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Joint Hypermobility and Joint Hypermobility Syndrome

Dedicated to my hypermobile patients, from whom I have learned so much. I've seen hypermobility syndrome, but you've lived it.

Many people have flexible or loose joints. They're the people, maybe like you, who did gymnastics or ballet when they were young and are "good" at yoga. Their joints ***move farther and more easily than most people's joints***, so they often can do tricks like bending their thumbs forward until they touch their fore-arms. Sometimes these people are called "double-jointed," and some may even have dislocated or popped their joints out of the socket. The medical term for joints that move too far is **hypermobility**, and the word for joints that are too loose and move too easily is **laxity**.

Experts estimate that up to 10% of the general population may have some degree of hypermobility, with women affected about three times more often than men. Most hypermobile people do not develop any problems from their loose joints, but some suffer chronic pain and other symptoms. Those who do suffer chronic joint pain and other symptoms related to their hypermobility or to the looseness of other tissues that often accompanies hypermobility have a condition called **joint hypermobility syndrome** (JHS).

Often, people who suffer from hypermobility syndrome are called hypo-chondriacs or lazy because they avoid many everyday activities, because these activities that cause them pain. Most of them don't look sick and, as a result, friends, colleagues, and even doctors can be unsympathetic. Furthermore, they may spend years unsuccessfully searching for the cause of their chronic pain and other symptoms because many doctors are unfamiliar with hypermobility syndrome and its complex set of symptoms. Such long delays and lack of understanding can lead to frustration (with doctors and with daily life), anger, anxiety, and depression.

Hypermobility

The **Beighton score** is used to measure a person's degree of hypermobility. One point is assigned for the ability to accomplish each of the following movements:

- bending your small finger back further than 90 degrees (1 point each side)
- bending your thumb forward to touch your forearm (1 point each side)
- hyper-extending your elbows and knees, that is bending them beyond a straight line (1 point each joint, each side)
- putting your palms flat on the floor without bending your knees (1 point)

Hypermobility – and hence the Beighton score – often decreases with age, as joints become less mobile, both as a natural result of aging and because loose joints are predisposed to premature osteoarthritis, the wear-and-tear form of arthritis.

Joint Hypermobility Syndrome

Joint hypermobility syndrome can include a wide and diverse array of symptoms, but the muscles and joints are most often affected, giving the syndrome its name. People with JHS often develop chronic joint pain and stiffness, most often in the larger joints; for example, the joints of the neck, shoulders, back, hips, and knees. However, smaller joints such as the ankles, wrists, and elbows often are affected as well. Joint pain often comes from the muscles and tendons around the joint rather than the joint itself, so that x-rays may be normal.

People with hypermobility syndrome may have a whole group of other conditions, in addition to joint problems, because of excessive stretchiness of other body tissues. For example, mitral valve prolapse and uterine prolapse, hernias, and gastroesophageal reflux disease (GERD) are more common in people with hypermobility syndrome.

The **Brighton criteria** generally are used to decide if a person with hypermobile joints also suffers from JHS.

Major Criteria
- Beighton score of 4 or more
- Joint pain affecting 4 or more joints and lasting more than 3 months

Minor Criteria
- Beighton score of 1, 2, or 3
- Back pain or pain in one to three joints lasting more than 3 months, or spinal spondylosis, spondylolysis, or spondylolisthesis

- Dislocation of more than one joint, or of one joint more than once
- 3 or more soft-tissue problems (e.g., tendonitis, bursitis)
- Tall, thin body shape
- Skin hyper-extensibility, stretch marks, thin skin, or abnormal scarring
- Drooping eyelids, nearsightedness
- Varicose veins, hernia, or uterine or rectal prolapsed
- Mitral valve prolapse

Any of the Following Required for Diagnosis
- 2 major criteria
- 1 major and two minor criteria
- 4 minor criteria
- 2 minor criteria and a clearly affected first-degree relative

And there are even more symptoms. Unexplained bruises often appear "out of nowhere." Many hypermobile people complain of dry mouth or constant thirst, often with a craving for salty foods. They are uncomfortable standing for long periods, so avoid lines and like to sit with their feet up. Many patients with JHS also have problems with their autonomic nervous system, the part of the nervous system that regulates circulation, breathing, and digestion. This can lead to symptoms such as lightheadedness, palpitations, and digestive problems, and probably plays a role in difficulty sleeping and overall fatigue, which also are common complaints.

Some experts believe that JHS is the same thing as the hypermobile type of **Ehlers-Danlos syndrome** (EDS), a condition also caused by extreme elasticity of body tissues. People with the hypermobile type of EDS have loose joints and soft, velvety skin, as well as other symptoms almost identical to those described here. For most people, the distinction between joint hypermobility, joint hypermobility syndrome, and the hypermobile type of Ehlers-Danlos is not an important one clinically, as treatments are similar. Exceptions include more severely affected patients, such as those who require braces or surgery to stabilize their joints, or those who have unusual symptoms such as weakness or loss of feeling in arms or legs, and those with certain eye problems or a family history of aneurysms, all of whom if possible should see a specialist with knowledge of EDS, in part to rule out other more serious types of EDS. Those considering having children may wish to see a clinical geneticist to see if they carry any of the known genes associated with Ehlers-Danlos, although many such genes have not yet been identified. **People with lax joints fall along a broad spectrum, from those with joint hypermobility but only mild or no related symptoms, to those moderately affected by JHS, to those more severely affected, many of whom truly have the hypermobile type of EDS**.

Joint Pain and Muscle Strains

Joint hypermobility syndrome, as the name implies, primarily affects the musculoskeletal system. Loose joints cause increased strain on nearby soft tissues (muscles, ligaments, tendons) that stabilize them. These soft tissues themselves often are overly lax, and because of their laxity and the increased strain on them, they are prone to tearing and spasm, leading to pain and stiffness around joints. The pain may or may not be clearly related to any specific activity. For some, any repetitive movement, such as walking, lifting, or carrying can be painful. Standing or sitting for any period of time can cause stiffness and pain, as can something as simple as cleaning a kitchen counter or bending down to pick up laundry.

Because of their role in stabilizing the trunk and the head, the neck and lower back are almost always affected. **Chronic neck strain** affects nearly every patient with JHS for two main reasons. First, the ligaments that are supposed to support the head are too loose and therefore cannot do their job well. The muscles of the neck are forced to do more of the work of supporting the head than they are meant to do, so they become strained. Second, most JHS patients have shoulders that are too loose, that is the "ball" of the upper arm is not held tightly in the "socket" of the shoulder. Because of the weakness of the shoulders, almost any activity that uses the arm, including reaching, pushing, pulling, and carrying, pulls not only on the shoulder but also on the neck. For these two reasons, neck muscles are constantly being strained, and what little healing may occur overnight is promptly undone the next day. Remarkably, this process occurs so gradually that many people with JHS do not even notice it, and when asked they may say, "My neck is fine," when in fact their necks are a mass of knotted soft tissue, soft tissue that does not feel soft at all!

Lower back pain also is very common in people with JHS, again for a number of reasons. As in the neck, the ligaments that should support and stabilize the spine and pelvis usually are too loose, putting extra strain on muscles to try to support the upper half of the body. Like the relationship of the shoulders to the neck, loose hips also put extra strain on the lower back to try to stabilize the pelvis. Among these muscles is the piriformis muscle, a small muscle at the base of the pelvis (in the buttock). When called upon to play a bigger part in supporting the pelvis than it is meant to, it can easily be strained. Once strained, it may tighten up and pinch the sciatic nerve, which runs directly beneath it. The resulting pain, called sciatica, can be felt in the buttock and often radiates down the back of the leg. This condition, sometimes called **piriformis syndrome,** is often mistaken for a pinched nerve from a ruptured disc in the spine.

People with JHS do have an increased tendency to have **disc problems**, sometimes at an early age, because the intervertebral discs that help cushion and support the spine may be less rigid than normal. Softer discs are more

prone to leak or rupture, allowing disc material to ooze out of the disc and pinch nearby nerves, causing pain. Disc problems in the neck cause pain down the arms, and discs in the lower back cause pain to be referred to the legs. Less often, tissues within the disc itself can break down, causing pain within the disc, which can be very difficult to treat.

Some people with hypermobility also develop **neuropathic pain**, which may be felt as burning, stinging, tingling, shooting, numbing, etc. Sometimes such pain is caused by disc problems, but often it is quite localized or does not follow the usual patterns of pinched nerves. Conventional nerve testing usually is normal, so these symptoms may be attributed to psychological rather than physical causes. This type of pain also can be particularly difficult to treat.

As mentioned earlier, osteoarthritis occurs more rapidly in loose joints. Therefore, arthritis in the neck and lower back is another frequent cause of neck and back pain and stiffness in JHS patients.

Hypermobility also commonly causes pain in the hips, shoulders, knees, and elbows. The shoulder in particular depends a great deal on its ligaments for support, and when the ligaments are loose, there is extra strain on the soft tissues of the shoulder. When these tear, **tendonitis** often develops. Similarly, hyper-extending the elbows can tear the tendons on the sides of the elbow. Pain in this area often is referred to as “tennis elbow” and “golfer’s elbow.” In addition, many people with JHS suffer frequent ankle sprains, which like shoulder and elbow injuries, may take a very long time to heal because they tend to get injured again and again while they are trying to heal.

Unstable hips often cause pain which, like neck pain, may go unnoticed for a long time, since the hip joint does not move around as much as the knee, shoulder, and ankle joints. Examination of the hip in people with JHS often causes them to cry out, “Ouch! I didn’t even know it hurt there.” Also, many people mistakenly describe pain from the back part of the hip joint as “back pain.”

The most common source of knee pain in hypermobile people is the cartilage between the kneecap and the knee. Because the soft tissues that are supposed to hold the kneecap in place are loose, the kneecap itself is often loose. After years of sliding around too much, the cartilage underneath the kneecap starts to wear down (a condition referred to as **chondromalacia**), causing pain – and sometimes a crunching or grinding noise – while kneeling, squatting, or climbing stairs. Osteoarthritis of the knee joint itself is not as common, but it can occur, especially in those who have done years of high-impact exercise or who are overweight.

Other joints that can be affected include the joints where the ribs meet the breastbone and where the ribs meet the vertebrae of the spine. Many people with JHS feel chest pain and tightness, and may even seek emergency care to

rule out heart disease, when the source of their symptoms is the joints of the rib cage, a condition called **costochondritis**, or inflammation of the rib cartilage.

In addition, the jaw, or temporomandibular joint (**TMJ**), is often affected by hypermobility. Just like other joints, looseness of this joint leads to strain on the muscles around it and wear of the cartilage in the joint. A variety of treatments are used for TMJ pain, but strengthening and stabilizing the joint offers the best hope for many for long-term relief.

Finally, there is an association between hypermobility and increased risk of **osteoporosis**, although it is not clear whether this is simply from inactivity because of pain or, more likely, that there is a specific defect in bone metabolism. The identification of receptors for adrenaline-like hormones in the bone suggests the possibility that loss of bone may even be related to autonomic nervous system dysfunction (see below).

Treatment of Musculoskeletal Symptoms

The treatment of musculoskeletal symptoms primarily consists of medication for pain relief, and exercise and physical therapy to relieve muscle spasm – in the short run – and strengthen the tissues around loose joints to stabilize them – in the long run. These are general treatment principles, which are more easily applied and more effective for some joints than others.

A brief word about **pain**: pain is not a good thing. "Toughing it out," "putting up with it," "learning to live with it," etc. are not productive approaches to dealing with chronic pain. "Medication can't be good for me," is similarly illogical. Pain is not good for you. Pain strains your system, wears you down, disturbs your sleep, and makes you irritable and even depressed. Medication that relieves pain often does a lot more; it often improves sleep, concentration, energy, and mood. Be hopeful that with appropriate treatment you will gradually need less and less pain medication, but take it when you need it.

Finding a doctor – whether a family practitioner, internist, orthopedist, rheumatologist, physiatrist, osteopathic physician, or any other physician – knowledgeable about hypermobility is critical. A physical therapist or personal trainer who is familiar with hypermobility is another valuable resource, especially in helping to develop an appropriate home exercise program. Many people with joint discomfort start exercising, only to find that their symptoms get worse. The basic rules of exercise I recommend are:

- **AVOID** high-impact exercises, such as sports that involve running, jumping, or physical contact. Swimming or water exercises, walking, Pilates, and Tai Chi are good choices. Some forms of yoga are OK, but others are not – see stretching, below.

- **AVOID** most forms of stretching that involve grabbing a joint and pulling or pushing on it to "loosen it up." Many people are reluctant to give up stretching, because it "feels so good." But in this case, pulling on tight muscles does not relax them, and the relief is only temporary. Stretching further strains and loosens joints. This is why many hypermobile people make their conditions worse by doing yoga. However, stretching hamstrings is OK and an important exception.

- **AVOID** heavy lifting, pulling, and pushing. Be particularly careful around the house and in the yard, where odd-shaped weights and unusual angles often lead to injury.

- **AVOID** hyper-extending your joints. That is, don't straighten your arms out fully so that your elbows lock, and don't straighten your knees to the point that they lock. This applies to carrying groceries or a gallon of water just as much as it does to doing exercises with weights.

- **DO** light resistance exercises, which are the cornerstone of strengthening and stabilization. Any degree of straining only causes injury. For weights, a good rule of thumb is that if you can't do 8 repetitions (reps) without straining, then the weight you have chosen is too heavy for that exercise. You should not consider increasing weight or resistance level until you can do two sets of 15 reps without straining. In general, more reps with a lighter weight are preferable to fewer reps with a heavier weight. Most people find no need for weights heavier than 10 pounds; hand weights of 2 or 3 pounds for some exercises and 5 pounds for others are usually sufficient.

- **DO** be persistent and consistent. You don't need to spend an hour in the gym. Even on days when you are "too tired" or "don't have time" to exercise, 5 minutes of light weights for shoulder strengthening and 5 minutes of isometrics for core strengthening every day will yield big benefits in the long run. No "two week cure" here!

Fibromyalgia

Fibromyalgia is a common diagnosis in people with JHS. Once muscles around loose joints become strained and painful with daily use they may become more and more painful, until chronic unrelenting pain begins to disrupt sleep and cause fatigue and depression. Each of these symptoms reinforces the others. For example, depression can disrupt sleep, cause fatigue, and increase sensitivity to pain, setting up a vicious cycle of pain, fatigue, poor sleep, and depression, which is the crux of fibromyalgia. Hypermobility also may predispose people to develop chronic fatigue syndrome, which has much in common with fibromyalgia.

Extra-articular Manifestations of Hypermobility

Problems affecting parts of the body other than the joints are referred to as the extra-articular manifestations of hypermobility. Lax joints are very often associated with increased tissue elasticity elsewhere in the body, especially in the blood vessels and digestive tract. In recent years, hypermobility also has been associated with a variety of autonomic nervous system problems.

JHS and the Autonomic Nervous System

The autonomic nervous system regulates all body processes that occur automatically, such as heart rate, blood pressure, breathing, and digestion. To compensate for stretchy blood vessels and increased venous pooling (too much blood collecting in over-stretched veins) most people with hypermobility appear to make extra adrenaline, which may account for the high-energy, always-on-the-go lifestyles of many hypermobile people. Unfortunately, if you get too tired, your body responds by making more adrenaline, so you keep going, not realizing how tired you really are. It appears that as you get more and more run down, your body gets more sensitive to adrenaline, so the small amount you have left can produce the same response a larger amount used to, so you still don't feel tired even when you are. Even when you do feel tired, you may continue to "push through" the fatigue, collapsing when the adrenaline wears off. Years of not feeling, ignoring, or pushing through fatigue may be one factor in the development of illnesses like chronic fatigue syndrome.

Many of the autonomic nervous system problems associated with hypermobility are characterized by an "over-response" to physical and emotional stresses, which often leads to fluctuations in heart rate and blood pressure, as well as digestive and respiratory symptoms. Sickness, pain, emotional stress, and even fatigue itself can raise adrenaline levels, and acute stresses can trigger adrenaline surges, leaving you jittery, anxious, and even more exhausted. Worse, such surges can trigger an excessive counter-response, causing nausea, sweating, lightheadedness, diarrhea, and of course even more fatigue. Even sensory stimuli, such as bright lights or loud noises, can trigger an exaggerated or over-response, causing sensitivity to light and sound.

Perhaps the most common symptom of autonomic nervous system dysfunction in hypermobile people is **orthostatic intolerance**, or lightheadedness on standing. Tilt-table testing often reveals abnormalities such as **neurally mediated hypo-tension** (NMH) or **postural orthostatic tachycardia syndrome** (POTS), fancy names for different ways in which the body fails to maintain a stable heart rate and blood pressure when a person stands up. Increasing salt and fluid intake, and avoiding caffeine and alcohol, which deplete the body of fluid, may reduce such symptoms. It also helps to keep your feet elevated, wear support hose, avoid prolonged standing, and of

course the obvious—if you get dizzy when you stand up quickly, don't stand up quickly!

Hypermobility and the Heart and Circulation

Because too much blood is pooling instead of circulating, people with JHS typically have cold hands and feet and low or low-normal blood pressure, in addition to lightheadedness on standing. Drops in blood pressure can trigger palpitations and racing and pounding of the heart. There is also an increased risk of migraine headaches, varicose veins, and hemorrhoids.

Although experts disagree on whether or not there is an increased risk of mitral valve prolapse in hypermobile people, many people with mitral valve prolapse are loose-jointed. Fortunately, for most people, mitral valve prolapse does not pose any serious risks. The term **mitral valve prolapse syndrome** has been used to describe the various autonomic and other symptoms that many people with mitral valve prolapse have, but in many cases, these symptoms are probably related to their underlying hypermobility.

Atherosclerosis (clogged arteries) is uncommon in people with hypermobility, in part probably because their blood pressures tend to be low and because most tend to have low cholesterol. However, unusual heart problems can occur. One of the most serious cardiovascular concerns in hypermobile people is an increased tendency for blood vessels to tear or even rupture, although this is primarily a concern for people with the more serious vascular type of Ehlers-Danlos syndrome.

Hypermobility and Headache

People with lax joints are predisposed to many different kinds of headaches. Migraine headaches are very common, in part because many migraines are triggered by fluctuations in hormone levels or blood pressure, which can be increased by autonomic problems. Headaches from chronic neck strain also are very common and can often turn into migraines. In addition, severe autonomic problems can cause a dehydration or "hangover"-like headache, possibly related to inadequate blood flow. Uncommonly, looseness of the muscles that control the eyes can cause difficulty focusing and eye strain headaches. TMJ problems can also cause headache.

Hypermobility and Digestion

Hypermobile people often have digestive problems, both in the upper and lower parts of the gastrointestinal tract. The esophagus and especially the tissues around it may be too stretchy, allowing stomach contents to come back up or "reflux" into the esophagus. Because the stomach contains acid, and the esophagus is not meant to hold acid, **acid reflux** causes heartburn in many

people with JHS. Furthermore, frequent reflux can cause serious burning and scarring of the esophagus, even increasing the risk of cancer of the esophagus. A stomach that is overly stretchy can cause food to stay there too long, a condition called **delayed gastric emptying**, which can cause patients to feel full quickly—and sometimes continue to feel full for many hours--and can also increase the risk of reflux. Autonomic problems can also affect digestion, e.g. causing digestion to occur too slowly or too quickly depending on the situation.

Intestines that stretch too easily increase the risk of constipation and bloating, and make gas much more painful. Many hypermobile people are diagnosed with **irritable bowel syndrome**, a diagnosis that often carries a negative connotation – that the patient isn't dealing with stress well or is really depressed and won't admit it – when in fact these symptoms, and their cause, are often physical and not psychological.

Tears of the abdominal wall muscles are another very common problem among people with hypermobility. The muscles themselves do not tear, rather the fibers connecting different muscles do, creating a gap between two muscles. Small segments of intestine can occasionally push up through these gaps, causing pain. As pressure backs up behind this "stuck" segment, pain gets worse and is felt in a larger area, but eventually resolves when the intestine that has pushed up into the layer of muscle falls back into place. The timing of the pain is quite random, depending only on the movement of bowel contents and the contractions of the intestines. The lack of any correlation to meals or bowel movements is one clue to the cause of the pain.

Unfortunately, these abdominal wall tears usually will not be found on a cursory physical exam, nor on x-rays, CT scans, or sonograms, and most physicians think they are rare and so do not look for them. Depending on where the tears are located, patients may be incorrectly diagnosed with reflux, ulcer, gallstones, ovarian cysts, diverticulitis, and most often, irritable bowel syndrome. Once patients understand the source of the pain, most can tolerate it, or find ways, such as changing position, to relieve it. Surgical repair is rarely necessary, except when true hernias occur, i.e. when intestine pushes through the abdominal wall muscles and stays there. The most important reasons to make this diagnosis are to prevent unnecessary testing and treatments for other incorrect diagnoses, and to reassure patients that they don't have something terribly wrong that hasn't shown up in the tests they've already had.

Hypermobility and Anxiety

The body's tendency to overreact to stresses by making too much adrenaline can lead others to think that hypermobile people are "too sensitive," "irritable," or "anxious." Patients themselves may notice this, saying, "I've always overreacted to little things. I can't help it." It is very important to recognize two things about this phenomenon. First, it is a physical reaction, so that counseling

usually will not be effective in treating this type of anxiety. Similarly, adrenaline highs and lows may be mistaken for the mood fluctuations of bipolar disorder, but mood-stabilizing medications usually are not indicated. When medication is required, beta blockers, which block adrenaline, may be as effective with fewer side effects than SSRI's like Prozac and Lexapro or benzodiazepines like Xanax and Valium. Second, while a feeling of anxiety can be produced by emotional stress, it is just as likely that such symptoms have a physical cause, most often fatigue, pain, or dehydration, and less commonly by a drop in blood sugar or blood pressure. Not surprisingly, researchers have found that anxiety and panic disorder are more common in hypermobile people.

Hypermobility and Sleep

Similarly, when hypermobile people try to fall asleep, the stimulating effect of their extra adrenaline may keep them awake. If they are able to fall asleep, they may continue to make too much adrenaline overnight, giving them a shallow, dream-filled sleep, so that they wake feeling unrefreshed. Pain further stimulates adrenaline, making restful sleep even more difficult. When studied in the sleep lab, they often have a relative and sometimes complete lack of deep sleep, and/or an increased number of sleep-disrupting "arousals." Poor sleep can cause irritability and fatigue, which in turn can trigger more adrenaline (to try to overcome the fatigue), which in turn can make sleep worse. This vicious cycle can eventually cause serious disability.

Like fatigue and pain, many patients are not aware of just how bad their sleep is. Although some people are aware of waking often or of having frequent very vivid dreams, many will insist that they "sleep fine," even while admitting that after sleeping 8 hours they don't feel rested when they get up. One obvious reason for this lack of awareness is, of course, that they're asleep, so they have no way of knowing that they're not getting enough deep sleep or having way too many arousals. Sleep studies done in a sleep lab are very helpful in demonstrating the nature and severity of sleep problems, and in ruling out other sleep problems like sleep apnea and periodic limb movements of sleep, which can coexist with hypermobility-related sleep problems. Recently home sleep monitors that can measure sleep stages (shallow, deep, REM) and arousals have become available. While not as good for initial diagnosis, since they don't detect apnea and limb movements, they are often helpful for monitoring and adjusting treatments over time.

Non-pharmacologic treatment of sleep problems begins with the basic rules of good sleep hygiene, such as trying to sleep the same hours each night, and using your bed only for sleep, not for work or watching television. Don't eat, exercise, or do anything stimulating too close to bedtime, and certainly don't have caffeine or alcohol in the evening. Be wary of medications, like over-the-counter decongestants, that can interfere with sleep.

When these measures fail, then medication is indicated, since virtually every system in the body is strained when you don't get a good night's sleep. Not sleeping well not only makes you tired and irritable and can affect your

mood, it also affects mental functions like memory and concentration, and has recently been shown to be a major contributor to weight gain in some people. Besides treatment for sleep apnea and limb movements when these are present, medications specifically for hypermobility-related sleep problems are often helpful.

As mentioned earlier, one possible explanation for the frequent arousals and lack of deep sleep is that patients are making too much adrenaline at night, just as they often are during the day. Some patients unfortunately seem to make too little during the day, waking tired and dragging through the day, only to get a "second wind" of energy (or a "first wind" for many!) at 9:00 or 10:00 at night, just as they are trying to wind down and get ready for bed. Heart rate monitors showing increased fluctuations in heart rate and occasional sudden increases in heart rate corresponding to arousals and awakenings lend support to this theory, as does the observation that medication to block or offset extra adrenaline helps many patients get a better night sleep. Adrenaline-blocking medications include various types of beta blockers, while medications like Valium and Ativan work partly by raising the levels of calming chemicals in the brain to offset the extra stimulating ones. Also, since chronic pain is so common in this patient group, appropriate pain medication at bedtime is often essential to achieving a restful night's sleep.

Hypermobility and the Genitourinary System

In women with hypermobility, the ligaments supporting the uterus can be weak, leading to an increased risk of **uterine prolapse**, a condition where the uterus "slips" and presses on the bladder. Weakness and excessive stretchiness of other tissues in the pelvis increase the risk of **cystocele** and **rectocele**, conditions in which the bladder and rectum, respectively, push on the walls of the vagina. Excessive stretchiness of the vagina itself and weakness of the muscles that support the pelvis can make intercourse painful for some women with hypermobility. Vulvodynia or **vaginismus**, a painful spasm of the vagina, also may occur more commonly in the setting of hypermobility. Weakness and laxity of the muscles of the pelvic floor also contribute to the onset of incontinence at an early age in some women with hypermobility.

Interstitial cystitis, which causes frequent and often painful urination, and often more diffuse pelvic pain as well, appears to occur with increased frequency in patients with hypermobility. A bladder that stretches too much may not empty completely, or may signal the brain that the bladder is full when it isn't. As with digestion, autonomic fluctuations can cause difficulty or frequency of urination. In addition, hypermobility may be associated with an increased risk of endometriosis (tissue that normally lines the uterus grows somewhere else, such as on the ovaries, behind the uterus, or on the bowels or bladder).

Pregnancy and delivery usually are not exceptionally difficult for hypermobile women. In fact, because circulating blood volume expands during

pregnancy, many women find that their circulatory symptoms, such as light-headedness and cold hands and feet, and for some even fatigue, are much better during pregnancy. On the other hand, some symptoms, such as heartburn, varicose veins, and hemorrhoids, can be worse during pregnancy. There may be an increased incidence of premature rupture of membranes and rapid (i.e. less than 4 hours) labor and delivery, and women with severe hypermobility are at risk for unusual complications.

Summary

Joint hypermobility is a very common condition, and many people with loose joints will have no related medical problems at all. Most people have just a few, such as achy joints and cold hands and feet, and they should not be overly concerned that they will go on to develop other complications. On the other hand, I have described some of the more common conditions associated with hypermobility, because those who do have many related problems may be relieved to understand that there is a unifying explanation for so many of the unusual things they have noticed about their bodies. Beyond the psychological relief of understanding their symptoms, with proper treatment most people with JHS can get significant relief from their physical symptoms, too.

My patients have long asked for something to share with their families, friends, and even physicians, to help educate others about their condition. I hope this paper helps fill this need.