Behçet's UK 2023 Conference and AGM

The 2023 edition of the Behçet's UK Conference and Annual General Meeting took place on Saturday 21 October 2023 at the Mercure Telford Central, Telford, Stevenage, and online. A total of 171 people registered to attend, of whom 71 registered to attend in person and 100 to attend online.

Welcome

Tony Thornburn, Chair of Behçet's UK, welcomed everyone to the 40th anniversary Conference and AGM. Baroness Ritchie of Downpatrick, Patron of Behçet's UK, welcomed everybody to the in-person and online event. She thanked the Chair and Trustees of Behçet's UK, as well as Dr Deva Situnayake and the staff of the Birmingham Centre of Excellence for organising the conference programme. Baroness Ritchie went on to thank all the clinicians who look after people with Behçet's at the three centres as well as elsewhere in England and the devolved nations, noting that it is just one of more than 7000 rare diseases. She emphasised the importance of capturing patients' data and encouraged patients to participate in clinical trials and registries whenever possible.

Behçet's UK AGM

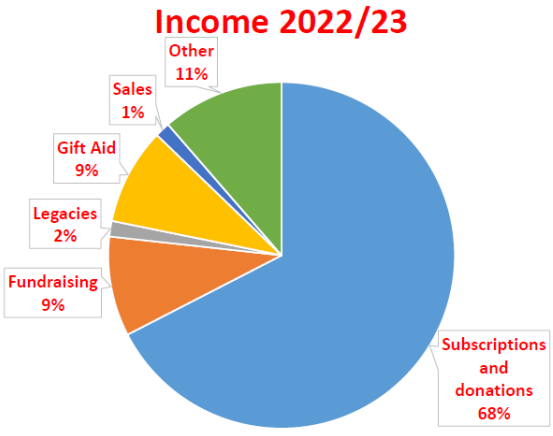
Tony Thornburn encouraged members to read the Annual Report, which is available on the Behçet's UK website. He thanked the administrative staff and all the Trustees for their work, noting Alan Lane's meticulous oversight of the accounts. He also thanked Behçet's Patient Centres, who were not represented at this year's conference, as well as all the members who had raised funds for Behçet's UK and the volunteers working on the Helpline and in support groups. Finally, Tony thanked Gemma Darlow for organising another highly successful Friends and Family Day in 2023, offering an important opportunity for members and their families to get together for a day of fun activities.

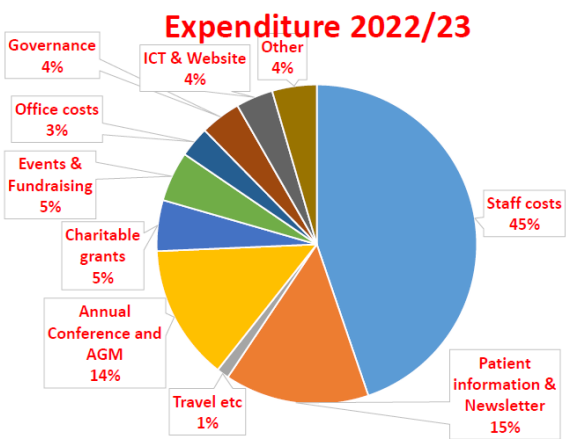
Looking forward, Tony said that the plan to establish a constituent management system to streamline interactions with members is ongoing. The opportunity to establish a specialist nurse post in Scotland is being actively pursued, and the hope is that this will be followed by posts in Wales and Northern Ireland. The British Society for Rheumatology/British Association of Dermatologists guideline for the

management of Behçet's is nearing completion and will lead to quality audits to drive consistency of care. Regarding research, Tony encouraged patients to consider participating in the clinical trial of secukinumab, which has now started. He summarised the history of the effort to establish a UK Behçet's patient registry, which began in 2017 and has been progressed by the data gathering exercise conducted in Birmingham. Tony concluded that a registry needs to be established, facilitated and funded (mainly) by Behçet's UK within a Limited Company by Guarantee, for the benefit of all and with an oversight committee comprising clinicians.

Alan Lane, Honorary Treasurer, reported that the benefits of standard membership include a welcome pack, quarterly newsletters, access to the Helpline, participation in local support groups, invitations to family days and free attendance at annual conferences. For the 13th year in a row, the membership subscription remains at £20 per year in 2023/24. In addition to 1254 standard members, there are now 32 junior members, 73 associates, 16 donors and 61 lapsed members. Almost all (1184, plus 17 awaiting diagnosis) members are patients, with 115 carers, 49 supporters, 48 medical staff and 20 overseas members.

The charity's total income for 2022/23 was £55,400. Two-thirds came from subscriptions and donations, with 9% from fundraising activities and another 9% from Gift Aid. Total expenditure was £66,237, 45% of which was accounted for by staff costs and 30% by patient information, newsletters and the annual conference. This represents a net deficit of £10,837, but the balance remains high at £144,413 as a result of two generous bequests. However, the patient registry and the specialist nurse for Scotland represent around £100,000 in planned spending, so continued fundraising efforts are essential.





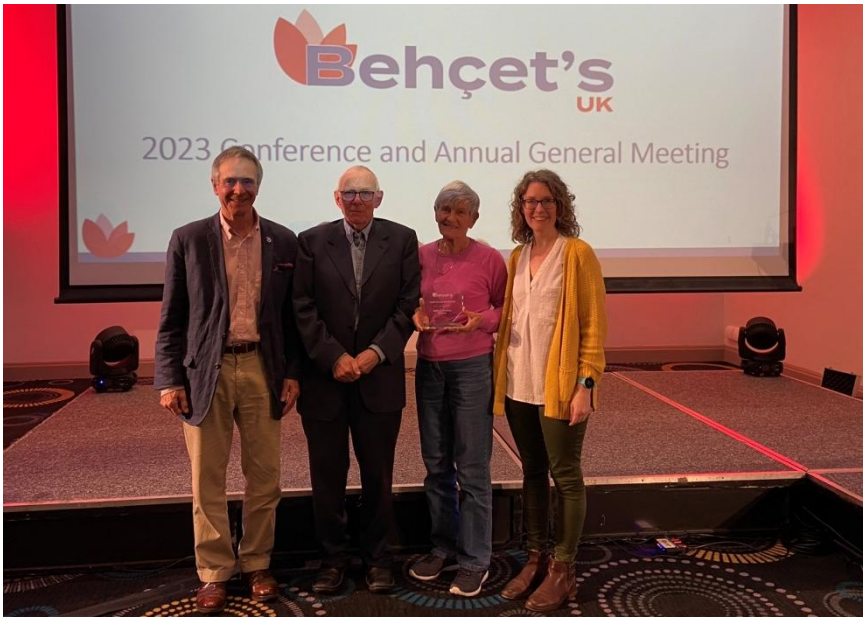
The research fund currently has a balance of £57,374 thanks largely to a generous donation. No money was spent in the past year, so this balance is available for projects planned in the next year.

Neil Williams, Behçet's UK Trustee, reiterated the importance of continued fundraising efforts to support the work of the charity. He mentioned the new merchandise to celebrate the 40th anniversary of Behçet's UK and the '40 in 40', 'Bake for Behçet's' and 'Life begins at 40' fundraising initiatives that have been running throughout the year. Neil thanked everybody who had taken part in fundraising events this year and asked people with ideas for fundraising to reach out to him and join the Fundraising Sub-committee. Other ways to help the charity's finances include buying Christmas cards or sending e-cards, shopping through online shopping partners, payroll giving and fund matching, donating through sales on eBay, playing the Weather Lottery, fundraising on Facebook and Instagram, and considering leaving Behçet's UK a legacy.

Rachael Humphreys, Behçet's UK Trustee, announced that the recipients of the Judith Buckle Award 2023 were Barrie and Georgina Seaman. They joined the Behçet's Syndrome Society (BSS) in 1985 after Barrie was diagnosed with the condition; the Society had around 60 members at the time. Georgina soon became a Trustee and the Information Officer, while Barrie was the Honorary Secretary from 1986 to 1999, Honorary Treasurer from 1999 to 2004 and a Trustee from 2004 to 2007. Together, they drove the Society forward during those years, with Barrie establishing and editing a twice-yearly newsletter and Georgina creating a range of information leaflets and single-handedly manning a telephone helpline for many years. Georgina's help, advice and knowledge changed lives, and she made sure that people diagnosed with Behçet's got the support and expert care they needed. In addition, Georgina set up the first BSS website and Barrie helped to organise several international patient conferences. Barrie's vision was to have specialist clinics with multidisciplinary care, an approach taken forward and seen to fruition by Jan Mather after she became Chair in 2006.

After accepting the award with Georgina, Barrie Seaman said that he and Georgina could remember Judith Buckle very well and worked with her in the early years of the

Society to increase its capacity to help people. He added that it was very heartening to see how well established and professional the charity now is.



Barry and Georgina Seaman with Tony Thornburn and Rachael Humphries

Tony Thornburn then announced the winner of the second 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 2024 award. The winner of the 2023 award was Clare Gibson, who has been a member since 2014 and has made several contributions to the newsletter. Tony said that Clare has been described as an inspiration of positivity who is characterised not by the loss of what she used to do but by embracing what she can do now. In the 2020 Bike for Behçet's/Run for Rare challenge, she set a target of 452 km and actually completed 904 km. She set up a support group and has great listening skills and empathy in her interactions with members. Most recently, she successfully represented Team GB in the Invictus Games.

In a short video, Clare said that it was a real honour to be nominated for the award and to know that her contributions were making a difference. A colleague accepted the award on her behalf.

Introduction and conference overview

Dr Deva Situnayake, Clinical Director at the Birmingham Behçet's Centre of Excellence and Consultant Rheumatologist, introduced the programme for the rest of the day. The theme of the conference was 'our data', resulting from conversations between Dr Situnayake and his son Daniel. For clinicians, understanding all the data on Behçet's and relating it to individual patients in the clinic can be challenging. The rate of growth in the field of machine learning and artificial intelligence is incredible, but these systems are only as good as the data that goes into them, and ensuring adequate safeguarding of patients' data is paramount.

Changing international landscape and challenges of Behçet's

Dr Situnayake began his presentation by asking whether Behçet's is a disease or a syndrome, concluding that in his opinion it is a syndrome. A syndrome is a recognisable complex of symptoms and physical findings that indicate a specific condition for which a direct cause is not necessarily understood. Only when a causative agent or process has been defined with a fairly high degree of certainty can a condition accurately be called a disease.

The main questions that people ask after being diagnosed with Behçet's are "Why did I get it?", "Could I pass it on?" and "How will it affect me?". A study among first-degree relatives of Behçet's patients in South Korea found that familial risks were greater within a generation (sibling–sibling) than between generations (parent–offspring), suggesting a prominent role of gene–environment interactions. Another Korean study found a more than 80% decline in the incidence of Behçet's between 2004 and 2017, along with changes in environmental exposures such as reduced infection rates and improved hygiene and socioeconomic conditions. Data collection is mandatory in South Korea, so medically certified and confirmed diagnoses are available for a population of 50 million people, along with kinship and family history information. The mean prevalence of Behçet's in the population was 26 per 100,000 with an annual incidence of 1.5 per 100,000. Almost 22,000 people were followed from 2002 to 2017, and 31,119 people developed Behçet's during this time. The male:female ratio was 0.53:1. Overall, familial cases accounted for 4% of all cases of Behçet's. Compared with a risk of 1 among people with no family history of Behçet's, the overall risk was 15 in those with a family history. This ranged from 13–15 in

people with an affected first-degree relative to 163 among those with more than one affected first-degree relative and 166 in people with an affected twin.

Dr Situnayake said that the answer to the second question, "Could I pass it on?", is yes, but the absolute risk is very low at 2.86 per 100,000 or 1 in 35,000. The Korean data raises other questions such as why women seem to be more susceptible and what environmental factors are interacting with the genome to trigger Behçet's. Certain features of Behçet's tend to group together into clusters, including an ocular/neurological cluster, a vascular/gastrointestinal cluster and mucocutaneous/arthritis cluster, with the latter being more common in women. Research in Japan has looked at changes in the proportion of these clusters over time, finding an increase in the gastrointestinal cluster and a decrease in the severity of ocular symptoms, which they suggested might be due to change towards a more Western lifestyle and diet. Gastrointestinal involvement in Behçet's is common in the UK (38–53%) and Japan (50–60%), moderately common in China (10%) and Taiwan (32%), but uncommon in Turkey, India and Saudi Arabia (3–4%).

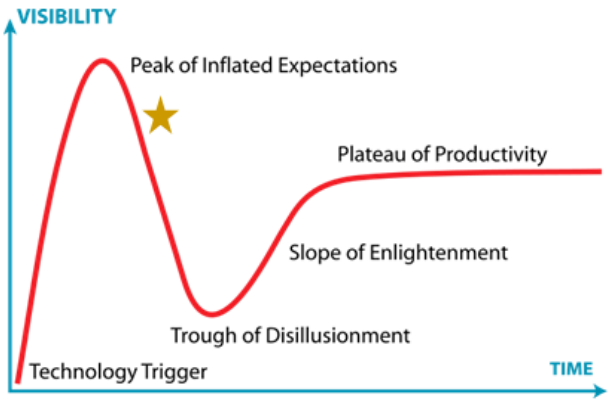
Genome–environmental interactions, which operate through epigenetic (gene regulatory) mechanisms that are heritable, might explain these regional differences as well as why one child in a family may get Behçet's when others do not. Registry data are not available for Behçet's patients in the UK, but some insight can be gained from data on inflammatory bowel disease (IBD) in Denmark. This data showed that a family history of ankylosing spondylitis and a personal history of antibiotic treatment during childhood increased the risk of IBD. This suggests a role for 'MHC-1-opathy' and the microbiome that may also be the case for Behçet's.

Getting to know the basics of machine learning and artificial intelligence (AI)

Daniel Situnayake, Head of Machine Learning at Edge Impulse, began his presentation on artificial intelligence (AI) by defining intelligence as understanding what is going on, predicting what might happen next and knowing the right thing to do, along with many other things. AI is exactly the same, but done using a computer. Examples include robot vacuum cleaners that can navigate around a house, chatbots and AI assistants that can have a conversation and reply to questions, web search engines and business software that can analyse complex statistics.

AI is made up of computer programs called algorithms. These can be very simple, such as a thermostat that turns the central heating on or off when a preset temperature is reached. Others, such as the robot cleaner, are much more complex. Algorithms are created by computer programmers or mathematicians, but some are so complex that they would be impossible to create by hand, even by an expert. For example, getting a thermostat to turn on the heating if a camera shows that the weather outside is cold sounds simple from a human perspective, but training an AI to do this is very difficult. This type of task requires machine learning (ML), which is way to create algorithms automatically. The first step is to collect data about the situation, and the ML model is then trained to make sense of it. In the example, this would involve feeding images of warm and cold weather into the ML model and adjusting it each time it gets the answer ('cold' or 'warm') wrong. This is done millions of times with lots of data until the model almost always gives the right result. An AI system can be built from many different algorithms (ML models and other types) connected together.

Benefits of AI include the speed and efficiency of computers at repetitive tasks, while ML can find hidden patterns within highly complex data and use them to make predictions. Drawbacks include that a lot of high quality data is needed to train the model, understanding how a trained algorithm works can be difficult, and AI systems lack common sense. Overall, AI is a helpful tool for understanding data and automating repetitive tasks, and it has already led to many real benefits, but it is not a magic wand that can be applied anywhere without sufficient thought and planning. A useful concept is the Gartner hype cycle, in which initial hype leads to a peak of expectations followed by a trough of disappointment such as we are starting to see with the new chatbots. Eventually, a plateau of productivity between the two extremes will be reached whereby AI is seen as a useful tool but its limitations are recognised.



AI – challenges, threats and opportunities in the NHS

Dr Anmol Arora, Academic Foundation Doctor at Cambridge University, said that AI is the ability of a computer to perform tasks usually associated with human intelligence, such as learning, reasoning and self-correction (that is, learning from its own mistakes). It has a wide range of uses in healthcare, including 'behind the scenes' roles such as bed allocation, as well as in clinical areas such as ophthalmology.

Human interpretation of a retinal fundus photo by a trained ophthalmologist can obtain quite a lot of information such as the approximate age of the person. However, in future, oculomics (the study of the association between changes or abnormalities in the eye and systemic health or disease states) will be able to provide information on age, sex, blood pressure, smoking status, diabetes status and cardiovascular risk, as well as the presence of ophthalmic disease. In this way, eyes can be seen as a window into wider health, providing a model for generalised disease detection. A huge AI model has been trained on publicly available datasets and linked to medical records, with the aim of diagnosing eye disease, predicting progression of eye disease and trying to predict systemic disease (e.g., stoke, heart attack or Parkinson's disease) using simple eye photos.

However, Dr Arora pointed out that there are many barriers to the use of AI in healthcare, including infrastructure and cost, availability of data, regulatory approval, accountability and stakeholder involvement. Any use of AI in healthcare needs to be acceptable to all the stakeholders involved, especially the public. Novel regulation is needed to ensure patient safety, as AI is different from drugs in that it is not static and evolves over time. Dr Arora concluded by stressing the importance of improving the quality of data through initiatives to increase the diversity and representation of datasets and build research data infrastructure so that data is kept safe but can be shared.

Ocular challenges in Behçet's: Birmingham Centre of Excellence experience

Mr Sreekanth Sreekantam, Consultant Ophthalmologist at Birmingham and Midland Eye Centre, described challenges in the diagnosis, treatment and monitoring of eye disease in Behçet's. The diagnosis is made clinically on the basis of pattern recognition and correlation with extra-ocular features, with the most common features

being anterior/posterior/pan-uveitis and retinal vasculitis. However, non-uveitic inflammatory features such as inflammation of the optic nerve or sclera have been reported in a minority (<5%) of patients.

Mr Sreekantam illustrated the challenges with the case of a 27-year-old man with irreversible loss of vision in his right eye, recurrent oral and genital ulceration, erythema nodosum and signs of possible hearing loss. An MRI scan of the brain showed inflammation with possible demyelination that was not typical of multiple sclerosis. Retinal images showed vessel thinning in the right eye consistent with a previous vascular event but no inflammatory features. Possible diagnoses include Behçet's, demyelinating disease and Susac syndrome. As these are all inflammatory conditions, systemic immunosuppression was started to protect his eye. He is being closely monitored for any new symptoms.

The EULAR recommendation for inflammatory eye disease is systemic immunosuppression along with corticosteroids. Immunosuppressive agents include conventional ones such as azathioprine or mycophenolate and biological agents such as interferon (no longer available in the UK) and anti-TNF agents, which are the drugs of first choice for ocular Behçet's. Local therapies such as steroid implants are useful to treat flares and prevent recurrences in patients on maximum immunosuppression.

To illustrate treatment challenges, Dr Sreekantam presented the case of a 21-year-old man with bilateral blurred vision, recurrent oral ulcers and an acneiform rash. Retinal imaging showed non-specific inflammatory deposits in the vitreous, but a fluorescein angiogram showed leakage from vessels in a pattern typical of Behçet's. The patient was started on oral corticosteroids and sent for screening tests for anti-TNF treatment. His quantiferon test was positive, which suggests contact with tuberculosis and is a contraindication to anti-TNF therapy, so he was referred urgently to a chest physician and treated for latent tuberculosis. His non-ocular symptoms were under control but he was experiencing side effects from the oral steroid, so he was given intravitreal steroid injections. However, this led to increased intraocular pressure which required additional topical and oral treatment. Once his tuberculosis had been treated, the patient was able to start anti-TNF treatment with infliximab. Interferon would have been the ideal treatment for this patient and is the treatment of choice in countries

where tuberculosis is endemic; it offers rapid control but its side effects can be quite severe.

Patients' response to treatment can be monitored using the SUN (Standardization of Uveitis Nomenclature) grading scale, but this is subjective. Retinal imaging is used for objective assessment, and regular fluorescein angiography is recommended to assess disease activity. In the patient above, this showed remaining inflammation after 6 months and he was switched to adalimumab. There was some improvement at 1 year, and azathioprine was added, leading to further improvement at 2 years, although some peripheral inflammation remained. Fluorescein angiography is very useful for monitoring treatment, but it is invasive, requiring injection of dye which carries a risk of anaphylaxis, and it requires multiple visits. Anterior chamber laser flare photometry has shown promising results in Turkey and may represent a non-invasive alternative.

Paediatric Behçet's and monogenic mimics

Dr Eslam Al-Abadi, Consultant Paediatric Rheumatologist at Birmingham Women's and Children's Hospital, began his presentation by considering the causes of mouth ulcers in children. Oral ulcers are seen in approximately 10% of children and 20% of adolescents, are usually painful, may be recurrent or persistent, and have many possible causes. They may be associated with other symptoms such as pallor, bruising, bone pain, weight loss, gastrointestinal features, infections, recurrent fever or sore throat, and multisystem inflammation (such as systemic lupus erythematosus or Behçet's). Dr El-Abadi said that his approach to investigation of recurrent aphthous ulceration included a full blood count, assessment for dietary deficiencies and infections, measurements of inflammatory markers, and specific genetic or other tests in selected cases.

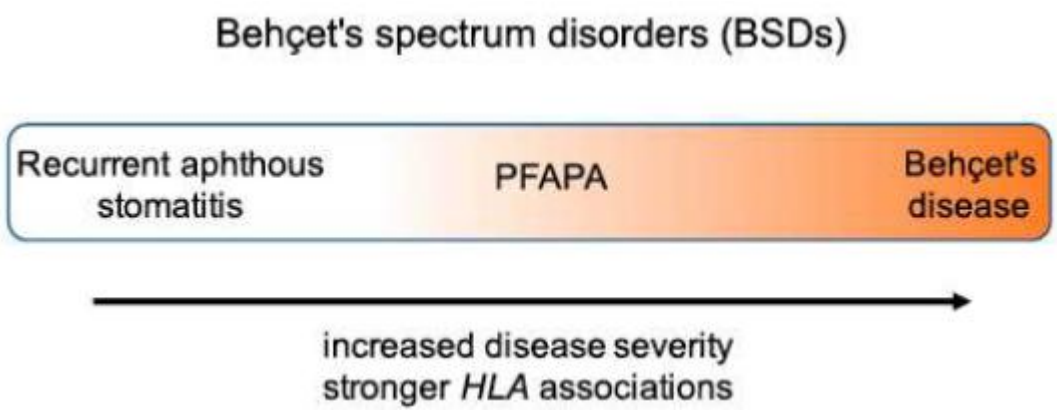
Dr El-Abadi described the case of a 15-year-old boy who had had recurrent mouth ulcers for 17 months, which had been treated with a variety of mouth washes and sprays. He then presented with a 2-week episode of multiple painful and distressing mouth and genital ulcers, as well as joint pain with no swelling. The patient was diagnosed with Behçet's after other possible diagnoses had been excluded, and he was started on topical and oral steroids plus colchicine. Four months later, he had a

recurrence of the ulceration and also blurred vision due to pan-uveitis, and he was started on an anti-TNF agent.

At the Birmingham Centre of Excellence, of 61 patients who reported having their first symptom of Behçet's before the age of 16, only 26 fulfilled the International Study Group (ISG) criteria before that age. Among these patients, the average age of onset of the first symptom was 9.5 years, with the second symptom appearing at an average age of 20. The average length of time between the first symptom and diagnosis of Behçet's was 19.6 years.

The epidemiology of Behçet's in children has been studied by Clare Pain and colleagues in Liverpool and London. The study found 56 confirmed cases of paediatric Behçet's according the International Criteria for Behçet's disease (ICBD), giving a prevalence of 4.2 per million children. The prevalence was 2.3 per million for children fulfilling the ISG criteria and 2.2 per million for the paediatric classification criteria (PEDBD). The median age at onset of symptoms, first presentation and diagnosis was 6.3, 10.6 and 11.7 years, respectively. Just over half the children (57%) were female, all had oral ulceration, 91% had genital ulceration and 54% had skin involvement. Ocular (18%), vascular (5%) and neurological involvement (4%) were less common. Three-quarters of the children required immunosuppressive drugs, and a quarter continued to have active disease despite medication.

Finally, common genetic susceptibility loci link PFAPA (periodic fever, aphthous stomatitis, pharyngitis, adenitis) syndrome, Behçet's and recurrent aphthous stomatitis (RAS). This has led to the concept of Behçet's spectrum of disorders, with RAS at one end, PFAPA in the middle and Behçet's at the other end.



Dr El-Abadi described a family in which the mother, two daughters and maternal grandfather all had some features of Behçet's while the father and two other siblings were healthy. Genetic investigations revealed A20 haploinsufficiency, which has some features in common with Behçet's but is caused by a mutation in a single gene. A study conducted in London found that of 11 cases of atypical Behçet's phenotypes in children aged under 5, nine had known disease-causing genetic mutations. These 'monogenic mimics' should be considered in young children with incomplete or atypical Behçet's features and a relevant family history.

International scientific update

Dr Graham Wallace, Senior Lecturer at the University of Birmingham, said that several studies of the microbiome in Behçet's suggested an association with a deficit of butyrate, a short-chain fatty acid (SCFA) released by certain bacteria into the gut that promotes a healthy gut lining and influences inflammation elsewhere in the body. Dietary sources of SCFAs include whole grains, fruits, nuts and vegetables, and bacteria in the gut help to breakdown dietary fibre to release SCFAs into the blood. The production of SCFAs such as butyrate by the gut microbiome can be modulated by probiotics, prebiotics, antibiotics, traditional immunosuppressive drugs, biologic drugs and faecal transplantation.

The MAMBA trial in Italy investigated whether a lacto-ovo-vegetarian diet enriched in substrates with potential for butyrate production or a Mediterranean diet supplemented with butyrate could be beneficial for the gut microbiota in Behçet's. After 3 months, the antioxidant capacity of patients' blood had improved; however, no effect was seen on the microbiota composition, suggesting that a longer intervention may be needed. Preliminary results from a larger follow-up study show a non-significant reduction in clinical disease manifestations as well as significant reductions in steroid use and global assessment improvement score for gastrointestinal symptoms. Dr Wallace described a case study of a 49-year-old female Behçet's patient with anterior uveitis who was given symbiotic (probiotic plus prebiotic) treatment with capsules containing seven bacterial species and fructo-oligosaccharide. After 9 months, her uveitis had improved (no attacks after the first 2 months), her inflammatory markers were suppressed and her immunosuppressive drugs had been

tapered off. Dr Wallace concluded that there is potential for dietary interventions in Behçet's.

Dr Wallace went on to summarise some other recent findings in Behçet's. A study in Turkey showed that measurement of femoral vein thickness can be helpful in the differential diagnosis of Behçet's in patients with limited disease manifestations, which may be particularly useful in patients presenting with venous thrombosis. Dr Wallace's group have had a paper accepted for publication which reports on a study among an international panel to identify defining features of ulcers in Behçet's in order to formulate guidelines for non-specialists. It is hoped that this will enable earlier diagnosis and quicker access to treatment. Turning finally to new treatments for Behçet's, Dr Wallace explained that secukinumab appears to be effective in dealing with mucocutaneous disease, while tocilizumab is better in patients with ocular and neuro-Behçet's refractory to anti-TNF agents.

Navigating chronic illness using metaphors as a compass

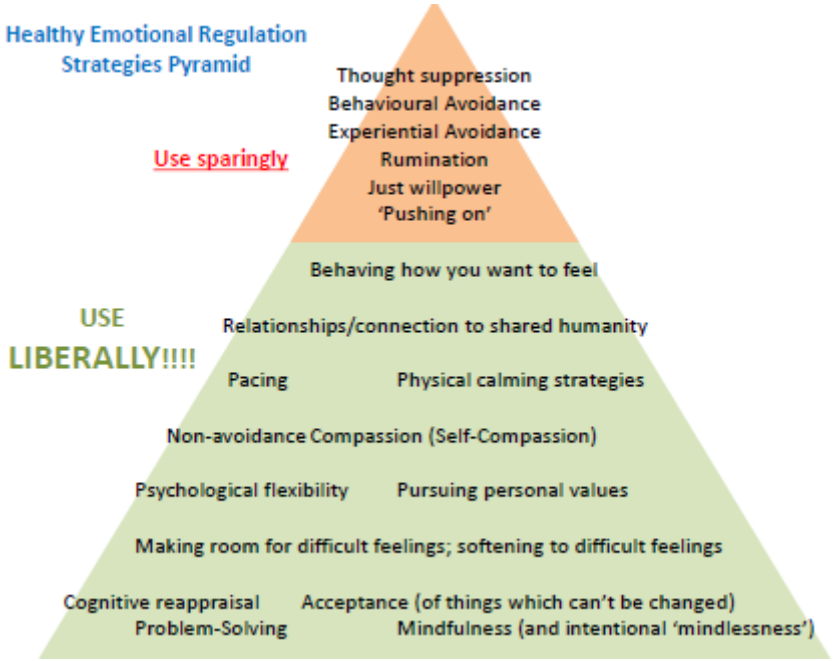
Dr Jonathan Harrold, Clinical Psychologist at the Birmingham Behçet's Centre of Excellence, said that people with long-term physical health conditions have an increased risk of mental health problems and emotional distress, including heightened stress, anxiety, trauma responses and grief reactions. This affects quality of life and the reasons for it are complicated and involve many factors, whether biological, emotional, psychological, environmental, behavioural or financial. He explained that avoidance or suppression of unwanted thoughts, emotions or experiences is not the answer and used three metaphors that might be helpful in dealing with such thoughts.

The first metaphor was a beach ball floating in the water near someone trying to enjoy a swim in the sea, which can be used to represent an unwanted emotion or experience. Trying to push the ball away or hold it under the water will not work in the long run, and the effort will spoil the swim. The solution is to be willing to allow the ball to exist and get on with enjoying the swim despite its presence. Acceptance of a problem can lead to the problem decreasing in importance.

For the next metaphor, Dr Harrold imagined being a bus driver who wants to go in a particular direction but cannot control the condition of the road or the traffic. Some of the passengers on the bus might be quiet while others are disruptive. The passengers

have names such as doubt, fear, resentment, criticism, encouragement, kindness or joy, and the ones nearest the driver or with the loudest voices try to influence the direction the bus goes in. Listening to them might make them quieter but can result in always going in the same direction. In this case, the solution is to make an effort to listen to some of the other passengers who find it more difficult to be heard. This will result in decisions made through positive choices rather than staying on autopilot.

Dr Harrold's final metaphor was the Japanese art of Kintsugi or golden joinery, whereby pieces of a broken ceramic item are joined back together with glue containing gold rather than being discarded or put back together invisibly. This way, the item becomes more robust and more precious than before it was broken. This can be seen as a metaphor for acceptance and personal development and healing, drawing attention to the damage and to the care that has gone into repairing it, and recognising the beauty of imperfections. In the same way, it is possible for people to repair themselves after adversity by using self-care and nurturing, with the gold represented by the strategies shown on the pyramid below.



Mining the England Hospital Episode Statistics (HES) and Clinical Practice Research Datalink (CPRD) – what have we found?

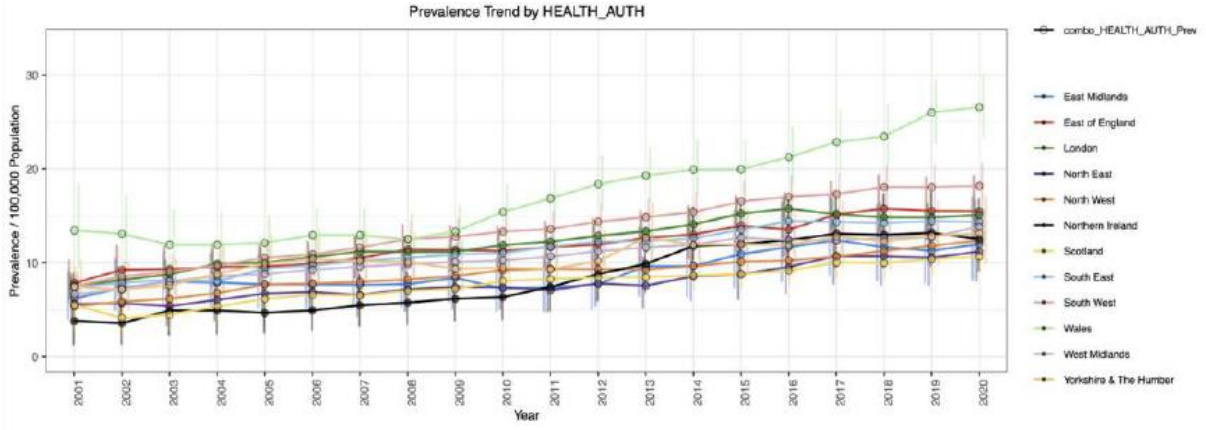
Dr Deva Situnayake returned to give the final presentation of the day. He explained that epidemiological studies are useful for quantifying and comparing disease rates in populations, as well as for looking at migration and questions such as nature versus

nurture. In a syndrome such as Behçet's, epidemiological studies can examine phenotype clusters and the evolution of clinical manifestations over time, compare disease outcomes, and consider differing pathogenesis and prognosis.

A meta-analysis of 31 studies presented at the International Conference on Behçet's Disease in 2014 found prevalence varying from 2 per 100,000 population in Northern Europe to 162 per 100,000 in Turkey. However, prevalence estimates vary widely depending on the type of study as well as geography. Migrants tend to adopt an intermediate risk between their original region and the one they have moved to, with a prevalence of 21 per 100,000 among Turks living in Germany.

The Health Improvement Network (THIN) is an anonymised primary care database containing data from 3.6 million people at >675 general practices in the UK. It includes data related to patient demographics, diagnoses, investigations, prescriptions and mortality, and is representative of the UK population as a whole. Research using THIN in Birmingham and Oxford reported a surprisingly high Behçet's prevalence of 14.6 per 100,000 in 2017, up from 8.3 per 100,000 in 2000, with a higher prevalence in women. The mortality risk was increased in people with Behçet's compared with the general population (adjusted hazard ratio 1.40), with a significantly increased risk of thrombotic disease that decreased after diagnosis of Behçet's. Other inflammatory conditions, such as IBD, psoriasis and rheumatoid arthritis, seemed to be overrepresented among the Behçet's cohort. Dr Situnayake said that there are overlapping genetic polymorphisms in these conditions, related to how the immune system responds to triggers at barriers such as the skin and the gut and sites of mechanical stress such as blood vessels and eyes.

Dr Situnayake went on to describe the project in Birmingham that explored the epidemiology of Behçet's from 2001 to 2022 in the UK using CPRD and linked HES datasets. An algorithm was developed to extract data for people with a diagnosis of Behçet's and compare them with controls. The prevalence was similar to that found in the THIN study, rising from 9.3 per 100,000 in 2006 to 14.7 per 100,000 in 2020. The increase was seen in all regions of England, with a more rapid increase in the South West and particularly in Wales. The prevalence seemed to be higher in people of mixed rate, and the increase was more rapid in women.



A retrospective validation study nested within the Birmingham Centre of Excellence included 4452 cases of Behçet's in HES between 2011 and 2019. Among these patients, 65% were female, 85% were Caucasian and the average age was between 30 and 49 years. The prevalence was about three times lower than in CPRD data and was higher in more deprived areas. A comparison of patients attending versus not attending the Centre of Excellence showed more comorbidities and greater use of infliximab in attenders. Non-Behçet's symptoms and gastrointestinal manifestations were more common in women, while ocular, vascular and cardiac manifestations were more common in men. Asian ethnicity was associated with the highest prevalence of ocular Behçet's. Mortality was lowest in the least deprived fifth, a comorbidity score of ≥5 increased the mortality risk, and infliximab treatment was associated with lower mortality. People with Behçet's had an increased risk of other MHC-1-related conditions such as IBD, ankylosing spondylitis and psoriatic arthropathy compared with the general population.

Dr Situnayake concluded by suggesting some topics for future research, including why the primary care record contains so many cases of Behçet's, why the prevalence is rising, and whether a pattern leading to potential Behçet's can be identified to shorten the time to diagnosis. A EULAR study group has been set up to identify overarching disease mechanisms in the 'MHC-1-opathies'.

Medical Panel Q & A

The day finished with a question and answer session with a Medical Panel comprising the staff of the Birmingham Behçet's Centre and the online speakers. In response to a question about the familial risk of Behçet's, Dr Situnayake stressed that the absolute risk is very low and that familial cases represent only 4% of all cases of Behçet's.

Responding to a question about diet, Dr Wallace said that no specific diet can be recommended, although the Mediterranean diet is beneficial for people with or without Behçet's. Replying to a question about migraine, Dr Situnayake explained that headaches can have many causes and are not usually related to Behçet's, although indirect factors such as tension and stress may be involved. Other questions related to the referral process for the Centres of Excellence and the role of GPs in treating gastrointestinal symptoms.

Clare Griffith, Editor