

**THE ROLE OF NARRATIVE MEDICINE IN THE MANAGEMENT OF GASTROINTESTINAL DISORDERS AND JOINT HYPERMOBILITY SYNDROME/ HYPERMOBILE EHLERS-DANLOS SYNDROME.**

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**ABSTRACT**

Joint hypermobility syndrome/Hypermobile Ehlers-Danlos Syndrome (JHS/hEDS) is a heritable connective tissue disorder affecting virtually every bodily system [Grahame, 2013; Castori et al, 2012]. Over the past 18 years there has been an explosion in the literature relating JHS/hEDS to gastrointestinal symptoms [Castori et al, 2015], with patients enduring symptoms from mouth to anus [Castori et al, 2015]. It would seem that the defective connective tissue and tissue laxity in JHS/hEDS causes GI symptoms in this patient cohort [Castori et al, 2015; Farmer & Aziz, 2010; Firkee et al, 2014; Zarate et al, 2010]. This leads to considerable stress in patients with JHS/hEDS, and a long catalogue of problems for which the need for narrative medicine becomes invaluable [Knight, 2015]. In this article, narrative medicine, where science meets humanities [Charon, 2006], is used as a way for the patient story to be seen and heard at the helm of a long and complex history of GI problems in relation to having JHS/hEDS, including highlighting awareness of the condition Anterior Cutaneous Nerve Entrapment Syndrome [ACNES] [Applegate, 2002, Koop et al, 2016].

**KEYWORDS:** Gastrointestinal problems, Ehlers-Danlos syndrome, joint hypermobility syndrome, narrative medicine

## **INTRODUCTION**

Joint Hypermobility Syndrome/Hypermobility Ehlers-Danlos Syndrome [JHS/hEDS] is now considered to be a multisystemic heritable disorder of connective tissue, which may be seen to affect most, if not all bodily systems [Castori, 2012; Grahame, 2013; Knight, 2013; Murray et al, 2013]. It is now becoming recognized and is diagnosed on the basis of clinical criteria such as the Brighton Criteria and Villefranche Criteria [Beighton et al, 1998]. Unfortunately there is a lack of true biological or genetic marker at the present time, despite ongoing research [Grahame, 2013]. The impact of Joint Hypermobility Syndrome/Hypermobility Ehlers-Danlos Syndrome goes well beyond having joints with a greater than normal range of movement, and associated skin manifestations [Castori et al; 2012; Murray et al, 2013; Tinkle, 2010]. A study by Murray et al [2013] involved an online survey including questions about pain and depression. 466 adults [90% female, and 10% male] answered the survey and in addition to reporting joint pain and hypermobility [99% respectively], also reported other symptoms including chronic fatigue [82%] and [96%] experiencing gastrointestinal (GI) symptoms. The current manuscript focuses on GI symptoms and JHS/hEDS. Since [at the time this manuscript was written], JHS and hEDS were considered by most experts to be the same conditions, the term hEDS will be used for the rest of the manuscript.

## **JOINT HYPERMOBILITY SYNDROME/EHLERS-DANLOS SYNDROME – HYPERMOBILITY TYPE AND GASTROINTESTINAL SYMPTOMS**

There has been an explosion in the literature on the topic of hEDS and gastrointestinal disorders, some 16 papers in the last 18 years [1987-2014 [Castori et al, 2015]. The prevalence of gastrointestinal disorders, such as Irritable Bowel Syndrome (IBS) were first noticed coincidentally in a Neurogastrointestinal Clinic by Zarate et al [2010]. By using the validated 5-Point Score used to diagnose generalised hypermobility [Hakim & Grahame, 2004], it was noted that patients presenting to a tertiary Neurogastroenterology clinic had a high prevalence of hypermobility [49%]. A rheumatologist further identified 17/25 [68%] as having symptomatic hEDS. Of those with hEDS and GI symptoms, 81% has abdominal pain 57% nausea, 48% reflux, and 38% constipation [Zarate et al, 2010].

Castori [2010], suggests that there is an 86% prevalence for GI problems in those with hEDS, with other studies considering it to be even higher than that [Murray, 2013]. Patients experience symptoms from the whole GI tract from mouth to anus, with problems including swallowing, reflux, IBS to chronic constipation and organ prolapses [Castori et al, 2015]. Very often patients will have a combination of several, possibly the full-range of GI problems [Castori et al, 2015; Farmer &

Aziz, 2010; Firkee et al, 2014; Zarate et al, 2010]. The symptoms can be very debilitating, and cause extreme distress [Castori et al, 2015; Firkee et al 2014].

## **NARRATIVE MEDICINE**

*“A patient saw a physician for the first time. The physician wanted to learn everything about the new patient, and listened attentively without interruption. The patient paused after a while and wept. When asked why, “No one ever let me do this before” was the response.” [Johna & Rahman, 2011, p.92].*

There is something fundamentally human about telling and sharing stories, in order to gain insight and understanding of another person's world. It is what we do every day. People have stories to tell, and want them to be heard [Adler, 2002; Hatem & Rider, 2004; Nowaczyk, 2012]. Charon [2007] writes that she first used the term 'narrative medicine' in 2000. She writes *“to refer to clinical practice fortified by narrative competence – the capacity to recognize, absorb, metabolize, interpret, and be moved by stories of illness. Simply, it is medicine practised by someone who knows what to do with stories”* [Charon, 2007]. Narrative medicine is where science meets humanities [Charon, 2006]. Simply put, the skills of narrative medicine, if employed readily by the medical profession, is invaluable – particularly in working with chronic and complex patients as those with HEDS. This is because these patients are frequently dismissed and many 'disbelieved' because of their myriad of symptoms, or because they appear 'difficult' [Bulbena et al., 2004; Baeza-Velasco et al., 2011; Grahame, 2013; Knight, 2015; Knight 2013; Shapiro & Ross, 2002]. It is therefore vital that they feel 'heard'. This, according to Charon, [2001] will not actually save consultation time, but will help the physician to understand the patient's plight and critically help patients to gain trust of the physician [Charon; 2001; Hydén; 1997; Johna & Rahman, 2011; Knight, 2013; Shapiro & Ross, 2002].

Charon, as cited in Peterkin (2012) begins her consultations by asking, *“What would you like me know about you?”* Peterkin's article makes some excellent recommendations for physicians including not interrupting the patient when they are speaking, and asking the patient to write a 1 page “impact of my illness” document that they bring with them to their consultation [Peterkin, 2012]. This makes great sense, and is something that I encourage my own clients to do in my role as a [complementary health] Bowen Technique practitioner. I also suggest my clients do this for appointments with other physicians. It is another form of use of narrative, and the feedback I receive (anecdotally) from such physicians as it not only saves time, but helps the patient to clarify their own story and the information that they want to impart, and how their illness affects them directly [Adler, 2002; Harter & Bochner, 2009; Peterkin, 2012].

The power of listening to the story should never be underestimated, as in Johna & Rahman (2011) patient's response of weeping [2011 p.92]. From my experience as a patient, it is the being understood and heard that is the most important part of the consultation. Indeed Kalitzkus & Matthiessen [2009] says, and I agree, that it can be a very healing experience both in listening to the stories of others and in telling one's own [Kalitzkus & Matthiessen, 2009]. Furthermore, Charon [2012] and Divinsky, [2007] also suggests that evidence is accruing that narrative medicine may help in reversal of burn-out in clinicians [Charon, 2012; Divinsky, 2007] – something that is important for physicians to consider who work with such a complex patient cohort as the hEDS patient [Lucas from Knight, 2013].

The following account is my own narrative medicine account of my personal journey in 'suffering' or experiencing gastrointestinal problems alongside my hEDS. I have done this in order that other physicians might understand and learn about the complexity of my story. As far as I am aware this is the first narrative medicine account on the specifics of gastrointestinal disorders and hEDS barring Knight, [2015], which was an article about the broader spectrum of the effects of hEDS.

### **MY JOURNEY WITH HYPERMOBILITY EHLERS-DANLOS SYNDROME AND GASTROINTESTINAL PROBLEMS.**

My symptoms began as early as 11 years old. My family practitioner said that I was suffering from colic, and that it was probably caused by growth spurt, along with my growing pains, also common in those with hEDS [Viswanathan & Khubchandani, 2008]. I was prescribed an anti-spasmodic drug, and the symptoms did seem to abate. At one point on a family vacation, it was questioned whether I might have appendicitis, owing to the severity of symptoms in the right iliac fossa. This was dismissed, and menstrual symptoms also ruled out.

Throughout my teenage years, my Gastrointestinal Symptoms (GI) progressed, worsening around menstruation, which, in my early twenties tallied with a diagnosis of endometriosis, diagnosed formally by laparoscopy, the gold standard measure of diagnosis [Kennedy and Gazvani, 2000]. The symptoms of endometriosis also often overlap GI symptoms, including Irritable Bowel Syndrome -IBS-type symptoms [Evans, 2005; Hamilton-Fairley, 2004; Henderson & Wood, 2000, Mears, 1996]. Although endometriosis is not yet clinically proven to be linked to hEDS, it does appear to be more common in those with hEDS [Tinkle, 2010]. A study by McIntosh et al [1995] involving 41 adult women with EDS, of which 27% had endometriosis [McIntosh et al, 1995]. A study by Hurst et al [2014] showed an extremely high prevalence [92.5%] of

gynaecological pain in women with Ehlers-Danlos Syndrome [Hurst et al, 2014], showing that more research is needed in the prevalence of endometriosis in the hEDS population.

By the time I had left home for university, I recall early problems with acid reflux/heart burn, a common problem for those with hEDS [Firkee,2014], for which my family practitioner gave me Gaviscon, which helped, and I was urged to avoid spicy foods. At a similar time, I started to encounter problems with constipation which hadn't been a problem until this point. Constipation is also a well-known problem for those with hEDS [Castori 2015; Firkee et al, 2014; Manning et al, 2003; Mohammed et al, 2010]. I recall visiting my family practitioner who issued me with medication, which I imagine would have been mild laxatives, (I cannot recall the specific medication). Eventually, both the acid reflux symptoms and constipation settled, although the constipation and then also bloating symptoms became worse around the time of menstruation, a diagnosis of endometriosis had then finally become confirmed, in my early twenties.

When I had left university and started work, I started to regularly experience chronic lower back pain, something that had begun in my late teens, but gradually worsened until it became a daily occurrence. Persistent pain is a substantial part of hEDS [Castori, 2012; Murray 2013; Rombaut et al, 2014; Sacheti et al, 1997]. It is known that symptoms of both endometriosis and IBS may also be related to lower back pain [Mears, 1996]. I had physiotherapy for my lower back pain which did not improve, and my gross muscular instability had yet to be pointed out, although one physiotherapy did comment 'my back was horrendous', and that I had a notable hinge at L5/S1. Eventually an MRI Scan showed a disc bulge and prolapse at L4/5. Years later, I went through very intensive course of physiotherapy that was to improve my muscular instability, lack of core control and proprioception which are notorious in the hEDS population [Castori, 2012; Ferrell & Ferrell, 2010; Simmonds & Keer, 2007].

In explaining my back pain, it may seem to the reader that I have drifted somewhat from a narrative on GI symptoms. Not so. I had a second laparoscopy for endometriosis which showed nothing (at this time). I was then finally referred to a Gastroenterologist, who diagnosed me with IBS, which can exacerbate both back pain and endometriosis [Mears, 1996]. By this time, some doctors were beginning to imply that my symptoms were psychosomatic, and the dangerous path of accusations of it being 'all in my head' ensued. It should be noted that many hEDS patients are frequently labelled with psychosomatic disorders, that their problems are 'all in their head' and that there is nothing wrong with them, which is a dangerous and detrimental path [Grahame, 2013; Knight, 2015 Shapiro & Ross, 2002]. Doctors must veer from such damaging labels, perhaps by, for example, using the skills of narrative medicine and empathy in order to engage with

the patient, or explain that the symptoms are more 'physiological/somatic' [Castori 2012; Knight, 2015; Nowacyk, 2012]. This is likely to result in a far more positive outcome for the patient, and the beginnings of a more therapeutic and trustworthy alliance between patient and physician [Adler, 2002; Charon, 2001; 2007; Knight, 2013; Nowaczyk, 2012].

Despite four more laparoscopies each showing endometriosis, my GI symptoms were not fully explored. Nor was I given any further help in their management. One gynecologist (actually insightfully, as it turned out), suggested I omit wheat from my diet. Somehow I soldiered on until 2011 when I was in my mid-thirties when I had been experiencing agonising pain on defecation. Eventually I could tolerate this no further and ended up going to my General Practitioner, GP, who said that I had an anal fissure, [something else that would be linked to hEDS and rectal evacuatory dysfunction RED – Mohammed et al, 2010], and would most probably need surgery. I was very upset by all this, notably incredibly embarrassed and ashamed. This is really the beginning of the major part of this narrative, and the investigations that followed, and series of numerous hospital admissions where I was still being labelled with psychosomatic problems. I was also diagnosed with Postural Orthostatic Tachycardia Syndrome (PoTS), which also impacts on the digestive system [Firkree et al, 2014; Grubb, 2010; Raj, 2006], but since anxiety can be a 'by-product' of PoTS owing to changes in blood pressure [Gazit et al, 2004], any implications to anxiety were adding to the damaging implied psychosomatic 'problems' despite anxiety also being linked to hEDS [Baeza-Velasco; 2011; Bulbena et al, 20014].

Perhaps it is no wonder if I was feeling depressed and anxious. I had a myriad of problems not only related to my hEDS, but digestive problems and now PoTS as well. Part of me often felt that the psychosomatic part was just a convenient way of doctors who just didn't know how to help or quite what to do. This seems to be a common problem for those with hEDS, because it is also known as an 'invisible illness' [Grahame, 2013; Knight, 2013; Knight 2015a; Murray et al, 2013; Tinkle, 2008].

## **INVESTIGATIONS**

I had already received my diagnosis of hEDS in 2009. In 2011, I was referred to a new Gastroenterologist. This doctor was of little help, other than suggesting (more as a passing shot) that I should consider the Low Fermentable, Oligo-Saccharides, Di-Saccharides, Mono-Saccharides Poly-Saccharides – Low-FODMAP - a diet that is very low in fermentable carbohydrates, and also an elimination diet –e.g. eliminating foods such as dairy, wheat/gluten [Williams, 2014]. A diet to be strictly adhered to for about 8 weeks, with food groups gradually reintroduced. The aim being to identify food group culprits, irritants [Williams, 2014]. At the same time I was sent to a colorectal surgeon in another (my local, and London teaching

hospital). At first I was seen by a junior doctor who sent me off to try Fybrogel which did not help my symptoms and made both symptoms and bloating far worse. I was then seen by a second colorectal surgeon who suggested I tried Movicol with a similar poor result. Finally I was seen by a colorectal consultant surgeon who said that my anal fissure would be repaired by him, and that I would need a 'Colon Transit Test' involving my swallowing one tablet each day which had different shapes. During this time I was not allowed laxatives as the purpose of the test was to see how quickly (or not) my gut was working. The outcome of the test was normal, showing that all three different shapes had cleared within the expected time-period. Next I had a colonoscopy. This showed sigmoid diverticular disease (diverticulosis at this point), a condition that appears to show prevalence in the hEDS population owing to tissue laxity [Firkee et al, 2014]. I also had a Magnetic Resonance Imaging Proctogram (MRI) scan which showed muscle dysynergy and that I was not coordinating my muscles on evacuation. I was then sent for Biofeedback Training. I complied with the training and change in toilet posture, but after a few months of training, the nurse gave up my training saying that I wasn't improving. By this time, I also had my anal fissure repaired, with Botox to relax the anal sphincter. The symptoms after this were unbearable, and I frequently cried on defecation. My belief, (when another colorectal surgeon tried the same technique some years later) was that I was stitched up so tight, I could not release at all.

By mid-2012, and now in my late thirties, about nine months after what was my first fissure repair, I started to have severe left iliac fossa pain. The pain was so severe that I went to the hospital (where I was being treated all this time) Emergency Room (ER). A plain x-ray showed that my colon was completely loaded. I was sent home with 32 Movicol sachets of which I used 17 in a 24 hour period. My symptoms were no better, and I returned to the ER a day later. Another plain x-ray showed that my colon was half-emptied. Blood tests showed infection, and a CT scan showed that I was having an episode of diverticulitis. Yet again, from my first visit to the ER, it would appear my symptoms and distress were ignored. I was admitted following the results of the bloods and CT-Scan. I was in hospital for 7 nights on double IV antibiotics, and my symptoms settled. I was discharged with further oral antibiotics.

Things continued to 'rumble on'. I still had great difficulties with evacuation, often having to eliminate an average of 5 times per day. It continued to be miserable for me and socially inconvenient, as I would also suffer from overflow diarrhoea. In January 2013 I had second admission for a diverticulitis flare up, which lasted 10 days. It should be noted that I was young for a diagnosis of diverticular disease, however, there is now some suggestion that diverticular disease may be associated with hEDS, but further research is needed (Firkee et al, 2014).

Again, things continued to be difficult for me. I was then simply 'written off' as having a Functional Bowel Disorder [FBD], for which there appears to be no treatment, only management. Again, this is a common condition in those who have hEDS [Castori et al; 2015; Castori et al, 2012; Firkee et al, 2014]. My problems really escalated following having a laparoscopic total hysterectomy in March 2014 for recurrent endometriosis.

### **CONSTANT ADMISSIONS AND DIETRY ADVICE**

From July-time I started to have regular and lengthy admissions to hospital. During the first one in July 2014, I was in hospital for 10 days. I was initially put into isolation rooms because it was feared I had an infection because of my overflow diarrhoea. I was fairly sure I was not infectious, but spent all my stay in two different wards in isolation rooms. During this time I was advised to go onto a low residue diet, and then about 4 days in I stopped eating altogether because I was feeling overfull, 'huge and bloated' and very constipated, and that eating was making me feel sick and further unwell. I did not have diverticulitis. MRI scans were normal. By the time I had refused food for 3 days, a dietician came to visit me. We discussed the merits of me trying the low FODMAP diet (Williams, 2014), and that my goal would also be to consume some nutritional drinks. These I tried, but promptly vomited them and continued to have diarrhoea. The nursing staff were beginning to despair, and I later learned that it was becoming implied that, yet again my problems were psychological and psychosomatic. On the tenth day, my colorectal surgeon discharged me. By this time I had managed to eat some plain rice. I left with the plan of following the low FODMAP diet by way of trial, and from this point onward (and to this day) remain wheat and gluten free. I have also lost 15kg over the past 12 months, and am now at my lowest weight since my early twenties. It would certainly seem that eliminating wheat and gluten improved my bloating symptoms, but still the problems of defecation remained. I then started to see a private nutritionist.

It should be noted that I was eating mainly self-prepared flapjack each day for breakfast. This was promptly removed from my diet, with much healthier, low-fat/sugar options, and a variety of different foods. Support was given for other meals, but despite the beginnings of weight-loss, my defecation and elimination problems remained. I also complained of regularly feeling overfull and often nauseous or close to vomiting, something that seems to be a common problem in patients with hEDS (Castori et al; 2015; Firkee et al, 2014). My nutritionist did not want me to remain for too long on the low FODMAP diet, as it was felt it did remove intake of various nutrients. However, it was certainly useful in identifying that wheat and gluten were clearly problem foods for me, along with certain pulses. I was gradually reintroduced to other foods, and advised on smaller portions to avoid sickness, and simple and easy to digest foods, along with a reduction in fat.

Positive outcomes of the diet change included weight-loss, and a reduction of my familial high cholesterol. However, diet did not change my abdominal pain.

In September, when I should have been going on vacation for a week to France, I ended up in ER again and was admitted to hospital for one week. Little was achieved during this admission. Again, investigations showed nothing remarkable, my pain was not a recurrence of diverticulitis, and my symptoms blamed on my FBD. My symptoms settled somewhat, and I was discharged. It was noted that I had another anal fissure, and plans were made for surgical treatment.

### **CONTINUED ADMISSIONS, SURGERY AND ACUTE ABDOMINAL PAIN**

In October I was then admitted to hospital for two weeks for abdominal pain. I was initially put on a morphine sulphate Patient Controlled Analgesia (PCA), and although this helped somewhat, I was then violently sick, and changed to another morphine drug, Oxycodone, and left on this drug for five full days with no intervention, or check up by the pain team. One week into my admission, I was transferred to another ward, but was given no further pain medication, other than the types of drugs I would normally take at home. Later during the week, I was contacted by Day Surgery admissions, and was told that a theatre slot had become available for repair of my anal fissure. This happened in the end to be with a different colorectal surgeon (to my usual one), and on a Saturday, whilst, as luck would have it, I was still an inpatient. This surgeon took a different approach, laying my fissure and also an anal fistula wound open, which made elimination easier, for a short time. I was discharged the day after surgery, having been accused by another patient of wanting to stay in hospital. It may have been a fly on the wall comment, but it was extremely upsetting. By this time I had missed numerous weeks from work, and was very distressed about my lack of income, now lack of vacation, and increasingly poor life quality.

### **DIAGNOSIS OF 'ACNES'**

Following this lengthy admission, I was sent to a different hospital with an expertise in hEDS. It should be mentioned that I had remained under the other hospital and had been tried on different drugs for my FBD. I attended the outpatient appointment and explained how I had very localised pain in my left iliac fossa, frequently ending up in hospital admissions. The consultant, and hEDS expert gastroenterologist performed some 'physiotherapy-type assessments' asking me to extend, flex and rotate my spine, including pushing and pulling against his limbs (showing resistant). I was diagnosed with Anterior Cutaneous Nerve Entrapment Syndrome (ACNES) or Chronic Abdominal Wall Pain, CAWP, this being like trigger point pain. I could have cried with relief as no one at my 'usual' hospital had managed to make this diagnosis, despite

numerous admissions. If only the previous hospital had considered more detailed physical examination and sought advice, my diagnosis might have been made sooner. My expert hEDS gastroenterologist alluded to a higher prevalence of ACNES in both hEDS and the fibromyalgia clinical population in his clinical experience (personal communication).

ACNES is actually a 'common' condition. The reported prevalence ranges from 3% in an unselected group of patients referred to a hospital gastroenterological service or admitted via the emergency department with any kind of chronic abdominal pain [Adibi P, Toghiani, 2012] to approximately 30% among patients in whom prior diagnostic evaluation for intra-abdominal diseases has not yielded any conclusive findings [Johlin & Buhac, 1996] from [Koop et al 2016]. However when patients present with abdominal pain without other clinically significant symptoms, ACNES should be high on the list of likely diagnosis [Applegate, 2002, Assen et al, 2013]. ACNES is not made better or worse with food or altered by bowel movements. It is often chronic, nagging and progressive and varies in intensity (Suleiman & Johnston, 2001). Indeed many patients go through numerous and costly investigations (as I did), with potential diagnosis of IBS and even being accused of having anxiety, hysteria and of malingering [Applegate, 2002; Koop et al, 2016]. It is not surprising that patients might worry because the pain can be extreme, which is no wonder when it is caused by nerve entrapment. Assen et al [2013] write that the pain is caused by the entrapment of an anterior cutaneous branch of one of more thoracic intercostal nerves [Assen et al, 2013, p.738].

Kopell & Thompson [1976] explain that the peripheral nerve entrapment occurs at sites where the nerve changes direction to enter a fibrous or osseofibrous tunnel or where the nerve passes over a muscular band [Kopell & Thompson as cited in Applegate, 2002, p.20]. Applegate [2002] believes that muscle contraction at these sites, (which are often very localised) may add additional 'insult' by direct compression and from muscular activity [Applegate, 2002]. Patients are often able to put their finger directly on to their pain with accuracy, although in my experience the pain, when it is at its most severe, often radiates across the whole abdomen and even up into the diaphragm. Nevertheless there is doubtlessly a point which can be 1-2cm in size, and is the epicentre cause of the pain [Applegate, 2002; Suleiman & Johnston, 2001].

A simple test devised by Carnett in 1926 determines diagnosis [Applegate, 2002; Sharpstone & Colin-Jones, 1993; Suleiman & Johnston; 2001]. The 'physiotherapy' type tests that I went through were the Carnett tests – tensing of the abdominal wall and increase in pain indicating a positive Carnett Sign, the patient often going 'that's it, that is where it is' [Applegate, 2002].

The management of ACNES involves a combination of local anaesthetic and steroid injections on the trigger point(s) [Applegate, 2002; Koop et al, 2016; Sharpston & Colin-Jones, 1993; Suleiman & Johnston, 2001]. At the time of writing this I have had two inpatient admissions requiring a Fentanyl PCA in order to control my pain, and now a total of six sets of local anaesthetic injections, including one with steroids, which can only be repeated six-monthly. I now no longer require inpatient admissions for ACNES, fully understand my pain, and further manage it with manual therapy; also a recommended treatment [Applegate, 2002]. Indeed on my day of diagnosis I began a course of manual therapy, and ordered a spiky ball to massage the affected area. The manual therapist had never encountered 'ACNCES' before, yet, was able to find the precise trigger points as indeed I was always able to locate, this borne out by the eventual series of local steroid and local anaesthetic injections to the aforementioned trigger points. Finally, one probable cause of ACNCES can be abdominal surgery, of which I have had several laparoscopies for treatment of endometriosis. It is quite likely that these indeed precipitated my ACNES (Applegate, 2002; Sharpstone & Colin-Jones, 1993). Physicians should be made well aware of ACNCES, which anecdotally according to my expert JHS/HEDS gastroenterologist suggested a higher prevalence of ACNES in both hEDS and the fibromyalgia clinical population, which requires further and ongoing research.

## **FURTHER FISTULAS AND ABSCESS**

For a brief time things seemed a little better (I was now 40 years old), until a week before Christmas 2014 when I started to feel feverish and had peri-anal pain. A GP put me on a course of antibiotics which were not working. A further discussion by phone to a GP prompted me to make a reluctant visit to the ER where bloods showed infection, I was placed on double IV antibiotics, and a diagnosis made of a peri-anal abscess. I spent the Christmas vacation in hospital, having seen my colorectal surgeon who wanted a repeat MRI scan before operating to clear the abscess in case of further anal fistula. I was then prepped for theatre, and 'forgotten' about. The next morning, I was told my consultant had gone away, and another surgeon, (who I did not know) would do my surgery. This doctor opted for Botox despite my reservation when I signed my consent form. However, I felt disempowered. Who was I to question the doctor on this matter? With insight, I wish I had challenged the decision, as I had to have an emergency repeat surgery following a latent MRI (post-surgery) that did in fact show another fistula. This surgery happened in February 2015. So I endured another surgery that might have been prevented if my consultant colorectal surgeon's wishes for an MRI had been carried out prior to the Christmas surgery. More NHS money wasted, not to mention another missed vacation and work-time.

## **PROLAPSES**

Following this surgery, it was suggested that I had a repeat MRI Proctogram, which is a video imaging used to assess bowel elimination using a radio-opaque gel for clarity. The results of this MRI Proctogram showed a radically different outcome to the Proctogram I had in 2012, which showed muscle dissynergy (which was not mentioned in the 2015 scan). Instead, the report showed that I had an entrocele, a cystocele, a sigmoidocele, a retrocele and rectal intussusception, the rectum descending 8cm below the coccxogeal line. It is known that there is a far greater prevalence of vaginal and anal prolapses and Rectal Evacuatory Disorders [RED], in the hEDS population, even amongst nulliparous women [Beighton et al, 2012; Farmer & Aziz, 2010; Mohammed et al, 2010; Norton et al, 1995].

My colorectal surgeon couldn't believe this new catalogue of problems. It was wondered whether I had a vaginal prolapse and the impact of the cystocele questioned. I was sent to Urogynecology for their opinion, but they concluded that no action be required at this time for the cystocele, or anterior vaginal prolapse at this stage, other than women's health physiotherapy, as recommended for this type of problem [Norton et al, 1995], and that it would be up to colorectal team to consider repairing the other catalogue of prolapses.

During this admission I met my (now) usual colorectal surgeon, we had a discussion on the merits of 'Ventral Mesh Rectoplexy surgery' to repair my rectal intussusception. We discussed whether this would, [or not], improve my symptoms of difficulties in bowel evacuation, but we considered that at least having my organs in the right position might improve things for me, along with further biofeedback training and ongoing dietary management.

## **REPAIR SURGERY**

Patients with hEDS are already prone to prolapses and because of their weakened collagenous structures [Carley & Schaffer, 2000; Mohammed 2010; Norton 1995) it was already discussed that this procedure may not hold as well in a patient such as myself, but worth trying to see if anatomically correcting the location of my organs would improve my situation. However it was also known that such surgery may also have a high failure rate in patients with hEDS (Tinkle, 2010).

Shortly before a planned surgical admission to correct my organ prolapses I had another attack of diverticulitis. My surgeons should have been informed, but weren't. I told them on the day of the surgery, and they said this might change their plans to lift my organs by mesh repair. Ultimately, this laparoscopic surgery ended up being diagnostic. The surgeons reported to finding inflamed pelvic adhesions, diverticular disease, 'obstructed defecation' and a large sigmoidocele with

'redundant' bowel, something that again, according to my expert JHS/HEDS Gastroenterologist appears to be more common in the hEDS population. I had laparoscopic surgery and a resection and removal of my sigmoid colon, and of my upper rectum, the bowel then stapled together. It is early days yet, and despite some initial constipation, largely corrected by increasing a gut motility drug, and adding dissolvable magnesium to my diet (to also aid constipation), I seem to have had an improvement in evacuation. I am continuing to use my biofeedback skills, and if I am very lucky, might have an improved long-term result, even if it doesn't hold quite as long because of my hEDS. At the time of going to press and despite five months of great improvement, I started to experience a recurrence of pre-surgical symptoms. The doctors are not entirely clear why this is (possibly because of my underlying Heds). A further scan showed that I had a 'striking' anorectal intussusception and in May 2017 I had mesh to repair this. Again, I was much improved following this second repair surgery, but about five months later my symptoms of incomplete evacuation returned, and I started needing to go to the toilet numerous times often with unsatisfactory evacuation. I occasionally became so constipated that my breath smelt of faeces and that I needed antibiotics to deal with resultant infection. In January 2018 I had a further MRI Proctogram scan which showed an anterior prolapse. As a result of this, and unless further mesh repair (that lasts) can be guaranteed, I am considering the possibility of a colostomy or stoma which sounds rather drastic, and could potentially be reversed, but it might last longer than the previous surgeries to repair my pelvic floor and straighten out my colon. At the point of writing, my case is being reviewed.

## **CONCLUSION**

This is a long story, involving a catalogue of problems at the helm of the heritable connective tissue disorder hEDS. The GI symptoms are wide-ranging for mouth to anus, and have caused me considerable distress. There is a considerably high prevalence of patients [approximately 86%], with hEDS who suffer from GI disorders [Castori, 2010]. The opportunity to be able to share the story from my point of view has been therapeutic, and also necessary as it may provide the medical professionals insight into how their decisions influence a patient's journey [Knight 2015, Knight, 2013]. The use of narrative medicine has been invaluable for this purpose, and I invite physicians to make full use of it, as a bridge between the scientific and the humanities [Charon, 2006]. It hardly needs mentioning that my patient journey continues, as does the need for improved communication between doctors and patients and patients and physicians, and the necessary ongoing research to improve the health and wellbeing for all of those with hEDS. Finally, physicians should be made aware of the condition 'ACNES' in relationship to persistent abdominal pain which presents with no obvious clinical cause [Applegate, 2002; Koop et al, 2016]. Further research would be needed to explore any links with ACNES and FM/hEDS.

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