Summary of ECD Global Alliance Internet Chat
17 May 2014

12 Attendees

- We were told that it was a national holiday in Norway, Independence Day. Our Norwegian member told us that she is doing well and it "feels great to be able to say that". She is taking 1 and 2 of Vemurafenib (otherwise known as V in the chat).

- Another of us has just spent a busy week at NIH. Little time was available to go and see anything. On 2 days he didn't get finished until 9pm. This member is on V. He has some questions as to whether it is working or not. According to his PET scan the disease is still spreading. He doesn't have the results to all of his tests yet. But, in the last two months, he has lost 40-50 percent movement in both shoulders. He is taking 3 and 3 V. He tried 4 and 4 but this made him very ill with pancreatitis. He has been told that if he was in the clinical study, he would possibly be withdrawn because of something it is doing to his heart. He is "borderline on something that has to do with the timing of how long it takes for my heart to beat". [This may well be his QT interval. This is a measure of the speed at which the electrical pulse, that coordinates your heartbeat, passes through the hearts "wiring"]. He has osteosclerosis in his joints and the docs don't know how to treat this.

- A member has bought plants today and needs to "puttem in the grnd". A member agreed with this because "it don't work if you puttem anywheres else". This member was also suffering from a headache at the time of the Chat, which was affecting his balance and vision.

- A member came on who has not been with us for a while. He has been on a cruise for the last month. It was great, but his "arms ached from all that rowing". His walking and balance are worse, and he has just thrown his back out, too! He has ruptured a disc. A CT scan shows no new growth of the disease. The treatment for his back is just rest. The Doc suggested surgery but he doesn't want to have any if he can help it.

- One of the members on the chat told us that she was the "Wife of" a member who has recently gotten the ECD diagnosis. She does the typing for him. [A number of other Chatters "revealed" that they were partners of patients, and that they helped out by doing the typing!] He started V 2 months ago, at a dose of 4 and 4, and has seen MAJOR vision improvement since starting the drug! The tumors in his orbits were squeezing the optic nerves to the point of losing vision completely, in right eye at times. The neuro-opthamologist was becoming alarmed. Then they re-biopsied the tumors, did several other test & scans, and finally diagnosed ECD. This news came as a particular shock since he was diagnosed at a different hospital, last year, with a totally different disease, and was having treatment for that!

- The neurologist of one member has written an article about her and Vemurafenib. She will distribute it amongst our group as soon as it gets published. The Doc might have problems getting it published if it is distributed before publication. This member has been on V for 5 months, and it has had a really good effect. Her nervous system and skeleton were affected by the ECD, and MRI and PET scans show massive improvement. The poor balance and stiffness that she had are slowly getting better.

- One member is slowly getting life together again after the recent death of his mother-in-law.

- We were asked whether we had problems with our hands. Many of us do have difficulties. This is why a lot of us need help for typing.

- A member said that his Doc didn't want him to try V. He is stable on his interferon treatment, and there are concerns about whether insurance would fund V. One of the things that the doc was worried about was dropping of the blood count due to the V, making him more susceptible to viruses and such.

- Members discussed whether they are going to attend the convention this year. A number said that they would be attending, and were looking forward to meeting up.

- A member has not been doing so well. He has had pneumonia and strep infections. He will be seeing a doc in the next week. Dr. Estrada gave him a treatment for the infections, but it doesn't seem to be working very well. He is unable to take the regular drugs for this because of his allergies, and the bad interactions with V.

  He is also not sure if V is working on him. The ECD has spread to his shoulders now. The PET scans show that it is still spreading, even though he has been on the V for a couple of months now. He is still waiting for the results of all the tests. They have earlier bone scans and MRIs to compare with.

- A member, and his wife, have recently attended a marriage workshop. His wife didn't feel that they had any problems but he wanted "to make sure it stayed great, like it is".

- A member came on who was suffering from a cold with everywhere aching.

- A member told us that she is making her first trip to NIH in the week of Aug 18. This member takes Kineret and methotrexate.

- A number of members said that they now seem to cry a lot now. This applied to both ECD subjects and to their caregiver, and they could be set off crying by very simple things. The caregivers were reminded that it is important for them to take care of themselves. It was recommended that they find someone who they can talk to about it. They shouldn't be taking on this caregiver role on their own. They were invited to contact Kathy at the support@erdheimchester.org email address, if they would like to be in contact with another caregiver of an ECD patient.

- A member said that "Doctors just don't give us any sense of reassurance. We always leave feeling dismayed". Almost any ECD patient can go to the NIH, and be enrolled in their study. You only need to contact Dr. Estrada-Veras to get started. You will see all sorts of specialists, all who have seen multiple ECD patients! They will run many, many tests and then sit down with you to tell you what they mean. The doctors will talk with you. For most, it is a great experience. Most patients and their family are glad they did it.

- A UK member who is on V told us some good news. His lung function tests are all showing improvement. PET scan showed thinning of the pleural effusions, and the only disease activity was in shin and a foot. He has not had the official results of his cardiac MRI, but the doc that did it, said that the growths on the heart, and aorta, had shrunk. He has had some TIAs (Transient Ischemic Attacks- like having a stroke, but the effects last for less than 24hours) which have slightly reduced his joy at feeling better! His blood pressure is high. He had a 24hour BP monitor on this week, and this was blowing up so hard that he had to stop after 12 hours. He is on medicine to make his blood clot less easily (Clopidogrel), and a statin to reduce blood lipids. He has had his renal function checked to see whether this is a cause for his high blood pressure. Despite these difficulties he said that his wife is coping amazingly well. He has to remember to always wear a hat when he goes out. His head starts to burn in about 5 secs (a side effect of V).

- The Prof that sees most of the UK members has told one member that he is going to a big ECD meeting in Sept. Kathy was aware that he intends to come to the GA conference and looks forward to meeting him. He seems to be ahead of the game through his other research. He is the UK head guy on a few blood diseases.

- For those that do not test positive for BRAF there is a drug coming out that may get another pathway, MEK.

- One member does not have support structures close to him. He is also a single parent, bringing up 2 boys, one of which has special needs.