

# Joint Hypermobility & the Link with CFIDS

Are you one of many CFIDS patients who have hypermobile joints? If so, you will be especially interested in this article, which explores **joint hypermobility** as a possible **predisposing factor** to developing CFIDS.

BY DR. ALAN POCINKI, GUEST CONTRIBUTOR

**M**any people have joints that are unusually flexible, or hypermobile. Often the degree of hypermobility is mild, and they don't even realize they are more flexible than others. In more extreme cases, individuals can amuse their friends by putting their feet behind their head or popping their thumbs out of joint.

I see many patients in my general medical practice with varying degrees of hypermobility, but in studying CFIDS patients since 1987 I have been struck by the presence of joint hypermobility in almost every one. To substantiate this clinical impression, I recently gave a survey about hypermobility-related symptoms to 20 consecutive patients seen in my office who met the 1994 CDC diagnostic criteria for CFIDS, and I examined their joints for hypermobility.

The prevalence of hypermobility-associated symptoms in these 20 consecutive CFIDS patients and age- and sex-matched controls (seen for unrelated medical conditions, such as asthma) is shown in the chart on the next page. The chart also includes each patient's Beighton score, a measure of the severity of joint hypermobility. The Beighton scale assigns one point each (right and left) for the ability to: pull back the fifth finger beyond 90 degrees; touch the thumb to the underside of the forearm; hyperextend the knees beyond 190 degrees; hyperextend the elbows beyond 190 degrees; plus a ninth possible point for the ability to put both palms flat on the floor while bent at the waist (without bending the knees).

One thing worth noting is that only 8 of the 20 patients had previously been told by a physician that their joints were unusually flexible. I recently saw a patient who is one of seven siblings who all developed CFIDS. They were studied at NIH in the 1980s in an unsuccessful attempt to find a CFIDS marker. He is now 50 and walks with a cane because of severe osteoarthritis and multiple spinal operations, yet when I explained the apparent association of hypermobility with CFIDS, his eyes bugged out and he exclaimed, "You



All photos in this article by David Love

Being able to amuse friends and family with tricks like these may be fun when hypermobile individuals are young, but the condition can lead to many unwelcome physical symptoms as people age.

mean could I like put my foot behind my head before I got sick? Of course I could. We all could!”

### What is joint hypermobility?

Hypermobility syndrome is a condition in which the joints are so flexible that they cause symptoms such as joint or muscle pain. Because of the looseness of the joints, there is increased strain on the surrounding soft tissues to stabilize the joints. These soft tissues are themselves often overly lax and, between their laxity and the increased strain on them, they are prone to tearing and spasm, leading to pain and stiffness around joints. (Many such painful sites correspond to the characteristic tender points of fibromyalgia, so it’s not surprising that hypermobility has also been associated with the development of fibromyalgia.)

The term hypermobility syndrome usually also suggests the presence of other symptoms beyond just joint and soft-tissue problems. Lax joints are often associated with increased tissue elasticity elsewhere, most prominently in the blood vessels. Overly stretchy veins dilate like balloons, filling up with too much blood, a condition known as venous pooling. Because much of their blood is “pooling” instead of circulating, individuals with this condition typically have cold hands and feet, low or low-normal blood pressure and lightheadedness on standing quickly.

Overly elastic vessels may also explain the predisposition hypermobile individuals have to varicose veins, hemorrhoids and migraine headaches. Problems with anxiety and disturbed sleep also appear more likely. The decreased rigidity of subcutaneous tissues offers diminished protection to the blood vessels underneath, so these patients usually report bruising easily.

## PREVALENCE OF HYPERMOBILITY-RELATED SYMPTOMS

SYMPTOMS	NUMBER OF CFIDS PATIENTS	NUMBER OF CONTROL PATIENTS
Ever diagnosed with CFS	20	0
Ever diagnosed with fibromyalgia	15	0
Ever dislocated a joint	7	0
Often get lightheaded on standing quickly	17	1
Bruise easily	17	6
Tend to have cold hands and/or feet	20	4
Often feel cold when others don’t	17	7
Neck often feels stiff	18	6
Joints often feel stiff, click, creak or grind	20	6
Ever diagnosed with migraine	13	3
Ever had varicose veins	9	4
Ever had hemorrhoids	12	3
Often crave salty foods	14	2
Diagnosed with mitral valve prolapse	7	0
Average Beighton score (up to 9 points)	6.3	0.5

The prevalence of features of joint hypermobility syndrome in my random sample of patients with CFIDS is quite high, suggesting the possibility of a pathophysiologic relationship between the two conditions. In fact, Peter Rowe and his colleagues at Johns Hopkins first described the association of CFIDS, orthostatic intolerance and hypermobility in 1999. David Goldstein has documented the extent of autonomic dysfunction in CFIDS patients studied at NIH, and others have described autonomic dysfunction in otherwise healthy patients with hypermobility. The combination of hypermobility and autonomic dysfunction appears not only to predispose certain individuals to develop CFIDS, but also accounts for most of its characteristic symptoms.

### The link to CFIDS symptoms

How might joint hypermobility and associated autonomic dysfunction account for the development of the symptoms of CFIDS? As mentioned above, pain in muscles and joints, without swelling or redness, may develop from the excessive strain that unstable joints put on the muscles around them. Venous pooling not only explains the orthostatic intolerance seen in many CFIDS patients, but diminished blood flow to the head and neck may contribute to sore throat by affecting blood flow to neck muscles, and it’s probably a major contributor to cognitive dysfunction—the “brain fog” many CFIDS patients describe.

Hypermobility patients are predisposed to at least three different types of headaches. Not only do they often suffer from migraines, but they get tension headaches from



**Dr. Alan Pocinki examines 23-year-old CFIDS patient Megan Gurney Lavedas, a former dancer and gymnast who has had CFIDS since she was 17. She is unusually flexible, with a Beighton score of 9 out of a possible 9 points. When Dr. Pocinki first saw her six years ago, her blood pressure was 70/40 and she could barely sit up without passing out. Today, thanks to intravenous fluids administered several times a week and medications, Megan has graduated from college and is teaching a dance class once a week.**

chronic strain of overtaxed neck muscles, which are trying to compensate for the laxity of ligaments supporting the head. They also are prone to dehydration- or hangover-type headaches from lack of blood flow to the brain.

To compensate for their poor vascular tone and increased venous pooling, most hypermobility patients appear to have increased adrenergic (or noradrenergic) tone. In other words, they make more of the body's "fight or flight" stimulating hormones, called catecholamines (or they may overrespond to normal amounts). Increased circulating catecholamines typically make these patients—when healthy—high-achieving, always-on-the-go individuals, just as so many CFIDS patients were before getting sick.

Sickness, pain or other stress can further raise catecholamine levels. With high levels to start, any physical or psychological stress that triggers a further increase makes levels way too high, leaving patients, as one recently said to me, "tired but wired." They may feel jittery and appear anxious.

Similarly, when they try to fall

asleep, the stimulating effect of the extra adrenaline may keep them awake. If they are able to fall asleep, patients may continue to make increased catecholamines overnight, giving them a shallow, dream-filled sleep, so they are easily wakened overnight and then feel unrefreshed.

Often patients will describe waking abruptly an hour or two after falling asleep, like they're "running a race," with their heart pounding and feeling "wide awake," with great difficulty getting back to sleep. Such episodes are often misdiagnosed as panic or anxiety attacks, when in fact their etiology is physiologic, not psychological. With the normal decline in blood pressure during sleep, some patients may actually become hypotensive enough to trigger a reflex catecholamine surge, waking them with a jolt. A drop in blood sugar can also be such a trigger. When I described this phenomenon to one patient treated unsuccessfully for years for panic and anxiety, he exclaimed, "That's exactly how I feel!"

In addition to problems with joints and circulation, hypermobile patients tend to have increased elas-

ticity of other tissues, including the gastrointestinal, genitourinary and respiratory tracts. Pain from stretch receptors that are too easily stimulated may well be the cause of many of the symptoms of irritable bowel syndrome, interstitial cystitis and vulvodynia. Increased pulmonary elasticity may cause airway collapse, triggering the reflex dyspnea—deep and/or rapid breathing and the sensation of not being able to get a full breath—that many CFIDS patients describe.

Thus, hypermobility and the physical features and dysautonomia often associated with it not only account for virtually all of the diagnostic features of CFIDS, but also explain some of the other symptoms often associated with the illness (including orthostatic intolerance), as well as some of the physiologic traits of patients before they got sick.

### **Symptom improvement**

If this model is valid, then correction of some of the underlying pathophysiology should result in improvement of symptoms. Although it's impossible to correct

entirely the problem of venous pooling, David Streeten found a decade ago that MAST trousers (like a giant blood pressure cuff) inflated around the lower extremities and abdomen to force pooled fluid back into circulation dramatically eliminated pain and cognitive dysfunction.

More simply, administration of intravenous fluids, to temporarily increase circulating volume, also greatly relieves symptoms in many patients.

Typical patient responses I hear are, "I feel so much better after the fluid" and "The difference between before and after the fluid is like night and day." I have been struck, as have many of my patients, by their observation that receiving fluids not only improves their fatigue, lightheadedness and overall sense of well-being, but their brain fog and muscle and joint pain diminish as well.

These observations suggest that other measures aimed at improving circulation may also help relieve some of the symptoms of CFIDS. In fact, many patients even find simple measures like wearing support hose and keeping their feet elevated whenever possible are helpful. An increased intake of salt and fluid, and avoidance of medications and foods that are dehydrating, such as alcohol and caffeine, help many patients feel better. Also valuable to some patients is the use of pharmacologic measures, such as fludrocortisone or nondeglycerized licorice, which increase salt and fluid retention.

The consensus of the 2003 NIH Workshop, "Neuro-Immune Mechanisms and Chronic Fatigue Syndrome," was that there is a

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"vulnerable population" at risk for CFIDS. It appears that patients with joint hypermobility syndrome are just such a vulnerable group. These individuals normally compensate for many of their physiologic problems, for instance with increased adrenergic tone to compensate for low

blood pressure and venous pooling. When confronted with certain triggers, however, such as acute physical illness,

major emotional stress or even specific environmental exposures, this compensation may be lost, and the symptoms of CFIDS may develop.

This model is an observational and therefore simplistic one. What exactly causes the disruption of the previous balance to precipitate the onset of CFIDS is unknown.

Although this model predicts an essential role for autonomic and/or hormonal (neuroendocrine) dysfunction, the precise cause must be elucidated by future research. ■

*Dr. Pocinki began studying CFIDS at the National Institutes of Health in 1987, and he continues to see CFIDS patients as a significant part of his general internal medicine practice in Washington, D.C., where he is an Assistant Clinical Professor of Medicine at George Washington University. He received a CFIDS ACTION Champion award in 2000 and last year was honored with the D.C. Medical Society's Distinguished Service Award.*

## TIPS FOR IMPROVING YOUR SYMPTOMS

- Wearing support hose and keeping your feet elevated whenever possible may sound like simple measures, but they can provide real benefits.
- Your doctor can administer intravenous fluids. Although it's a temporary fix, this does increase circulating volume and relieve symptoms in some patients.
- Many patients with hypermobile joints get relief simply by increasing their daily intake of fluids and salt.
- Cut back on foods that are dehydrating, such as alcohol and caffeine, or avoid them.
- Consider pharmacologic interventions like fludrocortisone, which increases salt and fluid retention.
- To alleviate venous pooling, try prescription high-pressure hose. Waist-high ones are the closest thing to MAST trousers, which are not widely available.
- While standing in line at the grocery store, frequently shift weight from leg to leg.
- Small doses of stimulating medications, such as pseudoephedrine and phentermine, can raise blood pressure and heart rate and improve circulation and energy.

## References

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