago de Compostela, Spain, to raise funds for Sarcoma Foundation of America. Through his blog and Instagram, he will document his journey, inviting everyone to follow along and support his cause. Every dollar raised will contribute to the underfunded research for a sarcoma cure. We commend his resilience and commitment to making a difference. Together, we can bring hope and healing to those affected by this rare disease. As of this article the fundraiser has passed \$10,000. [Read AJ's blog](#) or [donate to his campaign here.](#)



SFA Events

Sarcoma Awareness Month Children's Artwork Contest



Artwork Guidelines:

- Artwork must incorporate a yellow ribbon and/or sunflower
- Artwork can be any size and any medium
- Artist must be 18 years or younger at the time the artwork is submitted
- Submissions must be original artwork

Submission Rules:

- Artwork must be received by July 22, 2024
- Please include the artist's name, age, city and state of residence, and connection to sarcoma
- Please provide an adult contact person and contact information
- Artwork will not be returned to the artist

The winner will be announced at the end of July

The winning entry will be featured in SFA's newsletter and appear on SFA's 2024 holiday card

To enter or get more info scan the QR code or go to the url below:



curesarcoma.org/childrens-artwork-competition/



Race **to** Cure
SARCOMA

SARCOMA FOUNDATION OF AMERICA

REGISTER NOW for the 2024 Race to Cure Sarcoma series! These events not only help raise funds for much-needed sarcoma research but also unite the sarcoma community across the country and help to raise awareness about the needs of sarcoma patients.



BIRMINGHAM - MARCH 23

ATLANTA - APRIL 6



SOUTH FLORIDA - APRIL 20

BOSTON - APRIL 21

NEW YORK - APRIL 27

SAN FRANCISCO - MAY 11

SEATTLE - JUNE 22

CLEVELAND - JUNE 22

MILWAUKEE - JULY 13

NATIONAL VIRTUAL - JULY 20

WASHINGTON D.C. - JULY 20

LOUISVILLE - AUGUST 10

PHILADELPHIA - SEPTEMBER 14

SAN DIEGO - SEPTEMBER 21

CHICAGO - SEPTEMBER 28

NEW JERSEY - OCTOBER 6

ST. LOUIS - OCTOBER 12

LOS ANGELES - OCTOBER 12

DENVER - OCTOBER 26

REGISTER NOW

Our Contact Information

{{Organization Name}}

{{Organization Address}}

{{Organization Phone}}

{{Organization Website}}

{{Unsubscribe}}

