

## Summary of ECD Global Alliance Internet Chat on Jan 19, 2013

### 7 Attendees

- A relatively new member (diagnosed Aug 2012) came on, having just got back from a trip to the Mayo clinic. The member has been told that the ECD is in the brain, as well as in the bones. A visit to the NIH is planned for March. The knees are very painful. Kineret was started 3 weeks ago. This member wanted to know how you get to a stage where you need to use a wheelchair, and whether this is due to a general weakness, or because "your knees won't hold you up anymore". One member uses a walking frame, which has 3 wheels, or walking stick(s) when outside the house. This member also has powered disability scooters for outdoors. Something was needed, to help with walking, after a year or two (but not inside buildings). The member thinks that it would be good to be able to use an ordinary wheelchair too, when it would be the right sort of vehicle.

A member can manage to get around indoors, without help, but not outside without walking sticks.

And another member also needs to use sticks, because balance is so poor. A fall in the shower had caused a significant back injury.

- We were asked whether any of us had been found to have ECD lesions in the brain. All members had had such findings. The lesions had been found in the cerebellum, and pons. A member, whose wife died due to ECD, told us that when lesions were in the Pons, his wife had been able to walk around using a Walker (4 wheel, with handles) for some months, but then she became unable to stand at all, and also couldn't drive an electric scooter or wheelchair, and so needed to be pushed in a high-end wheelchair.
- We all agreed that no-one can say how fast the ECD will progress, or which part of the body it will affect. It progresses differently in different people. A member developed brain lesions and only survived for 15 months; others have significant effects from the ECD, but it doesn't seem to progress. One member has been in remission for 3 years, but, when it was active, the ECD caused blindness. Another member had brain lesions, found more than 5 years ago, but has been relatively stable with regard to these. But, for this member, lung problems have made for a very "exciting" time!!
- Persistent coughs were discussed. Food, or drink, going "down the wrong way", and "strong" smells such as chilli and black pepper are often to blame. A member has had a cough for, at least, seven years. Never been a smoker, and all the Drs have said that the lungs are "great".
- It was agreed that it was very important to have family, and friends, who can give support.

- Another new member came on whose husband has ECD. He has large organ involvement, ascites and renal failure, and needs to have 5-7 litres of ascites drained off each week. Mayo had made the diagnosis, and Dr McClain in Texas has been seen. The NIH will be visited in May. His treatment is interferon and Gleevec. A PET, done after 3 months of interferon, shows that things are either stable or have improved. He has really thick rinds around both kidneys, and it isn't known whether these are affecting the kidneys, or if it is due to the ascites. They are looking into going to Cleveland clinic, for a TIPS procedure, to stent the hepatic vein that goes to the inferior vena cava (IVC), and also an IVC stent. His vena cava is narrowed. Dr McClain feels that Gleevec may help the fibrosis in the vessels of his liver, but so far it hasn't. Anakinra will be started in a few weeks time, if there is no improvement.
- Retroperitoneal fibrosis is also a problem in another member, but the creatinine level is not raised. This member is already on Anakinra. It was difficult to take, at first, but then not so bad.
- The member, who recently died, had had ECD in the retroperitoneal area, and had needed a stent, in the right renal artery. Treatment with tamoxifen had controlled these lesions for some years.
- Members agreed that, one of the most frustrating things about this illness is, that the doctors only 'feel that it could work', when they try different drugs. As we all know, or have found out, this is the nature of ECD.