BSS 2013 Conference and AGM

The 2013 BSS Conference and Annual General Meeting took place at the Forest Hotel, Dorridge, Solihull, on 19 October.

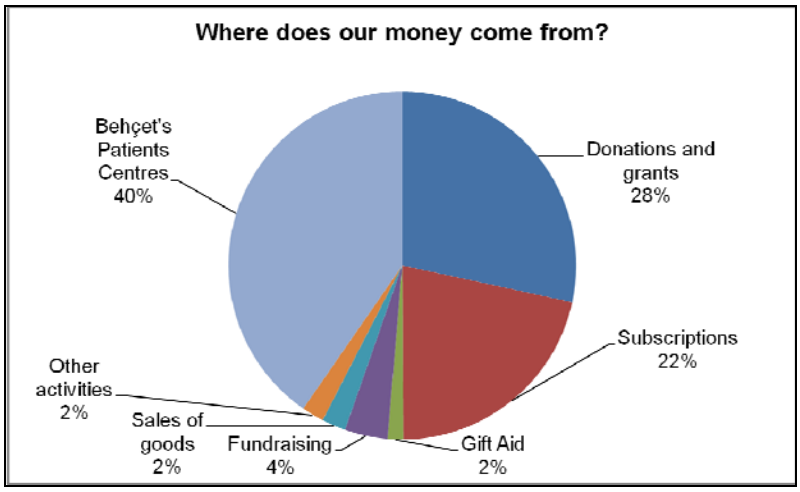
AGM essentials

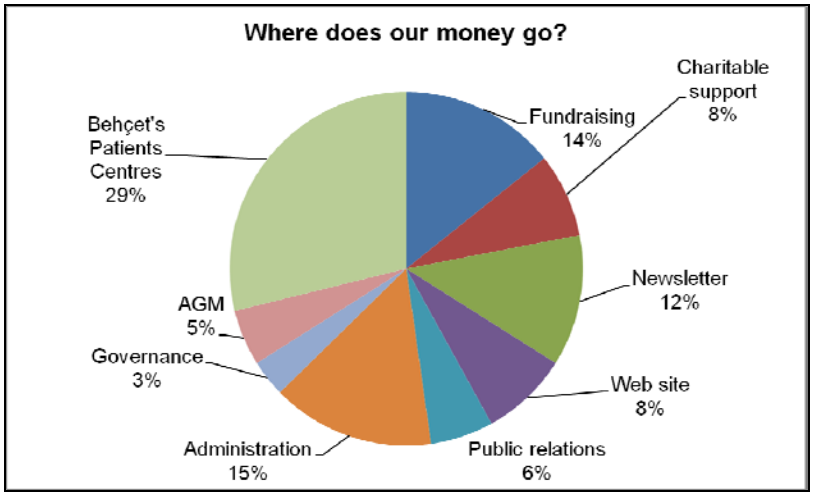
Jan Mather opened proceedings, remarking on the high attendance (over 100 people) this year. Alison Pemberton then presented the minutes of the previous AGM, which were accepted by the members present, and introduced the Trustees, including new Chair Alan Booth. She confirmed that the annual subscription will stay at £20 for 2013/14 and the Grant Aid limit will remain £750.

Treasurer's update

Alan Lane reported that the Society currently has 922 members. These comprise 661 full members, 3 junior members, 45 associates, 30 donors and 183 possibly lapsed members who will have to be deleted from the database to comply with data protection legislation. Of the 922 members, 728 are patients, 73 are carers/relatives, 47 are medical staff and 33 are supporters. Most members (757) live in England, with 66 in Wales, 50 in Scotland, 20 in Northern Ireland, 8 in the Channel Islands and 21 overseas.

The biggest source of money into the Society is the Behçet's Patients Centres (BPCs), followed by donations/grants, subscriptions and fundraising. The Centres also represent the biggest expenditure, followed by administration, fundraising, the newsletter and the website. Overall, the income for 2012/13 was £62,700 (compared with £54,900 for 2011/12) and the expenditure was £62,400 (compared with £44,500). The balance in the bank stands at £33,600, in line with the policy not to keep more than about 6 months' income in reserve. The research fund had built up to over £9000, and a grant of £9751 has been made to the UCL Institute of Child Health at Great Ormond Street Hospital to support research into Behçet's disease. However, the fund has already been replenished by funds raised by the Worshipful Company of Horners, and it currently stands at £5600.

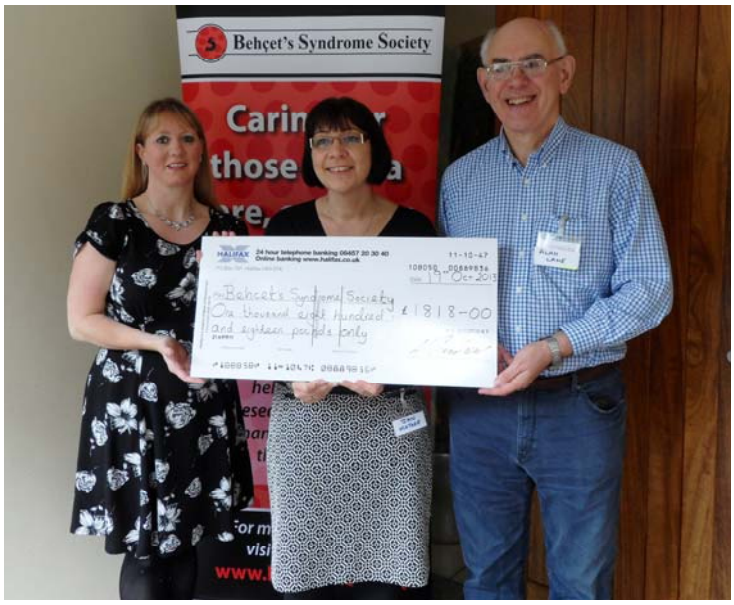




Director's update

Chris Phillips, who works for both the BSS and the BPCs, said that he gets many enquiries from patients and other charities. Medical queries from patients are sent (anonymously) to the Medical Advisory Panel for a response. The number of support groups is expanding, with 10 active groups and at least three more groups in the process of being established. The BSS is also involved in networking with other groups, such as National Voices, Rare Disease UK and Genetic Alliance UK, EURORDIS and the Specialist Health Care Alliance. Website forums linked to the Society are the BSS Facebook page, which has about 800 members, the Health Unlocked forum accessible via the BSS website, and the Rare Connect forum run by EURORDIS.

Fundraising highlights include about £2500 received from a supporter who has run car boot sales every month for the past 18 months, £500 from the Hadrian's Wall forage, £750 raised by swimming the equivalent of the Channel in a swimming pool and £30,000 over 7 years from one anonymous donor. Heidi Goodway, daughter of BSS founder Judith Buckle, presented Chris with a cheque for £1818 raised by doing a sky dive. A major upcoming event is the 'Tour de Behçet's', a cycle ride from London to Paris planned for September 2014. The idea is to leave from the London Centre on 17 September, arriving in Paris on the 20th and returning by Eurostar on the 21st. The cost of the challenge, which should be an amazing experience, will be £750.



Heidi Goodway, Jan Mather and Alan Lane with the cheque for £1818 presented to the Society by Heidi

Update on Centres of Excellence

Jan Mather, Chair of the BPC Directors, gave a brief history of National Commissioning, with funding being obtained for the three Centres of Excellence in 2012. All three centres have now been officially opened, and the BSS was present at each event. The three Support Workers are now all in place, and very good feedback has been received about the job they are doing. Since April, they have seen 79 patients in Birmingham, 73 in London and 42 in Liverpool; clinicians have seen 194, 248 and 357 patients, respectively. Future plans include:

- Exploring ways to cover Northern Ireland, Scotland and Wales
- Collecting information on patients in a database to be used as the basis for further research
- Using feedback from the centres to support a business case for further support worker coverage.

Chair's report

Jan then gave her report as outgoing BSS Chair. Her 7 years as Chair had included a lot of hard work and quite a few difficulties, but also many successes. Jan thanked the BSS Trustees and members, as well as her family and friends, for their support. She listed her top 10 highlights of her time as Chair:

10. Governance of the charity – updating the constitution and improving the membership database and annual reports
9. Fundraising – several successful events and the introduction of Society merchandise
8. The 2010 International Conference – being on the Organising Committee and organising the Patients Conference
7. Publicity – TV appearances, radio broadcasts, magazine articles, an article in the BMJ and attendance at various conferences and other events
6. Web presence – transformation of the website, social media presence and online donations and membership
5. Patient information – updating the patient factsheets and continual improvement of the newsletter
4. Research – publication of Society research and involvement in several research projects and clinical trials
3. Medical relationships – appointment of the Medical Panel, co-ordination of the Behçet's Forum and invitations to speak at medical training days and conferences
2. Helplines – reorganising of the Helpline and provision of handbook, training guide and updated telephony to the volunteers
1. Centres of Excellence – transformation of patient care in England, secure funding for biologic treatment and provision of Support Workers.

Kirsty Millard then paid tribute to Jan and the way the Society had been transformed during her time as Chair. She emphasised the important role of Jan's husband John, her son Sam and her parents. Kathryn Proudlock presented Jan with a large bunch of flowers, and Jan wished Alan Booth, Chris Phillips and all the Trustees success with continuing to develop the Society.

Managing a long-term condition

Dr Sophie Campbell, Clinical Health Psychologist at the Liverpool Centre, spoke about the wide-ranging challenges that Behçet's disease brings to patients and their families, and the resilience and determination needed to live life despite it. She aimed to offer some suggestions that might help people to cope. Long-term conditions have physical, psychological and social effects. In a condition such as Behçet's disease, the variability and the lack of a cure can be difficult to deal with.

Whereas disease involves changes to the body associated with objective findings common to those who have it, illness represents a person's subjective experience of being unwell. Distinguishing between these concepts may offer new ways of self-management. Medical management of disease is essential, but active self-management involves patients in taking the responsibility for parts of the illness under their own control. This can offer opportunities to improve quality of life and, depending on the level of involvement wanted, to become an expert patient. Self-management can lead to fewer or less severe symptoms, an increase in the amount one can do, an improved prognosis and feeling more in control of one's life.

Self-management may require changes to be made, which is not easy. People may be held back by the memory of what they used to do. The process of changes goes from reluctance through awareness, interest and implementation to commitment and finally integration. Coping with change requires a mixture of information and resources. Remembering previous successful changes may help. Patients can develop their own self-management plan (see Box 1), but implementing it can be difficult. Things that can get in the way include the pressures of just surviving, other people's expectations, perfectionism, pride and fear.

Box 1: Developing your own self-management plan

- Lifestyle choices can affect symptoms, for better or worse
- Obtain all the information possible about what may help
- It can be helpful to put together a written self-management plan, incorporating the factors that could help biologically, psychologically and socially
- Be prepared to test things out – no-one gets it right first time!
- Keep it flexible to cope with things like flares and exacerbations
- Share your plan with those around you (this helps them to understand what it is and why it is important)

Mind and body cannot be separated. Emotions can be powerful, but there is much that can be done to manage them or lessen their effect. Worry, anxiety and stress are common in chronic conditions, and may affect physical symptoms and coping. Fear is a natural reaction but can be tiring and reduce quality of life. Low mood and uncertainty are other common reactions. Tips to overcome or reduce the impact of these reactions are shown in Box 2. Inappropriate or outdated thinking can affect self-management and should be challenged by looking for the evidence and seeking other interpretations. It can be hard to accept the realities a chronic condition imposes. Acceptance is a dynamic, not a static, process – it doesn't mean you've given up. It is important to find the right balance between rest and activity, and not to leave seeking help until a crisis point is reached.

Box 2: Tips to deal with common reactions to chronic conditions			
Worry, anxiety and stress	*Fear*	*Low mood*	*Uncertainty*
Restrict worry to 'worry time' Concentrate on problem solving what you can Keep things in proportion Talk it over – this is a problem not a threat Distraction Relaxation	Accept that you may be afraid without criticising yourself Make contingency plans for the future; deal with the present Realise that it is unlikely that the whole collection of fears will happen	Remind yourself that this mood can be temporary Distraction and/or reward Seek help if it develops into depression	Learn to accept limited control over the future Become familiar with the discomfort of uncertainty Today is our only certainty; deal with what's happening now

Overview of Behçet's syndrome

Dr Deva Situnayake, Rheumatologist and Clinical Lead at the Birmingham Centre, began by saying that the gene for HLA-B51 is found in a higher proportion of the population in countries on the Silk Road (e.g. Iran and Turkey). People with Behçet's disease (BD) are 11 times more likely than the rest of the population to be positive for HLA-B51, and it is also a marker for severity of the condition. TNF gene polymorphism is also linked to the severity of BD, and factor V Leiden (a blood clotting gene) is also linked to BD (especially severe ocular disease). Most complications of BD are more common in men, although genital lesions and erythema nodosum are more common in women. The disease tends to be more severe in men, which is in contrast to most autoimmune diseases.

Autoimmunity results from a failure of the immune system to recognise 'self', allowing an immune response against a person's own cells and tissues. Autoimmune diseases such as rheumatoid arthritis and inflammatory bowel disease are very common, and are increasing. Most of these diseases are more common in women. Autoimmune diseases are rarely seen in areas with endemic infectious diseases, such as Africa and much of Asia. Environmental toxins and smoking are also thought to play a role. Autoimmunity is mediated by the adaptive immune system, involving T-cells, B-cells and antibodies. Auto-inflammation is governed by the innate immune system, involving a range of cells such as neutrophils and macrophages. It is characterised by recurrent episodes of systemic inflammation (e.g. fever), multiple tissue involvement and abnormal cytokine regulation (especially interleukin-1 and tumour necrosis factor [TNF]). BD has features of both autoimmunity and auto-inflammation and is probably a mixture of the two.

Various criteria for BD have been developed since 1946, but the most commonly used now are the 1990 International Study Group (ISG) criteria and the 2006 International Criteria for Behçet's Disease (ICBD). The ISG criteria require the presence of mouth ulcers plus two other manifestations; vascular complications are not included. The ICBD include vascular complications, and oral ulcers are not mandatory. Criteria alone cannot be used for diagnosis, and doctors need to take account of other factors such as severity of symptoms, family history and response to therapy.

Dr Situnayake gave a preview of the new international consensus recommendations for neuro-Behçet's disease, which are not yet published. The proportion of patients with neurological complications varies by country but is generally around 10%. Only about 10% of headaches in BD are attributable to neurological disease, so not all require further investigation. For example, migraines in BD patients should be treated in the same way as in any other patients. Neurological involvement in BD has characteristic presentation patterns, is investigated with MRI scanning and is potentially treatable.

Anti-TNF agents are increasingly used in the treatment of BD, but there are still questions regarding the risks versus the benefits and how long treatment should continue. Some information can be obtained from the BSRBR, a register of over 20,000 patients with rheumatoid arthritis treated with biological drugs. No significant increase in mortality or in cancer has been seen compared with patients taking older drugs. There is an increased risk of serious infection, including tuberculosis, especially in the first 6 months of treatment. The databases of the BPCs will accumulate data in patients with BD. A recent review article looked at the evidence for various treatments for different manifestations of BD and made the recommendations shown in Table 1. The BPCs have their own protocol based on a review of the literature, which will evolve as data are collected.

Table 1: Evidence-based approach to treatment of Behçet's disease

	Mucocutaneous	*Ocular*	*Vascular*	*Articular*	*Gastrointestinal*	*Neuro*
1st line	Colchicine	Steroids, ciclosporin, azathioprine	Steroids, azathioprine, cyclophos-phamide	Colchicine	Sulfasalazine, steroids	Steroids
2nd line	Steroids, dapsone, azathioprine, thalidomide	Interferon-α, anti-TNF	Anti-TNF	Azathioprine, steroids	Azathioprine	Azathioprine, cyclophos-phamide, anti-TNF, interferon-α
3rd line	Pentoxifyline, methotrexate, ciclosporin A, interferon-α, anti-TNF	Methotrexate, mycophenolate mofetil, cyclophos-phamide, rituximab	Anticoagulants, antiplatelets	Methotrexate, sulfasalazine, interferon-α, anti-TNF	Anti-TNF	Methotrexate, anticoagulants

Top tips for oral hygiene and mouth ulcers

Dr John Hamburger, Oral Specialist at the Birmingham Centre, said that a mouth ulcer is a breach in the epithelium exposing the underlying connective tissue – in other words, a hole in the lining of the mouth. Oral ulcers can result from trauma, infections, tumours or blisters, or can be idiopathic (such as the recurrent aphthous stomatitis [RAS] seen in BD). Ulcers are associated with many other conditions, including coeliac disease, inflammatory bowel disease and systemic lupus erythematosus. They can also be linked with the menstrual cycle, smoking cessation, stress or use of toothpastes containing sodium lauryl sulphate.

RAS affects up to 20% of the UK population and is characterised by recurrent and episodic blisters, usually with no identifiable cause. The multiple round or ovoid ulcers have well-defined margins, a yellow/grey homogenous base and an inflammatory halo; they are associated with varying degrees of pain. Around 80% are classified as minor, 5–10% as major and 5–10% as herpetiform (see Box 3); any of these may be seen in BD. The ulceration in BD can be confused with erythema multiforme or mucous membrane pemphigoid.

Box 3: Clinical types of recurrent aphthous ulceration		
Minor	*Major*	*Herpetiform*
Up to 1 cm diameter – usually around 5 mm Non-keratinised mucosa and dorsum of tongue Up to 10 in number – usually less Symptoms often mild Heal in 7–14 days Variable recurrence – monthly episodes common	>1 cm diameter 1–3 in number No site restriction Often distally in mouth Persist for 1–2 months Heal with scarring Very painful May necrose tissue Can mimic cancer	Multiple ulcers 1–2 mm but may coalesce to form large irregular ulcers No site restriction Healing approx 2 weeks More common in females Variable recurrence rate but often rapid NOT related to herpes virus

Investigation of mouth ulcers may involve blood tests such as a full blood count, serum iron and folic acid/vitamin B12, clinical chemistry and immunology. Biopsies are rarely done. Treatments include antibacterial mouthwashes (chlorhexidine, doxycycline), anti-inflammatory mouthwashes (Difflam), covering agents (Orabase, Iglu), topical anaesthetics, topical corticosteroids (mouthwashes or nasal/asthma sprays), systemic corticosteroids (± azathioprine) and immunosuppressant drugs (colchicine, thalidomide, anti-TNFs). A systematic review of 25 clinical trials of 21 different interventions reported in 2012 that no single treatment had been found to be effective. However, this may be due to poor methodology, and the review recognised that individual drugs appear to work in individual patients.

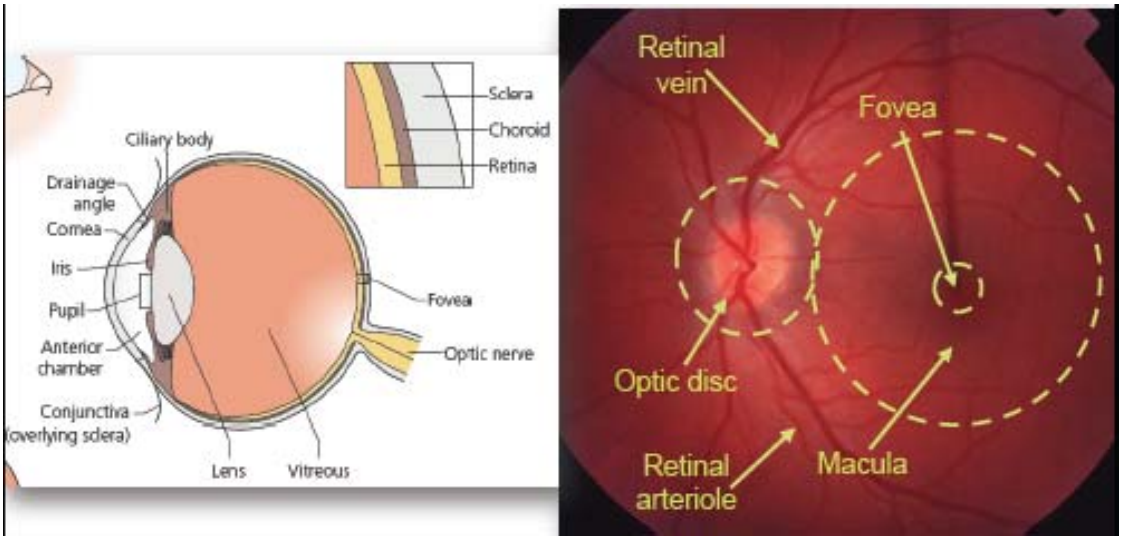
Poor oral hygiene contributes to increasing inflammation, leading to pain. It aggravates existing oral ulceration but is not a cause of ulcers, although sharp edges resulting from dental decay can cause ulcers. Increased bacteria in the mouth may cause secondary infection of ulcers, and more severe oral ulceration may possibly exacerbate other features of BD.

Electric toothbrushes may be useful for people who have difficulty using a manual toothbrush, but they can be very expensive. A review of 354 trials in more than 2500 patients found that only one type of electric toothbrush (the rotating oscillating toothbrush) showed a statistically significant benefit over manual toothbrushes, and the benefit was modest. It removed 7% more plaque and reduced gingivitis by 17%.

It is important to avoid frequent and/or sticky sweet foods. Anti-microbial mouthwashes may be of some benefit in improving oral hygiene. Effective tooth brushing and interdental cleaning with floss, electric flossers or interdental brushes are essential.

The eyes and Behçet's syndrome

Prof Phil Murray, Ophthalmologist at the Birmingham Centre, began by explaining the anatomy of the eye, with the cornea, iris, pupil, anterior chamber and lens at the front, the retina and optic nerve at the back, and the vitreous (jelly-like fluid) in the middle. The retina (the light sensitive layer) is surrounded by two other layers, the choroid and the sclera. The most important part of the retina for function is the macula, with the fovea at its centre.



In uveitis (intraocular inflammation), there is inflammation of the structures of the eye. Anterior uveitis involves inflammation of the iris and the anterior chamber and results in pain, redness and light sensitivity (photophobia). Posterior uveitis involves inflammation of the retina and choroid, resulting in floaters and loss of vision. Intermediate uveitis involves inflammation of the vitreous, while pan-uveitis refers to inflammation of all these structures.

Uveitis can be classified as infectious (e.g. bacterial, viral, fungal or parasitic), non-infectious (e.g. known systemic associations such as BD) or masquerade (e.g. tumour). The eyes are rarely the first system involved in BD (that most commonly being oral ulceration), but they are the next most common second system involved after genital ulceration. Diagnosis of uveitis is clinical, as blood tests are usually negative. In some BD patients, only one eye is affected.

The eye is examined with a slit lamp. In anterior uveitis, inflamed blood vessels in the iris leak albumen (a protein), which appears as dots. The albumen settles to form a hypopyon, which is very characteristic of BD. In posterior uveitis, the retinal veins, and eventually the arteries, become blocked leading to lack of nourishment and vision loss. Fluorescein angiography can be used to visualise the leakage of dye (and white blood cells) from the veins. Accumulation of white blood cells in the retina results in infiltrates, which are also characteristic of BD. Cystoid macular oedema, the main complication of uveitis, results from accumulation of fluid in the fovea. Optical coherence tomography (OCT) is a non-invasive method to see inflammation of the fovea, which destroys central vision. In end-stage uveitis, no vessels remain in the retina.

Treatments for uveitis in BD include topical, periocular, intravitreal (injection or device), oral or intravenous corticosteroids, and immunosuppressants such as methotrexate, azathioprine, ciclosporin, tacrolimus and mycophenolate. Biological drugs, such as anti-TNFs (infliximab, adalimumab) and interferon-alpha, appear to be the most effective therapies for Behçet's uveitis. Improvement in macular oedema can be demonstrated. There is very little good clinical trial

evidence for treatment of Behçet's eye disease, with most publications being of non-randomised studies and case reports.

Newer therapies include intravitreal implants, such as Ozurdex (dexamethasone) and Iluvien (fluocinolone), and XOMA 052 (gevokizumab), a recombinant humanised anti-interleukin-1β antibody. One study in seven patients with uveitis resistant to azathioprine and/or ciclosporin found that a single intravenous infusion of XOMA 052 led to complete resolution of intraocular inflammation in a median of 14 days, with a median duration of response of 49 days.

Final formalities

The day finished with the medical speakers answering questions from the audience. Next year's conference and AGM will be held on 18 October in Liverpool.

Clare Griffith, Editor