

Prologue

The Sophie Cameron Trust is quite a new organization – its story began in 1999. In February of that year, Sophie Cameron, who had just been elected Head Girl of the Royal High Senior School in Bath, returned from a holiday in Venice, Italy. She felt poorly, and several days later she was admitted to the Royal United Hospital in a delirious state. Over the subsequent weeks her condition worsened dramatically. She developed many of the classical symptoms of encephalitis lethargica (EL), including oculogyric crises, respiratory irregularity and coma, and was finally discharged home late in 1999 in a minimally aware state. Despite twenty-four-hour care from her family and various professionals, as well as intensive physiotherapy and other treatments, Sophie made little progress, and she died suddenly on 30th May 2006.

During Sophie's illness, her parents, Phillip and Judith, discovered that EL is currently a very rare condition, and it proved frustratingly difficult for them to identify reliable information about it. The many friends of the Cameron family also wanted to learn about EL and to offer practical help. This led them to form the Sophie Cameron Trust. The Trust committee soon realised that not only Sophie, but also others with EL might need assistance, and so the idea of a charitable trust arose. Individuals with the skills that would be needed, in science, medicine, law, accounting, fund-raising, public relations and management, were already involved, and were willing to donate their time to act as trustees or members of the fund-raising committee. So, in April 2002, the Trust was registered with the Charities Commission in the UK as charity No. 1092190. The aims of the Trust were set out in the bylaws, and are, briefly, to help people affected by EL and their families, to increase knowledge about EL and its implications among the medical profession and the general public, and to encourage research into the management and causes of EL.

Our first project, which arose directly out of Sophie's illness, was to explore existing treatments, and with the aid of matched funding from two other local charities, the Medlock Trust and the Brownsword Charitable Foundation, we set up a programme to investigate whether intensive physiotherapy could benefit patients in Sophie's position. Next, we needed to make contact with other EL patients – not easy because of medical confidentiality issues. While we considered how best to do this, we focused on raising the profile of EL among the medical profession. In particular, we wanted to see whether we could encourage a broad spectrum of research into EL without preconceptions as to what might be most valuable.

The requirements for developing a useful research programme seemed clear. Firstly, we needed to identify and engage with researchers who were active in the field. Secondly, we needed to discover whether funding on the relatively small scale that we could provide would prove sufficient to make a difference – we hoped that by providing small grants, we would be able to encourage research into this rare syndrome, stimulating projects that might otherwise never be started, and attracting funding from other sources to extend the projects that were successful. Then, at a conference on encephalitis, we met Dr. Russell Dale, who was the lead author of an article describing 20 modern cases of EL, entitled "Encephalitis lethargica syndrome: 20 new cases and evidence of basal ganglia autoimmunity", published in *Brain* in 2004.

Through Russell, we also met Dr. Gavin Giovannoni; with others, they were investigating whether EL might be part of a spectrum of autoimmune disorders. We received a great deal of help and encouragement from Gavin, and eventually we asked

him if he would act as (unpaid) Scientific Adviser to the Trust, because we knew that an expert neurologist's input would be needed to evaluate research proposals that we might be asked to fund. Happily, he agreed. His advice proved invaluable, and he has played a key role in helping us to develop the programme of research that is described on our website, www.thesophiecamerontrust.org.uk. He also set up the most important single project that the Trust is funding, a national surveillance programme for early identification and study of patients with EL in the UK. This is the only such programme currently operating in the world, and its purpose is to gain near-complete, long-term surveillance of EL across the UK. It involves the inclusion of EL on a monthly electronic questionnaire circulated to all neurologists and paediatric neurologists in the UK. We hope that this programme will eventually establish the incidence of EL, as well as providing epidemiological information, and perhaps insight into the causes of EL. In addition, blood, cerebrospinal fluid and other samples collected during clinical investigations for diagnostic purposes will be stored for future research, provided that patients or their families give their consent. All patients identified will also receive a letter giving them information about the Sophie Cameron Trust.

From the beginning, the Trust wanted to encourage international cooperation in research on EL, and we established contact with the editor and primary author of this book, Dr. Joel Vilensky, who kindly agreed to act as the Trust's "eyes and ears" for developments relating to EL in the U.S. and elsewhere in the world. We had the opportunity to meet him in 2008, when he gave a lecture sponsored by the Trust at the Institute of Neurology in London, attended by a number of eminent British neurologists. We were enormously impressed by his encyclopaedic knowledge about EL, and he generously provided an abridged version of his lecture for our website. Thus, when he requested funding for editorial assistance and translation of non-English-language material during the preparation of this book, we were keen to help. We felt that the book would not only provide a comprehensive resource for clinical professionals, but also would be useful to patients, family members and friends who wanted to understand their experience and place it in a broader context. In addition, we felt that the book would be a memorial for Sophie, and we greatly appreciate the fact that it is dedicated to her.

The Trustees would also like to take this opportunity to thank everyone who has contributed to the Trust; both those who have given their time and skills, and those who have contributed financially or in other ways. Some have preferred to remain anonymous, and others have been acknowledged on our website and in our newsletters, but particular mention here should go to the Royal High School, Bath, and its staff. They gave tremendous help and support to the Cameron family during and after Sophie's illness, as well as generously supporting the Trust by providing facilities for functions and organizing a variety of fundraising activities.

The final part of this Prologue should be dedicated to Judith Cameron, Sophie's mother. During Sophie's illness, Judith wrote a regular column for the Guardian newspaper which, focusing on the role of carers, provided an important forum for raising awareness of EL. The following paragraphs are taken from the article that Judith wrote following Sophie's death:

"At 8.55 pm on Tuesday, 30th May 2006, I was no longer the mother of four children, but three. My beautiful daughter, Sophie, suddenly died. Seven years earlier, aged only 17, she contracted a rare infection of the brain and a few weeks later, in Intensive Care, suffered a cardiac arrest. The oxygen starvation resulted in irrevocable brain damage and when she finally returned home, she needed 24 hour

care. Her condition became stable and we implemented a rigorous regime of physiotherapy to maintain her body in good physical shape. We prayed that, one day, either through drug or stem cell treatment, something would be discovered that could once again give her a life worth living ...

Today I am still working to remove all trace of her awful suffering and yet during those years, I tried hard only to concentrate on the present. It was too painful to dwell on the past and what might have been while I looked after the shadow of who my daughter once was. I was determined to do the best for the daughter I had. But of course Sophie was the little girl I taught to swim, the little girl who held my hand during the births of her younger brother and sister while her Dad took photos. She grew to be the only person in the household, other than me, who noticed when the bin needed emptying or the loo roll replaced. Only now am I slowly allowing myself to recollect the vibrant child I had for seventeen years.

Despite her severe disability, I had always assumed that Sophie would outlive us. I am sure that no mother is ever ready for her child to die. I hate to think that I will never ever see her again - never be able to cuddle or smell her, never be able to show her how much I love her. Any possibility of her getting better has gone. But I can feel that the weight of the seven years of Sophie's illness is starting to lift and I remember again my gorgeous, vivacious, sassy daughter. Although I still weep copiously, I am going to allow myself that privilege of remembering what a fantastic person she was. I will exult in who she was and continue to be proud of being her mother."

Richard Steele
Trustee

The Sophie Cameron Trust
www.thesophiecamerontrust.org.uk