

Summary of ECD Global Alliance Internet Chat

20 Feb 16

8 Attendees

- A member left a message to say that she has been well and is reducing her gabapentin dose. She will be down to 200 mg a day this week!! She is planning a fundraiser at Culvers of Highland on Monday, February 29th. If anyone is able to come that would be great! The address is 3950 Ridge Rd Highland, IN 46322.
- The member, who is now going to Dr. Kurzrock's clinic, has had an appointment at the University of California San Diego (UCSD). He had his regular check-up and lab work and is doing very well. He is running a mile a day on the treadmill. This is his instant therapy! He feels better than ever after the run and can't do without it. He feels that if he doesn't exercise, he needs a higher dose of Vemurafenib (V) to get the same outcome. Ever since he had ECD, his head is always "plugged up" because of all the lesions. As the lesions are shrinking, he is finding that running/exercising really improves his circulation and reduces ECD symptoms. He also told us that he has a feeling that V will be approved by FDA for ECD, within some point this year.
- Another member who has had V treatment said that she is also doing well with no pain and lots of energy. She is not tired anymore. She exercises for about an hour every day and sometimes manages 2 sessions on the same day! She only needs her walking stick for longer outside walks. She lives in Norway and there is not too much bright sun, but she is going to Florida next weekend and will have to "experiment with the sun there" (SPF50, hat and no bikini this year!).
- Another member who has been on V said that it was warm where they lived and she and her husband had met up with lots of friends and family from church at the park for lunch. She used sunscreen, wore a hat and protected her lips (which had got blistered recently). She gets in the pool daily and now can do strokes, as her shoulder heals, but now the left hip is giving her major problems. She did manage to walk around the block with her dog (a mere 65lb in weight!). Dr. Diamond thinks that all of her joint issues are from the V, not the ECD. She wants to exercise more, but her joints don't like it. She used to be very active. She is taking a dose of 2 in the morning and 2 in the evening, but she asked Dr. Diamond if she could drop to 1+1 while she is away in New Zealand for 10 days and he agreed. Joint pain in another V patient disappeared when the V dose was reduced.
- A Canadian member, who is trying to get put onto V, has not started it yet. His medical team is still checking in to it for him, since his insurance will not cover the cost. He had a good check-up recently and is staying stable. His latest CT scan, following on from one taken 8 months ago, has shown no increase in the mass that they saw before.
- A US member said that Genentech (the company that makes V) sent her a co-pay card that would cover up to \$25,000. Her insurance picks it up after \$3,000 in deductibles. She hadn't asked for the card, the company just sent it to her. Diplomat Pharmacy sends her the V every month via UPS. In January, the Diplomat Pharmacy in Flint, Michigan called her about sending more V. Then they said that if she wanted them to, they could use a co-pay card to cover the V for \$25 in January. She had said "OK" really fast! In February, they didn't ask because she had already met her deductible with doctor and physical therapy visits. (One member wanted to know where he could get that sort of card!!) What she didn't know was whether this happens

only in the US. A Canadian member wanted to know if this extended north of the border.

- The Sick Children's hospital in Toronto, Canada is doing a V trial with no age restriction, but the doctor of a member who would like to be in the trial, doesn't know if they will take him because he has had a kidney transplant. It was suggested that the member should contact the lead investigator himself to see if he can join the trial.
- Communication with doctors was discussed. A member said that she had sent Dr. Diamond an email and got an automatic reply saying that he was out of the country. She was very impressed by the fact that he still answered her on the same day! Her doctors in Norway definitely don't answer that quickly! It was said that Dr. Diamond is very good at responding to patients' emails. A member's Mayo doctors would even text with him, and give him their home numbers so he could call them anytime! One member has never been given an email address by one of her doctors, but her local oncologist uses her cell phone to text or call her. It was said that this sort of availability was good and "the way it's supposed to be" because a lot of time, patients are stuck with no answers. Getting a timely response really helps ease the patient's concerns, and it should be the best practice!
- Foot problems were the next item discussed. For one member, the calluses on her feet are getting so big, that Dr. Diamond says that she should have a podiatrist pare them. The worst place is on the ball of each foot under the fifth toe. She cannot walk without slippers or something on her feet because the calluses hit the floor first. She wears Speedo shoes when she is swimming. She has also developed "cysts" underneath the palms of both her hands. It was suggested that she reduce the dose of V temporarily to fix the problems with hands and feet first. She may drop to 1+1 before she flies to NZ on March 3rd. She will take a supply in case she needs more. A member gets her feet done every other month. Her toe nails have gotten thick from the V. Her toes are so active in her everyday activities that she has hard skin under her toes, but not under her heels. And another said that he is getting to the point that the hand and feet skin growth rate and rate of wear are similar, so no special treatments are needed. Although he does use a callus remover on his feet once in a while after a shower. He said that at one time, his finger skin was so thick that his "smart" phone's touch screen couldn't register his touches.
- A member who has been on V might be coming off it and starting on dabrafenib. This is because the V is "messing with" his liver enzymes, which are now "extremely high". Others had heard of the new drug, but were not aware of any published data. He has got approval from his insurer to make the change.
- It was said that Dr. Diamond thinks that newer kinase inhibitors are worth a try. They don't have as many side effects, although sun exposure is still bad. A member thinks that she may try something new in April.
- A member has just had an oncology appointment, and he has been told that all is steady, so the interferon is holding things. He said that interferon has been kind to him, since his diagnosis in 2007. He takes 3 mil units 3 times a week. It is the "first line" drug, but most people who use it don't like it. He has an endocrinologist appointment in the coming week.

- Another take on interferon was given. When a member was diagnosed at Duke last January, the doctor wanted to put her on Interferon. She said that "I politely declined". She said that she was going to refer herself to the National Institutes of Health (NIH), which she did.
- A member said that he knows of a man who only lives 20 minutes away from him, who has ECD and is on interferon. He is apparently stable now. He was diagnosed in 1998 and had to go through a lot before a diagnosis was made. The same member said that a friend of his wife's aunt has ECD, but it is thought that he doesn't know about the ECDGA.
- A member who has been waiting to start V now has the medicine in the house ready to go. There are quite a lot of restrictions related to it, such as diet food, cleanliness, etc. They have been given a special Hotline number to call if there are any problems. The dermatologist needs to be seen soon to get a base line assessment, and a mobility assessment is also needed. He is starting on a dose of 4+4. Everyone tolerates V differently. One member only lasted 3 days on 4+4, whereas another lasted 5 months!
- It was suggested that people should take vitamin D supplements. The members already using them were on different doses; 1000, 2000, and 4000icu a day.
 - One member has been put on vitamin B1 because his level was low. Another takes vitamin D and C. He had to stop taking B12 because he has a high level of platelets (these are found in the blood and involved in blood clotting).
 - One takes fish oil, CoQ10, and a B complex (which was recommended for her brain..."if I only had a brain").
 - Another thinks that there are several supplements that have helped him. He takes Fentanyl, interferon, bupropion and clonazepam. He said that the supplements have taken away most of the side-effects.
 - It was also pointed out that there have been a few recent studies saying that most vitamin supplements are useless.