

Scottish Centre for Complex Ehlers Danlos Syndrome (SCCEDs) Stage 2 Proposal

1. Description of service

The proposed service aims to establish a centre for the assessment, and management of patients with complex Ehler's Danlos Syndrome (hEDS) who required expertise that was not available locally or regionally in Scotland.

The centre would accept referrals from secondary care clinicians in the NHS Boards throughout Scotland, where it was felt that the patients' needs could not be adequately addressed locally.

The aim of the centre would be to offer a comprehensive assessment with a view to clarifying diagnostic uncertainty and suggesting a holistic package of care which could be delivered as close to the patients home with a view to meeting their needs.

Referrals into the service would be accepted from Consultant Rheumatologists across Scotland.

The criteria for referral will be:

- Clinical diagnosis of hEDS confirmed by a Consultant Rheumatologist with debilitating symptoms that persist despite an adequate trial of supportive therapy delivered locally.

Initial assessment

An initial assessment within an outpatient clinic will be undertaken by a Consultant Rheumatologist. During the 45-minute consultation, confirmation of diagnosis will be made and patient queries about the nature of their condition will be answered. The assessment will also include 2 comprehensive 1 hour consultations with skilled allied health professionals from physiotherapy and occupational therapy and in addition if felt appropriate input will be sought from clinical psychology. A treatment plan will be developed pulling together the expertise of the medical and allied professionals.

It is envisaged that the following day, the patient will have further 30-minute therapy sessions with the PT and OT to reinforce the treatment plan.

Initial review

At 3 months the patient will be reviewed by the Consultant Rheumatologist (30 minutes), Physiotherapist (30 minutes), Occupational Therapist (30 minutes) and (if necessary) the Clinical Psychologist.

Final review and discharge

A final review is anticipated at 6 months with patient then being discharged back to referring clinician local to patient.

Throughout the patient pathway the Centre will communicate with referring clinician and associated professionals to ensure that treatment recommendations can be delivered locally.

2. Provider & Location

NHS Lothian, Western General Hospital (WGH) in Edinburgh

3. Additional information requested at stage 1 (if applicable)

- Patient group to be defined more precisely
- Consideration should be given to including other non-inflammatory connective tissue disorders
- Input should be sought from rheumatology and orthopaedic colleagues
- Need for clear de-designation plan developed.

4. Patient need

There are few datasets on the prevalence of hEDS, but current estimates suggest it affects between 1 in 5000 to 1 in 10,000 people, which means that there may be between 500 and 1000 people in Scotland with the condition. Many people with hEDS do not require specialist input since the diagnosis can be made clinically based on history and examination and often no specific treatment is required.

However, a proportion of patients with hEDS present with chronic and disabling pain, recurrent dislocations, and kyphoscoliosis, along with various cardiovascular, gastrointestinal or genitourinary problems. These patients would be those defined as within the scope of a complex hEDS service.

NHS Lothian colleagues have audited the number of referrals with a diagnosis of EDS that have been treated by physiotherapy and/or occupational therapy at the Rheumatic Diseases Unit in Edinburgh. Extrapolating their data suggests that up to 350 hEDS patients in Scotland may need referred to Secondary Care per year, but it's likely that a smaller number will have complex hEDS as defined above and a smaller subset will require expertise not available locally or regionally.

5. Clinical effectiveness

A specialist national service – building regional capacity

This new national service will provide specialist assessment, and management of patients with complex Ehler's Danlos Syndrome (hEDS) whose needs and those of clinicians are not being adequately met at a local level. It is envisaged that the proposed service will share learning and build capacity in Rheumatology and related therapy services across Scotland. The service intends to work towards developing a 'Regional Hub and spoke' model of service. In this model, increasingly specialist levels of assessment could be undertaken in local units in the West and North regions of Scotland, thereby reducing over time the continuing caseload demand on the Edinburgh national specialist service.

6. Costs

The current service cost in Lothian is £133,000. The projected cost of the service is £584,000.

7. Support for proposal

A survey of Rheumatologists and Orthopaedic Surgeons indicates support for the proposal. Letters of support have been received by the Royal College of Physicians, Edinburgh and Ehlers Danlos Support UK. Additional letters are expected from the Scottish Committee for Orthopaedics and Trauma (SCOT), the Scottish Cardiac Society (SCS) and the Scottish Society of Gastroenterology (SSG).

Since NPPPRG, The Scottish Society of Rheumatology (SSR) Council however have declined to provide a letter of support for the proposal.

8. NSD assessment of proposal

The proposal as stands would address the lack of services for people with complex EDS. Stakeholder engagement has identified that the needs of many patients are not being addressed at present, there is diagnostic uncertainty and in some areas of Scotland expertise in the management of the condition is lacking.

The service would aim to provide diagnostic clarity and support to referring clinicians, holistic care and expertise for patients and recommendations as to continuing care needs.

At present a number of patients per year are referred to genetic services for EDS commissioned by NHS England and a number also require the input of rehabilitative services offered in either Bath or Stanmore. It is anticipated that as a result of the service there would be a lesser requirement to refer patients to these specialist services.

Given that the envisaged service would also provide education and aims to build capacity across Scotland, it is hoped that the service would also equip local teams with the knowledge to manage people with non-complex EDS and over time extend their scope to address the needs of those with Complex EDS. The engagement of regions and local units would be required explicitly for this, and a form of network management resource may be required to facilitate the development

| Criteria | Met |
|------------------------------------|-----------------------------------|
| Numbers | Yes |
| Equity | Yes |
| Evidence | Partial (evidence base is sparse) |
| Benefits of national commissioning | Yes |

The recent intimation by the Scottish Society of Rheumatology that they would be unwilling to support the application however casts doubt as to the proposed referral pathway in that in the main referrals to National Services are from local or regional specialists and are not routinely accepted from Primary Care. Neither the applicants nor NSD are of the opinion that direct referrals from Primary Care would be an appropriate use of resource and that some form of scrutiny and expert diagnosis would be required before referral.

9. NPPPRG Discussion

NPPPRG supported the continued development of a stage 3 application provided there was clarification of costs, patient numbers, model of service, inclusion of other specialties and with a view to measure quality of life.

It should however be noted that this recommendation predated the comments from SSR Council.

10. NSSC Consideration

NSSC is asked to review Stage 2 proposal and decide whether there is sufficient basis to proceed to Stage 3.

**NSSC Secretariat
September 2019**