Summary of ECD Global Alliance Internet Chat
03 Aug 2013

9 Attendees

- A new Chatter came on who was diagnosed, with ECD, this week. He lives New York. He told us that the doctors are now running a test looking for a specific genetic marker that will guide treatment (this is probably BRAF). He had, initially, been given the diagnosis of retroperitoneal fibrosis, which had affected his spleen, pancreas and stomach, but not involved the ureter. He had gone to John Hopkins to see a Dr. Scheel. He had suggested that his hip pain might be a sign of ECD (a Bone scan, and tests on his RPF surgery slides proved that it was ECD). He is awaiting the results of the test for the BRAF mutation. Later this month they are meeting to discuss treatment options. He asked if anyone had had any suggestions about what to do/ask during this visit.

  He told us that he was the father of 4 kids (13,11,5,3). He is married, 37 years old, and works as a Teacher (Math) and a Pastor (Presbyterian). He has hip and shin pain. The abdominal and side pain went away after RPF exploratory surgery and weight loss. He has noticed that his fingers/hands sometimes feel funny, that he seems to have to repeat himself more often than before, and that his feet feel heavier. His wife is worried about his ability to work, once he starts treatment. Members shared their "work experiences". One is still working fulltime, and is on no major treatment which makes her ill, so she is bouncing and wobbling around at work. Another had had to retire aged 45, and another had taken a 6 month leave of absence whilst taking the 2 CDA. A member, who is also a teacher, told us that he had been unable to work, after starting on treatment.

- It was said, by one member, that he may be able to pastor once a week, but teaching, and being on your feet all day, is another matter.

- A member told us that she would be going to San Diego, and will make the trip into "a little vacation".

- The widow of a member, who recently died, joined us. Members sympathized with her loss, and missed her husband's humour. She said that was joining us because, "You have all been so great with Phill". He had been having cladiribine (aka 2-CDA) treatment, which seemed to be going well, but he developed a cough, had trouble sleeping, and was very tired. It turned out that he had developed a DVT which led to a pulmonary embolism, which was fatal. His widow told us that he had found that he didn't know where his hands and feet were, unless he could see them. She wonders whether this due to ECD affecting his nerves.

- A member who has had a year of treatment with alpha-interferon has just had a PET scan, which has shown that this treatment was not working. Now the doctors have started a protocol of Methotrexate + Vinblastine + Prednisone. He is having pain in the joints of the arms and legs, since starting this new treatment. Another member takes methotrexate at a dose of 40mg a week, but hasn't ever had vinblastine, and only had prednisolone when his chest was very bad. A number of members had taken prednisolone (2 for more than 10years) It was agreed that "steroids make you fat".

- A member asked about BRAF. It is a genetic mutation that quite a lot of ECD patients have. It was first found in people with malignant melanoma (the worst sort of skin cancer). A drug, that was originally developed for use in melanoma patients, has been successfully used in hairy cell leukemia (which is closely related to ECD), and in a couple of ECD patients, with good results.

- A member told us that they would be going to Sloane Kettering hospital (one of the top cancer hospitals in the United States). This member is BRAF positive, and will be starting new medication. The doctor has said that there should be visible improvement in mobility in two weeks! This was the closest place to her that was accepting new patients. It will be a very expensive cure, to go to NYC often.

- A member is now not taking any medication for her ECD. The interferon has been stopped, and she is just waiting for her BRAF test result. This member has not tried any chemo, because the doctors say that she is not sick enough!

- A member said that she was, shortly, to start taking Anakrina, under the care of Dr Janku. Her husband has also been ill, having chemotherapy for cancer. He is doing well and there only 10 more weeks to go. She has been on methotrexate for many years at 20mg a week, and this is being carried on.