

Summary of ECD Global Alliance Chat

19 Dec 2015

11 Attendees

- A new member came on a few days before the Chat. He said that he was only 16, and wanted to get in touch with other children with ECD. His mother also came on asking about this, particularly asking whether there were any other ECD sufferers under 18 years of age.
- Members on the Chat were all doing well although one told us that he had sore ribs. He hoped that this was due to "a bad sleep" and nothing else.
- A member keeps "finishing" making her bourbon balls and then decides to make another batch of 6 dozen! [Summarizer's note; Must be like a Bourbon Ball "Groundhog day"!] She has made gingerbread dough to make gingerbread "boys" with her grandchildren!
- Today, she went in the local bakery to get special breads, and an older man looked at her and said "all of those goodies will make you fat." She had retorted that she is on a sure-fire weight loss diet called chemotherapy. He didn't really respond but she did hear a woman gasp!
- A member is now stable and not on any drugs for ECD. He sees his doctor in early January. He has MRI scans twice a year. No growth was showing on the last scan, but not much shrinkage either. He is BRAF+. He was treated with cladribine from July 2014 to Dec 2014. The cladribine helped stabilize his ECD. He was asked whether he had been troubled with lesions in his orbits. He has them behind his left eye. This has not been affecting his vision or caused bulging of the eye.
- A member has had a 2 week "chemo vacation" due to her dizziness and tremors, but has now been advised to go back to a dose of 2 in the morning + 2 in the evening of V by Dr. Diamond. Her shoulder problems have been helped by "aggressive" physiotherapy and the cortisone injection (into the joint). The V "vacation" was due to the shoulder issues, and Dr. D assumed that 3+3 was too much for her. She thinks that he has put her back on a lower dose of V because of the tremors and dizziness. The MRI of her brain showed that the pituitary and cerebellar lesions have improved a bit. "The tremors and dizziness are from ECD, chemo, or whatever!"
- The member who had both lymphoma and ECD diagnosed, gave details of her journey through the medical "maze". She had been on chemo for ECD for 18 months (Kineret and methotrexate; she had been on these for some while as a treatment for rheumatoid arthritis) then "a large knot" appeared on her left leg. The doctors thought that it was ECD coming through the bone. She had a rod put in the bone, and the knot removed. This was tested and it showed both ECD and Lymphoma. She has now restarted her ECD treatment, after having to stop for a while because she needed chemotherapy for the lymphoma. She will be having a PET scan in the early New Year to see how things are progressing.
- A member who has had V for a long while (after 2 years she has still not had a break from treatment), is now on 1+1, and is still doing well. She feels that she still keeps getting better. She is able to get around more, without her walking sticks. She can do more walking or exercise, and can really push herself. She managed to get her pulse up to 170 this week! She

was asked whether the scar tissue on her spine was getting any smaller. She said that she didn't know. The lesion was affecting the upper half of the 6th and 7th thoracic vertebrae.

- Another member had also had a lesion in his spine, in the bone (he said "on T and L". This presumably means Thoracic and Lumbar). As of his last scan it had pretty much gone, but it disappeared once before, and then returned. Another member has also had a spinal lesion but is not sure exactly where.
- The member with BRAF- ECD and problems with macular edema told us that she had taken a 10 day break from her medication (sorafenib). She has not had any edema in 7 weeks. Her vision is still blurry and wavy, but the edema has not been there. She is now back on sorafenib at a dose of 200mg daily. Her oncologist has told her to watch out for any side effects, and when they get bad, to stop the drug until the side effects get better.
- Visual issues were then discussed. One member told us that she has no measurable problem with her eyes, but about once a month, she suddenly has waviness in front of her eyes. The waviness is there whether she is looking at anything close, or far away. It usually lasts 15-30 minutes. She doesn't notice any wateriness of her eyes.
- Another member told us that she is troubled with watery eyes, and her eyes feel very irritated sometimes. She tends to have dry eyes, and uses eye lubricant, which helps a little. Another member usually puts OTC lubricant eye drops in when she wakes up. She gets dry eyes with the chemo and sometimes she "just can't read well".
- Other ways of reducing eye symptoms were discussed. One member wears a pair of wrap-around glasses to protect his eyes from dry air at home and even when he goes to sleep. This has helped a lot in preventing him from getting watery or irritated eyes. These are standalone wraparounds, and cost \$3 at Walmart (for the clear one). The arms need to be adjusted so that they won't be too tight. The glasses need to be sufficiently close to the eyes, and feel comfortable.
He also has an amber wraparound that he loves to wear to sleep in, and this can also be used outdoors. He wears the clear one during the day indoors.
The use of a warm compress (just a wet warm cloth) was suggested. Apply it on your eyes, a few times, when you wake up.
Finally, on this topic, a member said that she had started using fish oil (presumably as a diet supplement, not eye drops!!) at the suggestion of a doctor 2 years ago, and this had helped.
- Members discussed when they were next back to see their doctors and their current treatment plans. One, who goes to Memorial Sloan Kettering Cancer Center (MSKCC), said that she had been there twice at the beginning of this year and is going back in January. One is on prednisone (used for a problem other than the ECD) and is trying to wean this off very slowly. She goes back to MSKCC in early New Year.
- The Paris conference next year was mentioned. One member wants to go but will need to make a trip to New Zealand first. Her 22 year old son is interning in professional golf management on the North Island. [It's a hard life, isn't it?]

- After the Chat a member came on and said sorry that she had missed us. She and her husband (it is him who has ECD) wished us all a Merry Christmas. They should “know more” soon after an upcoming hospital visit.
- Another person came on after the Chat. She is from China and said that her father had been confirmed as having ECD. She said that she was pleased to have “found” the Global Alliance site.