Summary of ECD Global Alliance Internet Chat
31 Jan 2015

15 Attendees

*A number of members did sign in to chat, but then made no comments. I have included their logins in this list.

- A new member told us that his brother has ECD with a lot of bone involvement and tumor around his optic nerves. He tried Vemurafenib (V), but immediately got skin irritation & redness, photosensitivity, general swelling, & severe pain. He could barely move, even to lift a finger. The doctors stopped, reduced, and restarted the V, but he could never tolerate it for very many days. After the 3rd try he was removed from the study. He has since tried it again at just one pill a day, but couldn't manage even that. We were asked whether anyone else had had such a severe response, and what should be done now?

- A UK member came on to tell us that he couldn't be on the chat that night because he was going for a curry! He has been on V for some months now. His recent breathing test showed a normal lung capacity and lung function at 81%! A BIG improvement. PETCT showed nothing, with no active disease. That was said by the consultant looking at the images. He is awaiting the official report. He has been diagnosed with Type 2 diabetes. The pancreas gland was badly affected by the ECD, and the capacity to produce insulin is now lower.

- Another member who has been on V for a year has changed doctors, and has started a trial of a new drug, LGX818, or encorafenib. He now sees Dr. Braiteh, who, he thinks, used to work with Prof. Kurzrock at MD Anderson. "The V didn't work for me." He was admitted to the hospital with acute pancreatitis after taking just 3 doses of V. According to Dr. Braiteh, he then became immune to the drug's effects. The V probably was working for a while, but then quit. The last tests showed that the ECD is still very active in his legs. He has to take small steps when walking downhill. He also has continuing shoulder problems. He has just had surgery on the right one. A lot of scar tissue was removed, although he has no history of a shoulder injury.

  The new drug is aimed at BRAF600 mutations. The medicines just have LGX818 written on them. This drug was described as a "Super V". Another member told us that the new drug was called encorafenib. The member was asked whether he was also taking a MEK inhibitor. The answer to this was "no".

  When he started this drug he got scared because he started with the same symptoms, after the first dose, as when he took V. So far, he has "got over it". He feels a lot more tired. He can't get through the day without a couple of naps as it is. This is difficult when you are also the only caretaker for 2 boys.

- A member came on who does not normally get involved with ECD related stuff. He tries to forget that he has it. He thinks that this is a coping mechanism. He has had ECD for about 10 years, and lives in Maryland USA. His only physical symptom is due to his diabetes insipidus, but he has disease "pretty much all over".

He is 44 years old and has a wife and 2 children. He works full time for a government healthcare agency. His first treatment was dexamethasone and this had no effect. Then he was on interferon for 6 years, which halted the disease. He was prescribed a large dose of interferon (3 MU every day!). Three times a week was the recommendation at Johns Hopkins. He thinks that his local oncologist just got very aggressive, "but whatever, it worked". He had to discontinue it because he developed Interferon Induced Sarcoidosis after 6 years on the drug.

He stopped Interferon 3 years ago and has been on nothing since, until starting on a new trial. He has just left NIH, and is the first enrolled patient in Protocol 15-HG-006 under Dr. Estrada-Veras. This is testing a new drug combination; Dabrafenib and Trametinib. He has always wanted to take a conservative "do no harm" approach, and so he was reluctant to enroll in this trial since he was stable. However, he asked his local doctor for his thoughts, and he said that if there was a chance to actually knock this disease back, it should be taken.

This trial is being funded by the National Cancer Institute, and run through NIH. He is only on Day 3 of meds. The second medication is a MEK inhibitor which works on same molecular pathway as the BRAF inhibitor, just later in the process. He thinks that resistance develops to the BRAF inhibitors, and that this is the rationale for including a MEK inhibitor in the therapy. The only side-effect that he has noticed so far is dry lips.

He will have to go to NIH in months 1,2,4,6,8,10, and 12. Bethesda is a little over a one hour drive away, but lots of DC traffic can easily turn it into three hours!

He asked whether the chats were still being archived (summarizer's note: YES they are!!!), as he has not been getting them. He was advised to contact Kathy and to get put back on the mailing list.

- A member, who has been on V for 14 months now, told us that she is quite stable with no active disease. Poor balance is her main problem. She thought that she was going to need a wheelchair 1½ years ago but "now everything has changed".

- A member came on who had had an MRI in the previous week. He was relieved to find out that he "still has a brain!" His balance is not good. He has been on interferon since 2008 and this has stabilized him. He started on 10 MU 3x a week, and is now only using 3 MU 3x a wk. The doctors won't take him off interferon and try V, because he is doing so well. "Why fix what ain't broke!"