

Summary of ECD Global Alliance Internet Chat **Sept 27 2014**

8 Attendees

- Members all said that they had enjoyed the conference, and especially because they had the chance to meet face-to-face. The only "complaint" was that the conference had not been long enough!
- There were doctors present from all over the world; US, France, Italy, Israel, UK, and Australia. They were explaining what they have learned about ECD, and offering advice. The event is being summarized by a group of volunteers, and this summary will be sent out to everyone as soon as it is complete.
- The event in 2016 will be in Houston. The date has to be decided on, but it will most likely be in the Sept/Oct/Nov timeframe. If members have any suggestions on how to improve the conference for next year, or anything that they would definitely like to see presented or discussed, please would they let Kathy know (support@erdheim-chester.org).
- One member lives in Houston, and he and his wife are already looking for ways to help plan the conference next year.
- Some of the members, who are on vemurafenib (V), had also been at the conference last year. It was remarked upon that many seemed to be much improved since the previous year!
- Members talked about their experience of Anakinra therapy. One had started Anakinra just two weeks before. Her vision is getting a bit worse and she is not sure what to make of that. But overall, she has a bit more energy, and if the disease wasn't in the orbits and affecting her eyes, "I wouldn't know I had it." She wondered how long it takes for the Anakinra to show improvement. She does not have a red or pink eye, but has distorted and blurred vision. If the Anakinra doesn't begin to help her sight (or at least stop the progress of worsening) then it was recommended that she should let her medical team know!
- At the conference some of the doctors talked about Anakinra, and it certainly seems to help some patients. Some of the patients that have been having pain with their ECD, say that Anakinra helped with the pain almost immediately.
- A member, who attends Sloan Kettering and is on V, was asked how often he was attending. Once a month was the frequency, and this is thought to be standard.

- Another member used Anakinra, and was stable the whole time. The nurses kept telling him that of the three answers from tests, two were good (presumably the 3 answers are better, stable, or worse!). Stable is good. He is now on V, with slight improvement at first, followed by becoming stable again. He has not had many issues involving his vision so far. The main issue is with his heart where there is a tumor growing, although this is now shrinking. He had developed a pericardial effusion (a build-up of fluid in the sac around the heart) and had to have a "window" procedure performed. The docs took a biopsy from this tumor then, and also did a bone marrow biopsy. He was found to be BRAF positive so the Anakinra was stopped and replaced by V. This has been a better treatment for him. He said that Dr. Janku had returned from the first conference enthusiastic about treatment with V. He had said that "the results are too good to ignore". The member has now been on V for about a year.
- A member said that he had met and chatted with another member in the hall at Sloan Kettering! A lady was there who had Langerhans cell histiocytosis (LCH), BRAF + and does not show signs of active ECD. She is worrying that she will be taken off of the trial. So far, it is thought that Sloan has not taken anyone off the trial yet, except those people who have experienced unmanageable side effects. It was thought that if scans are not changing for 3-4 months, then this might be considered to be a suitable "endpoint". It was said that "it must be scary for her to be one of the first". Dr. Haroche, in Paris, is working to try to figure out when and how patients might be taken off V, and the Sloan team is in contact with him on this subject.
- One member had been taken off of V for about a month because the starting dose (4 in the am and 4 in the pm) caused many side effects. He was then restarted at 3 and 3 non-stop, and was much better. He has not heard anything about stopping the V. His symptoms/side effects are less severe than some, and Dr. Janku says that he would be fine with a treatment level of 2 and 2, also. He has no more skin lesions, but still has some foot pain.
- The brother of one member has recently gotten married. The member told us that the wedding had been "great". Regarding his ECD, he told us that he did not have any balance problems until he was put on Synthroid to treat a problem of an underactive thyroid gland. Until the ECD "cranks up again" he will be sticking with interferon, this has kept him stable for some years now. He now has to wear glasses because he has some double vision. He had his interferon dose lowered by Dr. Kurzrock in 2007. He is due for his regular MRI scan to assess the disease soon. He wonders why they do not do a bone biopsy on him to get a BRAF result. He said that he had already done the urine test, but that he had not heard about the result.