

## **Summary of ECD Global Alliance Internet Chat**

**21 May 2016**

8 Attendees

- The member who has recently started vemurafenib (V) said that he was very well. He has just had a follow up with the doctors who were very pleased with his progress. His heart is not "skipping a beat" anymore on the current combo of drugs. The doctor who sees him for his ECD hadn't seen him since February and was astonished at the changes.

On his previous visit he was in a wheelchair; and this time he was walking on his own feet. The follow up of his brain, heart, legs, etc. will happen in 6 months and physio will start in a month. They heard another ECD patient, who was wheelchair bound, was about to walk around the block with help from physio. "So there is hope yet."

- A member is about to give a couple of presentations at her workplace, one in the evening for friends, and the other during the day for patients and staff. She is going to speak about her "whole journey"; why she sometimes walks zigzag in the hallways, and her motivation to exercise. She is "kind of getting cold feet", but the invitations have been sent out. "So no turning back".
- Members discussed some of their eye problems. One has tumors in his orbits, but they are not causing visual problems. Another member had very serious bulging of his eyes and almost lost his vision in 2014. Vemurafenib "fixed" the bulging and swelling quickly. The eyes receded back quite a bit within months, but not to where they used to be. He has now been on a low dose of V for a while, and he is surprised that his eyes are still getting better over time.
- A new member from France told us that he has disease affecting his orbits, heart, kidneys, and some other places, too. He is on a dose of 1 pill in the morning and 1 in the evening of V. He started with 2+2, but only for a few days in October 2013. He took 1+1 until summer 2015. When he stopped V, the ECD flared up and he had to restart on 2+2, but he found this difficult. He was always tired. 1+1 V seems to be the right dose for him. He is OK if he has a nap each afternoon and avoids the sun. He sees Prof. Julien Haroche and is being admitted to hospital for a week every 3 months. He sees Prof. Haroche at the end of each week of hospitalization. The Prof. has given the member his phone number, so that if he has a problem and wants to speak with him, he can. "He's a good guy."  
The next time that he has to go to the hospital (Pitié-Salpêtrière in Paris) is in September, so he will stay for the symposium. In France, they don't have to pay for the medicine (60 tablets of V cost nearly 1,800€!), or the hospital. There is even help to pay for the transport!
- We were told that a member in the UK has posted on Rareconnect that V "pretty much took care of everything". He still has some scarring on the heart, but nothing else. He is on 1 V a day for maintenance.
- A member who has been on V for a while still has lesions in her brain, and a new one on her sternum after a year. She wonders whether the V is working now. In melanoma, it is common for V or D+T to stop working after a few months. In melanoma trials, some of the patients who developed

resistance to V would see their BRAF level come back up a few weeks before relapse, even when they were still on V or D&T! So far, ECD does not seem to react in this manner.

She had a PET scan in April. Dr. Diamond didn't seem concerned, but when she emailed him this week about dropping from 3 to 2 pills per day, he said that she should not do this until after she has had a PET in August.

Memorial Sloan Kettering (MSK) did the PETS in April and her insurance company won't pay. They said that she didn't need them! She thinks that unless this last PET gets paid for, MSK will not do another pro-bono. She was told that if MSK had no authorization for the scan, then she is not responsible for the charge. She said that MSK had tried to get it authorized, but it was denied. MSK will cover the cost this time, even though it was denied before the scans and also on appeal. She is going to get her attorney to say "hello" to her insurers again.

- Members spoke about rashes. One has developed a rash on her abdomen. She was told that it was probably due to the V. Another member said that every time she gets in the pool, she gets a rash on her abdomen. She wondered whether it could be due to the chlorine. She always showers afterwards, to get the chlorine off and then the rash dissipates.
- A member who exercises a lot, thinks that the rash is due to a reaction of V with the skin when you are warm/hot. He has more rashes after he runs, but he feels much better at the same time. His "theory" is that running generates heat and increases blood circulation, and that this might increase V's availability in the blood.
- Heat is always an issue if you're on V, but at the same time, it's an indication that V is working. However, you want the areas around the tumors to have the activities, not the skin.
- The reclassification of ECD as a "histiocytic neoplasm" by the World Health Organization (WHO) was talked about. This will probably make it easier to tell doctors and other people what illness we have!
- The Trovogene urine test for BRAF level was mentioned. This cost about \$1,700-2,000 retail. The urine test is established and insurance companies are reimbursing. A member who has recently started V was sent a package to test for urine, with specific instructions. The result came back very quickly.
- A member said that her sister has planned a trip for them to Wyoming and Washington State at the end of June. She said that she didn't "want to get stagnant. That's when bad stuff grows!" And another said that that was why he runs every day.
- After the Chat had finished a member left a message. He had missed the Chat because he was at "yet another Communion". He is now on Dabrafenib after having a rise in his liver enzymes when on V. He has been on it for about a month, has no side effects, and feels "great".