

Summary of ECD Global Alliance Internet Chat August 18, 2018

4 Attendees

- After the usual introductions, a member new to the chat said that she felt tired “as usual.” Since she is new, other members asked about her story and medication. She explained that she lives in Holland and was diagnosed with ECD about two years ago. She is BRAF+ and has been on dabrafenib (D) and mekinist (Mek) since then. Her doctor in Holland gets advice from Prof. Haroche in Paris. She has been to see Prof. Haroche once, just after the diagnosis was confirmed. She is doing pretty well on the medication, with some side effects in the beginning, but now it’s only tiredness.

She was diagnosed two years ago after a whole year of suffering and looking for a diagnosis. She’s had diabetes insipidus (DI) since 2007. Before starting treatment for ECD, she had fluid in the pericardium. Now this has gone. She just has the remnants of tumor around the big vessels leaving her heart. On her scans, some of the abnormalities have gotten smaller and others have disappeared. Her internist thought this cannot be considered as remission, and she is still taking the full dose of D+Mek.

- A U.S. member said that she had been on D and Mek. Dr. Diamond took her off the D in March, leaving her only on the Mek. Now, she is not on any chemo while she is having her heart complications investigated. She was taken off Mek the previous week by her local oncologist. She sees Dr. Diamond on September 6. Her fingers seem to be “rebellious” against the chemotherapy, as is her heart. She saw her local cardiologist because of chest pains and was put on the nitroglycerin patch. She will see her cardiologist again this week.

The T-wave on the echocardiogram (ECG) is going inverted and her left ventricle on the ECG is a “bit off.” She has added green tea to her diet hoping that will help her lose some water weight. Her internist “freaked out” when he took her blood pressure (BP) in both arms. There was a 30-point difference in the systolic BP between right and left arms. He thinks that she may have developed a subclavian artery stenosis.

In the past, she had a lesion around the outside of the aortic arch, but it is gone now. There was also a little lesion that was in the lower lobe of the right lung. That too is now gone. She wonders if this new “head stuff” is ECD, that is not showing up on the PET scan, or if it is a side effect of Mek. She hopes that the cardiologist, although new to ECD and is a MD with a PhD in women’s heart issues, will have some thoughts.

Her and her husband will be visiting Memorial Sloan Kettering Cancer Center in September to see Dr. Diamond and a pituitary specialist. They will fly in and out the same day.

- They will be meeting another member for lunch! The member they will be meeting is taking just one vemurafenib (V) a day. He has been in complete remission for three years now. This is based on his PET scans. He believes that some of the tumors are still there but are considered benign. It has been pointed out that although they have regressed, they may come back.

- A French member is due to go on a further visit (three weeks) to a spa in Rochefort sur mer. The spa treats rheumatism and skin diseases. It's known to treat psoriatic arthritis, which strangely resembles ECD. After he has this "cure" he has less pain for a while.
- A US member had to visit his local Home Depot with his wife to confirm their countertop order. They are in the middle of a kitchen remodel. The French member suggested a visit when the new kitchen has been completed.
- A member said that she has had some tumors in her cerebellum and brain stem, but they are never mentioned. She presumes that this is because they are so small. The only part of her brain commented on after her MRI is the pituitary. Her PET from two weeks ago is better with only the femurs being mentioned.

She had skin lesions for ten years before she was diagnosed with ECD, and that wasn't until her brain showed involvement. She also had started having leg pain. "But no one bothered to just do a simple X-ray, until I fussed." Members agreed that it was important to be fussy at times. When she was still working, she was a nurse and her husband is a doctor, so they know quite a lot of stuff. They know a lot more about ECD than most doctors in Lexington. "Of course, Dr. Diamond and Prof. Haroche know it all!"

She feels that she needs to make sure that everyone stays informed and that her scan and bloodwork results get sent to everyone. This is really tiring! Although they do have electronic records in the U.S. it doesn't mean that everyone looks at them. She uses Google to find scholarly articles. One day, she found that she was reading about herself in an article that Dr. Diamond had just published. And she was reading it in her orthopedics office!

- The member from Holland originally had to spend a lot of energy getting the cardiologist to communicate with the internist. Eventually she switched to a cardiologist that works in the same hospital as the internist.
- In France, electronic transmissions only work in the Salpetriere Hospital (the home of Prof. Haroche!). The results are sent to the member and his doctor by post and are only a summary. The results of tests in Paris are also sent to his rheumatologist in Strasbourg, who organized the very first tests by which ECD was uncovered.