

Summary of ECD Global Alliance Internet Chat

22 Nov 2014

12 Attendees

- A new member had logged on during the week before the Chat, asking for information. A reply was left for her and she was told that the Chat occurred on Saturdays at 3pm Eastern Standard Time (that's 8pm if you are in the UK). If she contacts Kathy Brewer [support@erdheim-chester.org] through the ECD Global Alliance website Kathy will send lots of info.
- A member posed a question about platelet counts. Platelets are cells that are found in the blood, and are vital for an effective blood clotting "system". For the previous 2 quarters his platelet count has been low at 137 and now 142. He asked whether this could be ECD related, and whether others had experienced the same. He has had 2 severe nose bleeds, and knows that these may be related to the low platelet count.
- The member who was originally diagnosed with Rosai Dorfmann Syndrome (RDD), and then after a biopsy had this changed to ECD, came on. She is now on Anakinra (Kineret). In 2012, she had treatment with Cladribine and then Clofarabine. This was when she was thought to have RDD. She started Anakinra 10 weeks ago, and will not have tests done to see if it's working for another 3-6 weeks. She has lesions in her orbits (affecting her vision), in the long bones, pelvis, skull, left kidney, and in the spine. She tested BRAF negative, so she couldn't go onto Vemurafenib (V). She sees Dr. Diamond from Sloan-Kettering (SK). Her eyes got worse after the last chat and she had to go back on steroids. The Lucentis did not work on her macular edema. One week her vision was 20/25 in the right eye, and the following week, having had a Lucentis injection, her vision was 20/300! Her very first symptoms were eye problems, and later disease was discovered elsewhere. None of the ophthalmologists knew what to do. Now she is seeing one at Sloan, but he is just monitoring things. She keeps getting macular edema. This affects her vision, and has caused some permanent damage. The Lucentis shots are not working, although steroids are, but "how long can you stay on steroids?" The retina specialist says that her optic nerves look fine. She was treated with cladribine (2CDA) when she was thought to have RDD, and this left her with a very depressed immune system. She also had another chemo drug, clofarabine, and it might have been this that lowered her CD4 counts, since it is much stronger than the 2CDA. Her CD4 counts continue to be low after a year and a half off those drugs. CD4s are part of your immune system, and help fight off the very serious kinds of bacteria.
- A member told us her story. She was diagnosed with ECD in Feb 2011, but had started having problems in 2005. She lives in Norway, is BRAF positive, and has been taking V since Dec 2013. Before this, she had been on interferon. She has twice been to the NIH, and has also been to the ECD patient & family gathering twice. Her Central Nervous System was affected and her long bones. She had a lesion on her spine, but that is almost gone now. Her walking has improved.
- Another member who is taking V came on. He lives in Minneapolis and goes to Mayo for some of his care, but he also goes to Sloan Kettering on either the first or second Thursday every month. At

SK, V is free because it is a location for the V trial. He is sure that he would have only gone to Mayo if it had been a location for the trial. He said "though it's costing a lot to travel to NY, I am lucky to have the best of both worlds."

Dr. Garrity was the ophthalmologist at Mayo who diagnosed his ECD by looking at his MRI, before the member had even seen him! He did the eye surgery to get a biopsy from behind the eyes back in May. Then the doctors worked to get him to SK so that he could be in the V trial. He used to have 2 large masses in the orbits. He hasn't tried any other drugs. V is working quite well across all areas, including spine, bones, brain, & orbits. He went back to Mayo to have his eyes checked. They have receded from the bulging at their peaks. His left eye has receded 5mm! "V is working ok for me."

- A member asked whether anyone had used, or heard of a drug called TNF - alpha antagonist (TNF stands for tumor necrosis factor). Her son's doctors' are talking about using it for his ECD, when his lungs are strong enough. They were damaged by another chemotherapy drug that he was given.
- Members discussed their plans regarding having a flu jab. Some said that they were not going to have one. One said that she had consulted an Infectious Disease specialist, who had recommended having the jab, although an oncologist had said not to have it! She will probably take the Infectious Disease doctor's advice.