

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

**28 May 2016**

11 Attendees

- Two of the chatters speak French as a first language, and their English is limited. They chatted between each other in French. I cannot translate this myself (schoolboy French was 40 years ago!!). Sometimes Google Translate was used, but not between them, so those bits of Chat are not here.
- The member who told us last week that she was getting involved with a local paper has given the interview. She goes to Mayo this week to see Dr. Habbermann to start on some kind of treatment for the ECD. She said she was excited to finally start treatment and nervous "all at the same time". So far, the ECD is just affecting her eyes. The eyelids are discolored and yellowish, very swollen and appear bruised. They hurt all the time like they have sand in them.  
She is BRAF negative, so some of the treatment options, such as vemurafenib (V), will not be available. She is also going to ask for her lower leg to be checked, which has been bothering her for years. It will be interesting to see what Dr. H is going to give her since she is BRAF-. She was advised to try and stay away from chemo.
- A new member came onto the Chat session. He lives on the Sunshine Coast in Queensland, Australia, just north of Brisbane. He says that it lives up to its name. "No better place to languish!" He had first thought that something was wrong when he spoilt a holiday by suddenly not being able to do normal things, such as walking, swimming, and staying awake. His shoulder and upper arm pains seem to be easing after 4 months without treatment. He does take 500mg of calcium with 400IU vitamin D most days.
- Bone remodeling was mentioned, although most members had not come across this term. The amount of vitamin D that people were taking was mentioned. One was taking 1,000 IU a day, another was taking 2,000 IU.
- We were reminded of the recent press release following the WHO reclassification of Erdheim-Chester Disease as a histiocytic neoplasm. This means that ECD is now considered to be a blood cancer. The doctors will be able to approach the cancer societies for more funding for research, and the health insurance companies can now classify treatment as cancer treatment. There shouldn't be much of a problem getting experimental drugs approved.
- One of the French speakers has just had a biopsy to confirm ECD. At the moment, he is treating himself "with plants". He said that avoiding pineapple might be good as it can destroy useful enzymes. After the biopsy results, he will be offered treatment. His hospital is in contact with Prof. Haroche in Paris. He said that he also has dry eyes, painful legs, back, and fingers. He has had leg pain for nearly 10 years, and the legs feel to be warmer than the rest of his body.
- In the information that comes with V there is a warning about the use of certain herbs and some herbal teas, etc.

- The member who was swapped from V to Trametinib (T) because of a rise in his liver enzymes said that he was doing well with no side effects. He has been doing more things at home; raking leaves, mulching, washing the cars, etc. While on V he would get a sunburn in minutes. Now he isn't burnt if he is out for 10-15 minutes. He has no rashes, just some keratosis. He sees a dermatologist every 3 months or so. He doesn't have to monitor his heart. (Heart issues are relevant to a member who may be transferred onto T). He takes 3 pills in the am and 3 in pm. This seemed a high dose to another member. The member on T thinks that he has been given this dose because he was on and off V for quite a long time. He said that although he didn't know his prognosis, he imagined that life expectancy was now a lot greater than the 50% 3 year mark that is in older literature.
- [Statistical lesson from the Summarizer; The 50% 3 year figure is the MEDIAN life expectancy and NOT the MEAN (=average). The median is a statistical term that means that 50% of patients have died by the third year after diagnosis. The other 50% may do so much better that they live a long, long time, and that will bring the MEAN expectancy up a lot. On the website there is an article called "The Median is not the Message" which explains all this. See - [http://cancerguide.org/median\\_not\\_msg.html](http://cancerguide.org/median_not_msg.html) ]
- A member, who goes to Memorial Sloan Kettering (MSK), had tests done at the end of March but doesn't have another appointment set up yet. She is still waiting for the T, having been off the sorafenib for a while. She still needs to take steroids for her eye problem (macular edema). She said that Dr. Diamond seems to be getting very busy. She recently had a scare after 3 areas of calcification were found on her mammogram and they had to be biopsied. Fortunately, they were all benign.
- One member has masses all over his legs, although his knees were the most painful part. V suppressed the pain as time went on, but his legs are still stiff in the morning. The leg problems make it hard to walk (along with his balance difficulties). He can walk now, with his wheelchair or with his walker. He has been taking small steps on his own for short distances. With the aid of the wheelchair, he can transfer more safely. He has his own caregivers in the morning, so the family does not have to worry about him "every single moment". Before treatment he was falling quite often. Before V he also had pain in his arm and shoulder and this is still an issue. He is waiting for physio to see if that may resolve things. The biopsy for the diagnosis was from his "hairy kidneys". He has recently walked down the stairs to check out the new gazebo on the patio. He had a chat with the neighbor, and was outside for over 1.5 hours. Then he walked up the stairs. There is a huge difference in how much he can do now.
- A member who is on V treatment said that he had a constant low level of pain in his hands, lower legs, and feet. He cannot make a fist, and has bumps and raised places in his hands and feet.

(T) = trametinib

(V) = vemurafenib