2007 Behçet’s Syndrome Society AGM

The 2007 Annual General Meeting took place at Aintree University Hospital, Liverpool, on 13 October.

Society business

After Chair Jan Mather had welcomed everyone to the meeting, Kai Li presented the accounts for the year ending 31 August 2007. He said that both income and expenditure had increased compared with the previous year, with the balance being slightly lower. Subscriptions had been received from around 500 members, as well as a single anonymous donation of £10,000. Grants totalling £7,500 had been received. The complete financial report is available on the website.

Richard West then introduced the Committee members, who were all duly elected. He appealed for volunteers to serve on the Committee or in other roles. Kathryn Proudlock gave an update on the Helpline, saying that the Telephone Helpline Association had made 70 recommendations, of which 42 were considered high priority. She thanked Jo Elvin, who had left the Helpline, and said that two new volunteers were being trained. More volunteers, especially men, are still needed.

Jan Mather presented an overview of the past 12 months, emphasising the success of the North West project and hoping that it can be rolled out elsewhere. CIA Solutions did not raise as much money as was hoped, but did obtain a grant of £7,000 for website development. A representative from the Committee attended the Liberal Democrat party conference and met Baroness Tonge. The three issues raised were lack of recognition by the medical profession, funding for attending hospital appointments and the lack of facilities for running the Society.

Turning to the upcoming 12 months, the main aim is the appointment of a Director to run the Society. The recruitment agency Armstrong Associates is being used. An advertisement has appeared, and four candidates are being interviewed. Another project is the updating of the Society’s information leaflets, in association with
medical specialists on the Behçet’s Forum. The new leaflets should be more acceptable to medical professionals.

Richard West said that fundraising would be more important than ever in the coming year, as the appointment of a Director will deplete the finances significantly. However, a major part of the new Director’s job will be to raise funds in a more structured and professional way.

**Living with Behçet’s disease**

Society member Antonio Pinto gave a talk on how Behçet’s disease has affected his life. His problems started with mouth ulcers in 1995, which he was told were caused by stress. Joint problems developed, and he was finally diagnosed in 1997. He has since attended many different hospitals in and around London and has had periods when was confined to a wheelchair. He does not tolerate immunosuppressive drugs.

Mr Pinto described how he had lost his friends, partner, job and quality of life as a result of his illness. However, in the past 4 years, he has learnt to cope better and is now able to predict flare-ups and increase his steroid dose accordingly. He said that other countries he has lived in (such as his native Portugal) do not have societies to support people with Behçet’s disease, so he feels very lucky to be living in the UK.

**Local support groups**

Alan Booth, a Society member living in Liverpool, lost his sight through Behçet’s disease. He said that in the 37 years since being diagnosed, he had only ever met two other people with Behçet’s disease. A work colleague saw the number of the Liverpool Support Group at the local eye hospital and passed it on. Mr Booth then spoke to the organiser of the group and attended his first meeting in February. He said that Behçet’s disease can be a very isolating condition, which even has to be explained to doctors. He had found it useful being with other people with the condition, chatting about their common experiences and gaining reassurance that his
problems were real. He encouraged members in other areas to set up local support groups.

Jan Mather added that the Society cannot pass on Members’ details, as permission for this was not asked for when the details were taken. In the future, the Society will be asking people for this permission via the website and newsletter.

**North West Project update**

Gill Grundy has been working 7½ hours a week as a Behçet’s Nurse for the past 2 years, supported by the Society. Her aims were to develop services to meet the demand for specialist care and to improve understanding and awareness of Behçet’s disease.

Achievements in the first year included setting up nurse-led clinics and a helpline manned by specialist nurses, establishment of a basic demographic database, publishing articles in *Nursing Standard* and *Aintree News*, and giving presentations at a local rheumatology meeting. Gill also carried out an audit of biological treatments (such as infliximab) for Behçet’s disease at Aintree; she presented the results in the main hall at the British Society for Rheumatology and at a regional anti-TNF meeting.

The database is the first one in the UK on Behçet’s disease and includes anonymised data on patient numbers, drugs, disease activity and responses to questionnaires on quality of life and depression. Patients on the database are reviewed in monthly nurse-led clinics.

During the second year of the project, Gill has been nominated for the Pfizer RCN award for innovations (for the database) and asked to write an article for the *RCN Bulletin*. Future plans include negotiations to continue the clinics after the funding finishes (for which the feedback so far has been positive), continuing to input data into the database, and carrying out more research. The database will be used to do annual audits of topics such as hospital admissions, drugs, adverse effects, reasons for changing medication, types of symptoms, time from symptom onset to diagnosis, and quality of life aspects of Behçet’s disease.
**Interferon Alpha trial**

Professor Sue Lightman gave a talk on the clinical trial she is running. She said that the drugs currently used in Behçet’s disease treat the symptoms but do not turn off the disease; they also have side-effects. There have been reports in the medical literature (in about 500 patients) suggesting that 6 months’ treatment with interferon alpha may actually switch the disease off, with an effect persisting for 3–5 years. This would allow doses of other drugs to be decreased.

Interferon alpha is an antiviral agent that also affects the immune system, reducing regulatory T-cells. It is used (at higher doses) in hepatitis C and some malignancies. It is very expensive and has some side-effects. The aim of the trial is to take Behçet’s patients on steroids and/or other immunosuppressants and allocate them randomly to having weekly injections of interferon alpha for 26 weeks or continuing as before. The patients in both groups will be followed up for 3 years, with questionnaires to be filled in by patients and doctors on symptoms experienced and drugs used. If the results are good, interferon alpha could become available to all Behçet’s patients.

The trial is running in several centres in London, and is now starting in the North West under Professor Moots. More volunteers are still needed. Prof Lightman stressed that interferon alpha is not an experimental drug and that it has been taken by many people, including those with Behçet’s disease. It does have some side-effects, including reaction to the injections, a flu-like syndrome, and reduced white blood cells and mood. The main objective is to be able to reduce the doses of other drugs needed.

**Ophthalmology talk**

Mr Kamal, an ophthalmologist at Aintree Hospital who runs joint clinics with Prof Moots, spoke about eye problems in people with Behçet’s disease. He said that about 70–85% of Behçet’s patients have eye involvement, usually in both eyes. About a quarter of men with Behçet’s disease present initially with ocular problems. Behçet’s
Eye problems have a relapsing–remitting course, with an average of 1.2 episodes a year, each lasting 2–3 weeks.

Eye problems can be at the front or back of the eye. Problems at the front of the eye include dry eye, punctate keratitis, uveitis, cataract, glaucoma and cranial nerve palsies. Problems at the back of the eye include posterior uveitis, retinal vasculitis, macular oedema, retinal haemorrhages, retinal detachment and optic atrophy. Symptoms at the front include blurred vision, grittiness, painful red eye, while those at the back include photophobia, floaters and gradual loss of central vision. Investigations that might be done include slit lamp examination, fundoscopy, fluorescence angiography, retinal scans and electrodiagnostics for nerve involvement.

Management consists of a multidisciplinary approach with both systemic and ocular treatments. Ocular treatments include artificial tears, topical steroids and intravitreal steroid injections under local anaesthetic. Anti-VEGF agents are new drugs that inhibit formation of new vessels and reduce the risk of bleeding in the eye. Intravitreal anti-TNF implants are in clinical trials. Surgical options include laser photocoagulation, cataract surgery, intravitreal steroid implants, vitrectomy, glaucoma drainage and retinal detachment repair. About half of patients can lose their vision within 5 years, but early identification and treatment can help to prevent this.

Rheumatology talk

Professor Moots then spoke about the development of Behçet’s disease care in Liverpool. He saw his first Behçet’s disease patient in 1997 and his second in 2000, and he now has more than 60 patients. Aintree takes regional and supra-regional referrals, many of them for confirmation of suspected Behçet’s disease. Care is shared with ophthalmology, obstetrics, neurology and dermatology, and there is a specialist nurse.

Ongoing research into Behçet’s disease in Liverpool includes the function of neutrophils (see below), the costs of illness and the interferon alpha clinical trial. The main challenge is to get more doctors to suspect Behçet’s disease and refer patients for confirmation of the diagnosis. Referrals from genitourinary medicine clinics are
important. The problem is that it is a rare disease with common symptoms, such as fatigue and headaches. Concerns about treatment include the best way to use biological therapies – for example, which one to use and for how long. Also, not all patients respond to these therapies, and this is one reason for the interferon alpha trial.

Neutrophils are white blood cells that protect the body against infection by gobbling up bacteria and toxic chemicals. They have been found in Behçet’s disease lesions, but it is not known whether this is a cause or an effect. Preliminary research in four patients has shown that neutrophils are dying spontaneously in some people with Behçet’s disease, and they also spontaneously secrete toxic chemicals in the absence of stimulation. Reduced cell surface receptors (CD16, CD32 and CD11b) have also been found. These results need to be confirmed in larger numbers, and drugs to inhibit neutrophils are in development.

Medical panel

The AGM finished with a medical panel consisting of Prof Moots, Mr Kamal and Gill Grundy. One question to the panel was about the side-effects of anti-TNF drugs. They replied that TNF is important in the defence against infection and cancer, and the long-term worry with these agents is an increased risk of infections such as tuberculosis. A central register has shown no excess of cancer so far. Another question was about the use of oral steroids, to which the panel responded that these have a lot of unwanted effects so intramuscular steroids are preferable, with intravenous ones in emergencies.

Asked about the prospects of a cure for Behçet’s disease, the panel said that it is hard to speculate about this until the cause is better understood. Long-term suppression without side-effects is a more realistic aim. Regarding alternative therapies, the panel stressed that no alternative therapies have been found to be effective but that complementary therapies can help people to cope with the disease and the effects of treatment.

In reply to a question about the possibility of passing Behçet’s disease on to one’s children, the panel said that it is not an inherited condition. Certain genetic markers
are more common in people with the disease, but it is very rare for the child of a patient to also have Behçet’s disease. Asked if total remission ever occurred, they said that this did sometimes happen, for example in women around the time of the menopause.

The panel were also asked questions about some old and new drugs. They said that they would rarely use thalidomide, as a dose that was effective would be likely to have serious side-effects. There is usually a better alternative, and most doctors would go straight to an anti-TNF drug. There is very little experience with rituximab (an anti-leukaemia drug) in Behçet’s disease, and it is as yet experimental. Combining biological agents is not recommended, as it increases the risk without increasing the benefits.

The final question was about stress and depression in relation to flares of Behçet’s disease. The panel said that it is a vicious circle, with Behçet’s causing stress which in turn leads to flares. Feelings are known to influence hormones, and these then affect the immune system.

Clare Griffith, Editor