

Summary of ECD Global Alliance Internet Chat **18 April 2015**

8 Attendees

- A member came on to say that he felt too poorly to chat today, but "on the mend".
- Two members who have both been on vemurafenib (V) discussed their experiences. One has been able to reduce her dose to 1 in the morning and 1 in the evening, and had been out walking for, probably 2.5 hours! The other member had "had the worst day since I started V!" His face was burnt and lips toasted on the previous weekend when he spent some time outside in the sun. He had thought that sunscreen, umbrella, etc. would protect him -- but NO! He now has blisters all over. His dose of V is higher than that needed for most "in remission". He doesn't like the idea of giving up the sun for the rest of his life. He will need to find a different maintenance drug!
- Interferon was mentioned. One member said that it had made her tired, but not better. Another had been on it for 9 years. "It was awful. But I think it kept me alive". This member has been on nothing since last September! Over the last 13 years he has "taken every conceivable scan multiple times". There seems to be no problem behind his eyes and a recent camera view of the ureter showed that the vemurafenib had greatly reduced the obstruction. After interferon, he went onto Kineret, and in January 2014 he started vemurafenib. V didn't seem to be having any positive effect, it was damaging his kidney, and so he stopped it last September. He has not taken any medication for ECD since then. His eyes are normal. There is fibrosis in the kidney area, that obstructs his left ureter, and he has a stent in the ureter to keep open the connection between the kidney and bladder.
In 2001, his eyes bulged out so much that he couldn't close his left eyelid. But soon after he started the interferon, they retracted to normal and have been normal ever since.
He was not part of a V clinical trial because he is in L.A. The doctors "simply prescribed it". He sees Dr. Michael Lill at Cedars-Sinai Medical Center. He started at 8 a day, but his creatinine level shot up and the dose was quickly reduced to 4 a day. If he was on a trial this would have meant that he would have had to stop the trial, but since he is "off-label" he is able to continue at a lower dose. If it turns out that there was a positive response to the vemurafenib, which MRI's didn't detect, he is close to resuming it. His doctors want to do a few more tests before he restarts. He is eager to do so.
His diagnosis was made at the Jules Stein Eye Institute at UCLA in late 2001. The article about interferon being effective in halting ECD came out about then. He went on interferon in January 2002. His eyes retracted to normal within three weeks. MRI's showed the disease behind his eyes had been greatly reduced by the interferon. "Interferon has brutal side effects, but its impact on the disease is powerful too."
- The other member told us that V has done wonders for him, "but it has also brought along a ton of trouble". This is mainly pain and inconvenience from the side effects, and also the costs of travel, and lodging, and a ton of bills. The other member suggested that maybe he should drop out of the trial and get the drug by prescription. His main doctor at Mayo is a pioneer, and authority, in interferon -- and he "refused" to give him interferon for his ECD.

- A member came on who is on a specific treatment for ECD. She has had high blood pressure and some other side effects. She goes to NY on the 27th for a PET and MRI to see if the drug has done anything. She still has macular edema. A test called an OCT, performed by the retina specialist, detects this, and it is basically leakage into the macula that causes blurred vision and eventually can cause vision loss if left untreated. The only thing that helps her is steroids.
- Timetables at treatment/ investigation centers were discussed. Some members have a very full and busy timetable, others have an easier time of it!
- Two members came on after the Chat had finished. One is in DC at her daughter and son-in-law's, and will be admitted to NIH tomorrow.
- The other member told us that she has been feeling ill for about a week, and on that day, she finally had some energy, so she was outside doing what she could!