Behçet’s UK 2022 Conference and AGM

The 2022 edition of the Behçet’s UK Conference and Annual General Meeting took place on Saturday 15 October at the Holiday Inn, Stevenage, and online. A total of 193 people registered to attend, of whom 54 attended in person and at least 56 attended online. About half (48%) of those attending in person were members.

Welcome

Tony Thornburn, Chair of Behçet’s UK, welcomed everyone to the 2022 Conference and AGM. Baroness Ritchie of Downpatrick, Patron of Behçet’s UK, welcomed everybody to the in-person and online event, pointing out that this is the likely format for the future. She thanked the Chair and Trustees of Behçet’s UK, as well as Prof Farida Fortune and the staff of the London Centre of Excellence for organising the conference programme.

Behçet’s UK AGM

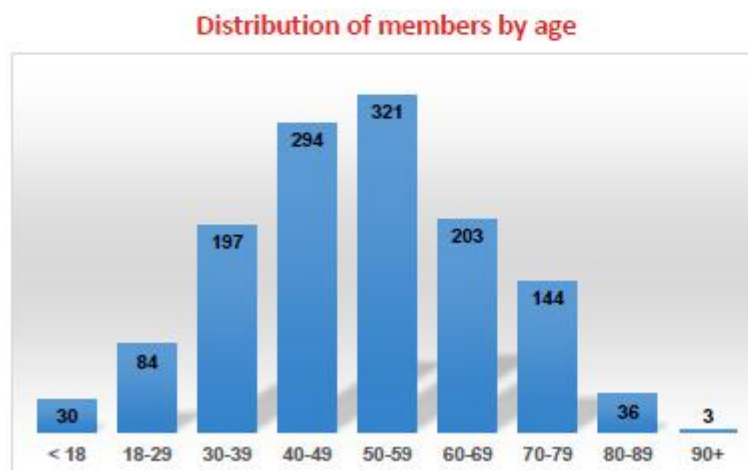
Tony Thornburn encouraged members to read the Annual Report, which is available on the Behçet’s UK website. He thanked the administrative staff and all the trustees for their work, mentioning Alan Lane’s excellent oversight of the accounts. He also thanked the organisers of the various face-to-face and online support groups. The quarterly newsletters reflect another successful year for the charity, with an emphasis on research and the fantastic fundraising by members and supporters. The wider visibility of Behçet’s continues, with an increased social media presence, input into the UK Rare Diseases Strategy Action Plan and engagement with all-party political groups, including in the devolved nations. Behçet’s UK works closely with the Behçet’s Patient Centres and the Medical Advisory Panel, and work is ongoing to update the medical factsheets.

Looking forward, Tony mentioned the plan to establish a constituent management system to streamline interactions with members. Another aim is to improve outcomes for Behçet’s patients in the devolved nations, including establishing a specialist nurse in Scotland to help with coordination of care. The Centres of Excellence are involved with the British Society for Rheumatology and the British Association of Dermatologists in the development of a UK-specific, NICE-accredited guideline for the management of Behçet’s. Regarding research, following the completion of the

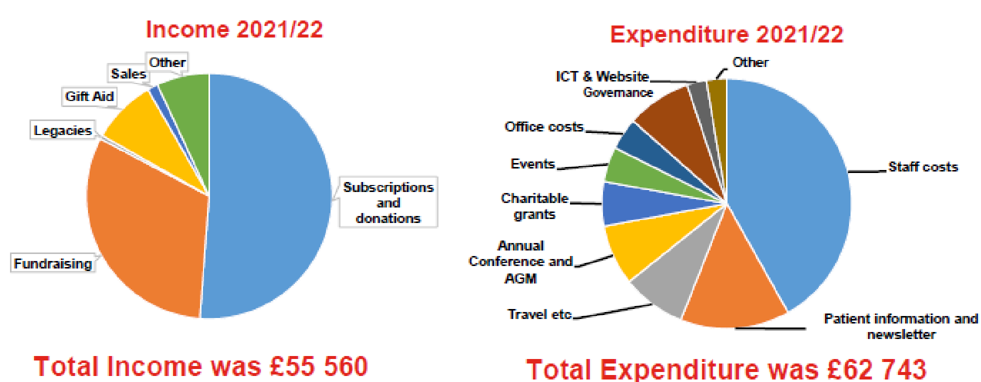
Bio-Behçet's trial comparing interferon-alpha and infliximab, Prof Robert Moots has secured £700,000 in funding from Novartis for a major clinical trial of secukinumab, a biologic drug used effectively in several inflammatory conditions. The epidemiological research by Dr Priyanka Chandratre in Birmingham is on target and will form a baseline for future research as well as feeding into a patient registry. A study on monogenic mimics of Behçet's, being conducted by Alice Burleigh under the supervision of Prof Paul Brogan at the Institute of Child Health in London, will provide insights into the genetics of Behçet's-like diseases.

Finally, Tony said that the 2022 Friends and Family Day had been a great success. Behçet's UK hopes to extend the opportunity to Scotland, Wales and Northern Ireland in the future.

Alan Lane, Honorary Treasurer, reported that the membership of the Society has continued to increase steadily, now standing at 1236. The benefits of standard membership include a quarterly newsletter, access to the helpline, participation in support groups, invitations to family days and free attendance at annual conferences. In addition to the standard members, there are 30 junior members, 77 associates, 21 donors and 79 lapsed members. Almost all (1184) members are patients, with 121 carers, 52 supporters, 52 medical staff and 20 overseas members. Most members are aged 40–60 years, and most (1172) live in England. For the 12th year in a row, the membership subscription remains at £20 per year in 2022/23. A typical grant award is capped at £500, but up to £1000 can be awarded in exceptional circumstances.



The charity's total income for 2021/22 was £55,560. Just over a half came from subscriptions and donations, with another third from fundraising activities and nearly 10% from Gift Aid. Total expenditure was £62,743, 42% of which was accounted for by staff costs. Although this represents an operating deficit, the balance remains high at £155,250 as a result of a large legacy; the ideal balance would be between 6 and 12 months' normal income (~£40–80,000). However, some large items of expenditure are planned, including a patient registry and a specialist nurse for Scotland, so continued fundraising efforts are essential. Finally, the research fund currently has a small balance of £7855.



Neil Williams, Behçet's UK Trustee, reported that the amazing fundraising efforts this year brought in £17,500 for the charity. Several events relating to the 40th anniversary of Behçet's UK are planned for 2023, including '40 in 40', 'Bake for Behçet's' and 'Life begins at 40'. Other ways to help the charity's finances include buying merchandise and Christmas cards, shopping through Amazon Smile and other shopping partners, donating through sales on eBay, playing the weather lottery, fundraising on Facebook and considering leaving Behçet's UK a legacy. Neil asked people with ideas for fundraising to reach out to him and consider joining the newly formed Fundraising Committee.

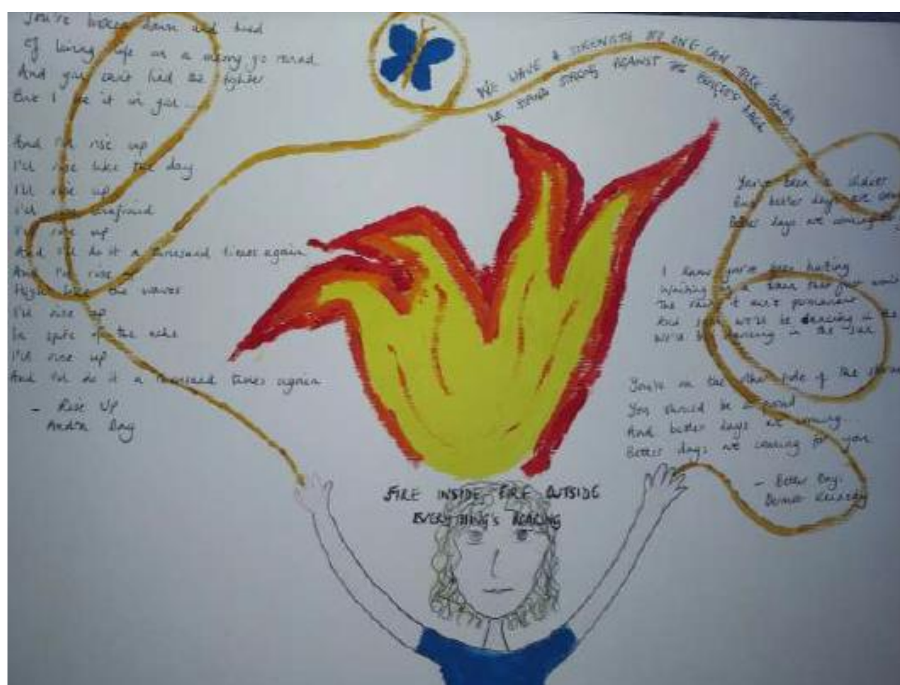
Rachael Humphreys, Behçet's UK Trustee, announced that the recipient of the Judith Buckle Award 2022 was Richard West. Richard was diagnosed with Behçet's by Prof Dorian Haskard in the 1990s and, like many others, spoke to Georgina Seaman at what was then the Behçet's Syndrome Society and realised that he was not alone. Wanting to do the same for other patients, he became a Trustee in 2001 and went on to be Secretary and Vice Chair of the Society. He has made a major contribution to improving care for people with Behçet's, raising the profile of the

condition across Europe as a patient representative at EURORDIS and acting as a patient representative on the EULAR Guidelines Committee. He was instrumental in organising international patient conferences in London and Paris, and helped to set up the Centres of Excellence in the UK.

Richard thanked everybody involved in selecting him for the award, saying that he has always enjoyed talking to people about Behçet's. He emphasised the importance of the inclusion of support coordinators and psychologists at the Centres of Excellence, pointing out that this is not available to people with other rare diseases. He said that treatment of Behçet's has advanced a lot in recent years, as has Behçet's UK.

Tony then announced a new initiative to recognise the contribution of individual members who have been instrumental in supporting anyone in the Behçet's UK community, improving their quality of life, independence and dignity. The first Behçet's UK Members' Award was awarded posthumously to Annette Robinson, who ran support groups in London and East Anglia as well as a private Facebook site where more than 400 people could share personal information. The award was accepted by Annette's husband Brian, who said that she was very fond of all her friends in the Behçet's community and that it had meant a lot to her to be able to talk to people with the same condition that she had.

To finish the AGM, Rachael spoke about the Breathe Creative project, in which Behçet's UK members were invited to register for a creative project to explore their lived experience of Behçet's through the arts, with the end goal being an animated film. Behçet's UK worked with Breathe Creative, an organisation that supports health and wellbeing through the arts; Katja Stiller, a trained therapeutic arts facilitator; BAFTA award winning animator Jane Hubbard; and musician Jacob Meadowcroft. During the 10-week process, the 24 members who registered an interest split into two groups to explore and reflect on their diagnosis, care, treatment and experiences in a safe space through creative writing, art, poetry and music. They then worked on the script, which involved a balancing act to incorporate the darkness and struggle of living with Behçet's, but also the hope, strength and resilience. The end result (which can be viewed on the Behçet's UK website) is a raw, vulnerable and emotive film that reflects the lived experience of Behçet's.



Behçet's Patients Support

John Mather, BPS Operations Manager, said that the team wanted to reach out to patients and listen to their issues and concerns. He asked the patients attending (in-person or online) who have visited or had contact with one of the Centres of Excellence to contact him during the day and let him know what went well, what did not go well and how things could be improved.

COVID-19 vaccination

Prof Farida Fortune, Clinical Director of the London Behçet's Centre of Excellence, began her presentation by reminding everyone that COVID-19 has not gone away. She added that while it is sometimes referred to as the 'great equaliser' it has actually had the opposite effect, increasing poverty and inequalities. People aged under 65 who live in the poorest 10% of areas in England were almost four times more likely to die from COVID-19 than the wealthiest people. The death rate has decreased significantly with vaccination, and it is important that everyone who is eligible is vaccinated against both COVID-19 and flu.

Since March 2020, clinics at the London Centre have been a combination of face-to-face and telephone appointments. Face-to-face appointments are important for Behçet's patients, so the London team did their best to keep the centre open throughout the pandemic. COVID-19 symptoms are now much milder than they were in 2020 and are very like those of a cold or flu. The most common symptoms in people with Behçet's in 2022 are cough, fever, fatigue, joint pain, flu-like symptoms and shortness of breath. COVID-19 symptoms do not appear to differ between the various phenotypes of Behçet's. Patients from the Mediterranean and Middle East are more likely to have had COVID-19 than others, but the rate of infection in Behçet's patients overall is lower than in the general population and long COVID has been rare. Shielding was beneficial for people with Behçet's.

Response to COVID-19 vaccination was similar in male and female patients and among those with and without Behçet's disease activity. The response was better in younger patients and in those who received the Pfizer rather than the AstraZeneca vaccine. Response was lower in patients with neuro-Behçet's and in those taking versus not taking medication. Care needs to be taken in patients on high-dose prednisolone, cyclosporine A, methotrexate or cyclophosphamide, which prevent

antibody production; treatment needs to be planned around vaccination. Prof Fortune reiterated that the potential dangers of COVID-19 outweigh the risks of vaccination.

Dealing with pain in Behçet's

Prof Ali Jawad, Consultant Rheumatologist at the London Behçet's Centre, began by explaining that there are four types of pain. Acute, or nociceptive, is caused by injury or inflammation pain and lasts less than 3 months, while chronic pain can be nerve-related (neuropathic) or functional (neuroplastic). Acute pain settles with or without treatment, but chronic pain is very difficult to treat. There are many causes of pain in patients with Behçet's, including ulcers, skin lesions, headache or migraine, arthritis and arthralgia (joint pain), and there may also be unexplained pain that is difficult to treat.

Joint pain without swelling is called arthralgia and is very common in many diseases, including COVID-19 and flu. It occurs in half of Behçet's patients and is related to active disease manifestations elsewhere in the body, usually easing when they do. It responds to colchicine or anti-inflammatory tablets such as ibuprofen. Joint pain with swelling is due to arthritis and affects the knees, ankles, hands and wrists. It usually has an acute onset and resolves in a few days or weeks. It can be treated by removing the fluid from the joint and injecting steroid. Joint pain is not very often the main problem in Behçet's, and joint damage is very rare. Joint pain unrelated to Behçet's may be caused by problems such as hypermobile joints or scoliosis in patients under 40 and by menopause, early osteoarthritis, degeneration of the spine or lack of exercise in older patients.

Up to 40% of Behçet's patients have widespread body pain or fibromyalgia that is unrelated to disease activity and does not respond to analgesics or steroids. It is associated with fatigue, poor sleep, headache, irritable bowel and bladder, painful periods and exercise intolerance. Tests are normal. Management involves staying as active as possible, eating a balanced diet and getting enough vitamin D, and practising good sleep hygiene; Tai chi has been shown to be helpful. It is important to avoid opiates such as codeine or tramadol, as these do not help and are very addictive, but a small dose of an antidepressant such as amitriptyline may help some patients by improving sleep.

Understanding neurological complications in Behçet's

Dr Desmond Kidd, Consultant Neurologist at the London Behçet's Centre, explained that neurological complications of Behçet's include inflammation within the brain, the veins and the arteries, as well as headaches and cognitive/psychological symptoms. Neurological involvement has been reported as occurring in 3–20% of people with Behçet's, usually in the first few years. Meningoencephalitis (inflammation of the brain and the surrounding membranes) most often involves the brainstem but can also involve the hemisphere, spinal cord or cranial nerves (including the optic nerves). Magnetic resonance imaging (MRI) shows characteristic lesions, which may be tumefactive (resembling a tumour). Inflammation within veins can lead to venous sinus thrombosis, cortical vein thrombosis, intracranial hypertension, headache, visual changes and, rarely, stroke-like events. Inflammation within arteries is very rare but can cause headache, stroke-like events, aneurysm formation or brain haemorrhage.

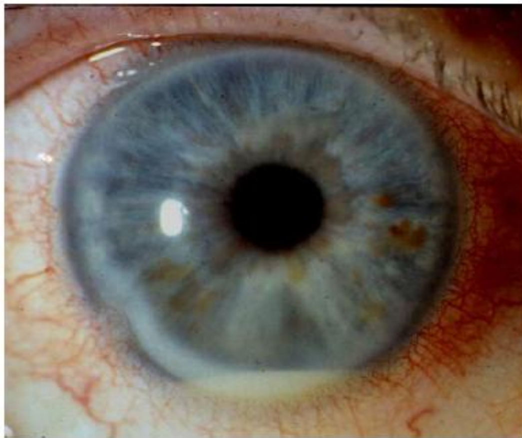
Headache is common in people with Behçet's; the prevalence is >80%, with 98% being vascular-type headaches. Debilitating symptoms are common, and treatment is often inadequate. MRI scans are normal, as are cerebrospinal fluid pressure and constituents. Migraine preventatives, anticonvulsants and triptans are used in the treatment of Behçet's headaches, but codeine should not be used. Dr Kidd finished by mentioning that 'psycho-neuro-Behçet's symptoms include cognitive dysfunction, fatigue, anxiety and depression, and (very rarely) psychotic symptoms.

Eye problems in Behçet's

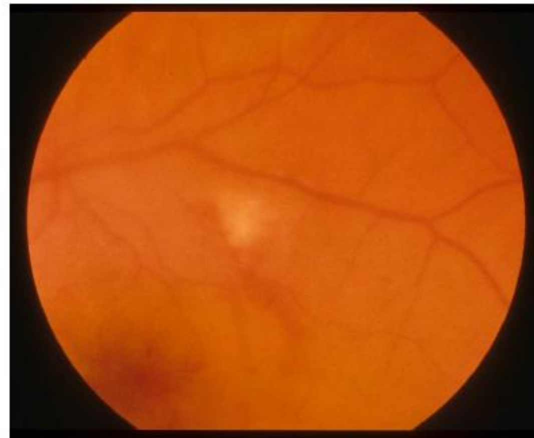
Mr Marwan Ghabra, Consultant Ophthalmologist at the London Behçet's Centre, said that ocular involvement is one of the principal manifestations of Behçet's. It occurs in around 50% of patients overall, with rates as high as 70% among young male patients. Eye disease usually develops within 3 years of oral ulcers first appearing and is the initial symptom in around 20% of patients. In early disease, the only complaint may be a slight impairment of vision associated with few floaters, but some patients complain of redness, pain, sensitivity to light and tearing.

Hypopyon (the presence of pus cells in the anterior chamber of the eye) is seen in a third of cases of anterior uveitis in Behçet's. Anterior uveitis may resolve spontaneously or can progress to glaucoma. Acute anterior uveitis alone usually has a good outcome. Posterior uveitis is a more serious condition and can involve vitritis,

retinal infiltrates, recurrent ischaemic retinal vein occlusions, macular oedema/ischaemia, and neovasularisation/vitreous haemorrhage whereby new vessels form which are abnormal and can bleed and leak. Late ocular complications of Behçet's include widespread changes to the retinal pigment epithelium, fibrosis of vessels, secondary optic atrophy, rubeotic glaucoma and phthisis resulting in a shrunken, non-functional eye.



Hypopyon in Behcet's disease



Intra retinal infiltrate in Behcet's disease

Prognostic factors that are taken into account when selecting treatment for eye disease include the patient's sex and age, disease duration, site of Behçet's symptoms and frequency of ocular recurrences. Anterior uveitis may not need treatment, and if it does then topical treatment is usually sufficient. Posterior uveitis on the other hand almost always needs systemic treatment; 80–90% of eyes will be blind within 4 years if left untreated. The EULAR recommendations state that uveitis involving the posterior segment should be treated with a regimen containing immunosuppressive (azathioprine, cyclosporine A) or biologic agents (interferon alpha, anti-TNF-alpha). Systemic corticosteroids should be used only in combination with immunosuppressive agents. Treatment should prompt and closely monitored. First-line treatment of uveitis at the Centres of Excellence is effective in 80% of patients, although 50% later relapse.

Mr Ghabra concluded by saying that patients need to be educated so that they seek help quickly. Behçet's is still a blinding condition if not treated promptly and effectively. Conventional treatment is largely effective in Behçet's uveitis, and there is a role for infliximab/adalimumab in difficult cases.

Psychology and Behçet's

Dr Steve Higgins, Clinical Psychologist at the London Behçet's Centre, spoke about the psychological aspects of living with Behçet's. It is important to determine whether a patient's presentation is a result of their physical state or their social and environmental context. It is rare that someone presents with a purely psychological concern. People's early experiences shape how they cope with adversity, and their medical journey can be long and frustrating. Diagnostic delay can lead to medical trauma, which can arise from repeated minor healthcare events or difficult consultations, as well as from major medical events. A feeling of not being believed or that nobody can help can be very damaging. Diagnosis leads to help and treatment, but not a cure, so patients need to adjust to living with a long-term condition.

Learning to live with a chronic condition can be likened to a process of grief and loss, with stages of denial, anger, bargaining and depression. However, the final stage is better described as acceptability than acceptance. One has to adjust to living with the symptoms, as well as with the effect on family life, work and socialising. Some people 'mask' themselves and do not let other people see how much they are affected. Having a chronic condition changes your sense of who you are; it is important to make sense of your experiences and realise that the problem is not who you are but your circumstances. In the case of an autoimmune disease such as Behçet's, some of the psychological, behavioural and functional changes are caused by the immune system's inflammatory response. Flares can lead to increased pain sensitivity and fatigue, low mood and mild cognitive impairment such as memory problems.

In terms of ways of helping, family and personal relationships are very important, and wider communities and support networks can also be of great benefit. Psychological therapy mainly involves self-help strategies such as mindfulness-based cognitive therapy to help cope with stress, acceptance and commitment therapy to help build a meaningful life and compassion-focused therapy for developing a better relationship with oneself and others. Self-help books are also available to help overcome body image problems and intimacy issues, as well as to manage chronic pain and fatigue.

Is this Behçet's?

Dr Bindi Gokani, Specialist Staff Doctor at the London Behçet's Centre, explained that history and symptoms, physical examination and investigations are used to determine whether a person has Behçet's. Sometimes the process is easy and the answer is clear, but often it is very complex. Many areas of the body are affected by Behçet's, but the symptoms are common to others as well. Blood tests can give information about nutritional deficiencies, infections and other conditions, as well as much other information. As diets change, the risk of nutritional deficiencies increases. Common deficiencies include iron, vitamin B12, folate and vitamin D, which can cause symptoms similar to those seen in Behçet's.

Dr Gokani described several cases seen in the London Behçet's clinic. A woman in her early 60s presented with oral and genital ulcers, fatigue, joint pain and skin lesions. She had iron deficiency anaemia, and her symptoms improved after it was treated. Iron deficiency anaemia can also exacerbate symptoms of Behçet's. Vitamin D deficiency is also common in Behçet's and causes fatigue, headache, mood changes, muscle cramps, and bone and joint pain. Another female patient, in her mid-70s, presented with oral, genital and skin lesions, as well as weight loss. She was found to have lichen planus, and her symptoms were much improved after treatment. Patients with lichen planus are quite commonly referred to the Behçet's clinic. A 64-year-old woman's presentation of oral and genital ulcers and skin lesions did not quite fit the picture of Behçet's; after blood tests and a biopsy, she was diagnosed with pemphigous, one of a group of immunobullous diseases.

Among younger patients, a 37-year-old woman had a history of oral ulcers, swollen lips and genital ulcers, but had no lesions when she was seen in the clinic. A thorough history and examination provided clues to the diagnosis of erythema multiforme, which was successfully treated. An 8-year-old girl, meanwhile, who presented with oral and genital ulcers, skin lesions, leg pain and fever, was found on genetic testing to have Majeed syndrome, a rare disease characterised by recurrent episodes of fever and inflammation in the bones and skin.

Dr Gokani concluded that Behçet's can be difficult to diagnose, as there are many conditions with similar symptoms. Establishing a diagnosis requires a team approach and careful attention to the history, examination and any test results.

Behçet's Patients Support

John Mather returned to present the feedback received during the day. Key themes included:

- Clinic logistics (can be difficult to find)
- Shared waiting areas (reception staff not always aware of Behçet's)
- Clinic administration (post-clinic support/information, notification of new/cancelled appointments)
- Supportiveness of staff (medical, psychological and non-clinical all very good)
- Communication (checking in with patients waiting to be seen)
- Waiting times (to see consultant, have blood taken, collect prescription)
- Medical information sharing (impact of Behçet's on other medical procedures)
- Out of clinic support (how to contact clinic, where to go for support)
- Processes (information flow, prescriptions, follow-up appointments)

John said that the team would first look for any 'quick wins' and then develop medium- and long-term goals to improve patients' journey through the clinic.

Medication and compliance

Dr Sarah Sacoor, Specialist Doctor in Behçet's at the London Centre, offered some practical tips on taking medication. It is important to take medication as instructed because it manages symptoms and prevents severe complications of Behçet's such as vision loss, blood clots and neurological disease. Patients take their medication because it was prescribed by their doctor, they understand the need for it and are aware that stopping it will result in their symptoms worsening, and they are worried about severe disease. They may stop their medication because they have side effects, forget to take it or run out of it. They may feel better and think they don't need it anymore, or they may be worried about immune suppression or COVID-19. Other reasons include having an infection, not being sure how long to take the medication for and being pregnant.

Biologic drugs such as infliximab are used for severe disease and are usually prescribed for 6–12 months. Most patients continue their other drugs as well and have regular blood tests; it is important to collect the test results, as they will not be available to the Behçet's clinic. Steroids such as prednisolone are rescue drugs given for acute episodes of disease; long-term use can cause adverse effects, but medical advice should be sought before stopping them. Colchicine is a first-line anti-inflammatory drug; high doses can cause gastrointestinal side effects, but the lower doses used in Behçet's are well tolerated. It is safe in pregnancy (although it can cause sickness) and during breastfeeding. Other anti-inflammatory drugs such as azathioprine, mycophenolate mofetil and cyclosporine take several weeks to start having an effect, and regular blood tests are needed. Azathioprine can cause side effects initially but is safe in pregnancy. Both men and women should not conceive while taking mycophenolate mofetil. GPs may not prescribe these drugs, but the centres can prescribe them if requests are sent in advance along with the most recent blood test results.

Topical treatments include Benzyderm gel, which can be applied three or four times a day to relieve mouth ulcers, and Triorasol mouthwash, which is very effective for mouth ulcers and also reduces scarring. The powder is dissolved in warm water and used for 2 minutes to rinse the whole mouth four times a day; no food or drink should be taken for an hour afterwards. Difflam mouthwash can be used before eating or tooth brushing to reduce pain.

<u>Medical Panel Q & A</u>

The day finished with a question and answer session with a Medical Panel comprising the staff of the London Behçet's Centre. The panel was asked if it is safe to have vaccinations for COVID-19 and flu at the same time, to which the answer was yes, as long as you do not react badly to jabs. Asked about the evidence for cold water therapy in Behçet's, the panel said that it is sometimes used in cancer patients but there is no evidence for any benefit in Behçet's. In response to a question about the best drug to take during pregnancy, the panel agreed that azathioprine is a good option.

Clare Griffith, Editor