The Hypermobility Syndromes Association (HMSA) wishes to respond to its Members concerns following the 17th February 2017 publication of the The Ehlers-Danlos Society document entitled "2017 International Classification of EDS - Your Questions Answered".

In the following statement and information we aim to bring our Members as up-to-date as possible with the HMSA’s position, understanding, and the steps we need to take in continuing to support the hypermobility community.
• Foremost the HMSA is here to support the **whole** hypermobility community, regardless of diagnosis.

• This includes those with Hypermobility Spectrum Disorders (HSD), Ehlers-Danlos syndrome (EDS), Marfan syndrome, Stickler syndrome, Osteogenesis Imperfecta (OI), and rarer disorders.

• Any changes that arise from the 2017 Criteria for EDS do not change this fundamental principle at the heart of the HMSA’s vision and work.

• The HMSA will continue to work in a constructive way with other organisations and groups across the globe.
• We will continue to share our wealth of experience, providing the most up to date information and advice.

• We are aware that the 2017 Criteria for EDS is soon to be published, alongside a series of articles and guidelines related to disorders that affect many people with hypermobility-related disorders.

• One important aspect of the 2017 Criteria for EDS is that it will more clearly define a number of new variants of EDS that have been identified clinically and genetically over the decades since the 1997 Ville Franche Criteria.

• The HMSA will make every effort to reflect the information on new variants of EDS, and the changes in the terminology.
• Also the HMSA understands that the work being published through the International Consortium recognises, in detail, the breadth of different symptoms and disorders experienced by hypermobile individuals.

• We hope it will assist all in accessing the help they need, and the HMSA over the coming months, will be updating its own web information and new editions of our literature accordingly. This is a mammoth task but we will ensure the HMSA Information Standard accreditation is maintained.

• With regard to Hypermobile EDS, we also understand that a new Criteria has been developed and that only the term hEDS should be used.

• We also understand that if someone does not fulfill the criteria for hEDS or other more rare variants of this, or other syndromes, the term Hypermobility Spectrum Disorder (HSD) is now advised, and the term Joint Hypermobility Syndrome (JHS) is to be dropped.
• The HMSA has been working with people affected by what are now known as Hypermobility Spectrum Disorders for 25 years, and believes it is best placed to support people with this diagnosis.

• By sharing our knowledge and expertise we can best meet the needs of the HSD population.

• The HMSA will continue to raise awareness of the HSD population, lobby for better recognition, earlier diagnosis, and access to treatment.

• The HMSA is a driving force for this population.
• Until the full criteria, papers and guidelines are published, it is not possible to fully appreciate how the proposed changes might affect those who have already been diagnosed. Once these are made available we will be working with all our partners to determine how best we support anyone who thinks they are affected by this change.

• What we cannot endorse in any way is the commentary about which disorder is the more serious, severe, or deserving of care. We consider this divisive and completely unhelpful.

• The HMSA stands ‘with and for all’ hypermobile patients.
• Finally, we are aware of some of the issues that our Members may face in obtaining a diagnosis and treatment, due to the implications that the changes in the use of the terms HSD and hEDS may have on medical insurance. This is a serious issue which we intend to address in the near future.

• The HMSA will, in the coming weeks, offer advice and information for both people with a diagnosis, and the health/medical organisations’ charged with providing diagnosis, treatment and management regarding the needs of those with these conditions.

• The HMSA is uniquely placed to be able to meet both the patient and the professional populations needs.
• The HMSA will continue to be transparent and open to approaches from other organisations in regards to this work.

• The HMSA will welcome questions from Members of the HMSA and the wider hypermobility syndromes community, on any topic which they feel has not been addressed adequately or anything that they do not feel they understand.

• We will inform our Members of any potential changes to this Statement of Position.

Ms. Donna Wicks, CEO
Mr. Robert Studholme-Smith, Chair
Mrs. Claire Smith, Project and Partnership Director

Please send any correspondence to;

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In the next couple of pages Dr Hakim, our Chief Medical Advisor and a Trustee of the HMSA, describes briefly the thoughts behind the need for a change to the terminology.

We acknowledge that we are not able to cite the hEDS classification criteria until these have been published.

Expected date of publication is 15th March 2017
The key problem has been distinguishing JHS from Hypermobile EDS – we have described this and the publications relating to this in our HMSA literature before.

In July 2016 we shared the following thoughts. Look at the case examples below: using the Brighton Criteria for JHS Persons 2 to 6 could be given a diagnosis of JHS

**CASES**

Person 1: Hypermobile, Simple injury, Recovers Normally.
Person 2: Hypermobile, Joint pain, Nil else.
Person 3: Hypermobile, Joint pain, Soft tissue injury, Normal skin stretch, Mild scars.
Person 4: Hypermobile, Joint pain, Mild skin elasticity, Marfanoid shape.
Person 5: Hypermobile, Joint pain, Skin elasticity, Papyraceous Scar, Family history.
Person 6: Hypermobile, Joint pain, Skin elasticity, Scars, Joint subluxations, Family history.

**BUT, which of these individuals would also meet the 1997 Villefranche criteria for hEDS?**

As you travel from Left to Right in the examples above, it becomes more and more possible to consider Hypermobile EDS as the diagnosis. This is why there is confusion between JHS and hEDS, and why the terms are used interchangeably in some cases where a person meets both criteria.

**FROM**
Person 1: Who does not have hEDS

**TO**
Person 6: Who does have hEDS
So can we say that JHS is the same as hEDS? Yes in some cases, but surely not in all? JHS captures lots of different people with differing degrees of signs and symptoms. However to reduce confusion between whether someone has JHS or hEDS it was agreed internationally that JHS would be dropped and the term hEDS used. The Ehlers-Danlos Society made this public later in the summer last year. But…

i. **Question:** Where do we draw the line as to what hEDS is?  
**Answer:** by establishing a clear set of criteria for the diagnosis – the criteria based on the scientific evidence, expert opinion, and international agreement. This is what the consortium and The Ehlers-Danlos Society has done, and will be published shortly.

ii. **Question:** How do we describe the concerns of those that do not then have hEDS or a more rare condition, but have hypermobility-related issues?  
**Answer:** describe the fact that other people have hypermobility-related musculoskeletal concerns, and recognise that these too can appear on a spectrum of severity both in terms of the physical signs, severity of symptoms, and associated concerns such as fatigue etc. Give this a name that reflects that. This is described in the paper by Castori et al. Feb 2017 referenced at the end of this document.
The International Consortium with The Ehlers-Danlos Society has sought to answer these questions.

On the far Left the person is hypermobile and well with nothing else to find; on the Right of the dash line are people with hEDS as defined by the new criteria. To the Left of the dashed line i.e. those who do not meet the criteria (or have any other condition) we now use the term Hypermobility Spectrum Disorder (HSD) – people with their own sets of problems due to their hypermobility but who do not have hEDS.
So what about all the other things many individuals have – POTS, mast cell, bowel problems etc.?

• First, it is reassuring to know that all the key areas have been looked at by many experts and lay people from across the world within the International Consortium. Guidelines are being produced that describe how all of them may be associated with EDS, and how they should be assessed and treated.

• Second, there is evidence of association with EDS and a number of disorders, but what has not yet been shown for most of them is whether these are *directly caused* by having hEDS. Also, for a number of them it has not yet been shown exactly how common they are in the whole hEDS population compared to the general population, or other groups of people with chronic medical conditions.
• The HMSA understands that the guidelines being published will, in part, describe the research needed to enhance our understanding of the relationship between these disorders and hEDS. We are able to cite 2 of them in the references by way of example as these have been published
A final comment:

The HMSA understands that all the work of the consortium will be published on or around the 15th March 2017. However several papers are already available online through the American Journal of Medical Genetics and might help our Members understand more fully the comments made above. Unfortunately the HMSA is not able to disseminate these - a standard copyright rule that applies to all the Journals’ publications.


