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Living with the hypermobility syndrome

The Hypermobility Syndrome Association (HMSA) is a support group run by and for people who have the hypermobility syndrome (HMS) and their families. We receive many letters and telephone calls every day from people who require not just moral support but also urgent help to find effective treatment and answers to many questions about the condition. One common denominator our members share is that they feel that most medical practitioners (including rheumatologists) do not understand the impact that HMS has on the lives of patients, nor is the medical community able to provide effective treatment for them. We have found that the HMS is a complex disorder that is difficult both for medical practitioners to treat and for patients to control, and that there is poor communication between the two groups. Determining why this is so may open the way for the patient and the doctor to find new and effective methods of management. To this end, I hope to accomplish three things in this paper: first, to describe what HMS is like for patients and their families; secondly, to express what our members feel about the treatment they receive; and thirdly, to propose some suggestions for improving the situation.

With such a complex disorder it is often difficult for patients to tell their doctors the impact that HMS has on their lives. For the benefit of both doctor and patient, I will try to explain how HMS can affect lives. First, however, some general comments about HMS. HMS varies widely in terms of the severity of the symptoms experienced by patients, but also varies in individual patients from one day to the next. The patients fall into two broad groups. Those least affected may suffer periods of pain and injury, usually lasting several weeks. After effective treatment, they can be relatively symptom-free for some time. For patients in the other group, who are more severely affected, each day is a struggle against pain and injury. Our members from both groups report deterioration over time, particularly after pregnancy. This is contrary to much in the literature, which suggests that symptoms decrease with age. The reported stiffening with age does not always bring less pain. On the contrary, many of our members have increasingly painful symptoms as age advances.

HMS is difficult to diagnose because the patients look well and present differently from each other. Some are very mobile, while others who were once mobile are no longer so. In addition, they often do not respond well to the currently prescribed treatment. Pain is the most common symptom reported to us. For patients it comes in varying degrees, and is often quite unbearable. We are often asked for specific suggestions for pain relief and effective management. However, we currently have no proven effective treatment to offer the majority of patients. It is my experience that medical practitioners are loath to give general advice, particularly with such a variable condition. Severe HMS can colour an individual's attitude to his or her body and whole life. Life and all activities become linked with pain, and perhaps more debilitating than the pain itself is the fear of pain.

The following profile describes those who are more severely affected. Not everyone with HMS will fit this description, but many do. For those most affected by HMS, one could say (as is often said for rheumatoid arthritis) that life itself becomes a painful endurance. Everyday activities of life carry the price of pain: these include brushing teeth, getting dressed, shopping for food, doing the laundry, any repetitive movement, including chopping, typing, ironing and walking, and especially lifting and carrying.

Holding down a job or looking after a family is often barely possible, and is sometimes impossible. Frequent absenteeism from work due to pain and injury labels the HMS patient as lazy or problematical and may halt advancement at work. It is not uncommon for an HMS patient to have tried several careers. Obviously, with the problems of daily life just outlined, caring for children and running a household is as difficult for an HMS patient as is working in full-time employment. The effects of HMS on the family and relationships can be devastating. Many patients tell me that even being touched can cause them pain. Partners often become frustrated when patients are unable to participate in family activities.

I have described the life of many of our members. Patients often have trouble communicating this to their doctors. To aid in your understanding, I would like to describe what it may be like in the rheumatology clinic patient's chair. This description does not apply to all, and I use the female person because the majority of our members are female. I believe these descriptions may apply equally to the male patient. It is based on the reported cumulative experience of our members.

If the patient has not yet been diagnosed as suffering from the HMS, the chances are that she may have

been made to believe that her pain is psychosomatic, and in some cases that she may have injured herself on purpose. This scenario is most common when the patient may not recall an 'accident', which would normally account for such an injury, or when her description of how an injury occurred could not account for such a severe injury in a normal patient. She may have high expectations, having waited many weeks for an appointment to see the consultant and having invested in the hope that the specialist might be able to help. She may be depressed as a result of her pain and disability. In addition, she may seem desperate, willing to do anything to be free from the pain. The patient may appear to be angry at all medical professionals after years of inadequate or inappropriate care. She may be defensive and anxious to prove that she needs help, and she may therefore appear to be exaggerating her problem. Finally, she may be unclear what symptoms to relate, as the problem has often been present throughout her life and she has begun to wonder if it is a normal part of her life.

Another area of special consideration to bear in mind is that HMS is a familial syndrome. I mean this in two ways: obviously with a heritable disorder there are certain familial considerations, such as genetic counselling; in addition, it is a familial syndrome in the sense that it can have a particularly destructive effect on a family.

First, many patients express fear about passing down HMS to their offspring; a life of pain is not a legacy that one would want to pass on to one's children. In addition, many women realize that they will not be able to cope adequately with the demands of motherhood or pregnancy. Secondly, if a parent is affected with HMS the following problems may arise within the family. Children may feel insecure and anxious about the parent's health, because both parent and child are unsure of when the pain may become overpowering. Therefore, the parenting ideal of continuity and stability may become impossible to sustain. In common with other heritable disorders, a complex family dynamic may take place: the child sees the effects of the disorder on the adult, and becomes fearful and resentful when he or she is forced to come to terms with a potentially problematic future with the condition.

If, however, a child is the family member with HMS, the problems are different. For example, should a child be made aware of a chronic disorder when no effective and accepted treatment is currently widely available? What are the psychological effects of being a medically monitored child? How will HMS affect life at school? For example, should the school be informed? Should the child take physical education? Should the child carry a schoolbag? How will the chairs and desks affect the child? Should the child have special advice on footwear? Should the child be given special exercises, for example postural exercise programmes, to prevent problems later on in adulthood? It is clear that there is an urgent need for research into an accepted and effective treatment/management for paediatric and adolescent patients, particularly as children are being diagnosed in greater numbers and at a younger age.

What do the patients feel about current treatment? In the course of being examined by a rheumatologist, many of our members have reported being asked to perform their 'party tricks' for doctors and medical students. The doctor may not recognize that the original mobility is often decreased through pain and over time. Moreover, the performance of these tricks will cause pain physically and emotionally, which can set the patient back for weeks, if not longer. All patients recognize the need for rheumatologists to measure their degree of mobility, but the attitude and manner described can only lower the patient's confidence in the medical profession.

Many patients report that they often receive only diagnosis and reassurance as treatment from rheumatologists. This may be compared with the placebo effect, in that it is initially helpful but its positive effects only last a limited time. For other patients, reassurance can have a negative effect and can be confusing, as it does not equate to their own experience with HMS.

In many cases, after the initial diagnosis the patient struggles on alone until he or she is forced to seek out further treatment because the symptoms have continued or become more severe. This usually takes the form of non-specialized physiotherapy and pain medication. Many of our members have reported that this kind of physiotherapy has often made their condition worse.

We are often asked for the names of rheumatologists and physiotherapists who specialize in the condition because the treatment the patient has received so far has not been helpful. For the many patients who contact us, there is no widely available or universally acknowledged effective programme of exercise or physiotherapy treatment. Many of our members have struggled for years with medical treatment that has made them worse, not better. General practitioners often do not recognize the need to send their patients to specialized clinics. Patients are left feeling isolated by their pain and by the lack of effective medical treatment.

Self-help and patient-led management is currently the best approach for the long-term treatment of HMS. Unfortunately, patient-led management and self-help are particularly problematical for the HMS patient because HMS is a disorder that robs patients of control over their lives. All chronic illness and chronic pain cause feelings of loss of control. The problem is exacerbated with HMS because there is not always a clear link between an activity and the onset of pain. Therefore, it is difficult for the patient to lower the level of pain by behavioural modification. The fear of pain is often a greater contributory factor in patient decline than is the pain itself. This fear, combined with the lack of a clear cause-and-effect relationship between an event and subsequent pain, works to lower the patient's strength through progressive physical inactivity. In addition, it makes some patients insecure and unsure of their body's limits and can promote self-blame and poor self-esteem.

For successful self-management, it is important for patients to feel some internal sense of control, to feel that they can change their lives for the better. This self-empowerment is often made more difficult because many HMS patients go from one doctor or alternative practitioner to another in search of help. Many of our members have complained of having to see a different physiotherapist each time they are referred for treatment, often with serious consequences relating to lack of continuity and conflicting advice. Another aspect of this problem is that because pain often occurs some time after the activity that caused it, exercise programmes are difficult to maintain. The lack of regular appropriate exercise, combined with self-blame, may lead to weight gain for some patients. The resultant lowering of self-esteem fuels the downward spiral, putting both patient and doctor in a difficult position. Doctors correctly suggest to an already vulnerable patient that the weight gain is making the problem worse. This fuels the self-blame and accentuates the downward spiral.

I now take this opportunity to suggest three words, three 'R's to improve treatment and remind you of the special needs of HMS patients: recognize, research and respond.

Recognize the impact of HMS on the lives of patients. Recognize the mind-set of the patient who lives in pain and in fear of pain. Chronic pain can affect cognition and clarity, which limits good communication between patient and doctor. Recognize that the whole family needs to be treated as this disorder affects every aspect of daily and family life.

Research into the disorder and determine the cause of this crippling pain. Research into the development of strategies of management, including physiotherapy and drug and cognitive therapies. Research into patterns of symptoms to determine why, for example, many people who are very mobile have no pain while others who are less mobile are suffering. Or, for example, why some women with HMS experience less pain during pregnancy whilst others experience a great deal more pain than is usual for them when they are not pregnant.

Respond to the needs of the patient, and work with the patient.

Doctors are beginning to recognize the role that patient support groups may have in helping them to research into various disorders. There is a flow of information from the patient to the doctor, but the flow is often in one direction only. A patient needs to be empowered with information to get better. I would urge rheumatologists and others not just to publish in medical journals and then wait for the knowledge to trickle down to the patients by haphazard means. It is my experience that too little information is the cause of anxiety among patients, not too much.

In summary, be partners with your patients in their treatment and perhaps in time your waiting rooms will be less crowded.

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