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BRIEF ANSWERS TO SPECIFIC CLINICAL QUESTIONS

Q: Can fibromyalgia and chronic fatigue syndrome be cured by surgery?

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A: A RECENT, HIGHLY publicized theory linking brainstem and spinal malformations with fibromyalgia and chronic fatigue syndrome is an interesting but unproven hypothesis. Thus, although the theory emphasizes the need for careful neurologic evaluation of all fibromyalgia and chronic fatigue syndrome patients, it is far from clear whether surgery is an appropriate treatment.

HOW THE CONTROVERSY BEGAN

During the past year neurosurgeons Michael J. Rosner and Dan S. Heffez caused a great deal of excitement in the mass media and on the Internet by suggesting that they found a surgical treatment for fibromyalgia and chronic fatigue syndrome.

In a *Wall Street Journal* article[1] and on a subsequent television broadcast on ABC's *20/20 Friday* program, Rosner and Heffez presented preliminary data linking these conditions to the Chiari malformations (protrusion of the brainstem downward against the spinal cord) and cervical myelopathy. In an Internet interview, responding to a question about the extent of symptom relief after surgery in patients with fibromyalgia or chronic fatigue syndrome, Heffez replied that "every symptom can be improved to a greater or lesser degree,"[2] implying that the surgery was very successful.

Unfortunately, no peer-reviewed manuscripts have yet been published that confirm any relationship of Chiari malformation or cervical myelopathy to fibromyalgia or chronic fatigue syndrome, nor has anyone yet published any data about the outcomes of patients after surgery.

THE CHIARI MALFORMATIONS

Chiari I and Chiari II malformations are congenital hindbrain dysgeneses, often diagnosed in infancy and early childhood. They involve protrusion of the lower portion of the brainstem into the spinal canal, resulting in varying degrees of spinal cord impingement. Chiari I, the displacement of the cerebellar tonsils into the upper cervical canal, presents more often in adulthood, is less serious, and is often amenable to surgical intervention. Chiari II is characterized by displacement of the cerebellar tonsils, with related caudal dislocation of one or all of the following: the medulla, the vermis, and the fourth ventricle with kinking of the medullary cervical junction. Syringomyelia (cavitation of the central portion of the spinal cord) can be variably present in both.[3]

Symptoms associated with these malformations are related to the degree of anatomic abnormality. Signs and symptoms were reported in eight representative series comprising 769 patients.[3–10] Presenting symptoms were cranial nerve abnormalities (50% to 70%), limb weakness (30% to 60%), sensory abnormalities (50%), headache (50%), neck pain (50%), and ataxia (40%).

At physical examination, atrophy with hyporeflexia, generally in the upper extremities, was noted in approximately 35% of patients. Hyperreflexia in the lower extremities, often associated with ataxia, was found in 30% to 50% of patients, with the Babinski sign present in 15% to 65%. Anatomic diagnosis was confirmed by computed tomography or magnetic resonance imaging (MRI).

FIBROMYALGIA AND CHRONIC FATIGUE

Both fibromyalgia and chronic fatigue syndrome are characterized by widespread pain, present in 80% to 97% of cases.[11–13] Fatigue,

At present, any link is at best only an interesting hypothesis

sleep disturbance, morning stiffness, and depression occur in most patients. In addition, 30% to 50% of patients report headache, diffuse paresthesias, impaired cognition, intermittent constipation and diarrhea, subjective muscle weakness, urinary frequency, orthostasis, subjective joint swelling, or subjective swelling of the neck glands.

Fibromyalgia is common, occurring in 3.4% to 4.9% of women and 0.5% to 1.6% of men, based on two reports.[14,15] Approximately 80% of patients with fibromyalgia also have symptoms of chronic fatigue syndrome, suggesting that there is little difference between these two syndromes.

SIMILARITIES AND DIFFERENCES AMONG THE DISORDERS

Patients with Chiari malformation, cervical myelopathy, or both share some symptoms with patients with fibromyalgia and chronic fatigue syndrome, most notably headache, paresthesias, and weakness. But there are important differences: the weakness associated with Chiari malformations is objective, whereas in fibromyalgia and chronic fatigue syndrome it is subjective; and the diffuse pain, fatigue, and disturbed sleep prominent in fibromyalgia and chronic fatigue syndrome are rare in Chiari malformations and cervical myelopathy.

A small subset of patients may have both conditions by chance alone

Symptom ascertainment bias, however, may play some role: the neurosurgeons treating Chiari malformations did not routinely ask about the subjective symptoms of fibromyalgia or report these in the neurosurgical literature. It is also important to remember that in two large radiologic series, despite MRI findings of herniation of the cerebellar tonsils, 30% to 64% of patients had no typical neurologic symptoms. Might these patients have had fibromyalgia or chronic fatigue syndrome symptoms that simply were not recorded?[8–10]

RESPONSE TO SURGERY: A DIFFERENCE OF OPINION

According to Heffez, "8% of patients have not observed any meaningful benefits that they can recognize," but added that "no patient has been made worse by any of the surgeries" performed.[2] The two surgeons are very enthusiastic about the outcome. The exact number of patients treated, however, is uncertain, but as cited in the Wall Street Journal,[1] Heffez and John D. Weingart, MD, of Johns Hopkins "have done about 75 of these surgeries, and Dr. Rosner an estimated 250."

Reports in the surgical literature regarding outcomes of standard repair of Chiari malformations are less enthusiastic. Most patients with Chiari malformation undergo posterior fossa decompression, and those with cervical stenosis undergo laminectomy.[3–5] Overall, at 2-year follow-up, 50% are clearly improved, 20% are worse, and 30% are unchanged. Upper extremity weakness and dysesthesias, often related to central cord lesions, are most resistant to improvement.

IS THERE A LINK BETWEEN CHIARI MALFORMATIONS AND FIBROMYALGIA/CHRONIC FATIGUE?

That a small proportion of patients with fibromyalgia and chronic fatigue syndrome may have anatomic neurologic syndromes is possible. The Chiari malformations are very rare: A total of 147 patients were seen over 6 years at a tertiary care center specializing in cervical cranial malformations.[9] Contrast that number with the frequency of chronic pain in the general population, which has been demonstrated to be 10.6% for diffuse pain and 20.1% for regional pain,[14] and one must conclude either that only a very small subset of fibromyalgia and chronic fatigue syndrome patients also have a Chiari malformation, or that this cervical malformation is drastically underreported in the general population. Given current estimates of the frequency of both conditions, it is highly unlikely that an anatomic malformation is responsible for symptoms in the majority of patients with chronic pain and fatigue.

That a small subset of patients have both conditions by chance alone is also possible. Certainly, patients with a chronic progressive neurologic condition that goes undiagnosed for years will experience great stress, a key etiologic factor associated with fibromyalgia and chronic fatigue syndrome.[16] If surgery

improves the symptoms of the anatomic syndrome, it will also improve stress and, thus, the symptoms of fibromyalgia and chronic fatigue syndrome. Rosner and Heffez emphasize that gainful improvement of fibromyalgia and chronic fatigue symptoms may take many months, in keeping with the hypothesis of secondary benefit.

On the other hand, some patients with lesions on MRI have no typical neurologic symptoms,[9] and it is possible that these lesions are of etiologic significance by unknown mechanisms in a small subset of fibromyalgia and chronic fatigue syndrome patients. A pilot prevalence study has been published in abstract form in which MRI was performed in 38 patients with fibromyalgia and in 23 healthy controls.[17] Tonsilar herniation was observed in 8 (31%) of the 26 fibromyalgia patients who had technically adequate studies and in 11 (73%) of the 15 controls who had adequate studies, suggesting a high asymptomatic prevalence of this anatomic finding in the general population which was not higher in fibromyalgia.

Further prospective investigations might be indicated, but given the high frequency of chronic pain, fibromyalgia, and fatigue in the general population and given the high sensitivity and low specificity of MRI and its prohibitive expense, only patients with fibromyalgia or chronic fatigue syndrome who have quantifiable neurologic symptoms such as upper extremity weakness or muscle atrophy, upper extremity hyporeflexia, objective ataxia, lower extremity hyperreflexia, or a positive Babinski sign should be enrolled in an initial MRI pilot study.

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