Hypermobility: national feedback

Page 1-8: Main themes we've heard from patients.

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Introduction:

After Healthwatch Calderdale's work into hypermobility syndromes was raised locally, nationally via an adjournment debate in the House of Commons and on social media nationally, we have been contacted directly by people across the country sharing their own experiences of NHS care for hypermobility syndromes.

While we cannot represent people individually outside of the Calderdale area, we decided to collate the national feedback as an update to our work as it shows the themes we discovered in our work in Yorkshire and the Humber are being experienced in other parts of the country.

The detail below comes from direct contact from people since autumn 2019. It represents views of patients in Essex, Oxfordshire, Teesside, Brighton, West Sussex, Hartlepool, East Yorkshire, Kingston, Warwickshire, London, Sunderland, North West, South West, London, Kent, Humberside and York.
We have further been contacted on social media from people in Liverpool, Cheshire, Manchester, County Durham, Middlesbrough, Redcar, Stockton, Rochdale, Herefordshire and Plymouth, in addition to the above named places.

A number of comments come from members of the Ehlers-Danlos Teesside patient-led group who have shared with us the speech they gave to the Scrutiny Panel local to them.

Main themes from national contacts:

- Why are local Healthwatch organisations elsewhere are unable to explore hypermobility?
- GP lack of knowledge of condition
- Difficulties obtaining referrals
- Misdiagnosis
- Poor access to care
- Negative impact on wider health
- Private assessments
- Impact on life
- Lack of joined-up support for children
- Ideas

Local Healthwatch unable to explore hypermobility:

Warwickshire: One person contacting us asked her local Healthwatch in the Midlands to explore the issues around hypermobility. She was told staff did not have the resources to explore it, either independently or with other Healthwatch local to them.
She was told: "…*We do not think the work needs to be duplicated in (Healthwatch area) as Healthwatch Calderdale have worked in partnership with 15 other Healthwatch and written a very informative report. With their permission we will use this report to influence commissioners in the West Midlands."*

Another person from the South West told us: *"I can say without a shadow of a doubt that your local findings are mirrored elsewhere in the country. The themes highlighted in your reports are ones I hear on a daily basis and have experienced myself."*

GPs lack of knowledge of condition:

North West: One woman told us that her GP talked about mental health when she first presented her symptoms. She struggled on for months experiencing much pain, before asking for a second opinion due to her experience the first time. On the second occasion, her mental health was once again explored ahead of her presenting symptoms.

Sunderland: *"I have had several different GPs who have left my GP practice and I currently do not have one who is familiar with my condition".*

South London: *"There is an absolute lack of training and knowledge on this condition so unless the patient studies it they are unlikely to get diagnosed and even less likely to access proper treatment and service. I was dismissed several times by my GP for problems related to my hypermobility, and for some I still have not been able to access treatment".*

York: "I'm lucky that I have always had GPs that believed in the symptoms and when I was younger my GP was very supportive in trying to help – although he didn't know much about the condition."

Humberside: "Doctors not understanding or connecting symptoms, advising things when presenting with pain, like "come back in 6 months", "buy a splint" etc." surgeon only interested in the specific joint, no holistic care, it is still a disjointed service."

East Sussex: "GP had not heard of it. Persuaded him to refer me to London EDS clinic. They tested my flexibility and failed me as I was too stiff, in spite of being a 58year old male and having clearly hyper mobile hands, stretchy skin, a hernia and high palette/dental issues.
POTS: Three GP's had not heard of it. Had tilt table test and confirmed I had POTs. No treatment offered. Specialist said "we don't know much about it"
Mast Cell: Not one of several GP's or specialists believed in it or had heard of it. No treatment offered.

Teesside 1: “I spent my whole life in and out of doctor’s offices. I’ve always been met with a lot of stumped medical professionals and I felt like I had to take control of the situation myself.”

Teesside 2: ‘She’s just fussy,’ – when I couldn’t eat due to nausea and stomach cramps after eating so I would refuse all foods as a 2-year-old child, understandably causing immeasurable stress to my parents.

Teesside 3: “I was abnormal as a child and saw many, many, health professionals. Not one picked up on my hypermobility. I have a life limiting condition that isn’t understood. I wish to live in a world where everyone understood. Pain, always in pain, and doctors not understanding why.”

Teesside 4: “A common response is, "oh you're a bit bendy, yeah?". Which conveniently ignores the various other symptoms that have spanned the last 20 years and seen me ushered into various consultants, wards, doctors, surgeries, nurses, specialists, prescriptions, all while trying to alleviate a single symptom while my health declines further.”

Difficulties obtaining a referral:

South London: “*I started having symptoms in my teens which were put down as growing pains or just dismissed. I've been trying to find what was wrong with me for years and I have countless medical appointments and test behind me. As it is often the case, I felt I had to become "my own doctor", as I was constantly dismissed, not believed, told to get on, that it was just anxiety or all in my head/psychosomatic (as if by telling me that, magically, it would just disappear.”*

South West: One woman was in her 40s by the time she got a diagnosis which fitted her symptoms. She described her experience as *“40 years of completely incorrect treatment”* adding *“prior to diagnosis I was treated as though I was a hypochondriac (even though I reported the same issues repeatedly - pain, dislocations, fatigue, gastro issues etc.) and repeatedly fobbed off with ‘you must be stressed/depressed’.”*

North West: one person fought for a referral to a neurologist, then later found the clinician had put on the referral: "obvious self-harm scars on left arm" – scars which were decades old. She waited over 12 months to see the neurologist and was admitted to the Accident and Emergency Department prior to that due to her condition. Once she did see the neurologist her symptoms were taken seriously, but she felt the pathway to getting there was challenging. She was also “accidentally” discharged from neurology so had to be re-referred and wait another 9 months.

South London: *“What I wanted investigated was mast cell activation syndrome (MCAS), and I found out from Facebook that the Ehlers Danlos syndrome (EDS) toolkit for general practitioners (GPs) mentioned MCAS. I had never heard of EDS before, but reading it made instantly so much sense. I went to see my GP and told*

him I wanted to be evaluated for EDS. He wasn't convinced at all, but he did test my hypermobility and agreed to refer me. He tried to refer me to University College London Hospitals (UCLH) but it was refused as I was out of area and the specialist service is a tertiary referral."

Misdiagnosis:
North West: *"One clinician logged an ailment on the system as the patient having "a funny turn".*

North West: GP told one woman her magnetic resonance imaging (MRI) scan was normal, but having learnt about hypermobility & craniocervical instability 18 months later, the patient requested a copy of her MRI and saw it was abnormal in various ways, plus her brainstem was being compressed, which was missed.

Teesside1: "Why did it take 28 years for a health professional to recognise me? I can't afford for me or my kids to get a private diagnosis, which is devastating, otherwise I would've been diagnosed a very long time ago."

Teesside 2: "I've been asking for EDS to be considered and investigations to be made for several years, with no success and very little support from anywhere within the NHS. I am currently still fighting for a diagnosis. Which I have been fighting for since 2015."

Teesside 3: One patient says there are patients who have been forcibly sectioned by a local gastro doctor because they cannot eat due to gastroparesis (a common co-morbid condition that paralyses the stomach). This Doctor removed their feeding tubes and deemed them anorexic. They have also placed a new style feeding tube into one Teesside member and ended up breaking all their ribs.

Essex: "…at that hospital they were adamant that my pain was simply an injury that had 'healed' and my pain was due to pain gates still being open, and my 'fear' of moving, I argued that this was not the case. I was right they were wrong but too blinkered to see what was starring them in the face… they only saw what they wanted to see. It was 25 years ago and it still angers me now."

Poor access to care:
South West: Getting care/treatment after a diagnosis wasn't straightforward for one woman, who said: *"There is no local specialist service and consequently because of the issues I was facing and negative outcomes from previous inappropriate treatment I was referred to London."* She said the London-based specialist is overwhelmed with referrals so are unable to take on more patients.

North West: a woman "pleaded" for physiotherapy for her neck and shoulders to manage the pain and keep the muscle strength, but none was received.

Sunderland: *"I am constantly in pain, have digestive and neurological issues that have not been investigated, cannot stand without pronounced heart rate increases causing dizziness, vision blackouts and possible fainting, then overheating,*

flushing and sickness which can only be relieved by lying down, and severe fatigue".

"If I attempt to "push" through my symptoms, my immune system weakens causing me to catch viruses and infections, often multiple at one time, which exacerbates my other conditions and becomes very difficult to recover from. I feel as if I have exhausted every avenue for seeking help with my conditions, many of which have not been monitored at all since diagnosis."

South London: "I have been on physiotherapy services which made me worse. I have gotten worse since my diagnosis but cannot access any physiotherapy nor specialist services as my GP feels I've had "too many".

Teesside: "Based on the experiences of others who also have the condition I don't feel it beneficial to seek or pursue an official diagnosis given the lack of pathways and patient care available. Instead I just manage the symptoms unsupported by the NHS, which has included repeated dislocations as a child and currently includes muscle wastage and severe chronic pain."

"A proper care pathway is so desperately needed for sufferers of people with Ehlers-Danlos Syndromes and hypermobility spectrum disorders because right now it's fragmented at best and quite frankly is terrible. Even things like counselling aren't offered and we have so much to deal with its overwhelming to say the least. If we had a proper care pathway people would be diagnosed earlier instead of being sent for individual problems to be dealt with such as referral for knee problem, referral for back problem and so on. When it's fragmented like this it's not picked up on quick enough leading to us suffering more and for longer and this in turn is a bigger burden on the NHS."

Essex: "I naively thought that once I got the EDS diagnosis, getting access to the correct treatment would be straightforward, but this has simply not been the case. Everything is a battle; most medical professionals are ignorant about EDS and hypermobility. There is no joined up thinking, I have to get referred here there and everywhere, wait months to see anyone in hope that I will be lucky enough to see someone who knows something about this condition. My GP is sympathetic, but also does not know what best to do to try to help me, and I am becoming more and more exhausted and jaded by it all."

Negative impact on wider health:

North East: One woman told us that even after her diagnosis, any other medical issue is then "assumed" to be linked to her hypermobility. The woman said: "*This is poor and shoddy care and further upsets the feelings of sufferers who frankly in the majority rather than minority of cases have been fobbed off their entire lives and treated like hypochondriacs. This is a national scandal and absolutely needs to change.*"

Sunderland: Individual was told that because she was assumed to have Hypermobile EDS, a life-threatening condition, which she also has, is not being explored with genetic testing.

Private assessments:

North West: due to poor feedback about NHS care, one woman was told she would have to be seen out of area, which would unlikely to be funded, so she faces paying privately for tests. She expects the initial cost to be £1,650.

Essex: In spite of many years of what I can now see are typical hypermobile EDS symptoms, I was not diagnosed until (patient was in 50s), and only then because of a chance meeting with someone with EDS and coincidentally a passing remark from a pain management consultant (private) when I was concerned about continuing symptoms in spite of treatment... "well you are hypermobile after all..." to which I replied "Am I?". I consequently sought diagnosis through a private rheumatologist, fortunately we had medical insurance.

Essex: "I have been getting some help from an osteopath, not available from the NHS of course, I have to pay for it. The osteopath is positive I have scoliosis, but this again has not been picked up by any NHS doctors or physios. When I mentioned it to one of the junior NHS physio he dismissed the idea, saying that he very much doubted it as it "would have been picked up by now"."

Impact on life:

Sunderland: one woman has been unable to work and was told by a postural orthostatic tachycardia syndrome (POTS) specialist physiotherapist to use a wheelchair at all times outside. She receives Personal Independence Payment (PIP) at the lower level of daily living allowance but do not receive anything for the mobility component. She receives no other benefits.

North West: another individual, who sought advice outside of the NHS, was able to manage her pain to the extend she has less seizures and is able to go outside daily, something she was unable to do prior. She can walk once a fortnight, prior to this she hadn't walked in a year.

York: *"I spent most of my life following the boom & bust model - and wondering why I couldn't do as much as I wanted to! But at that time, as I worked freelance, I could still manage to do a job that I can no longer do, provided that I took lots of time off between jobs. But a lot of the time I felt exhausted and tired as well as in increasing pain round a large amount of my body."*

Teesside: The dose and the volume of medications made once patient so unwell it caused a bowel obstruction. The same patient had to give up studying half way through second year because of their condition not being managed.

Another patient told us: ***"I'm tired and I don't know how to fight the system anymore. My house is inaccessible to me. My wheelchair is inaccessible to me. I have no independence anymore. I have to rely on others, and I hate it."***

Essex: "I had suffered many, many years of pain, fatigue etc. I had chronic fatigue in the 1980s and had thorough investigations for RSI problems in the early 1990s, as a result of which I lost my job, I have never worked full time since."

Lack of joined-up support for children:

Warwickshire: One mother says she struggled to access joined up healthcare and a clear referral pathway for her teenage son. She also says they struggled to access social care and mental health support, plus the Education, Health and Care Plan (EHCP) was not "fit for purpose" and her child was missing education due to lack of support as a result of unsupported needs.

South West: a mother, who has hypermobility herself, found it hard to get a diagnosis and care for her children when they were displaying symptoms. She said they were "passed from pillar to post" adding *"Diagnosis needs to happen early to mitigate cumulative effects of the condition."*

"My middle son is also showing many signs and when I brought this up with his paediatrician, he has since been discharged because he didn't have a clue what it was."

Ideas:

People called for a *"better treatment pathway within the NHS".*

One person said: *"As care and symptom management, ideally, calls for such a multidisciplinary approach, it seems to me that it is unclear to GPs and consultants who should be managing and collating this care. A better system of handover between GPs would also be very useful, as it's very difficult at the moment to find a GP, especially one familiar with EDS, who remains at one practice for any length of time."*

Essex: "It has to change. We need multi-disciplinary EDS clinics to help us with our various problems. So many people are suffering like I am and the NHS is doing little to help us."

Another felt regional centres of excellence would be a useful idea: "It would be good to have a dedicated clinic for people with a hypermobility syndrome in the same way as there are diabetes clinics."

The following comments all came from a presentation presented to Teesside Scrutiny Panel: "I would like to see children of people with EDS automatically screened by knowledgeable professionals for the condition."

"There should be a standardised pathway for all EDS/HSD patients. That way both medical professionals and patients know where they stand.

"We have a very long way to go but the first step to that will be making our health professionals understand, be trained to treat us effectively, and be diagnosed quickly to enable us to get the correct care."

"A proper care pathway is so desperately needed for sufferers of people with Ehlers-Danlos Syndromes and hypermobility spectrum disorders because right now

it's fragmented at best and quite frankly is terrible. If we had a proper care pathway people would be diagnosed earlier instead of being sent for individual problems to be dealt with such as referral for knee problem, referral for back problem and so on. When it's fragmented like this it's not picked up on quick enough leading to us suffering more and for longer and this in turn is a bigger burden on the NHS."

The Ehlers-Danlos Society have set up a program called 'Project ECHO', where leading clinicians and patients from all over the world can video chat online and information share.

January 2020 update: case studies of 2 personal experiences to obtain a diagnosis and treatment/support.

Brighton: Three years ago T's journey to diagnosis began. Here T shares his journey with us:

After many appointments with 2 different GPs regarding chest pains and breathing issues I was having, I was sent to be assessed by a Cardiologist who noticed how loose/elastic my skin. This led him to raise the possibility that I may have EDS. He explained he would refer me to a Rheumatologist at The Royal Sussex County Hospital in Brighton.

After the initial referral was made in January 2018, I was seen in March/April 2018 where the Doctor did concur that that there was the possibility I had EDS but kept repeatedly telling me that she had seen worse cases than mine.

Despite those comments the doctor did refer me to Guys and St Thomas in London for referral who promptly wrote back informing me that I was not currently sick enough to be seen by them.

I was re-referred back to Brighton for a follow up appointment. At this appointment the doctor examined me, ticking the necessary boxes, asking relevant questions etc. eventually reaching the diagnosis. The doctor informed me that she would be seeing me again in 3-4 months' time to monitor my condition etc. but in the mean time I would be referred to:

- Gastroenterology for further research into my weight loss/stomach issues;
- Occupational therapy for my hands/legs as well as pain management;
- Referred to UCL for Genetics test/to be seen by a specialist.

After 3 long years for being told I'm not sick enough, that there is nothing really wrong with me etc. I was finally feeling a little more positive but the positivity did not last long.

At the follow up appointment despite informing the Doctor I saw about pains, difficulties walking, and a change in my gait etc. I was discharged

from the department as I had not yet experienced dislocations of any nature.
The pain management team simply informed me that painkillers don't affect EDS so it's all about training my mind to ignore the pain.

The physio/OT for my hands was great and very helpful however I had to wait 45 weeks to get an appointment to be seen by a separate physiotherapist for my legs. After 3 separate sessions he discharged me and felt it was best if I self-managed what I can do and when I'm able to do them.

While this was all going on I was also seen by cardiology and respiratory departments to try and find the cause of my ongoing breathlessness, dizziness and chest pains, as well as the possibility my EDS had led to me developing POT Syndrome.

It was recommended by Physiotherapy, Cardiology, Respiratory and my O/T that for both my physical as well as mental well-being that I did in fact need to be referred to the UCL. The doctor sent off the UCL referral letter only to find out that they are not accepting any more referrals until August next year.

I do understand that there are people out there with conditions much worse than me but this is my life. It might not matter much to them but it matters to me.

I also found it very disconcerting when I have been seen by medical experts I have had to explain what EDS is as they have never heard of it. One doctor Goggled the condition and its symptoms in front of me as he had never heard of it.

I have actually reached the stage where until I end up in hospital I will not be revisiting my GP or anyone as it feels like a waste of time. I know there is no cure for EDS but a little professional help or belief would be nice in the meantime.

Oxfordshire, Coventry and Birmingham: patient with experiences of NHS in 3 areas of England:

I was diagnosed aged 27 despite being symptomatic since the day I was born.
I was born with dislocated hips and had frequent problems with them. It wasn't picked up by GPs or early years specialists.

I was also extremely ill my entire life, my immune system was non-existent, for example I had whooping cough at least 3 times despite all my vaccinations. I continued to have no energy, no stamina, no physical ability in terms of muscle tone at school, I was so bendy and floppy. Like a sack of tissue with nothing holding me up.

I was frequently at hospital for joint dislocations and ligament injuries from minor accidents after being forced to engage in the PE classes at school, especially running which physically disabled me every time. I always had tonsillitis - pretty much every 6 weeks - I was never offered a tonsillectomy. I was on antibiotics easily half the year since I was born.

When the NHS dentist visits began, it became clear I was totally resistant to anaesthesia - but I was told I was making a fuss/an anxious child and eventually began to accept fillings, tooth extractions and brace orthodontist appointments being able to feel every inch of pain.

As a young adult I developed Pericarditis and Intracranial hypertension. I was fobbed off suffering in total agony and hell for 2 years until I lost the use of my left side of my body and was admitted to hospital.
The NHS team at the hospital were mainly trainee doctors so many attempts at a lumbar puncture went wrong. (Also being anaesthetic resistant the pain was astonishing) When I eventually had one I developed a huge CSF leak (very common in hEDS) I was told my extreme symptoms were depression, diagnosed with IIH and told to lose weight. I leaked so hard for so long I would have seizure episodes if I lifted my head even half an inch from the ground. The pain was excruciating.

I suffered a miscarriage with many complications resulting in 2 operations.

All this time I continued to ask doctors (mainly my GP) to look at the big picture; why am I so sickly? Why is my B12 low? Why am I deficient in everything on blood tests? Why can't I thrive despite being actively healthy? Why do I have so many variations of illnesses? Why don't I get a proper fever when I have severe infections (sepsis after the miscarriage) why is my hair falling out? Why am I so flexible and why can't I build muscle? Nothing ever came of it.

Two years ago, I was struck down by extreme reactive arthritis after acquiring an infection. I was tested for everything from Lupus to rheumatoid arthritis. I was eventually referred to a rheumatologist a year later. The doctor there was shocked I hadn't had aspirations of my joints and treated for reactive arthritis. During that time I thought I would die, the symptoms and the pain were so bad. I was in a sleep like state, shivering, with blue lips for almost 3 months.

I was then sent further to a geneticist and another rheumatologist who both diagnosed hEDS and every single thing I had suffered became clear and they were nice and helpful.

I now have severe cervical spine instability, I am partially paralysed from the neck down, and have lost my entire life, career.
I am seeing a surgeon next week who will fuse my neck and skull together - but we will have to somehow raise funds for that because although the NHS neurosurgeons have seen the state of my neck and symptoms, they cannot and will not operate on people with a hypermobilty diagnosis - because the evidence and studies aren't there (apparently), despite there being literally thousands of us in this situation.

I pray future hypermobility generations get doctors who actually understand these

conditions affects so many (people) and causes so many other ‘illnesses’ that you cannot look at one thing separately.

I never had “so many” problems, I had one condition which manifested in all these different ways and with the right knowledge and treatment I know for certain I would not be in the position I am today.