

A13/S(HSS)a

**2013/14 NHS STANDARD CONTRACT
FOR BEHÇET'S SYNDROME SERVICE (ADULTS AND ADOLESCENTS)**

PARTICULARS, SCHEDULE 2 – THE SERVICES, A – SERVICE SPECIFICATION

Service Specification No.	A13/S(HSS)a
Service	Behçet's Syndrome Service (Adults and Adolescents)
Commissioner Lead	
Provider Lead	
Period	12 months
Date of Review	

1. Population Needs

1.1 National/local context and evidence base

Management of Behçet's Syndrome (BS) in England has historically been uncoordinated and highly variable with huge variation in expertise in diagnosis, treatment strategies, medication used and duration of therapy. With a median time to diagnosis as long as 12 years, there is considerable potentially preventable end organ damage such as blindness suffered, with evidence from existing patients that unfortunately supports this concern - adding an urgency to the need for this service. As a consequence of the disease, more than two thirds of adults diagnosed with Behçet's receive some form of illness benefit and families may suffer from the trauma associated with the development of genital ulceration in children.

International evidence-based (EULAR) guidelines for treatment of Behçet's Syndrome based on systematic literature reviews have been established – including data on Number Needed to Treat (NNT) and effect size for response to medications.

This has identified a place for higher cost biologic agents (such as Tumor necrosis factor (TNF) inhibitors and interferons) in situations of intolerance or inefficacy to the standard drugs. However, since that review, the field has advanced significantly with further evidence accruing from UK and international authors to inform best practice. Effective early management of the aggressive uveitis in Behçet's Syndrome is essential to prevent blindness. The effectiveness of cyclosporine A has been superseded by results from the TNF inhibitors (comparison of infliximab versus cyclosporine during the initial 6-month treatment period in Behçet's disease).

There is some evidence to support the use of non-biologic agent Mycophenolate mofetil (MMF) in severe Behçet's Syndrome for eye or other organ complications

with patients moving to a biologic agent in situations of inefficacy or intolerance of MMF (Mycophenolate mofetil for ocular inflammation; Ocular Immunology & Inflammation; Mycophenolate mofetil for the treatment of uveitis).

2. Scope

2.1 Aims and objectives of service

The aim of this service is to ensure that patients of all ages suffering from Behçet's Syndrome can access timely diagnosis and receive optimal treatment equitably across the country.

The service will comprise three centres, utilising currently available medical expertise and maximising geographical access. The service will ensure that patients receive a definitive diagnosis, or exclusion of, Behçet's Syndrome in line with international diagnostic guidance, and access a range of key specialists in a 'one stop' process, providing the best care from diagnosis for a chronic, rare, disease. This model will serve not only to improve the experience of service by the patient but also clinical outcomes through best practice approaches to diagnosis and treatment.

Behçet's Syndrome in England is rare, symptoms are wide ranging and historically the lack of a clinical diagnostic test, has led to a prolonged time to diagnosis and the attendant risk of irreversible major organ damage, such as blindness.

The centres are based at Barts Health NHS Trust; City Hospital NHS Trust, Birmingham and Aintree University Hospitals NHS Foundation Trust, Liverpool. Each hospital has specialists with relevant expertise and will provide multidisciplinary teams that work together within each hospital and with the other Centres to further improve the care of Behçet's patients. The Behçet's Syndrome Society also has a presence in each centre to assist with the psychological support required when dealing with a rare, chronic condition.

The target population is all of England with a suspected and confirmed diagnosis of Behçet's Syndrome. The prevalence of Behçet's in the UK is actually unknown but estimates put it at 0.64 per 100,000 of the population.

The objective of the service is to ensure that patients of all ages with Behçet's Syndrome receive an accurate diagnosis in a timely manner and optimal clinical management delivered on a shared-care basis with the centres acting as hubs. Behçet's Syndrome is rare and requires care from multiple clinical specialties. Most medical professionals see only a few cases during their careers.

The purposes and goals of the service are therefore to offer early diagnosis and appropriate treatment for patients with Behçet's Syndrome in England, preventing irreparable damage to organs during the time otherwise taken to receive an accurate diagnosis and ensure patients enjoy the best quality of life possible.

2.2 Service description/care pathway

Behçet's Syndrome is a rare inflammatory multi-system disorder of unknown cause, typically characterised by recurrent oral aphthous ulcers, genital ulcers, uveitis and skin lesions. However it has a high potential to cause blindness or lead to life threatening complications when, for example, the central nervous system or major blood vessels are involved.

Whilst some of the common manifestations are self-limiting after variable periods, other problems can lead to irreversible damage without timely and appropriate therapeutic intervention. For example, the development of uveitis can cause blindness and vasculitis and thrombotic activity to central nervous system involvement including stroke or death – typically in young people.

On receiving a referral, the Behçet's specialist nurse at the centre will contact the patient by phone to take preliminary details. On arrival at the centre, the patient will be greeted by the specialist nurse. Initial tests will be undertaken including weight, height, blood pressure and samples of blood and urine. Appointments will then begin with the ophthalmologist, rheumatologist and oral specialist. Following the completion of these appointments, a review session is undertaken with all the specialists to agree the most appropriate treatment pathway for the patient. This is then discussed with the patient. Before leaving the clinic, the patient will have a further opportunity to speak to the specialist nurse and also the opportunity to book an appointment with the support worker if further support and signposting is required.

The centres provide a one-stop comprehensive clinic to allow patients to obtain rapid access to diagnosis and treatment for this complex disease. A newly diagnosed patient may attend the clinic up to three times in the following 12 months. Patients with an existing diagnosis will receive an annual check up, unless a flare is experienced, when further contacts with the clinic may be required.

The service includes:

- clinical consultation with experts representing multidisciplinary specialties at each clinic
- rapid referral to other related specialists where required
- telephone access to a specialist nurse both before and after appointments
- representation from the patient organisation, the Behçet's Syndrome Society, to improve information in the form of new patient leaflets, coverage in a newsletter and on the website and coordination of support groups
- support worker to provide non-medical support particularly to newly diagnosed patients.

Prior to the day of appointment, the patient will have an initial phone call from the specialist nurse, to gather information about their health since the last appointment and symptoms precipitating the referral. On entering the clinic, a face-to-face appointment takes place with the nurse, along with preliminary investigations that will include blood and urine tests. The patient will then consult the specialists. When the

specialists have assessed the patient, the team decides on appropriate

investigations and a care pathway and discuss this with the patient. The specialist nurse will support the patient through investigations and may perform supplementary tests under instruction from the multi-disciplinary team (MDT). The nurse will also ensure that correct epidemiological and demographic data is collected. A support worker service will be offered to the patient.

The product of the service is accurate, early diagnosis, provision of full information to patients and optimal treatment to manage the condition and minimise flares.

The provider will work with the service commissioners to ensure sufficient considerations are given to communications with patients and with the wider NHS to ensure that the service is publicised to improve patient access.

Risk Management

It is widely recognised that an effectively planned, organised and controlled approach to the risk management process is the cornerstone of sound management practice, which aims to anticipate and wherever possible prevent, or manage risks to patients, staff, visitors and the organisation. Good risk management awareness and practice embedded in the service is an essential success factor in ensuring that risks are managed systematically and consistently.

All centres are currently compliant with or are working towards:

- CQUINS national core standards
- 16 new essential standards for quality and safety
- national risk management standard level 2

Each centre will identify risks and manage these to improve patient care and there are formal arrangements in place to support the standards above. The centres also work on the principle of promoting a policy of openness and accountability and by effective communication, which will extend both within their centre and across the service.

By commissioning three centres, there is the opportunity for peer review from which shared learning can take place thereby mitigating risk.

Accessibility/acceptability

The centres will offer a comparable service to everyone in England who either has a confirmed or suspected diagnosis of Behçet's Syndrome. In the literature and at the centres, equal access to the service regardless of age, culture, disability or gender will be promoted. Full provision will be made for assessment and care of children, who will be seen in the centre accompanied by the consultant paediatrician and paediatric nurse in charge of their care. Transitional clinics will be performed with adult specialists undertaking clinics alongside paediatricians, to ensure a seamless transfer of care.

To promote to all, advertising will take place in differing forms including:

- briefings for clinicians and researchers at the Behçet's Forum (an informal meeting of clinicians interested in Behçet's)
- mailshots advertising the Behçet's Centre to the members of the Behçet's Syndrome Society which is the largest database of patients in England & Scotland
- The Behçet's Syndrome Society website and helpline will advertise the centres by providing information about the centres, how to contact them, what their purpose is etc. The website reaches a large audience, many of whom are searching on their symptoms without a diagnosis
- Choose and Book – the Behçet's Centres will be registered on this system so that GMPs and other referring parties can make an informed referral.

2.3 Population covered

This service will cover patients registered with an English General Practitioner. Patients from Wales, Scotland and Northern Ireland will not be part of this commissioned service and the trusts will ensure separate arrangements are in place for patients from these and other non-EU referrers.

The centres have been chosen due to the geographical spread throughout England. With the transport network, patients in all areas of England should be able to access at least one of the centres.

2.4 Any acceptance and exclusion criteria

Patients will be referred by hospital consultants or General Practitioners within the NHS in England.

Several categories of patient will contact the centres:

- existing patients: will visit the centre annually for a review
- existing patients undergoing a 'flare' may contact the centre more frequently
- new patients: will visit the centre to receive a diagnosis or an exclusion from a diagnosis. If diagnosed with BS, they will visit the centre on further occasions during their first year. If a diagnosis of BS is excluded, they will not visit the centre again.

Patient pathway Appendix 1

Drugs protocol and pathway Appendix 2

Referral criteria, sources and routes

The centres will provide both a diagnostic service and ongoing care for all patients with either a confirmed or suspected diagnosis.

Referrals will be accepted from colleagues in primary and secondary care where Behçet's Syndrome is suspected. The specialist nurse will triage the patient by

phone on receiving the referral.

Exclusion criteria

The centres will accept referrals where Behçet's Syndrome is suspected based on the International Diagnostic Criteria. There are no specific exclusion criteria for referral.

Response time & detail and prioritisation

New referrals will be prioritised according to level of disease activity and threat of major organ damage (e.g. reduced vision or neurological features). A patient with an organ-threatening flare will be seen within 2-3 days. A new patient will be seen within 2-3 weeks depending on an initial assessment of disease activity from the specialist nurse.

Discharge Criteria and Planning

As there is currently no cure for Behçet's Syndrome, patients will require symptomatic treatment depending on clinical signs and symptoms and the severity of these in addition to whether the patient is experiencing a flare. All patients will be kept under periodic review.

Patients who are referred with suspected Behçet's Syndrome that is subsequently not confirmed will be discharged following their initial or subsequent appointment unless a significant element of doubt remains and it is deemed clinically appropriate for further review.

2.5 Interdependencies with other services

The providers of the National Behçet's Service are Barts Health NHS Trust; City Hospital NHS Trust, Birmingham and Aintree University Hospital NHS Foundation Trust, Liverpool.

Behçet's Syndrome is a multi-system disease that can affect almost any organ or multiple organs throughout the body and affects the patients in 'flares' of activity.

From the Behçets Syndrome Society (BSS) Survey Report diagnosis typically takes 12 years. Due to the multi-systemic nature of the condition, referrals are expected from rheumatology, ophthalmology, dermatology, gastroenterology, oral specialists and gynecology clinics. Referrals are also expected from primary care, dentists and sexual health professionals.

The clinicians in the centres will provide the most significant portion of the service,

but will have agreed procedures for care closer to home. This will work through managing joint protocols for delivery of treatment and medication or to support medical teams through the complex management of inpatients through shared care.

The type of treatment offered will depend on symptom control, which may be highly variable in organ system involvement and flares which may be recurrent or new and which vary in severity. In these cases treatment may range from topical anti-inflammatory medication to systemic immunosuppressive drugs through to the biological drugs.

2.6 Relevant networks and screening programmes

There is currently no clinical test for Behçet's Syndrome precluding any screening programmes.

There is an informal network of medical professionals with an interest in Behçet's Syndrome called the Behçet's Forum. This meets annually and currently has a small active membership that undertakes research into Behçet's Syndrome. As part of the centre proposal the forum will annually have a shared national meeting with patients.

3. Applicable Service Standards

3.1 Applicable national standards e.g. NICE, Royal College

The specialists and nurses in the centres will continue with continuous professional development. Within the centres, there is a commitment to the sharing of knowledge and development as follows:

- Targeted Centre Education – this will be delivered by the specialist consultants in each of the centres on a quarterly basis to the other professionals working in the centre to ensure a base level of understanding;
- Behçet's Centre Conference (one held at each centre annually) – the content of this day is delivered by staff within each centre and the Behçet's Syndrome Society to GPs, nurses, optometrists, counsellors, students and consultants from surrounding Clinical Commissioning Groups (CCGs). The purpose is to increase the awareness and understanding of Behçet's Syndrome to aid appropriate referrals;
- National Behçet's Research Network – this is aimed at all researchers and clinicians nationally and internationally with an interest in Behçet's Syndrome with the purpose of sharing research output and improving clinical practice as appropriate.

All the participating consultants will be subject to annual appraisal, linked to revalidation through the trusts identified responsible officer. They will all be up to date with mandatory training.

The centres will provide a variety of information to patients and carers. Most will be published on the centre's website along with an information pack which will be presented to all new patients during their first appointment. The pack will include:

- centre information;
- clinical information about Behçet's syndrome;
- information about the Behçet's Syndrome Society;
- specific leaflets on each symptom written by medical experts;
- information describing the Support Groups and how they may be helpful.

The centres will also have a page or part thereof in the Behçet's Syndrome Society quarterly newsletter to highlight news and contact information. They will have a specific area established on the society's website at www.behcets.org.uk

Information will also be created for referring medics to advertise the centres on the website.

The support worker will provide sign posting information in relation to the benefits system and around the NHS for further services.

Service Standards

Area 1: Patient-Centred Care

Standard	Description	Compliance Measured By
1.1	Both patients & carers will be provided with comprehensive information and support in order to make informed decisions about their treatment choices.	Review of process. Review of information documentation. Notes review. Patient consultation.
1.2	Information will be made available in a wide range of formats and on more than one occasion. It will be clear, concise, and culturally sensitive and evidence based where possible.	Review of information documentation. Notes review. Patient consultation.
1.3	A range of psychological and social support services will be offered to meet the needs of patients and carers through both the support worker and other specialists.	Availability and presence of support worker and psychologist. Review of training. Take up rates for these services.
1.5	Patient and carers' self-help and support groups are encouraged and all patients referred to the designated centre will be provided with information about them as	Number of support groups organised by support worker. Number of attendees within the group. Availability of information leaflets.

	appropriate.	
1.6	Patients and referring clinicians will have access to the specialist nurse for advice, information and support that is appropriate to the urgency of the needs of the patient.	Specialist nurse triage outcomes
1.7	Patients, carers and their General Practitioners will have access to the specialist nurse in the first instance and dependent on the assessment, other members of the multi-disciplinary team to discuss specific problems or concerns.	Specialist nurse usage Patient satisfaction surveys
1.8	The views of patients, carers and staff will be regularly and formally sought and the results openly available.	Patient satisfaction surveys. Review management minutes to confirm results discussed and acted upon if necessary.
1.9	There will be formal arrangements for addressing complaints and other comments made by patients, carers and staff at other times.	Review complaint pathway.
1.10	Staff in the multi-disciplinary team will have training and be supported in using communication skills. There will be a policy for breaking bad news.	Review policy. Review training.

Area 2: The Specialist Multi-Disciplinary Teams (MDT)

Standard	Description	Compliance Measured By
2.1	<p>The core multi-disciplinary team for care of patients with Behçet's Syndrome will include the lead clinician, specialist nurse, an ophthalmologist, an oral specialist a neurologist, a dermatologist, the support worker and administration support. Other specialists (e.g. psychologist, gastroenterologist and genitor-urinary specialist) may attend according to the needs of the different aspects of the service or will have fast-track agreements.</p> <p>The attendance and activities of the MDT should be recorded and maintained in a register.</p>	<p>Team: Short CVs including qualifications, specialist training and duration of Behçet's experience</p> <p>View register of MDT meetings</p>
2.2	There will be evidence of continued professional development and	Appraisal and CPD record

	attendance at national and international Behçet's meetings by the clinicians within the MDT.	
2.3	Lead Clinician The lead clinician must have adequate time in his/her job plan to devote to the Behçet's service.	View job plans of each clinician

Area 3: Access, Assessment and Diagnosis

Standard	Description	Compliance Measured By
3.1	The centre will offer clinic appointments to new patients within 2-6 weeks based upon disease activity.	Report from centres
3.2	For patients on regular follow up, all assessments where the clinician is resident in the clinic will be completed at one visit.	Audit of clinic visits
3.3	For patients who have no confirmed diagnosis of Behçet's but where doubt remains, will receive a diagnosis within 12 months	Audit of clinic visits

Area 4: access, assessment and diagnosis

Standard	Description	Compliance Measured By
4.1	The centre will have a well-defined programme for staff training.	Training programme and information
4.2	Consultant core members of the MDT will attend at least one Behçet's meeting or conference per year.	CPD/comparable certification
4.3	Consultant core members from each centre will meet and share best practice with each other on an annual basis	CPD/comparable certification
4.4	The specialist nurses will attend at least two relevant study days per year.	CPD/comparable certification and or personal training record

Area 5: Research and Development

Standard	Description	Compliance Measured By
5.1	Research Strategy The centre to have a research strategy and programme which documents: current and planned activity, the resources needed to support activity and objectives for development.	Research strategy document Progress against objectives in the Annual Report

5.2	<p>Research Governance There will be evidence of compliance with research governance requirements:</p> <ol style="list-style-type: none"> 1. systems are in place to ensure that an appropriate member of staff is notified of, and has approved, all research in the organisation 2. systems are in place to ensure all ongoing research has ethics committee approval 3. arrangements are in place to ensure that someone acceptable is responsible for making sure that informed consent and procedures in the protocol approved by the ethics committee are being adhered to 4. evidence of compliance with the Mental Capacity Act 2005 which provides safeguards for a person who lacks capacity to consent to research. 	<p>NHS Trust Research Governance Policy and Procedures in place, plus evidence of trust and REC approval of all research activity</p> <p>Written procedures are agreed and systems are in place to audit compliance</p>
5.3	<p>Research Outputs and Outcomes There will be regular participation in national and international Behçet's conferences.</p>	Summary of activity in Annual Report
5.4	Clinical practice will be informed by regular review of research evidence derived from local, national or international research activity, subject to availability of resources.	Examples documented in Annual Report
5.5	Peer-reviewed publications on Behçet's will be reported annually to the National Commissioning Group.	Publications listed in Annual Report

Area 6: Communication with Primary & Secondary Care Services

Standard	Description	Compliance Measured By
6.1	Referring physicians must receive adequate feedback after they refer patients.	Audit of feedback to referring clinicians letters
6.2	After each visit to the follow up clinic, a letter with details of investigations and changes in medication must be sent promptly to the referring clinician.	Audit of clinic letters

6.3	GMP, GDP or another specialist GPs, hospital doctors and other health care professionals with questions about any aspect of Behçet's care will be able to contact the MDT for advice.	Satisfaction survey of users of the service Usage record of nurse specialists.
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Area 7: Management & Organisation

Standard	Description	Compliance Measured By
7.1	Leadership and Accountability There will be clear accountable leadership of the Behçet's Centres with a lead clinician notified to NHS England. There will be adequate time available to the lead clinician and manager to perform this role.	A management structure that clearly demonstrates lines of accountability within the service and within the trust. Job plans and job descriptions outlining responsibilities and sessional time allocated.
7.2	Lead Clinician and Manager Responsibilities The lead clinician will have overall responsibility for ensuring staff are fully aware of the standards against which the Centre will be assessed and that mechanisms are in place to achieve compliance with the standards.	Clinical governance annual and other reports.
7.3	Evidence Based Services / Protocols There will be evidence based service protocols, which are reviewed and updated regularly covering all aspects of the service. These will be available to all new staff joining the service and will form part of a formal induction to the Behçet's centres.	Documented protocols made available. Record of induction of new staff
7.4	Workforce Planning There will be effective and sustainable workforce planning covering all professional disciplines forming part of the multidisciplinary team. All staff will have regular appraisal and agreed professional development plans.	Published senior and junior staff rotas. Record of appraisal and professional development plans
7.5	Business Conduct There will be regular business	Record of attendance and

	meetings, not less than four times each year, within the centre to address issues specific to the Behçet's centres. This will include financial reporting, activity reporting, education, audit, and clinical governance and research issues.	minutes of meetings made available.
7.6	<p>Resource Use</p> <p>There will be clear accounting for all income to the Trust that is for the delivery of the service in accordance with the fiscal guidance set out by NHS England.</p> <p>This will include finance directly managed by the Behçet's Centres and that which is managed by the finance infrastructure within the Trust.</p>	Trust Financial Reports NHS England Review
7.7	<p>Data Collection</p> <p>Robust arrangements will be in place for timely and accurate collection of activity and outcome data. Data will be made available to NHS England under agreed reporting mechanisms.</p>	Data transfer to NHS England

Area 8: Audit

Standard	Description	Compliance Measured By
8.1	There will be a regular programme of clinical audit.	Annual audit report
8.2	The agreed clinical outcomes will be reported to NHS England by 1 st September each year for all patients seen face-to-face in the 12 months ending on 31 st March.	External reports

4. Key Service Outcomes

The desired high level outcomes of the service are: accurate, definitive diagnosis of Behçet's Syndrome to reduce the current average 12 year diagnosis period; access to therapy with the most effective treatments; avoidance of inappropriate therapies and reduction in treatment related morbidity and mortality; improved quality of life.

The service will provide an annual report on the following measures:

Clinical outcome:

- number of flares per patient;
- visual acuity;
- arthritis pain score;
- For newly diagnosed patients only: Time to diagnosis (defined from time of first contact with a doctor for a Behçet's related symptom).

Quality Performance Indicator	Threshold	Method of measurement	Consequence of breach	Report Due
Reduction in diagnostic time	Average of 6 years to diagnosis	Patient survey for diagnostic time	n/a	Annually
Improvement in Mucocutaneous lesions	Baseline survey from Behçet's Syndrome Society	Patient survey	n/a	Annually
Reduction in pain due to arthritis/arthropathy	50% reduction over the 5 year period from baseline survey	Patient survey	n/a	Measured annually, target over 5 years
Improved Quality of Life	Improvement from current baseline survey	Patient survey	n/a	Annually
Reduction in the number of flares	Reduction in the number of flares	Patient survey	n/a	Annually
Reduction in the loss of visual acuity	Reduction in the loss of visual acuity	Patient survey	n/a	Annually
Improved access to Support Groups	Larger % of patients taking part in a support group	Patient survey	n/a	Annually
Improved signposting for patients	Improved knowledge in patients	Patient survey	n/a	Annually
Improved information	Improved knowledge in	Patient survey	n/a	Annually

	patients			
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5. Location of Provider Premises

Location(s) of service delivery

The centres will be located throughout England at the following hospitals:

- London (Bart's Health NHS Trust)

- Birmingham (City Hospital NHS Trust)
- Liverpool (Aintree University Hospitals NHS Foundation Trust)

Days/hours of operation

The clinics will be held at the following frequencies:

Liverpool and Birmingham – four clinics per month

London – six clinics per month.

A 24 hour on call service will be available to all patients.

The specialist nurse will be available by phone at any time during working hours and will advise the appropriate course of action. Outside of working hours, the Behçet's Syndrome Society provides a helpline.

The support worker will be available both in the clinic hours and to visit the patient's house if required.

Sub-contractors

The majority of services will be sourced from within the trust named, but the following sub-contractors will be used:

- Liverpool:
 - Oral Medicine - Liverpool Dental Hospital
 - Support Worker - the Behçet's Syndrome Society
- Birmingham:
 - Oral Medicine – Birmingham Dental Hospital
 - Support Worker – the Behçet's Syndrome Society
- London:
 - Support Worker – the Behçet's Syndrome Society

All centres are currently compliant with or are working towards:

- CQUINS national core standards
- 16 new essential standards for quality and safety
- National risk management standard level 2

Adopted