Hypermobility Syndromes Association,
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Registered in England and Wales (1011063)

Hypermobility Syndromes: Guidance for commissioners using the Kent Model (adults and adolescents)

EXECUTIVE SUMMARY

Hypermobility is a term used to describe the ability to move joints beyond the normal range of movement. It may present in just a few joints or can be widespread. This is usually of no medical consequence and commonly does not give rise to symptoms. For some it can be an advantage, e.g., athletes, dancers and musicians. However, for a small but significant percentage of the population hypermobility may be associated with a variety of symptoms (**Hypermobility Syndromes**).

There is now compelling evidence that hypermobility is linked to Chronic Widespread Pain (CWP), Fatigue, Fibromyalgia, Irritable Bowel Syndrome (IBS), Postural orthostatic Tachycardia Syndrome (PoTS), Autonomic Nervous System Dysfunction and a variety of psychological and neurodevelopmental conditions including Anxiety. Currently many patients are mislabelled as having “Medically Unexplained Symptoms”.

Conditions include the **Hypermobility Spectrum Disorder/hypermobile EDS (HSD/hEDS)** (formerly known as Hypermobility syndrome). There are the rare forms of Heritable Disorders of Connective Tissue (HDCT) including for example: Ehlers-Danlos syndrome, Marfan’s syndrome and Osteogenesis Imperfecta.

Understanding of these conditions and comorbidities is poor in primary and secondary care and this leads to unnecessary consultations, incorrect diagnoses and investigation, inappropriate prescribing (particularly opioids) and other medical interventions which are ineffective, expensive or harmful. Medications are frequently ineffective in these conditions.

Importantly, a variety of interventions have been shown to be effective in alleviating symptomatology and pain, this includes specialist physiotherapy, OT, podiatry and therapies such as Tai Chi, the Alexander Technique and specific exercise programs. Psychological interventions including mindfulness and Mindful Self Compassion improve patient wellbeing.

Correct early diagnosis leads to simple interventions which can often make a significant difference. Late or missed diagnosis has significant negative consequences. Surveys have shown that on average the delay from presentation to diagnosis is 10 to 14 years.

Immediate action is required because the main specialist centre in London has closed it’s doors to referrals whilst a more sustainable arrangement is sought. The Kent Model seeks to improve education and patient wellbeing using existing resources. This paper describes how the Kent Model can help improve local services, thereby improving the patient, doctor’s and health professional‘s experience.

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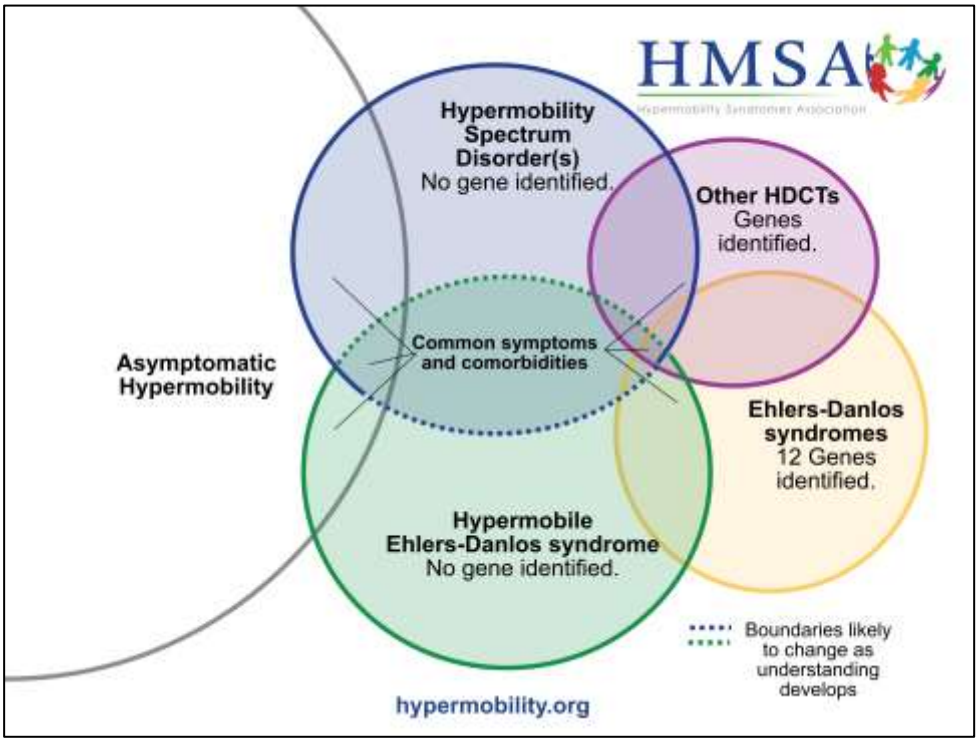
Hypermobility is the term used to describe the ability to move joints beyond the normal range of movement. Joint hypermobility is common in the general population. It may be present in just a few joints or it may be widespread. It is most common in childhood and adolescence, in females, and Asian and Afro-Caribbean races. It tends to lessen with age. In many people joint hypermobility is of no medical consequence and commonly does not give rise to symptoms. Hypermobility can even be considered an advantage, for example athletes, gymnasts, dancers and musicians might specifically be selected because of their extra range of movement.

What are the hypermobility syndromes?

For a small percentage of the population, instead of being advantageous, hypermobility may be associated with joint and ligament injuries, pain, fatigue and other symptoms. Hypermobility can also be a sign of a more significant underlying condition, which are often passed down through the generations. These conditions are known as Hereditary Disorders of Connective Tissue (HDCT). Joint Hypermobility Syndrome, and heritable disorders of connective tissue including Ehlers-Danlos syndromes, Marfan's syndrome, Osteogenesis Imperfecta, Stickler syndrome and other hypermobility associated conditions were reclassified into four broad groups illustrated below:



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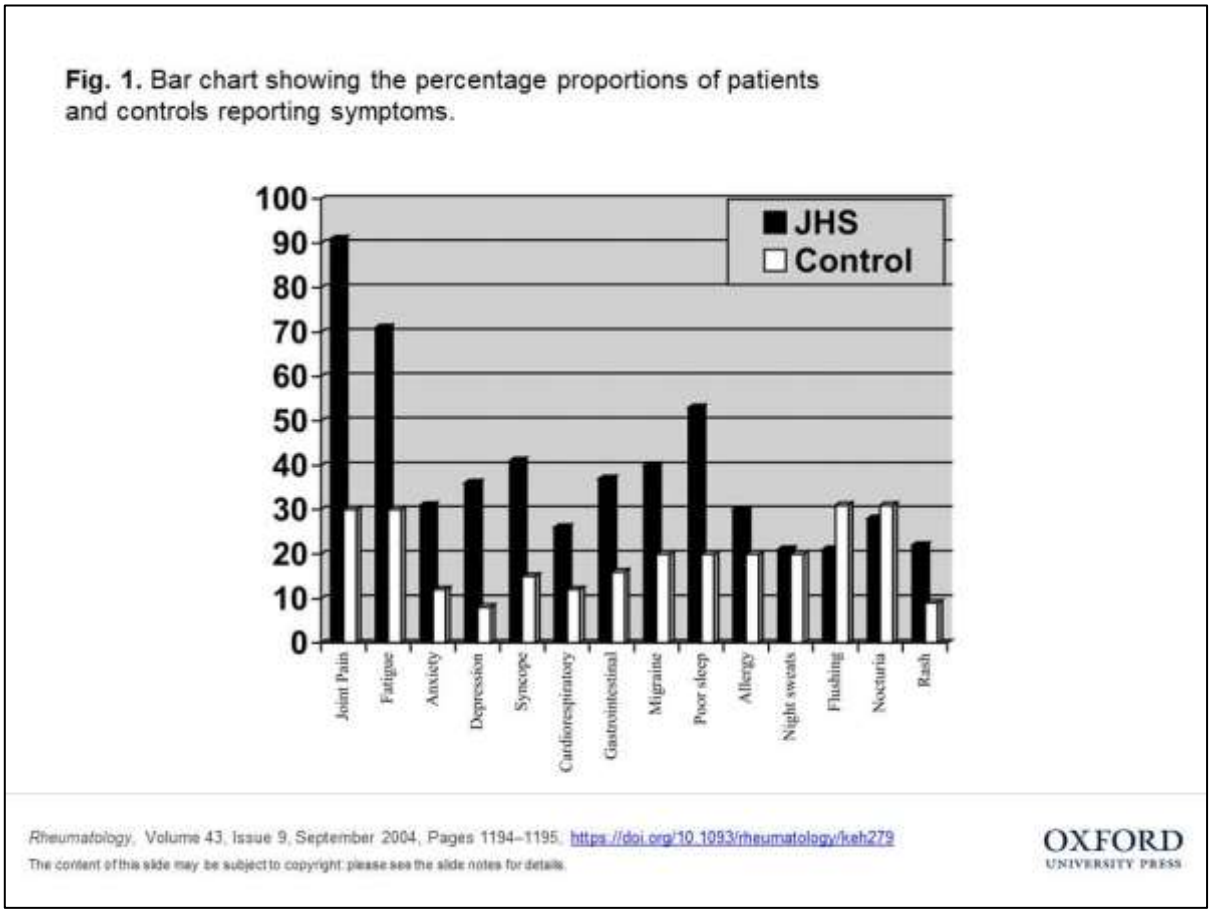
BACKGROUND

Hypermobility syndromes are long term health conditions in their own right but they are frequently associated with a wide range of comorbidities. As a result, multiple disciplines are involved in the diagnosis, treatment and management of people with hypermobility syndromes, across a range of primary, secondary and tertiary services, assisted by third sector organisations, such as the HMSA and other charities.

Symptoms experienced by this group of patients

The chart below outlines the widespread nature of the symptoms experienced by patients with joint hypermobility syndromes. There is a marked difference from the normal population. The situation is similar internationally.



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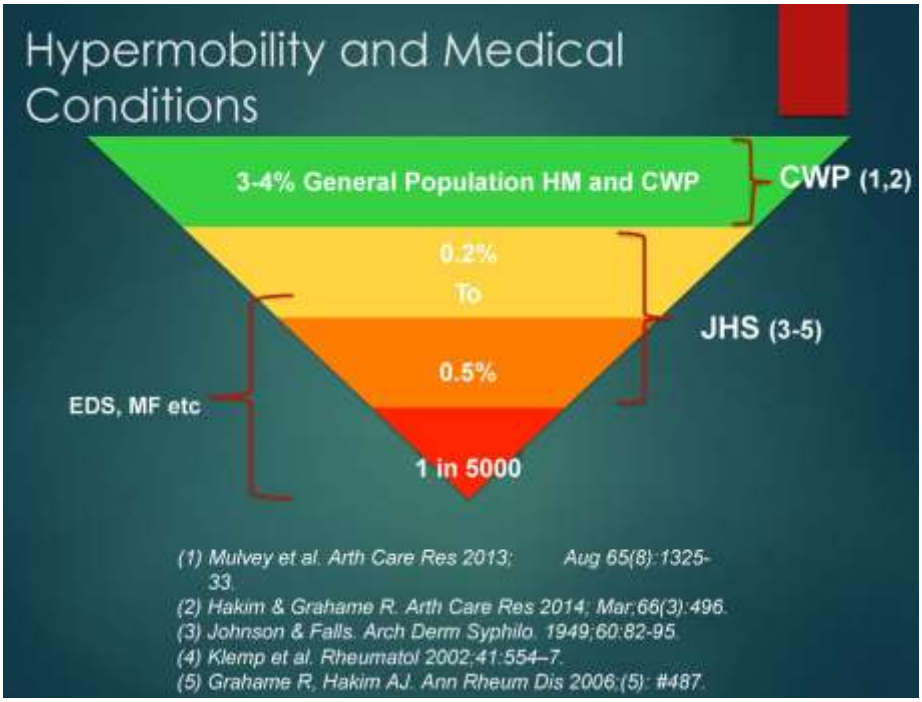
PREVALENCE AND IMPORTANT ASSOCIATION WITH OTHER CONDITIONS/ SYMPTOMS

Joint Hypermobility syndromes are associated with:

- Functional GI symptoms including diarrhoea and constipation.
- Fibromyalgia and fatigue syndromes

- Chronic widespread pain
- Postural orthostatic Tachycardia Syndrome
- Autonomic dysfunction
- Anxiety disorders and depression
- Irritable bladder
- Pelvis and pelvic floor problems
- Neurodevelopmental disorders, including ADHD and Autism
- Poor would healing
- Easy bruising

The inverted triangle below shows the evidence based prevalence for chronic widespread pain (CWP), Hypermobility syndrome HMS(under the old classification) and rare conditions such as Ehlers –Danlos and Marfan's. References included in slide.



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ASSESSMENT OF THE CURRENT SITUATION

Nationally, from a patient perspective, these services are often disjointed, with lack of service provider knowledge and no clear pathway available. Patients report that they feel disbelieved, frustrated and lack confidence in the professionals they see at appointments. 55% of the HMSA members who participated in a snapshot survey in 2012, reported that it took 10 years or longer to get a diagnosis. In the same survey, good care experiences were significantly improved when delivered by specialist services. This appears to be because of validation of symptoms and condition specific advice. This can actually be replicated locally with the right education, largely in primary care.

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Lack of early diagnosis and access to treatment has a significant impact on the level of functional ability, increasing psychosocial issues with a wider impact on family, friends, employers etc. Many patients find that they end up retiring on medical ill-health grounds or have to leave their employment because of worsening symptoms. Many younger patients struggle to stay in education and do not continue further education as planned. A significant number of patients with a hypermobility syndrome are unemployed, and on DWP benefits. With changes to the benefit system in the UK, many patients find they are under pressure to return to, or gain employment, despite having a lack of skills needed to manage their symptoms and awareness of what can be done to make employment or further education achievable.

This information has all been reinforced by the results of a recent BMJ paper studying Hypermobility in Wales.[1]

Key findings in this paper included that:

- A mean of 14 years elapses between the first clinical manifestations and the actual diagnosis.
- For 25% of patients, this delay lasts over 28 years.
- A misdiagnosis was given to 56% of patients resulting in inappropriate treatment in 70% of the patients
- For 86% of the patients, the delay in diagnosis was considered responsible for deleterious consequences."

In a nutshell, there is a significant lack of resources for teaching people how to live well with their hypermobility syndromes, the secondary specialists services are not joined up and often do not recognise the associated comorbidities, and there is a severe shortage of tertiary provision, all hampering effective condition management.

Incorporating third sector organisations into education services and service delivery can assist with meeting the needs of this population of patients without the need to refer out of area.

Current options open to GP's include the following

1. Referral to Rheumatology.
2. Referral to the Community Chronic Pain Service
3. Referral to University College Hospital National Hypermobility Unit (via Secondary care) – *currently not accepting referrals*
4. Referral to Stanmore's Hypermobility and Chronic Pain Service (via secondary care referral only)

[1] https://bmjopen.bmj.com/content/9/11/e031365)

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The Usual Experience (unsatisfactory!)

The overall effect of this is that, in relation to hypermobility syndromes, medical professionals are struggling to:

- diagnose patients with hypermobility syndromes
- know what to do with a patient after diagnosis
- give appropriate management advice in the time available
- give appropriate treatment
- know when to refer them to other services – and where to refer them

This in turn means further deterioration of symptoms pending appropriate diagnosis, treatment and management, leading to an even greater strain on the system:

- more GP appointments
- increased referrals to local services (primary, secondary and tertiary)
- increased referrals to 'out of area' services
- increased clinical investigations
- patients suffer negative outcomes as a result of late diagnosis

Given how common HSD and hEDS are believed to be, this cannot be a sustainable ‘normal pathway’ for all.

In addition, the GP may not have the knowledge and skills to confidently work with this patient group even after diagnosis has been made, and would benefit from access to advice and support from additional local professionals and patient charities like the HMSA.

It's time for this to change!

RECOMMENDATIONS

Endorse and implement the Kent model improving services through education using existing resources. Aims of the Kent Model

- To increase education and improve outcomes for people with hypermobility syndromes.
- To give access to appropriate treatment and management support to the population of people in the region who have hypermobility syndromes, and create confidence in the available support.
- Increase knowledge, awareness and resources for local medical, health and social care professionals.
- To build a network of professionals with an interest in working more closely with the charity in order to develop a better service.
- To reduce the number of people who would need to be referred ‘out of area’ and thereby reduce the cost to the NHS
- To build up an ‘HMSA Care Web’ of individuals, who would be willing to act as a local professional with sound basic understanding of the conditions.
- Undertake regular service evaluation.

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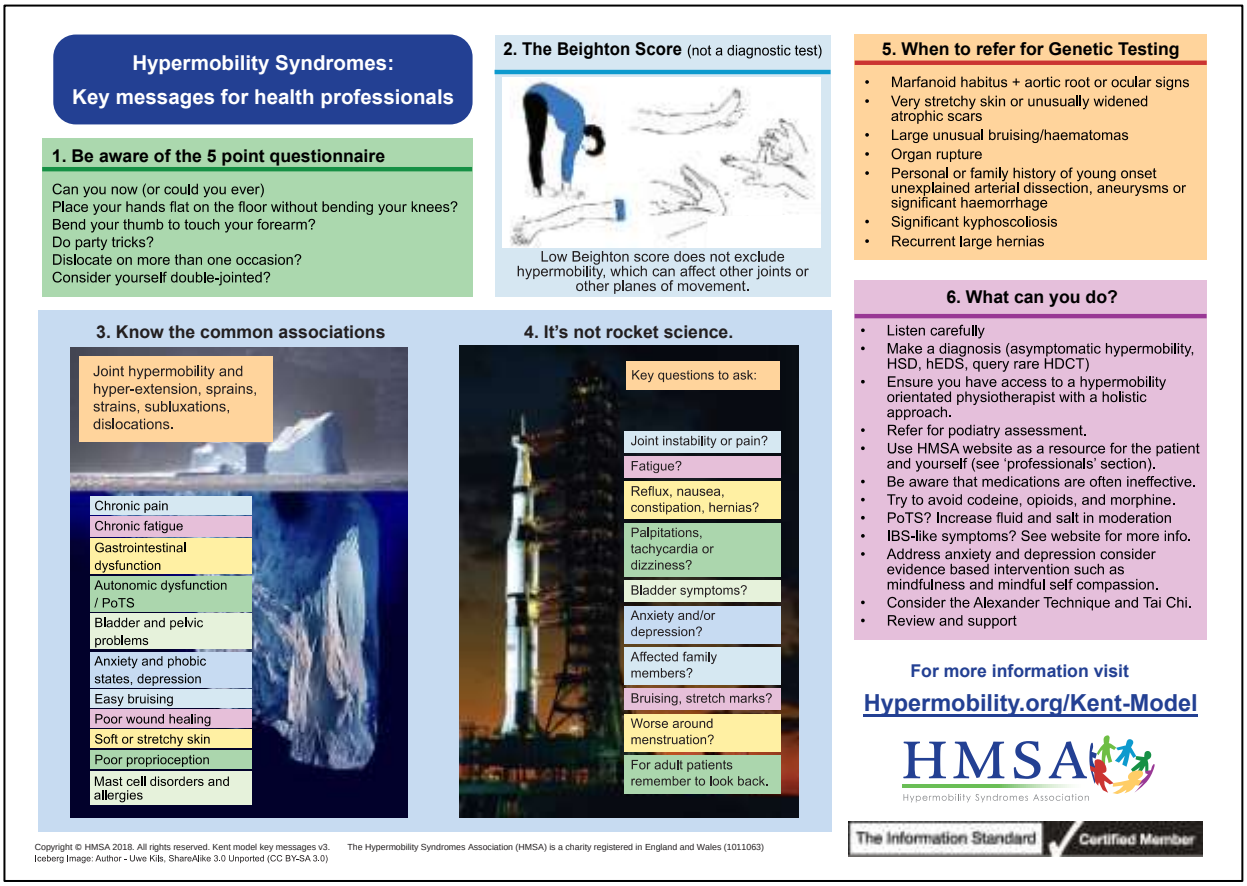
It is our belief that the vast majority of patients with a hypermobility syndrome can be diagnosed and managed IN PRIMARY CARE, with referral to secondary level only dependent on comorbidities and complexities, or to access various services such as physio, OT, podiatry etc.

For some people with either very complex issues or those with suspected rarer types of hypermobility syndrome, tertiary service input may be of benefit, and indeed be the correct means to proceed for the primary and secondary service professionals involved – the Kent Model resources will help professionals to recognise when referrals are needed.

The Kent Model aims to give professionals confidence in their skills and knowledge, as well as in the services they may refer the patient to, and simultaneously give patients the confidence that their needs have been understood, potential risks appraised and comorbidities recognised, leading to access to the best evidence based practice in treatment and management, either in their local area or in a few cases at recognised centres of excellence.

Much of the groundwork for this project has already been undertaken in East Kent through a series of Masterclasses and GP meetings using PLT sessions in Ashford, Canterbury and Folkestone. More than 300 GP's have received education and a similar number of health professionals including rheumatologists and physios.

The summary recommendations are combined into the 6 point handout (below) which is complementary to the HMSA webpages(link).which outlines the totality of the project and is linked to the extensive patient resources elsewhere on the website, which focuses on patient empowerment and self management:



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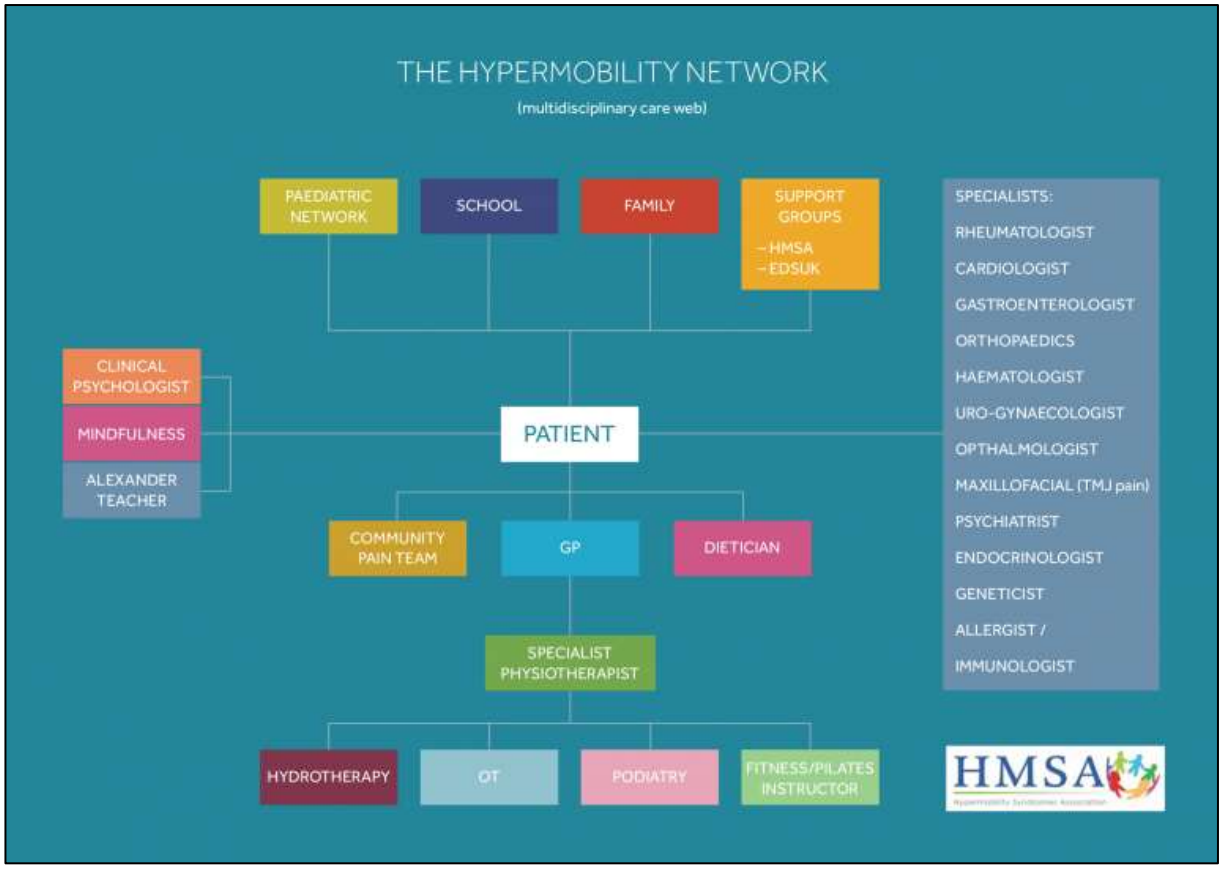
A CARE WEB has been outlined to emphasise the multidisciplinary nature of long term management for some patients.



ISSUES FOR LOCAL DISCUSSION

1. Mechanisms for education

Primary care Networks, for example, Practices are grouped into 16 PCN's in East Kent. These could form the platform for locally integrated services.

2. The Role of the local Rheumatologist

- Rheumatologists have a key role in identifying and excluding inflammatory joint disease.
- Hypermobility causes joint pain, mild joint swelling and stiffness.
- Patients with RA may have co existent hypermobility
- Acute presentations e.g.: PoTS, Gastrointestinal problems
- Awareness of MAST cell activation
- Need to identify and plan management for complex HSD/hEDS
- Identification of patients who need to be referred to specialist centres.
- Consideration of the role of the Rheumatology Nurse specialist regarding follow up
- Recognising joint hypermobility in patients attending early arthritis clinics

3. Development of physical therapy services and appropriate fitness regimes

Standard physiotherapy guidance may not work in this group of patients and can make things worse. Specialist physiotherapy techniques need to be more widely available though education. Dr Jane Simmonds and colleagues from the Institute for child Health associated with Great Ormond St Hospital run regular physiotherapy courses (including master classes with practical workshops).

Modified Pilates techniques available on line are currently being evaluated (Jeannie Di Bon).

4. Role of centres of excellence in London University College hospital and Stanmore

- University College Hospital provides a comprehensive diagnostic, investigative and rehabilitation service, accessible via secondary care. This service has limited capacity and is currently closed to referrals pending review and development of a more sustainable model. This is where the Kent Model helps with developing local services through education, freeing up capacity in London for the more complex cases.
- Stanmore provides a Nationally renowned pain management program accessible via secondary care. Key elements of this service could be reproduced locally through education.

Resources and references

- RCGP Spotlight toolkit on Ehlers-Danlos syndromes (https://www.rcgp.org.uk/clinical-and-research/resources/toolkits/ehlers-danlos-syndromes-toolkit.aspx)
- HMSA publications: The ‘living with hypermobility’ guide
- American Journal of Medical Genetics. March 2017. Special Edition
- HMSA Kent Model webpages

This document was created by Dr Philip Bull FRCP, Consultant Rheumatologist and Honorary Senior Lecturer with assistance from the HMSA team, including Donna Wicks and Hannah Ensor.

With thanks to Dr Alan Hakim, Consultant Rheumatologist. Dr Hanadi Kazkaz, Consultant Rheumatologist in charge of the Hypermobility Unit, UCH London and Dr Jane Simmonds, Consultant Physiotherapist at the Institute of Child Health Great Ormond St.