

Behçet's UK 2024 Conference and AGM

The 2024 edition of the Behçet's UK Conference and Annual General Meeting took place on Saturday 19 October 2024 at the Holiday Inn Liverpool City Centre Hotel and online. A total of 145 people registered to attend, of whom 60 registered to attend in person and 85 to attend online.

Welcome

Tony Thornburn, Chair of Behçet's UK, welcomed everyone to the 2024 Conference and AGM. Baroness Margaret Ritchie of Downpatrick, Patron of Behçet's UK, thanked Prof Rob Moots and the Liverpool Centre of Excellence team, as well as Gemma Darlow, for organising the conference. She also thanked all the clinicians in the centres of excellence and elsewhere who care for people with Behçet's, and those involved in developing the new British Association of Dermatologists (BAD)/British Society for Rheumatology (BSR) living guideline for Behçet's, which should increase knowledge among non-specialist clinicians. Finally, Baroness Ritchie highlighted the importance of a patient registry and encouraged everyone to allow their data to be used confidentially for research purposes when requested.

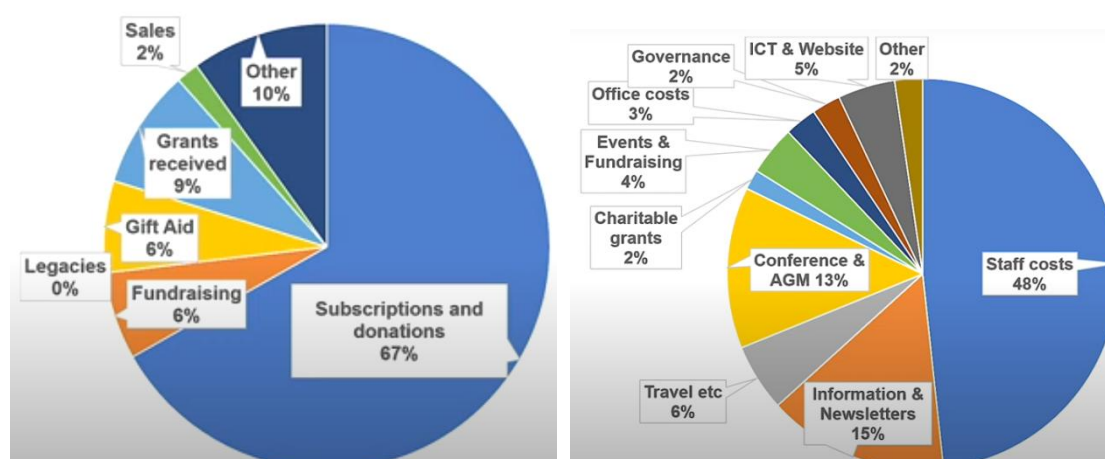
Behçet's UK AGM

Tony Thornburn encouraged members to read the Annual Report, which is available on the Behçet's UK website. He thanked the officers, trustees and administrators for their work. After mentioning the regular newsletters, Behçet's Patients Centres and the Support Coordinators, the amazing fundraising by Behçet's UK members, and the helpline and support groups, Tony highlighted the wider visibility of the charity and the success of the Friends and Family Day. He pointed to the BAD/BSR guideline and the secukinumab trial as notable recent developments. Looking forward, Tony noted the potential for increased efficiency from a constituent relationship management system, the expected impact of the new guideline, the hope for improved outcomes in the devolved nations, and the potential for a patient registry and research focus.

Alan Lane, Honorary Treasurer, reported that Behçet's UK now has 1271 standard members, of whom 1134 are patients. Benefits of standard membership include a welcome pack, quarterly newsletters, access to the helpline, participation in local support groups, invitations to family days, free attendance at the annual conference

and the ability to vote on any resolutions. The number of members continues to rise, which may be due to more people with Behçet's being identified.

The charity's total income for 2023/24 was £46,980, with 67% coming from subscriptions and donations. Unusually, 9% of the income came from a grant from the BAD, but no legacies were received. The expenditure for the year was £69,311, with staff costs accounting for 48%, information and newsletters for 15%, and the Conference and AGM for 13%.



The General Fund and Research Fund stood at £144,413 and £65,228, respectively, at the end of 2022/23; after the transfer of £50,000 from each into a new Projects Fund ('pump-priming' for a patient registry and a specialist nurse in Scotland), the balances at the end of 2023/24 were £74,309 and £17,217, respectively. Projections for the end of 2024/25 are £39,000 for the General Fund and £0 for the other two funds. Alan reported that the annual subscription for members not exempt from paying has been £20 for the past 15 years; the trustees are proposing to increase this to £30 in September 2025, which is slightly below the rate of inflation. This needs to be approved by a vote of the members attending the AGM. Alan added that an increase in fundraising activities will also be needed for the charity to achieve its objectives.

Neil Williams, Behçet's UK Trustee, began by saying that Behçet's UK needs money for the daily running of the charity, the research projects it is involved with and the large projects it is planning. Neil thanked members for their incredible fundraising efforts in the past year. He asked people to get in touch with him if they could consider joining the Fundraising Committee. Other ways to raise money for Behçet's UK include becoming a member; making a donation; running fundraising events;

buying merchandise and Christmas cards; using online shopping partners (easyfundraising and Give as you Live); using payroll giving, match funding and gift aid; donating while selling on eBay; fundraising through Facebook or Instagram; participating in the Weather Lottery; or leaving a legacy.



Rachael Humphreys, Behçet's UK Trustee, announced that the recipient of the Judith Buckle Award 2024 was Kai Li, who became a Trustee of the Behçet's Syndrome Society in 1999 and Honorary Treasurer in 2004, dedicating 10 years to the charity. His expertise as a financial controller and business advisor provided a solid foundation and a safe pair of hands for the charity's accounts. Kai was involved in numerous fundraising efforts over the years,

including giving a presentation at his local school. Originally from Hong Kong, Kai is described as a wonderful family man, fun-loving, jovial and generous with his time.

Jan Mather accepted the award on Kai's behalf as he was unwell and unable to attend in person. In an email, Kai said that he was one of the few members to have met Judith Buckle in person in the late 1980s. He added that it had been a privilege and an honour to witness and be part of the process of the transformation of the humble beginning of the charity to the professional organisation that Behçet's UK is now.



Tony Thornburn then announced the winner of the third annual Behçet's UK Members' Award, which recognises the contribution of individual members to the Behçet's UK community, adding that nominations are now open for the 2025 award. The winner of the 2024 award was John Mather, who recently retired as Operations Manager of the Behçet's Patient

Centres after 10 years in a role that he initially took on as a temporary measure. John

professionally invested much time in building the team of Behçet's Support Coordinators and was part of the interview panel to appoint Bec in Birmingham and Jackie in Liverpool; only Jean in London pre-dates him. John has also spent much time and effort fundraising, including the London to Paris bike ride, climbing Ben Nevis and organising the Yorkshire 3 Peaks Challenge, in addition to providing much support for a partner with Behçet's. Accepting the award, John said that his was always a supporting role. His roles in relation to Behçet's were first supporting someone with Behçet's, then supporting the Chair of Behçet's UK and selling Christmas cards at AGMs, and finally managing the three Behçet's centres and the support coordinators.

Introduction and conference overview

Prof Robert Moots, Clinical Director at the Liverpool Behçet's Centre of Excellence, Consultant Rheumatologist, and Chair of the Behçet's UK Medical Advisory Panel, introduced the programme for the rest of the day, saying that it included excellent speakers on important topics.

Driving forward care through clinical trials

Prof Moots began his presentation by listing the challenges in treating Behçet's, including the rarity of the disease, the many different organs affected, the different clinicians involved in care, the variable (often poor) quality of clinical trials, the lack of publications (e.g., in paediatrics, reproductive health and psychological effects), and the variable availability of treatments. Over the years, the side-effects of therapy have often been worse than the disease. Prof Moots added that the treatment algorithm used at the three centres is now obsolete, but minimal evidence is available to update it. To do this, clinical trials specific to UK patients are needed.

The BioBehçet's trial of Roferon (interferon) versus infliximab was the first (and so far the only) head-to-head trial of two biologics in Behçet's. Both drugs were effective in reducing disease activity and the side-effect profiles were comparable. The results have been published in *Rheumatology* and show that conducting a randomised controlled trial in Behçet's in the UK is possible. However, Roche withdrew Roferon from the market as it was not cost-effective. Now, a trial of secukinumab (an anti-IL17 agent used in arthritis and some skin conditions) has been designed to the highest standards. The cost of £1 million is being funded by Novartis,

and the trial has saved the NHS at least £250,000 in drug costs. Progress in recruiting the 64 patients needed for analysis has been slow, and an extension was requested. With additional financial support from Novartis, PHARMEExcel and Behçet's UK, recruitment is now complete. Most patients taking secukinumab in the trial are keen to remain on it. If the trial is successful, the NHS will fund secukinumab for Behçet's patients in England.

Prof Moots finished by speaking about the BAD/BSR living guideline, which will be updated as and when new evidence becomes available and aims to ensure evidence-based care for all patients. The guideline contains 87 recommendations, including management by a multidisciplinary team, assessing organ involvement and targeting treatment accordingly, some key recommendations on reproductive health and, for the first time, recommendations on psychological management.

Work and Behçet's

Jackie Pooler, Support Coordinator at the Liverpool Behçet's Centre of Excellence, said that patients are increasingly asking for workplace-related support. People with Behçet's work in all sectors and ask for advice about current employment, training, studying, starting work, volunteering and unemployment. Problems at work can be related to the invisible nature of Behçet's, the fluctuating symptoms, trivialising of symptoms such as mouth ulcers, and apparent 'favouritism' leading to isolation from colleagues. Jackie stressed that people do not have to disclose their diagnosis to an employer, but if they want to do so the support coordinators can provide supporting letters and Behçet's UK leaflets. The Equality Act 2010 provides protection against discrimination and requires employers to make reasonable adjustments such as different shift patterns, equipment and flexibility regarding time off for appointments. Patients are encouraged to use resources such as the employer's occupational health or human resources teams. The support coordinators can also signpost patients to sources of specialist advice such as Citizens Advice or ACAS (the Advisory, Conciliation and Arbitration Service), or people can self-refer to a trade union for advice.

For people re-entering the workplace, the support coordinators can give advice on confidence building and on what can and cannot be asked in interviews. Jobcentre Plus offers a Work and Health Programme, as well as disability employment advisors

and work coaches, and help with preparation of CVs, job applications and interview skills. In addition, the government's Access to Work Scheme is a personalised service to help people with a health condition or disability get or stay in work by providing money towards the cost of special equipment at work or travel expenses. Citizens Advice can advise on in-work benefits to help with childcare costs and Working Tax Credits.

Experience of registries helping rare diseases

Martin Twycross, Industry Liaison Manager at Dendrite Clinical Systems Ltd, explained that a registry is simply a database to collect real-world (ideally longitudinal) clinical data on a particular disease, and that Dendrite has been providing registries (>200 worldwide) for over 30 years. Registries increase understanding of the disease and lead to better quality of life and outcomes for patients. They are useful for informing regulatory bodies, especially when clinical trials are impractical. Registries can be used to collect data on demographics, medical history and investigations, comorbidities, treatment and follow-up. Patient-reported outcome measures (PROMs) can be collected automatically via text message or email and can be very simple (e.g., smiley faces). Using PROMs reduces the bias that can occur when data is collected by a health professional.

Martin went on to give examples of different types of registries. A 'classic' registry for Waldenström macroglobulinaemia (WM) was founded by a bequest, has been running since 2016 and includes 1640 patients with this blood cancer. The rate of return of PROMs is 73%. The first report from the registry, produced in 2018, led to an alteration of the disease profile from elderly white men to 35–40% women and starting in the 30s. The registry is supported by Janssen, which has a drug approved for use in WM, with the data showing that 70% of patients respond well.

An innovative 'support network' registry for Castleman disease (a lymphoproliferative disorder) was set up in 2023 with 'seed' funding by a drug company. A doctor making a diagnosis of Castleman disease can send an email to obtain urgent advice if needed and participate in monthly case review calls. This has led to more consistent diagnosis and treatment at a national level. The registry currently contains 45 patients with a confirmed diagnosis. Finally, in April 2024, a novel 'patient-led' registry was launched for alkaptonuria, a very rare genetic

condition (~100 cases in the UK), and currently includes 66 patients in 26 countries. Each patient keeps a journal, providing a first-person perspective of how this multisystem disease develops over time.

Several factors are needed for a registry to be successful, including clear objectives, appropriate data fields to support the objectives, ease of use, complete and accurate data input, high quality outputs, good representation of patients and compliance with General Data Protection Regulation (GDPR) requirements.

Joint pain in Behçet's

Dr Jagdish Nair, Consultant Rheumatologist at the Liverpool Behçet's Centre of Excellence, said that up to 90% of Behçet's patients have joint pain caused by inflammatory arthritis, enthesitis (inflammation at sites where tendons and ligaments attach to bones) and sacroiliitis, mechanical pain (osteoarthritis/degenerative arthritis) or fibromyalgia (chronic widespread pain). Between 20% and 50% of patients have inflammatory arthritis, which includes non-infective arthritis, septic arthritis, reactive arthritis and gout/pseudo-gout. Inflammatory arthritis can be monoarthritis (affecting one joint), oligoarthritis (affecting 2–4 joints) or polyarthritis (affecting many joints). The inflammation is usually non-destructive, causing no damage to the bones.

Enthesitis and sacroiliitis are uncommon, with enthesitis affecting the hips, knees and ankles and sacroiliitis affecting the sacroiliac joint linking the pelvis and lower spine. Mechanical pain caused by osteoarthritis/degenerative arthritis mainly affects the weight-bearing joints, while fibromyalgia, which causes widespread pain and tenderness as well as fatigue and cognitive problems, can occur in people with a genetic predisposition. A study of patients attending the Liverpool Behçet's Centre found that widespread pain was more common in those with high disease activity.

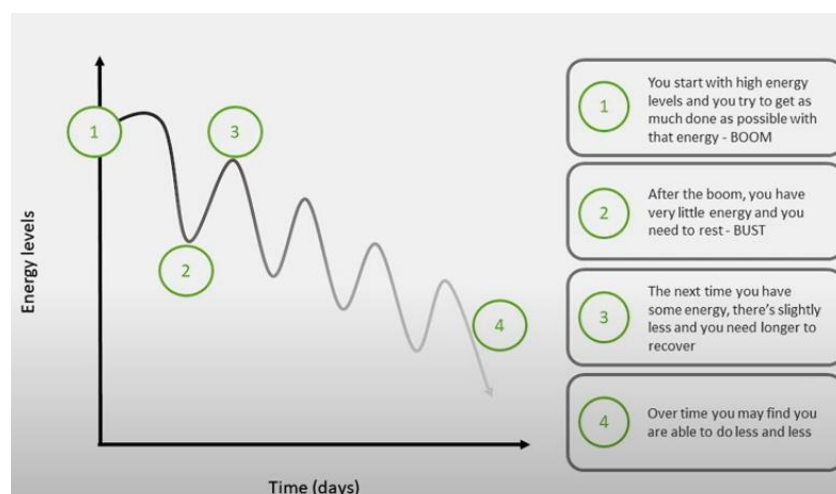
Assessment of joint pain involves history-taking, clinical examination and investigations. The latter include blood tests, X-rays, scans such as ultrasound and magnetic resonance imaging (MRI) to show active inflammation, and joint aspiration where necessary. Dr Nair explained that there is no single approach to management of joint pain. Inflammatory joint pain can be treated with analgesics, non-steroidal anti-inflammatory drugs or immunomodulators (e.g., colchicine), or with a steroid injection, as well as with physiotherapy and occupational therapy (OT). Degenerative

joint pain can be treated with analgesics and physiotherapy/OT. Fibromyalgia can be treated with physiotherapy/OT, graded physical activity and cognitive-behavioural therapy. For all types of joint pain, psychological management may be useful and workplace adjustments may be needed.

Fatigue in Behçet's

Dr Suzanne Crozier, Clinical Psychologist at the Liverpool Behçet's Centre of Excellence, said that Behçet's is associated with higher rates of anxiety and depression and lower quality of life. The disease is often invisible and the route to diagnosis can be long. The unpredictable nature of the symptoms can lead to a sense of uncertainty and have an impact on patients' work and social lives. Most people with Behçet's experience fatigue, and this can have a big effect on quality of life. Fatigue is very different from tiredness, and communicating its impact can be difficult.

The definition of fatigue includes cognitive aspects (so-called brain fog), decreased mood and lack of relief by sleep. The cause of chronic fatigue in Behçet's can be primary, due to the inflammatory disease process, or secondary as a result of emotional distress, pain, disturbed sleep, side effects of medication, inactivity or overactivity, infections, diet or environmental factors. Psychologists use the biopsychosocial model, which gives a holistic view of fatigue including biological, psychological and social aspects. The mind is important in fatigue, with a complex interplay between thoughts, emotions, behaviour and physical sensations. A boom and bust cycle can emerge whereby people try to do too much when they have more energy and then experience a crash, leading to an increase in fatigue and a reduction in energy levels over time.



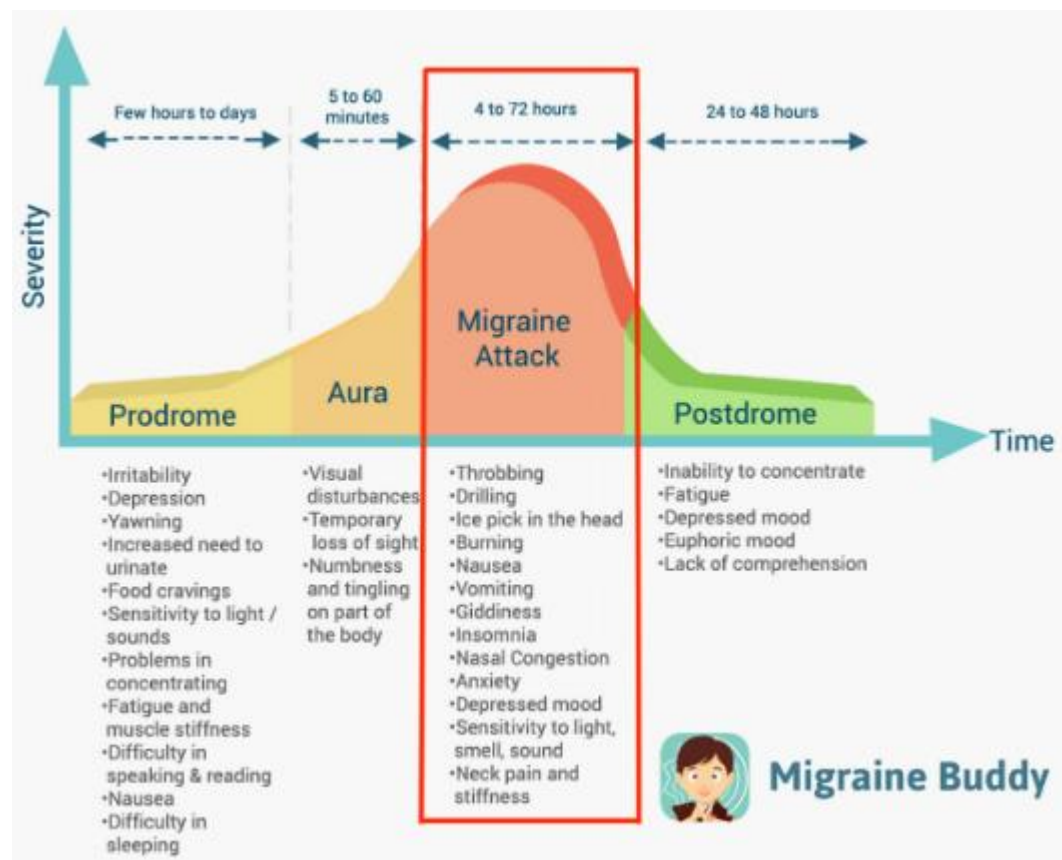
Dr Crozier explained that psychologists use talking therapies to help patients adjust to their diagnosis and problems such as anxiety, depression, chronic pain and fatigue, offering listening and a space to process the impact of living with a chronic condition. Education and sense-making are also used, helping people to understand how their fatigue affects them and what can help them. Pacing (energy conservation) is a useful technique that can help people to learn their own limits and plan accordingly. People can be helped to identify their personal values and live in accordance with them rather than trying to do everything, and increasing self-efficacy can offer a sense of control. Other strategies and skills that people can learn include ways to manage unhelpful thoughts and mindful awareness of emotions. Fatigue can lead to higher levels of self-criticism and low self-esteem; compassion-focused therapy can help people to increase self-compassion and manage difficult emotions.

Headaches in Behçet's

Dr Mona Ghadiri-Sani, Consultant Neurologist at the Liverpool Behçet's Centre of Excellence, explained that neurology is the study of the brain, spinal cord and nerves, and Behçet's can affect all of these. Around 10% of people with Behçet's have neuroBehçet's, and inflammation of the brain, brainstem or spine is seen in about 2% of these. Neuro-Behçet's is investigated with scans, blood tests, lumbar puncture for spinal fluid analysis, and nerve tests. However, headache (usually migraine) is the most common symptom seen by neurologists. Migraine is common in the general population (the third most common health condition in the world), and is much more common than neuro-Behçet's in people with Behçet's. It is more than just a headache, with symptoms such as light, noise or smell sensitivity and dizziness being common, as well as cognitive effects.

Between December 2021 and June 2022, of 24 patients seen for a neurology consultation at the Liverpool centre, 20 reported headaches, most commonly chronic migraine and medication overuse headache. Of 18 patients with chronic migraine, only 13 had received preventive drugs such as propranolol or pregabalin. To avoid medication overuse headaches, other causes of pain (e.g., joint pain or mouth ulcers) should preferably be managed without using painkillers. Migraine has a high socioeconomic impact and is a major global cause of disability and loss of

productivity. A migraine attack consists of several phases, comprising a prodrome, aura, the attack itself and a postdrome phase.



CGRP (calcitonin gene-related peptide), a pain-signalling neuropeptide and potent vasodilator, is increased during an attack and is a mediator of migraine. Caffeine and painkillers increase CGRP and so should be avoided by people prone to migraines. Management of chronic migraine consists of lifestyle approaches (sleep hygiene, regular meals, good hydration, avoidance of caffeine and analgesics), preventive drugs, acute therapy and management of comorbidities; advanced care may include cannabinoids, Botox and anti-CGRP monoclonal antibodies. A migraine probiotic can also be useful. Episodic migraine can progress to chronic migraine, and early intervention to prevent this is needed.

Oral health and Behçet's

Dr Emma Morgan, Academic Clinical Fellow and Speciality Trainee in Paediatric Dentistry in Liverpool, began by saying that oral ulcers are very painful and have a big impact on speaking, eating, tooth brushing, etc. They are common in the population and can be minor, major or herpetiform. Minor ulcers are small and last 1–

2 weeks, while major ulcers are >10 mm, last for several weeks and cause scarring. Herpetiform ulcers are the least common type, consist of multiple small ulcers in one area of the mouth (e.g., the tongue) and last 1–2 weeks. Ulcers can be caused by trauma, infection, stress, hormonal disturbances, or immunological, gastrointestinal or haematological conditions. Features that cause concern and should prompt attendance at the dentist or GP include persistence, lack of healing, firmness, bleeding and new symptoms such as numbness. Home management of mouth ulcers involves diet (e.g., avoiding acidic or spicy foods), toothpaste (SLS-free), pain relief (e.g., Difflam spray), oral hygiene (e.g., salty mouthwash). Treatments that can be prescribed include steroid or antibiotic mouthwash, numbing spray, systemic steroids, immunosuppressive agents and anti-inflammatory drugs.

Dr Morgan mentioned that joint pain can also affect the mouth and jaw, with pain in the temporomandibular joint or surrounding muscles. This can be managed with a softer diet, pain relief with anti-inflammatory agents, jaw exercises or a bite guard. Specialist management might involve further assessment with an MRI scan and treatment with steroid injections, Botox (for muscle pain) and wider systemic therapy.

Discussing prevention of oral health problems in children, Dr Morgan said that bottles should be avoided after a child reaches 1 year of age, and children should start seeing a dentist around this time. Tooth brushing with at least 1000 ppm fluoride toothpaste should start when the first tooth appears, and sugar-containing foods and drinks should be avoided at bedtime. Children aged 3–6 years should be supported to brush their teeth, especially at night, and should have 6-monthly dental check-ups with fluoride varnish application. After the age of 7, children should be brushing twice daily with adult toothpaste and can use interdental brushes, as well as continuing to have 6-monthly dental check-ups.

Behçet's and the eye

Dr Nima Ghadiri, Consultant Medical Ophthalmologist at the Liverpool Behçet's Centre of Excellence, said that the eyes can be the window into diseases of the whole body. He explained that eye disease affects around 75% of people with Behçet's, two-thirds of whom have uveitis, which is vision-threatening in up to 25%; uveitis is more common and severe in males. Uveitis is inflammation of the uvea (the middle layer of the wall of the eye) and can occur in the front (anterior uveitis), middle (intermediate

uveitis) or back of the eye (posterior uveitis) or affect the whole eye (panuveitis). Behçet's uveitis is usually of sudden onset and limited duration, is recurrent and affects both eyes (one at a time).

Symptoms of anterior uveitis include light sensitivity, red eye and headache. Hypopyon (an accumulation of pus in the lower part of the eye) is uncommon but requires prompt treatment; it resolves quickly. Posterior uveitis (retinitis) is more dangerous and often has fewer symptoms. It is painless but there may be reduced vision, haze and floaters. Other eye problems in people with Behçet's include retinal vasculitis (caused by leaking blood vessels) and optic neuropathy (damage to the optic nerve), which occurs in around 25% of those with ocular Behçet's and leads to reduced colour vision and visual field. Dry eye is common and may be caused by blepharitis, inflammation of the eyelid caused by a bacterial infection.

Diagnosis of eye problems involves testing of visual acuity, colour vision and visual field and a slit lamp examination of the back of the eye. Multimodal imaging includes fluorescein angiography to show retinal vasculitis and optical coherence tomography to visualise retinal oedema. Uveitis is reversible, so early diagnosis and treatment is important to prevent permanent damage. Problems that can occur include synechiae (abnormal adhesions following inflammation), glaucoma (raised intraocular pressure that can be caused by steroid eye drops) and development of cataracts. Treatment of ocular Behçet's ranges from eye drops through immunosuppressants to targeted biological therapies, with surgical interventions such as vitrectomy also available. Dr Ghadiri concluded by saying that new imaging methods will improve diagnosis and new treatment pathways are being investigated.

Medical Panel Q&A

The day finished with a question and answer session with a Medical Panel comprising the day's speakers and Dr Graham Wallace, Senior Lecturer at the University of Birmingham (online). In reply to a question about dry mouth, Dr Morgan said that it is a complex condition that is common in people with autoimmune diseases; the available saliva substitutes are unsatisfactory and people often eat a lot of sweets to relieve the problem. Dr Nair replied to a question about the difference between fibromyalgia and widespread pain due to Behçet's by saying that a thorough medical history and examination should be able to distinguish widespread pain caused by

inflammation and that resulting from an abnormal response to nerve stimulation. Asked whether people susceptible to migraines can drink decaffeinated coffee, Dr Ghadiri-Sani confirmed that a few cups a day should be fine. Regarding plasma exchange for neuro-Behçet's, she said that it might have a place in some cases but most of the studies are very old. In response to a question about the potential for anti-IL13 agents, Dr Wallace said that IL13 does not seem to have much of a role in Behçet's and is more involved in antibody production. In response to a question about why work is important, Jackie Pooler mentioned the economic benefits as well as the therapeutic benefits of mixing with other people. Asked what the main benefit of a registry is, Martin Twycross said that it could increase understanding of the disease and of the efficacy of different treatments. Progress has been slow for Behçet's, but the next steps are to agree the main objectives of the registry and what data needs to be collected to achieve these.

Regarding the likely upcoming advances in eye disease, Dr Ghadiri said that diagnostics and prognostics using artificial intelligence, imaging and 'big data' have enormous potential. For oral medicine, Dr Morgan hoped that the role of the mouth in overall disease will be increasingly recognised, with dentists becoming part of the multidisciplinary team. Dr Nair said that greater understanding of the pathogenesis of Behçet's should lead to more personalised therapy for joint and other problems, while Dr Ghadiri-Sani said that the new migraine treatments such as anti-CGRP agents can be revolutionary. Jackie Pooler would like to see greater recognition of the cost of attending a Centre of Excellence for some patients and the need for satellite centres in the devolved nations. Martin Twycross hoped to see progress on the establishment of a Behçet's patient registry. Finally, Dr Wallace predicted that increased knowledge of the phenotype clusters in Behçet's would result in more targeted treatment, and the role of the microbiome and specifically butyrate offers a potential new therapeutic approach.

Clare Griffith, Editor