Summary of ECD Global Alliance Internet Chat
26 Jul 2014

4 Attendees

- Before the Chat a new member had come on. She was seeking advice/information because her dad was recently diagnosed with ECD. She is finding this really hard to come to terms with, because she has read some of the out-of-date information available on the internet. The doctors haven't given a prognosis, and he doesn't want to know; but she does want to know. She wants to be able to prepare herself. She asked "Does anyone know what to look out for in someone with this disease?"

- The Chat itself was, again, this week a very small, select group. One said that the others "must be all at the beach!" The "select group" kept it "short and sweet".

- A Member left us a message before the Chat that she would be at a family reunion and unable to make the chat. She has been having horrible shoulder pain. She has had an MRI and had the joint injected with drugs. These are still not working, but the doctor is hopeful. They said that she does not have a tear that they can see, but it may be hidden. If she continues to have the pain she will need to go back. Other than that she is doing alright!

- Another new member from Canada came on. He had been diagnosed in the previous week. He asked some general questions. He has heard about the NIH study and is hoping to get on it. He told us that Vemurfenib is licensed in Canada, but he doesn't know whether insurance will cover it.

- The other member on the Chat said that he had been diagnosed 9 years ago. He is currently fairly stable, but had to retire aged 45, 7 years ago. He said that he had met another ECD patient (for the very first time!!), earlier this year. He then gave a brief summary of his treatment over the 9 years. He went through interferon therapy, and then 2 CDA (cladiribine). For the last few years, he has been taking methotrexate (a good old fashioned drug). He only has to take it once a week, at a dose of 40milligrams. He said that his regular doctor says that he now has 15 ECD patients, and that the member was his very first! This doc will be attending the ECDGA conference later this year.

- The newly diagnosed member had been on methotrexate a couple of years before, for retroperitoneal fibrosis, but had not found it easy. He starts on cladiribine (2CDA) this week. He will have it for 5 days in a row every 4 weeks, by IV infusion over 2 hours. He will have an anti-sickness drug in pill form. He has been told that this treatment is "quite tolerable". He had a bone marrow biopsy taken, and this confirmed ECD with a positive BRAF V600 mutation. The other 2 members had both been diagnosed after having had a biopsy of abnormal tissue around the kidneys ("hairy kidneys"). BRAF hadn't been invented all those years ago!

- The other member had also had cladiribine for 5 days every 4 weeks, but his cladiribine was given as injections under the skin. The anti-sickness drug was given IV, and the worst bit of the whole thing was the placing of intravenous needle each day. He never felt sick, so he stopped having it. In the UK, you are not

allowed to go home from hospital with anything stuck into a vein. So you have to be “speared” afresh each day if you are having the cladiribine as a day case.

- A Vemurafenib success story was on the Chat. She started V last August. She had been diagnosed in 2000, having had symptoms since the 1980s. She lost a fantastic amount of weight (retained fluid), and got mobile again. She said that it was like getting a new life! “Life is so much better this summer than last! I'm grateful for it every day.”