Summary of ECD Global Alliance Internet Chat
13 Jun 2015

12 Attendees

- In the days between the Chats a message was left by a new member. Her husband has had ECD for 6 years, and she said that they were "going through some tough times now".

- A member who was troubled with knee pain has been found to be deficient in vitamin D. He suggests that if any ECD patients are troubled by aching knees, hips, or other bones and they haven't been tested for their vitamin D level, they should ask for it to be done.
  This member has been going to a place called Whitby (the one in the Dracula story) most weekends. This is on the North East Coast of England, and they don't have internet coverage there so he hasn't been on the Chats.

- The ECDGA has been contacted by Dr. Achille Aouba in France, who is interested in doing a retrospective study of the use of anakinra as an ECD treatment. This study will not be responsible for administering treatment, only looking at the records of patients who have been previously treated with anakinra. An email should be sent soon with more information, but if you are interested, please contact Kathy at support@erdheim-chester.org.

- A member is starting her second month on vemurafenib (V) at a dose of 2 in the morning and 2 in the evening. She is tired and joints, like her fingers, wrists, and knees hurt every day (but not all at once). So, overall, she thinks that V must be having an effect.

- A member reminded us that V hadn't worked for him. He is BRAF positive. He had started treatment with interferon, but the disease just progressed. He was put on V, but ended up in the hospital three days later. He was the second person ever to develop acute pancreatitis due to V. A couple of weeks after this he restarted at a low dose and built back up to 4+4. After being on it for almost a year, it was decided that V wasn't working. He is now on an experimental drug called lgx818, or encorafenib. He has been in a trial for about 4 months.
  He is still in lots of pain, tired all the time, and "unable to get things accomplished". His latest tests state that the disease is stable, but active. He is experiencing several side-effects with this new drug. He has lost almost all of his hair (ALL OVER), and he is having problems regulating his body temperature. He has noticed that he doesn't sweat much anymore.
  Two weeks ago the doctor made the comment that he would put him on another drug, if there was one, but there isn't. Because the doctor mentioned this he thinks that, just as with V, this trial drug is also not working.
  His doctor, Dr. Braiteh has told him that he intends to go to this year's conference. The member, himself, doesn't think that he will be able to afford to go. Dr. Braiteh is in Las Vegas NV, and he used to work with Dr. Kurzrock at MD Anderson, before she moved to San Diego.

- The member, who has just moved to San Diego, has already been to see Dr. Kurzrock. He had been on a V trial already but his disease is now in remission. The meeting with Dr. K well, and he will see her again at the end of August. He actually saw her, and not one of her staff, and she reviewed all his records and provided an independent opinion. She was very thorough, and had requested that all scans, lab work, pathology reports, BRAF sequencing tests, etc. be sent to her office before the appointment.

- A member who has just started V (with fantastic results!) told us that he was "doing great". He is on a dose of 4+4, and has had "no side effects".

- Kathy explained why her Chat name was Garyskat. Her late husband (who had had ECD diagnosed postmortem) used to call her "Kat". When the Chat room was started, she was uncertain about using her true identity, and so she chose "GarysKat", and it has stuck.

- Members on, or who have been on, V discussed side-effects. One got very blistered lips, a week ago, from indirect sunlight, so she is now "very, very careful". She has learned! She also has a fair amount of red bumps on her legs, arms, etc. (not a rash). One of the other patients had also had this. The first member said that she was "swimming" in a warm water indoor pool about 3 times/week, then showering the chlorine off, and using lotion to keep her skin moist. AND she stays OUT of the sun, ALL DAY. (Summarizer's note; She's learnt alright!)

- The member, whose daughter has ECD, came on. Because her sight has been damaged so badly, the chat isn't easy for her. On the previous day, they had visited a recumbent bike shop. She, and her husband, already have a tandem bike, and they go out around town. They do a few miles a day, and are working their way up to more. Her balance is also a problem, but she is in remission. While they were at the recumbent bike shop, they found a pedal-powered wheelchair, which can be steered! If one has leg issues, it might not be the right thing, but if it's a balance problem and one has the leg strength, it is great. It costs $3300! They live in SE Pennsylvania, and she gets her treatment at The Hospital of the University of PA in Philadelphia. A LONG list of specialists work/worked on her case: oncology, neuro-ophthalmology, ophthalmology, cardiology, and more. She has been to the NIH twice, and they would like her to go again.

  He said (the father of the ECD patient that is) that he is "fine. Just getting older and not being as active as a retired guy should be".

- Testing for BRAF was talked about. Most patients only need a single test. Some have it done more than once, on the same tissue sample to confirm the result. For one member, 2 separate samples had been tested.

- Then the procedures for some of the trials for drugs developed since V, were mentioned. Our member who failed V, and may be failing on a further Rx, was asked if he had inquired whether he could be on one of the other NIH clinical trials (although they are not supposed to take anyone who has been on another of the BRAF inhibitors (kinase inhibitors)).

  The member, who might need to be on such a trial, reminded us that he lives on the West Coast, and the trials are all on the East Coast. We were again reminded that it's "absolutely free", except for your time. However, he is a single parent with a special needs child, and has been told that, since he took V (even though this was off label, and not in one of the trials) he doesn't qualify.

- One member said that she had been at NIH for a week at the end of April, and didn't qualify for the two "new" kinase inhibitors (Dabrafenib and Trametinib), because the lesions in her brain were too small, but perhaps they would try them on him.

- A member who had been to the NIH said that all of the doctors that she had seen, were all so respectful and knowledgeable (and nice!). Members agreed that the time at NIH was very busy.

- The ECD history study was mentioned. A member, who could not be part of a trial, said that "At least I will get to go back there next March or April".

- A UK member came on, who lives, for most of the time, on a Greek island. She has not been able to chat "for a long time", because the time difference makes it difficult (it is late in the evening) and she tends to "need early nights these days". Her most recent PET scan, after eighteen months on methotrexate, showed that her bones had improved a little, so she is feeling more hopeful now, if not physically much better. She has new pain killers that are working, but they make her feel sick. "You can't win with these drugs!" She still needs a stick to walk some days, but she is swimming every day in the "lovely" Greek sea, which is the best therapy. She did make quite a few typos, but blamed it on the ouzo that she had had earlier ("I know that I will sleep well!").

- Another UK patient admitted that he was "having a beer", too. He is working hard as he and his wife have decided to move house, and need to make it look good for the estate agent's (realtor's) pictures.

  He is thinking of moving to North Lincolnshire, which won't be far from Derbyshire where another UK member lives. They will be able to meet up, and both of them want to do so! They have already met, when she has flown in from Greece for her clinic appointments (the North UK Chapter of the ECDGA!). She has met the other member's dog, and asked "How is that lovely Doggie?" She was told that he "Keeps us all busy and he's got such characterful little ways".