

Summary of ECD Global Alliance Internet Chat on 05 October 2013

9 Attendees

- A patient mentioned that Physical Therapy has been helping her greatly. She is meeting, with a lawyer, in a little over a week to see about disability.
- A member, whose spouse has recently been diagnosed, and been on interferon for just a few weeks, asked about side-effects.

One member had been on interferon for 9 years. He thinks that "it kept me alive. But the side effects can be fierce". Two and a half years ago, he switched from interferon to Kineret. This drug has been just as effective for him, without the side effects. The principal side effect from interferon was fatigue, and heavy flu-like symptoms, about 24 hours after taking it. But over the years, the fibrosis behind his eyes was actually reduced by the interferon.

Another member had found the interferon OK to take, but it didn't calm down the ECD. He didn't have the fatigue that most people get. He said that "Perhaps it was only water! NO side-effects and NO effects".

- A member, who has been having trouble with back pain, is going to the Doc this week, and having injections in his back.
- Stabilization and remission were discussed. To diagnose this various scans can be used, for instance, MRI scans every so often, of the areas where the disease has been detected. PET-CT scans that didn't show any active disease areas, for one member, suggested remission, but he is still taking once weekly methotrexate tablets (which is chemotherapy).
- Another member has been told that she is in remission (and is now only taking a low dose of DDAVP for DI (Diabetes Insipidus). Her oncologist said this in '09, when all of her tumors/lesions were gone or reduced to scar tissue. She was diagnosed aged 27, and is now nearly 34. She had had tumors on her optic nerves, a tumor on her heart that caused fluid build-up, and she had lesions on her long bones in both legs and her left humerus. She had DI years before she showed any signs of ECD. She lost her sight when her blood pressure dropped to 70/40, due to the fluid collection around her heart. A pericardial window was made and her heart has been fine. But she lost all the sight in her right eye and most in her left. She has balance problems.
- The mother of a member was standing in for her daughter who has ECD. Her daughter is OK but had just gone to the sporting goods store with her brother. She

has been very tired, and her back has also been bothering her. She is on Kineret now. The Docs have her on many medicines and the side effects are tiredness. The Docs did notice that her rash looked a little better, when they were doing her spinal tap, which she has had done before. The docs said that "her levels were high" and removed some fluid. They aren't sure if that is the reason for her dizziness and headaches, but the tap seems to have helped. She had some problems in 2006, was diagnosed with ECD in 2010, and is now 20 years old.

- Members agreed the bad thing about kineret is having to have a daily shot. They said that, the best site, for the shot, was the abdominal wall (stomach). One member has "divided" his abdomen into four quadrants, and rotates the daily shots among them.
- We are getting new patients to go and see our "in the know" docs. A US member said that THE OTHER person in NC with ECD is seeing the SAME oncologist, and, back in the Mother Country, 2 new patients have started to see the Doc, in Leeds, that a member has been seeing for some years for his care.
- It was remarked that, it is odd having a disease, which is so rare, that most people, including doctors, have not heard of. A member believes, for example, that he is the only patient in Los Angeles. [Editor's note: There are multiple patients in the Los Angeles area.] A UK member has never met an ECD patient, face to face, in 8 years, and before he became ill HE WAS ONE OF THOSE DOCS THAT HAD NEVER EVEN HEARD OF ECD IN THE FIRST PLACE!!
- One US member has met with two other ECD patients, one from Norway (twice!) and one from Pittsburgh. This should all change for several members of the group after the San Diego meeting.
- Kathy Brewer would know if any other patients in other countries are registered with the Global Alliance. If there are other registered patients, Kathy will send them an E-mail to ask whether they would like to be in contact with anyone else, and then send details. Also members can look on the website at the "Patient to Patient"- Case Log.

[Editor's note: See

<http://www.erdheim-chester.org/PatientToPatient.html>

and select "Case Locations" to see the country/state of residence for all patients registered with the ECD Global Alliance.]

- Problems with benefits were discussed. It was said that living on SSI was not a luxurious life, and you aren't allowed to accumulate more than \$2000. One member has had to stop working due to being in so much pain, but her applications for benefits are being denied. She is seeing a lawyer in 2 weeks, to see if this will help in getting approval.

[Editor's note: For those who are filing for SSI or SSDI in the US, please tell your contact that ECD is on the Compassionate Allowances List (CAL) for the Social Security Administration.

See - <https://secure.ssa.gov/apps10/poms.nsf/lnx/0423022947>. The Compassionate Allowances (CAL) initiative is a way to expedite the processing of SSDI and SSI disability claims for applicants whose medical conditions are so severe that their conditions obviously meet Social Security's definition of disability. It is not a separate program from SSA's two disability programs, SSDI and SSI.

For a simple explanation of some of the disability options and how to get started applying, see: <http://www.erdheim-chester.org/DataFiles/PresentationsAndArticles/Disability%20help.pdf>.]

- A member, who has been having sudden episodes of vertigo, had an MRI in the previous week.
- A member came on, whose son has ECD, diagnosed after 2 biopsies, in November 2012. He will be 22 in December. He was born in the US. They live in NY. His mother was born in Trinidad. He has lots of fatigue, long bone pain, and pains from his shoulders that radiate down his arms, lower back pain, and belly pain. He is also troubled by pain and discomfort caused by lymph nodes his neck. He is not working, or in school. He is now on Kineret, and able to be more physical. He used to spend most of his days in bed, sick. He started with a back pain, last year, that wouldn't ease up. There had been nothing prior to that. The docs were thinking about lymphoma.

This had been the case with another patient as well. Her neurologist thought that she might have had lymphoma, and was very concerned about her tests. This member had found a lump on her head, at the site where a baby's soft spot would be. This tumor was Langerhans Cell Histiocytosis (LCH). MRI showed tumors behind each eye. Her pituitary, cerebellum, and bones became involved, and there was one tumour on her heart. Small growths on her eyelids were noticed by an ophthalmologist and that gave the clue to ECD.

The tumours behind the eyes damaged the optic nerves and caused blindness. She used to cry every day, for hours, about how she couldn't see anymore. She finally got past that, a giant step, with the help of a psychologist friend, who, for a long time, saw her at least twice a week.

- Members discussed the large number of doctors involved in their care. One member sees specialists in; Endocrinology, Cardio-thoracic surgery, Neurosurgery, Neuro-Ophthalmology, Ophthalmology, Oncology, Cardiology, Radiology, Psychology, and a General Practitioner. This list helped get her to get SSI! They were blown away by all of those specialties.

- Another member says that she keeps a book of the cards, of all the Docs that her daughter sees, to carry with her. And, she keeps a list of all the health and Doc appointments, about 11 pages! Her Doc keeps in touch with the NIH, and has also been in contact with a doctor in Paris. Kineret appears to be working better than the interferon for her, but it is trouble having to have daily injections.