Summary of ECD Global Alliance Internet Chat
31 May 2014

15 Attendees

- Some of the attendees had come onto the chat pages before the chat actually started.

- A member left us a message, saying that he had fallen at home and injured his back, and so would not be chatting [since then he has had a short hospital stay, probably has a broken rib, and is back at home. Message; in future, you need to drink more water with it!].

- Members discussed plans to go to the NIH. There were 4 chatters planning to go during the rest of spring and summer of this year. Members wishing to go to the NIH need to contact Dr. Estrada Veras via email. Go to the ECD Global Alliance page or click http://clinicaltrials.gov/ct2/show/NCT01417520?term=erdheim+chester&rank=1 for the clinical study details. The NIH historically has paid for the flights from within the US and the hotel stay for a companion. "They are very nice and accommodating and the doctors are super sweet and helpful" said one member.

- A member told us that her husband is on V, but doesn't feel "any sense of direction with our doctors who are so unfamiliar with ECD". The wife of another member who is taking V said that the drug's effects seem to have plateaued a little. He has been taking it for 10 weeks. At first his vision improved remarkably, as the swellings around his optic nerves went down. He has pseudo orbital tumors behind both eyes. Now he feels that the right eye is starting to bulge a bit.

- A member negative for BRAF is on interferon, but this doesn't seem to stop the increase in bone pain.

- Another member was taking interferon, and it helped with her bone pain at the beginning, but she became accustomed to it quickly, so she changed to Anakinra. This isn't helping with the pain, but it does seem to help with her rash and her brain lesions.

- 2CDA (cladribine) had been mentioned to one member, but no-one on the Chat had had experience with this.

- V (vemurafenib) is the drug that is being used in a clinical trial for BRAF positive patients, and being used off label by some patients. Members had heard that there was a new drug being tested for BRAF negative individuals.

- Members discussed their need to take analgesic (pain-killing) medication. Some people felt "fuzzy" or sick on them. Aleve (naproxen) was recommended by some. This is a non-opioid drug and so it doesn't have those side-effects.

- A number of members suffer with deep bone pain, some of whom are on V, and some of whom aren't. This pain is probably related to the ECD. A member found that when he cut back on steroids, the pain got worse, although he did still have some results from Aleve. Opiates drugs mask the pain. Aleve is an anti-inflammatory, which is what you need if there is inflammation.

- One member sees a pain management specialist at Northwestern in Chicago and is trying various combinations of medicines.

- A member is tapering off his steroids at a rate of 1mg a week. He has been on it for more than a year and wants it "out of his life"! He has got down as far as 18mg daily. He also has foot neuropathy, which means that there are times when he can't feel one (or both) of them. This makes balance issues even more challenging!

- A member has deep bone pain. This was hard on her knees. She had been a catcher in softball. Since she stopped she has bad pain in her low back, and right shoulder. She didn't notice much difference when she was on steroids. She uses Advil and Aleve but these are "no real help". Her pain team is at the University of Chicago, and they are stumped. They "try so much" and it doesn't help. She got injections in her back, and these helped greatly, but only for 2 days, when they had told her that the effect should last at least 2 weeks.

- We were asked whether any of us suffered from eye tumors. One member has been struggling, for a year now, with constant headaches, blurred vision, and balance problems. He is worried that the orbital tumors may be returning. He has a brain MRI on the 11th of June, and one from March to compare it with.

- Balance issues were discussed. A member said that she walks around like she is "wasted" all the time because of her balance problems. She said that her friends start making some really funny jokes about how much she has had to drink. She has had problems with excess spinal fluid, but the docs are not sure of the cause of her imbalance. She has had her heart and inner ear checked out, but nothing has been found. Her vision is 20/20.

- Another member has had problems with optic nerve tumors. He told us that his vision was, sometimes "really funky". He has had an eye exam recently. The doc said that his field of vision has actually improved this time. However, he is still seeing the optician soon about glasses to help double vision. When he needs to change his fentanyl (a strong pain-killer absorbed through the skin from a patch) the vision sometimes gets worse. It was suggested that maybe the pain-killing medicine relaxes the muscles around the optic nerve. One member sometimes uses a small dose of Valium (Diazepam), and this can help, but he hates feeling 'out of it' due to the medicines.

- A member told us that his "Magical ECD Mystery Tour" had begun in March, with a visit to the ER with pink eyes. He had many trips to eye doctors, and an MRI scan. He was misdiagnosed, and told that he had lymphoma, with brain and orbit involvement. No one could take a biopsy until he went to the Mayo Clinic. [Mayo's radiologist speculated that it was ECD before he even arrived for orbit surgery, to get a biopsy!]. He was at Mayo for 3 days, and had all kinds of tests/scans. He went back a couple more times for more MRIs. He also has ECD in spine, knees, shins, etc. Several weeks later, he was tested BRAF V600E mutation positive and he decided to enter the clinical trial on V at Sloan-Kettering. He has been on 4 and 4 for a week. But some side effects are starting; a very dry skin, and swelling of the hands and feet. He thinks that he will likely have to reduce the dose next week.

His optic nerves were so compressed that he had pink eyes all the time. Within days of starting V they were better. He said that he was curious about interferon as he almost started with it. Another Chatter has been on it since halfway through 2007. It was a new treatment then. His progress is assessed with regular MRIs. Insurance coverage for interferon for ECD is not straightforward. “Everything is an EXPERIMENTAL drug”! Interferon is used “until it stops working”.

You need to be BRAF positive to be taking V. Your BRAF status is worth knowing because currently BRAF+ patients have more treatment options available.

- One member reports that there is a clinical trial in Italy using a combination of interferon and V for melanoma (the most serious type of skin cancer). Apparently, there is new evidence showing that the two will work better than either one alone for melanoma.

- A member has just finished her first month of interferon therapy, and has not noticed any improvement at all. She doesn't know her BRAF status. Interferon, in general, takes a while to produce results. With V, which is specifically for BRAF mutation, you can see some improvements quickly. She has ECD in her bones: maxillary bones to start with, but growing all over her body for 3 months (legs, arms, hands, thoracic bones). Scans did not show any CNS or cardiac involvement.