

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

### **10 Dec 2016**

11 Attendees

- A member told us that he had recently had a phlebotomy (drawing of blood). They drew off a pint and then "threw it away"! He thinks that this may be due to the extra testosterone that he is taking, making his blood too thick. Removing blood like this helps to make the blood "thinner" as the body fills the space, that is made by the removal, with serum and not more blood cells.
- A member on vemurafenib (V) has had a release of her trigger finger on the left hand. The wound from the hand surgery is not healing too well, and she wonders if this is because she is taking V. Since she is left handed, she said that she "might not type well". She goes to see Dr. Diamond this week, has a PET/CT scan, and sees the dermatologist. She is hoping that bad weather does not keep her from flying to NYC. She leaves at 6:30 am and has a PET/CT booked for 10:45!  
She is beginning to think that the V "is screwing me more than ECD". She is now down to a dose of one in the morning and one in the evening, but her joints are a mess. She had a steroid shot in her shoulder a couple weeks ago, but her shoulder is still "wacko".  
A member suggested that she should ask Dr. Diamond to switch her to dabrafenib (D). She said that he may make the switch without her needing to ask! She saw a cardiologist this week because her blood pressure is wildly up and down. The cardiologist picked up a bruit (a noise due to turbulence in the blood flow) in the left carotid artery (in her neck).  
She has been asked to join the new ECD registry that is being set up in Canada, and is going to do so.
- A new member also thought that taking V is worse than having ECD, except that without V he wouldn't be able to worry about it, "because I wouldn't be here!" He found that knots under the skin seemed to come and go in the first year, but now are gone. He has noticed that certain symptoms come such as arthralgia [joint pain], and often, after a year, they go. He told us that his blood pressure (BP) is also totally erratic, but he thinks that this is due to stress.  
He asked again about neuropathy. He has had Type 2 diabetes for 25 years (this often causes a peripheral neuropathy), but he feels that that is not the whole reason. He was diagnosed at the end of 2012, after a fruitless year and a half of investigations. He got on the internet and "miraculously" got in contact with Dr. Scheel, who, at that time, was one of the only doctors in the US who had many ECD patients. The thing that set him off down the ECD trail was that he had been told by his radiologist that he had retroperitoneal fibrosis. He has also been using Kineret since early 2012. He goes to John Hopkins hospital for treatment.
- Another member had had some peripheral neuropathy in her legs before she was diagnosed with ECD.
- The tumor-like lumps and knots that occur in ECD were discussed. One member had an ECD tumor in her spine, but the V worked its magic and it went away. Another said that he had growths all over, although he has been stable on interferon for years. He said that when he has a bone scan, it lights up like a Christmas tree!
- A new member asked about such growths. He has one, the size of his little finger, on his cheekbone. He asked about peripheral ones growing between bone and skin.

- One member said that she has "knots" in her skin that come and go. These are mostly on her hands and feet, none on her face. She also has arthralgias (painful joints) all the time.
- Members discussed the differences between Vemurafenib (V) and Dabrafenib (D). One member was started on V, but then was changed to D, because his liver enzymes rose while on the V. He tolerated V at 4+4 for five and a half months. He said that he has noticed reduced muscle strength. "I couldn't do more than 10 push-ups." He has got a little bumpiness on the sides and backs of his arms.
- Dabrafenib is used for BRAFV600E positive ECD, just like V. They seem to be functional equivalents. For one member, the main issue is that it is taking so long for V to get FDA approval. And since there is no trial for D alone for ECD, how could it ever get approved? [Editor's note: There is a trial ongoing at the NIH for the use of Dabrafenib and Trametinib in combination for the treatment of ECD. It is hoped that this trial will be an important step in getting this combination treatment for ECD approved by the FDA.]
- Members discussed which doctors they see and where. One said that he had just learnt that his local oncologist has not spoken to Mayo Clinic since his diagnosis in July, and his local oncologist had never heard of ECD before then. It was said that we should not rely too much on "official communications", but be our own advocates. The member, whose own doctor has not been communicated with, is going to Mayo later this month and plans to clear up a few misconceptions of who is doing what. "This is my life and I need to make sure I am treated appropriately."
- Another member said that in order to be his ECD doctor, the doctor must listen to him, and let him tell the doctor his point of view.
- It was suggested that experienced ECD patients should author a paper on what to expect - since it appears that "the wheel is re-invented" every time someone is diagnosed. One member said that she likes to write. She also has a medical background, so she would know what to include, and what is just too complicated to include.
- Eyes were the next topic. One member said that her eyes go blurry after reading or typing for just a little while. Also, her eyes are so dry when she wakes up, that she can't open them without eye drops (over the counter (OTC), not prescription).  
No one else had this symptom, but others said that it was important to "relax" your eyes often. It was said that you should apply a cold or warm compress to relieve the eyes (try using a warm wet cloth on the eyes for 5 mins in the morning and before bed time). This helps to open up the glands, which produce the secretions that lubricate the eyes. A member had used more cold applications when he was on a high dose of V, due to the heat it generated.
- After the Chat, a member came on with an update. She is doing well apart from some stiff joints. She knows that she has had many simple procedures that have helped a lot, and many things that didn't do a thing. She said, "We just have to keep hope!"