

## Summary of ECD Global Alliance chat held on Saturday 25<sup>th</sup> September 2010

6 Present:

- A new member was welcomed to the chat. Their spouse was diagnosed with ECD a month ago. Initial symptoms included pain in the knees and arms. The patient became paralyzed on their left side and was admitted to hospital. They received a diagnosis after being in hospital for three weeks. They are at home now but still partially paralyzed. Tests showed that the patient has a large mass on the right frontal lobe of the brain and some interior right brain involvement.
- Other members on the chat discussed information with regard to medical history and treatments with the new member.
  - Patient 1. This patient was diagnosed in 1995 and has a growth on the pituitary gland that has been monitored since 1992. They received a diagnosis of ECD after brain surgery. The patient has received radiation treatment and has been treated with prednisolone. They have not received any other treatments and are still on 10mg/day of steroids.
  - Patient 2. This patient was diagnosed in 1994, after two and a half years with symptoms. A CAT scan revealed fluid present in the heart. They have been treated with 2CdA for 18 months. This appears to have slowed disease progression.
  - Patient 3. This patient has heart involvement and has suffered from accumulation of fluid in the pericardium. Pericardial window surgery was performed which alleviated the fluid build up. The patient has received six cycles of 2CdA which appears to have stopped the growths. The patient feels they have benefited from supplements and vitamins.
  - Patient 4 . The patient had to stop work due to ECD in 2007 They have received radiation treatment as well as interferon, fentanyl patches and clonazepam.
- Some new emerging treatments were discussed: Gleevec and anakinra (kineret). There is little information on these treatments available at present but there are indications that one or more 'ECD knowledgeable' doctors are interested in them. Gleevec has been prescribed by Dr.

Kurzrock and a paper has been published by Dr. Aouba (Paris) on anakinra as a treatment option. Both these doctors are listed on the ECD website.

- There are links to a number of full-length articles on ECD on the website. There are other articles which cannot be put onto the website because of copyright issues. When these seem particularly interesting and relevant, an announcement is made to the group, so that members may obtain them if required. If anyone finds an article that is interesting and is not on the website, please let the group know.
- The incidence of ECD was discussed. It is unknown how many patients are presently being treated for ECD. The ECD Global Alliance currently has 63 patients from eight countries. As the website is in English this limits access to patients from non-English speaking countries.
- It was discussed that all drugs used to treat ECD are considered 'off label', meaning that they aren't marketed or tested against ECD. Experience suggests that they help however.
- It is mentioned in a number of articles on ECD that there have been less than 350 cases of ECD described in the literature since 1930. This figure may be misleading and actually refers to the number of cases published in the literature, rather than the total number of patients who have received a diagnosis.
- There was a discussion on the difficulty of diagnosing ECD. Lack of awareness may mean that, despite, many investigations, a diagnosis is not made. Raising awareness is difficult as symptoms vary and the disease does not 'fit' naturally within one specialty. One member's spouse underwent numerous investigations and saw many eminent doctors, but the diagnosis of ECD was only made at autopsy.
- The next chat will be held on Saturday 2<sup>nd</sup> October 2010, at 3pm EST.