

Orthostatic Intolerance in CFS ☐

David S. Bell, MD, FAAP (oud voorzitter van de adviescommissie “Chronic Fatigue Syndrome” van het Amerikaanse ministerie van Volksgezondheid, USA)

Quest #68, 2005

[Dr. Bell is a world renown authority in ME/CFS. His present areas of research are the autonomic nervous system, the role of ADH and blood volume, orthostatic intolerance, and the education of young people.]

Chronic Fatigue Syndrome (CFS) is a multi-symptom illness with a wide range of severity, able to cause significant disability. For an excellent comprehensive review please see John & Oleske (1). The illness has been surrounded in controversy for years, particularly because the degree of disability is not predicted by the relatively normal physical examination and routine laboratory findings. Even the name “chronic fatigue syndrome,” contributes to the controversy by implying that simple fatigue or tiredness is the central disabling symptom. In this paper, I would like to suggest that the central and most disabling symptom of CFS is not fatigue but the symptom of orthostatic intolerance.

Disability is defined as an alteration of an individual’s capacity to meet personal, social, or occupational demands because of an impairment (2). Criteria for the **research diagnosis** of CFS were first published in 1988 (3) and revised in 1994 (4) and again recently (5). Essentially, these criteria require a new onset of activity limiting fatigue which is not caused by ongoing exertion and is not relieved by appropriate rest. Thus, by definition, CFS requires at least some degree of disability. Patients must also have at least four of the following eight symptoms: cognitive dysfunction; recurrent sore throat; tender cervical or axillary lymph nodes; muscle pain; multi-joint pain; headache of new pattern; unrefreshing sleep; and post exertional malaise lasting more than twenty-four hours. **Clinical criteria** published by the Canadian Consensus Panel (6) describe ME/CFS in more detail and include orthostatic intolerance.

For those patients unable to work or attend school due to CFS, they express difficulty in explaining the exact reason, other than to say they “feel too sick.” Of course, employers or schools have a difficult time understanding, as illness is generally expected to resolve within a short time. It is the use of the term ‘fatigue’ which causes the greatest confusion in regard to disability status. Technically, fatigue is a state of recovery, and this does not occur in persons with CFS. Occupational medicine physicians may argue that it is appropriate to work with fatigue, with the assumption that normal fatigue, like normal muscle weakness, will respond to increased activity. While this is appropriate for normal fatigue, it is usually not the case with CFS.

The symptom of fatigue as experienced in CFS is quite different from the shared common experience of fatigue. Patients use several terms in an attempt to describe this symptom, including “weakness”, “heaviness”, “exhaustion”, and “sleepiness”. This experienced sensation is better served by the term ‘orthostatic intolerance’ meaning limitation of sustained upright activity. Orthostatic intolerance is defined as the inability to tolerate, over time, the upright position, either sitting, standing, or walking. The symptoms are at least partially relieved by recumbency [lying down](7).

The term orthostatic intolerance is used in two ways: it is a symptom confused with simple fatigue, and it is an umbrella term for more specifically defined conditions such as postural orthostatic tachycardia, orthostatic hypotension, delayed orthostatic hypotension, neurally mediated hypotension, and orthostatic narrowing of the pulse pressure. **Abnormalities in the autonomic nervous system underlie both the symptom of orthostatic intolerance and its defined subtypes, and are active areas of current research.**

The symptom of orthostatic intolerance causing limitation of sustained upright activity is the central disabling symptom of CFS. After a period of time in the upright position, a person with CFS becomes overwhelmed with “fatigue,” pain, confusion and other symptoms requiring the patient to lie down. Symptoms such as sore throat, lymph node pain, muscle and joint pain are not in themselves orthostatic, but in my experience, are exacerbated by prolonged standing or sitting. I feel that the cognitive symptoms of CFS are orthostatic in nature, but this has not been tested.

In CFS, activities such as light walking may be better tolerated than quiet standing, most likely because of the circulatory effects of muscle contraction. However, even these activities are limited in persons with CFS. In general, sitting is better tolerated than standing, but both are considered orthostatic stress. The symptom of worsening or malaise after exertion in the diagnostic criteria (4) is caused primarily by orthostatic stress. Recumbency, with or without sleep, is the action which relieves this discomfort.

As a symptom, orthostatic intolerance may be described just as any other symptom. Proof of the existence of this symptom is always difficult, just as the presence of pain is difficult to prove. Attempts to prove orthostatic intolerance with a tilt table tests have lead to categorization of the subtypes described above. However, persons with CFS may have a normal tilt table test despite severe symptomatic orthostatic intolerance (8). It is for this reason that quiet standing in the office with behavioral observations combined with pulse and blood pressure monitoring has been suggested and normal values established (9). Furthermore, quiet standing more closely resembles normal or daily orthostatic stress.

The misunderstandings surrounding CFS have led to substantial suffering of patients over and above the symptoms imposed by the illness. One potential way for these misunderstandings to be addressed is to employ the more accurate term of orthostatic intolerance to describe the central disabling symptom of the illness.

References

1. John JJ, Oleske, eds. “A Consensus Manual for the Primary Care & Management of Chronic Fatigue Syndrome”, *The Academy of Medicine of New Jersey & The New Jersey Department of Health and Senior Services*, Lawrenceville, NJ, 2002.
2. American Medical Association. “Guide to the Evaluation of Permanent Impairment. 5th ed.” Chicago Ill, 1995.
3. Holmes GP, Kaplan JE, Gantz NA, et al. Chronic fatigue syndrome, a working case definition. *Ann Intern Med* 1988;108:387-389.
4. Fukuda K, Straus SE, Hickie I, et al. International Chronic Fatigue Syndrome Study Group. The chronic fatigue syndrome: a comprehensive approach to its definition and study. *Ann Intern Med.* 1994;121:953-959.
5. Reeves WC, Lloyd A, Vernon SD, Klimas N, Jason LA, Bleijenberg G, Evengard B, White PD, Nisenbaum R, Unger ER, et al. Identification of ambiguities in the 1994 chronic fatigue syndrome research case definition and recommendations for resolution. *BMC Health Services Research.* 2003;3:25.
6. Carruthers B, Jain A, DeMeirlier K, Peterson D, Klimas N, Lerner A, et al. Myalgic encephalomyelitis/chronic fatigue syndrome: Clinical working case definition. diagnostic and treatment protocols. *J Chronic Fatigue Syndrome.* 2003;11(1):1-12.
7. Stewart J. Orthostatic intolerance: a review with application to the chronic fatigue syndrome. *J Chronic Fatigue Syndrome.* 2001;8:45-64.
8. Gerrity TR, Bates J, Bell DS, Chrousos G, Furst G, Hedrick T, Hurwitz B, Kula RW, Levine SM, Moore RC, Schondorf R. Chronic fatigue syndrome: what role does the autonomic nervous system play in the pathophysiology of this complex illness? *Neuroimmunomodulation.* 2002-3;10:134-141.
9. Stewart JM. Orthostatic intolerance: A review with application to chronic fatigue syndrome. *J CFS.* 2001; 8:45-64.
10. Streeten DH. “Orthostatic Disorders of the Circulation”. *Plenum* New York. 1987:116.