

Summary of ECD Global Alliance Internet Chat

15 Nov 2014

5 Attendees

- In the days leading up to the Chat, a member came on whose husband has ECD. He has been in hospital for the previous 14 days, and has been very ill. He had an injury to his foot, in March, which did not heal. Ten days ago he had to have his little toe amputated.
- A member came on who lives in Florida. She has been on Anakinra injections for ECD since September. At first, she was diagnosed as having Rosai Dorfmann Syndrome. She had 2CDA (cladribine) at first, and then Clofarabine. She has been stable since 2012, but has had to stop these as they are damaging her immune system. She was on 2CDA for two months, and then the doctor decided to switch it to Clofarabine. She finds that dexamethasone (a powerful steroid drug) helps her vision, but she is "never on it long enough". She also gets shots of Lucentis in her eyes, because she has macular edema. She has ECD in her long bones and left kidney, as well as in the orbits and her skull. In 2012, the doctors thought that she had Rosai-Dorfman disease, and pathology was sent to Texas Children and NIH, and they confirmed this diagnosis. But in 2013, the doctor changed the diagnosis to ECD. In July of this year, she went to Sloan-Kettering, where a further biopsy was taken from her left tibia. The pathology this time favored ECD. She is 61 and a retired middle school teacher. She sees Dr. Eli Diamond for her ECD treatment. He was at the conference in Bethesda, MD this year.
- A member came on who has been using interferon 3 times a week for some time. He has recently had lots of tests and is about to start treatment with Vemurafenib. He was diagnosed in 2005, and his current doctor is Dr. Bauman at the Dayton VA Hospital (until he retires later this year). After all the recent testing the doctors decided that he should use oxygen, because of his difficulty breathing while sleeping at night and during the day at times of high exertion. This has made a real difference. He sleeps much better in the night, and wakes more rested. After daytime exertion he "gets back to normal" much faster. The ECD is in his long bones, and around his heart, lungs, kidneys, and pancreas.
- The other member told his story. He was diagnosed about 10 years ago. He had to retire 7 years ago (only 45 years old), from his job as a family physician. "The patients thought that the doctor was drunk!" He had treatment with interferon and 2CDA but they "didn't work!" He was then treated with steroids and methotrexate. He has been taking methotrexate (only 40mg once a week) for a few years now. It is an old chemotherapy drug. The steroids started at a dose of 120mg daily. They were gradually reduced over 6 months and then stopped. He has been stable since, and is possibly in "remission"! A PET scan will be done to see if there is any active disease. Initially his disease was in his brain (cerebellum), lungs, around his heart, in his sinuses and in his spinal bones. Even if he is in "remission", he will still be slurry and wobbly!! He will not be able to return to work-"Goodbye Tension. Hello Pension!"

- The father of a young woman with ECD came on. She is also in "remission", although her balance is terrible, and her sight was severely damaged by the ECD. She was treated with LOTS of prednisone, but what really "did the trick" was chemo with 2CDA. All she takes now is a low dose of DDAVP for diabetes insipidus.

She had first become ill in the fall of '05, with the discovery of a tumor on the top of her head, where a baby's soft spot would be. This was removed and tumors on her optic nerves were irradiated. The radiotherapy didn't work and those tumors grew. Tests, CT scans, and MRI's revealed lesions on the long bones in her legs, and upper left arm. She had a tumor on her heart, diabetes insipidus, and a more general decline. She had prednisone first for a while to try to reduce the tumors on her optic nerves, so that she could see. She had chemo for 7 months, 5 days each month with 2CDA and after that, interferon. All the tumors are now scar tissue, or gone and her health is generally good. She will be 35 on 12/31. She attends a "VERY GOOD" hospital, the Hospital of the University of Pennsylvania, and sees several doctors there. Her oncologist is the "captain of the ship".

Editor's note: "Remission" has not formally defined for ECD. In the context of this discussion, it is believed that the chatters used the word "remission" to indicate there are no active ECD sites known in the body.