Summary of ECD Global Alliance Internet Chat
18 Oct 2014

7 Attendees

- A new member came on to tell us about her mother-in-law who was diagnosed with ECD almost 2 years ago. The member was seeking information to help support her husband and sister-in-law, and also to support and guide the medical professionals involved in her care. In this case, the doctors began with a treatment of chemotherapy and prednisone. The member said that the chemotherapy had affected her adrenal glands, to the point that her body is not producing enough cortisol (an ex-doctor writes; the prednisone is more likely to have been the culprit. The adrenal glands don't have any work to do while a person is taking prednisone so they "go to sleep", and then can't produce enough cortisol when it is needed). The steroid has produced a terrible problem with her wanting to spend money (it's that ex-doctor again; steroids can produce changes in mood and behavior). She is currently in the hospital, awaiting endoscopy to determine why her stomach is hurting so much. The doctors have requested previous biopsies, to see if she may have the "mutation"-BRAF needed to take a new drug-vemurafenib (V).

- Two members who are taking V discussed their treatment and progress. One is on the V trial at MD Anderson, and the other is on the trial at Sloan-Kettering (this requires the member to take a monthly flight to NYC).

- One member told us that a slight improvement was noticed at first, then stability thereafter. His present dose is now 3 pills in the am and 3 in the pm. He had to stop taking V for about a month, because of side effects when his dose was 4 and 4. He restarted at 3 and 3 and has been much better. He also has heart involvement. The tumor on his heart shrank at first, but is now stable. He has scans on a regular basis (every 5 or 6 weeks), the most important seems to be the cardiac MRI, as this is the only way that the tumor on his heart can be seen.

- The other member on V has MRI or PET-CT scans. He is giving urine samples for tests to monitor BRAF levels. He was told that this level has dropped quite a bit since he started on V. All his tumors, bones, and spine are responding to V. "The only thing is traveling to NYC monthly, with overnight stay every other month." Another member said that she was very fortunate to have great doctors so near to her home.

- A member told us that it was said, at Bethesda's event in September, that therapy for BRAF negative patients is the new thrust of research.

- A member told us that he has had several test recently in preparation for starting V.

- A member talked about his daughter who has ECD. She is doing pretty well. She just takes DDAVP for diabetes insipidus. She has an MRI at least once a year, but these have been coming up with good news for a few years now. She tests negative for BRAF. Her best treatment was with cladribine (2cda). Her oncologist chose this treatment because she thought that the ECD growths were somewhat like hairy cell leukemia. Her father said that her sight, which she lost due to the ECD, might be restored with some stem cell work. “We've been told that could happen in her lifetime.” It was said by a member that “she was an inspiration!”

- Politics raised its (ugly) head again. Someone told us that another member will be running for Mayor of Bethesda! She thinks that she may even win with all our support! A member said that she had his vote, and another wants to move house “just so I can vote for you”! ☺

- A member told us that he forwards the chat summaries to 8 or 9 medical people, almost every time. He said that he knows that some of them read them, because he occasionally gets a comment about them.

- An ECD awareness event was mentioned. This event took place at a televised high school football game, which will help get the word out about ECD!