Dear Primary Care

Somerset Community Pain Service and the Rheumatology team have teamed up to provide general practice in Somerset with some information to help manage patients with non-inflammatory rheumatology conditions (fibromyalgia, hypermobility). This sits alongside existing NICE guidelines and other tools including the GPtoolkit from Ehlers Danlos Support UK.

Fibromyalgia

A diagnosis of fibromyalgia is valid irrespective of other diagnoses. A diagnosis of fibromyalgia *does not* exclude the presence of other clinically important illnesses.

There is no confirmatory blood test for fibromyalgia. Testing to rule out other pathologies should be kept to a minimum and may include the following blood tests: FBC, U&E, LFT, TSH, ESR, and CRP. Any further tests should be based on clinical suspicion for other conditions and may include RF, ANA, ANCA, CK, Bone profile, Vitamin D, PTH and considering obstructive sleep apnoea.
Please also use the scoring system (appendix 1) which is aimed to provide guidance in making the diagnosis.

Hypermobile Ehlers Danlos Syndrome (hEDS)

The GPtoolkit for EDS is a useful tool to support diagnosis and management of EDS. Appendix 2 contains the criteria for hEDS which can be used to confirm the diagnosis. Once diagnosed, the patient should be given information about EDS and its management.

Exercise has the strongest evidence base and should be first line for long-term management of hEDS. This should include graded aerobic exercise and resistance training. There is limited evidence to support the use of analgesia, anti-inflammatory drugs or other pharmacological interventions. Cognitive behavioural therapy has proven to be effective in people with mood disorders or poor coping strategies.

Management should be in primary care or via onward referral to physiotherapy and/or local IAPT and pain service as appropriate.

A diagnosis of hEDS is valid irrespective of other diagnoses. There is considerable clinical overlap with fibromyalgia and chronic fatigue syndrome.

With a consensus of practice, we hope that patients with fibromyalgia and/ or hEDS will be able to access the most appropriate care more quickly, with onward referral to the Somerset Community Pain Management Service (SCPMS) as necessary.

Best wishes

Dr Suzanne Carty (Consultant Anaesthetist, clinical lead for SCPMS)
Dr Kerry Aston (Consultant Rheumatologist)

Appendix 1 – Fibromyalgia Diagnostic Criteria

<table>
<tr><td colspan="2">Part A
Pain in the last week</td><td>Part B
Symptoms in the last week</td></tr>
<tr><td colspan="2">Region</td><td>Symptom</td></tr>
<tr><td>Left upper region

□ Left Jaw
□ Left shoulder girdle
□ Left upper arm
□ Left lower arm</td><td>Right upper region

□ Right jaw
□ Right shoulder girdle
□ Right upper arm
□ Right lower arm</td><td>Grade the severity of the symptom*

Fatigue 0□ 1□ 2□ 3□
Waking unrefreshed 0□ 1□ 2□ 3□
Cognitive Symptoms 0□ 1□ 2□ 3□</td></tr>
<tr><td>Left lower region

□ Left hip (buttock, trochanter
□ Left upper leg
□ Left lower leg</td><td>Right lower region

□ Right hip (buttock, trochanter
□ Right upper leg
□ Right lower leg</td><td>Have the following symptoms been bothersome in the last month? (Tick if yes)

□ Headache
□ Abdominal Pain
□ Depression</td></tr>
<tr><td>Axial region

□ Neck
□ Upper back
□ Lower back
□ Chest
□ Abdomen</td><td>Total Points:

____ /19 = Widespread pain index (WPI)</td><td>Total points:

____ /12 =Symptom severity score (SSS)</td></tr>
<tr><td colspan="3">Fibromyalgia severity score: Widespread pain index + Symptom Severity score = ____ / 31</td></tr>
<tr><td colspan="3">Fibromyalgia Diagnosis:
A. Pain and symptom score thresholds
WPI ≥7 plus SSS ≥5
Or
WPI ≥4 plus SSS ≥9
AND
B. Generalised pain present in 4 of the above 5 regions (excluding jaw, chest and abdomen)</td></tr>
<tr><td colspan="3">*0 = no problems; 1= slight or mild problems, generally mild or intermittent; 2= moderate considerable problems, often present and/or at moderate levels; 3 = severe pervasive continuous life disturbing problems</td></tr>
<tr><td colspan="3">Taken from:- Wolfe F, Clauw DJ, Fitzcharles MA et al.
2016 Revisions to the 2010/2011 fibromyalgia diagnostic criteria. Semin Arthritis Rheum 2016;46(3):319–329.</td></tr>
</table>

Appendix 2: 2017 Diagnostic criteria for hypermobile EDS

Adapted from Malfait et al. Am J Med Genet C Semin Med Genet. 175(1): 8–26

Criteria: All 3 must be met

CRITERION 1:
Generalized Joint Hypermobility (GHJ)
Must meet Beighton Score for age

Age	Beighton Score (see below)
Prepubescent or adolescent	≥6
Pubescent up until age 50	≥5
Over age 50	≥4
Patients with acquired joint limitations (eg injury or surgery)	BS 1 point under age requirements AND a positive 5PQ (see below)

CRITERION 2:
At least 2 features must be present:

Feature A: Systemic manifestations of inherited connective tissue disorder (CTD) (need ≥5)

1. Unusually soft/velvety skin
2. Mild skin hyperextensibility
3. Unexplained striae distensae/rubrae
4. Bilateral piezogenic papules of heel
5. Recurrent/multiple abdominal hernia
6. Atrophic scaring in ≥2 sites
7. Pelvic floor, rectal, and/or uterine prolapse in children, men or nulliparous women
8. Dental crowding and high or narrow palate
9. Arachnodactyly
10. Arm span-to-height ≥1.05
11. Mitral valve prolapse
12. Aortic root dilatation with Z score > +2

Feature B: Family history
(1 or more first degree relatives must meet criteria)

Feature C: Musculoskeletal complications (need ≥1)

1. MSK pain in ≥2 limbs, recurring daily for ≥3 months
2. Chronic widespread pain for ≥3 months
3. Recurrent joint dislocations or frank joint instability, in the absence of trauma (a or b)
a. ≥3 atraumatic dislocations in same joint or ≥2 more atraumatic dislocations in two difference joints occurring at different times
b. Medical confirmation of joint instability at two or more sites not related to trauma

CRITERION 3: All 3 prerequisites must be met

1. Absence of unusual skin fragility.
2. Exclusion of other heritable and acquired connective tissue disorders. In patients with an acquired connective tissue disorder, additional diagnosis of hEDS requires meeting both Features A and B of Criterion 2. Feature C of Criterion 2 cannot be counted in this situation.
3. Exclusion of alternative diagnoses that may also include joint hypermobility by means of hypotonia and/or connective tissue laxity.

Beighton Hypermobility Score.

Adapted from Beighton et al. Ann Rheum Dis. 32(5):413–418

Task:

- Passively dorsiflex 5th finger beyond 90°
- Passively place thumb adjacent to anterior aspect of forearm
- Hyperextend elbow joint beyond 10°
- Hyperextend knee joint beyond 10°
- Lay palms flat on the floor without bending the knees

Key:

1 point for the ability to perform each task (1 point per side of body, when applicable) Max 9 points

5PQ Five-Point Questionnaire for identifying hypermobility.

Adapted from Hakim and Grahame Int J Clin Pract. 57(3):163–166

Questions:

1. Can you now (or could you ever) place your hands flat on the floor without bending your knees?
2. Can you now (or could you ever) bend your thumb to touch your forearm?

3. As a child, did you amuse your friends by contorting your body into strange shapes, or could you do the splits? 4. As a child or teenager, did your shoulder or knee cap dislocate on more than one occasion? 5. Do you consider yourself double jointed?
Key:
Two or more “yes” answers suggests joint hypermobility.

NB A diagnosis of generalised joint hypermobility (without EDS) can be made from criterion 1. Management is essentially the same.