

Proposal for modifications to ICD-10-CM for Chronic Fatigue Syndrome, Myalgic Encephalomyelitis, and Postviral fatigue syndrome

Submitted by International Association for Chronic Fatigue Syndrome/Myalgic Encephalomyelitis
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The International Association for Chronic Fatigue Syndrome/ Myalgic Encephalomyelitis (IACFS/ME), based in the US and the largest international group of professionals dedicated to researching and taking care of people affected by this medical condition, recommends:

- a) Removing “Chronic fatigue syndrome, NOS” from R53.82 “Chronic fatigue, unspecified”, in the Symptoms and Signs chapter
- b) Adding “Chronic fatigue syndrome” to the neurological chapter at G93.3
- c) Modifying the G93.3 title term to “Postviral fatigue syndrome, Chronic fatigue syndrome, and Myalgic encephalomyelitis”
- d) Adding separate G93.3 subcodes for the terms “Chronic fatigue syndrome,” “Myalgic encephalomyelitis,” and “Postviral fatigue syndrome”
- e) Revising existing exclusions as needed and adding exclusion for “chronic fatigue.”
- f) Removing the word “benign” from “benign myalgic encephalomyelitis” so the term used is “myalgic encephalomyelitis”

The existing ICD-10-CM classification for these terms, especially the classification of chronic fatigue syndrome with unspecified chronic fatigue, does not reflect current scientific knowledge, best clinical practices, or the 2015 report of the National Academy of Medicine concerning this condition. It is also inconsistent with international World Health Organization ICD standards set in ICD-10, as detailed in the corresponding rationale below for each recommendation. This can have a significant negative impact on issues like tracking of morbidity/ mortality, healthcare resource planning, appropriate reimbursement for and documentation of clinical care, provisioning of workplace/ school accommodations, and determination of disability benefits. Proposed modifications are also shown at the end of this document in the Requested Modifications section.

Rationale for each of these recommendations:

a) Separating chronic fatigue syndrome from chronic fatigue, unspecified: For the last 3 decades in the United States, **chronic fatigue syndrome (CFS) has been recognized as an individual diagnostic entity in its own right and not merely an individual symptom. Every CFS case definition that has been used in the United States includes symptoms beyond**

only chronic fatigue.¹ For example, the most used diagnostic case definition, Fukuda 1994, requires severe, disabling fatigue of at least 6 months accompanied by at least 4 out of 8 other symptoms (e.g. muscle pain, unrefreshing sleep, problems with concentration, sore throat, etc.). Consequently, it is medically inaccurate to classify CFS under “chronic fatigue, unspecified.” Doing this is the equivalent, for example, of classifying asthma under “cough, unspecified” merely because coughing may be one symptom of asthma.

Reinforcing this point, a 2015 report by the National Academy of Medicine (NAM) on myalgic encephalomyelitis/chronic fatigue syndrome (ME/CFS) noted that ME/CFS is different than medically unexplained chronic fatigue, that the level of fatigue is “more profound, more devastating, and longer lasting than that observed in patients with other fatiguing disorders,” and that “this complex illness presentation entails much more than the chronic presence of fatigue.”²

Given that CFS is the diagnostic code used in the United States for the disease ME/CFS, it is important that CFS not be reduced to one of its symptoms or use the same code as the symptom of chronic fatigue. We recommend that CFS no longer share a code with chronic fatigue, unspecified.

b) Moving CFS to the neurological chapter: There is **substantial scientific evidence of neurological impairment in ME/CFS.**³ Consequently, the World Health Organization and all countries except for the United States **classify CFS under G93.3 in the neurological chapter of ICD-10**, along with “Postviral fatigue syndrome” and “Myalgic encephalomyelitis.” Furthermore, in developing ICD-11, the World Health Organization explicitly recommended that, “...in the absence of compelling evidence mandating a change, legacy should trump with regard to the question of moving certain conditions to new chapters”.⁴ WHO staff have stated that **chronic fatigue syndrome will not be placed in the Symptoms and Signs chapter in the forthcoming ICD-11.**⁵

Further, in ICD-10, the term is “chronic fatigue syndrome,” not “chronic fatigue syndrome, NOS.” The rationale given by NCHS in 2011 for adding the term “NOS” (not otherwise specified) to chronic fatigue syndrome in ICD-10-CM was that it indicates that CFS is “not specified as being due to a past viral infection.”⁶ However, as discussed below, CFS definitions do not preclude a viral onset.

Thus, in accordance with scientific findings and international standards, we recommend placing CFS under G93.3 in the Neurological chapter. We also recommend removing the term “NOS” from “chronic fatigue syndrome” since the rationale for its addition is not correct and more specific versions of CFS have not been defined.

c) Modifying the G93.3 title term: Part of the stated rationale for not following the ICD-10 and classifying CFS at G93.3 in the neurological chapter of ICD-10-CM was the view that ME is postviral while the term “Chronic fatigue syndrome” was intended for cases where “the physician has not established a post viral link.”⁷ However, ME definitions explicitly include non-viral precipitants such as bacteria and parasites and CFS definitions allow viral precipitants. Further, while ME and CFS often occur after an infection or infection-like episode, a variety of other triggers such as immunization, pregnancy, surgery, and physical trauma have also been observed.

Thus, **“Postviral fatigue syndrome” as the title of the G93.3 entity, fails to accurately convey the breadth of these conditions.** We recommend “Postviral fatigue syndrome, Chronic fatigue syndrome and Myalgic encephalomyelitis” to correctly accommodate the range

of precipitants.

d) Separate subcodes for ME, CFS and PVFS: In reviewing and recommending new diagnostic criteria for myalgic encephalomyelitis/ chronic fatigue syndrome (ME/CFS), the 2015 NAM report noted significant differences between CFS and ME definitions, stating, “Historically, however, **the diagnostic criteria for ME have required the presence of specific or different symptoms from those required by the diagnostic criteria for CFS; thus, a diagnosis of CFS is not equivalent to a diagnosis of ME.**”⁸ For instance, the Fukuda 1994 definition does not require post-exertional malaise, cognitive dysfunction or unrefreshing sleep, all required by the ME definitions and the new NAM criteria.

Further, postviral fatigue syndrome is an ill-defined term that has been used when symptoms have been present for periods less than 6 months whereas ME and CFS typically require symptom presence of more than 6 months.

Thus, the three diagnoses refer to different entities. We suggest the G93.3 title be amended to “Postviral fatigue syndrome, chronic fatigue syndrome and myalgic encephalomyelitis” with separate subcodes (G93.30, G93.31, G93.32) for each condition.

e) Exclusions: The following existing exclusions need to be revised to reflect the proposed classification and terminology changes: Excludes1 at G93.3 (Chronic fatigue syndrome NOS), Excludes1 at R53.82 (Postviral fatigue syndrome), Excludes2 at G04 (Encephalitis, myelitis and encephalomyelitis) and Excludes1 at A85 (Other viral encephalitis, not elsewhere classified.) An Excludes is also required for R53.82 (Chronic fatigue, unspecified).

f) Removal of the term “benign:” The term “benign” was originally included under the assumption that patients do not die. But further research has demonstrated that patients can die of the complications of ME/CFS⁹ and that the condition results in a high medical burden for patients. The NAM report noted that ME/CFS can be very debilitating and “frequently and dramatically limits the activities of affected patients.”¹⁰ People affected by ME/CFS experience, on average, lower health-related quality of life than people afflicted by major depression, multiple sclerosis, rheumatoid arthritis, and certain types of cancers.¹¹ Using the term “benign” downplays the effect of ME/CFS on patients; we recommend removing it from the tabular listing and adding to the index if needed to maintain backward compatibility.

Requested Modifications

Modifications to Tabular Listing:

	R53.8	Other malaise and fatigue
	R53.82	Chronic fatigue, unspecified
Delete	R53.82	Chronic fatigue syndrome NOS
Delete		Excludes1: Postviral fatigue syndrome (G93.3)
Add		Excludes1: Postviral fatigue syndrome, chronic fatigue syndrome and myalgic encephalomyelitis (G93.3)
	G93	Other disorders of brain
Revise	G93.3	Postviral fatigue syndrome, chronic fatigue syndrome and myalgic encephalomyelitis
Add	G93.30	Postviral fatigue syndrome
Delete	G93.3	Benign myalgic encephalomyelitis

Add	G93.31	Myalgic encephalomyelitis
Add	G93.32	Chronic fatigue syndrome
Delete		Excludes1: Chronic fatigue syndrome NOS (R53.82)
Add		Excludes1: Chronic fatigue (R53.82)

Other impacted ICD terms:

	A85: Other viral encephalitis, not elsewhere classified
Revise (title, code)	Excludes1: Benign Myalgic encephalomyelitis (G93.31)
	G04: Encephalitis, myelitis and encephalomyelitis
Revise (title, code)	Excludes2: Benign Myalgic encephalomyelitis (G93.31)

Modifications to Index Listing:

Delete	R53.82 Chronic fatigue syndrome
Add (if needed)	G93.31 Myalgic encephalomyelitis (benign)

Background: What is ME/CFS?

According to a 2015 report by the National Academy of Medicine (NAM), myalgic encephalomyelitis/chronic fatigue syndrome is “a serious, chronic, complex, and multisystem disease” characterized by impairment in the neurological, immunological, autonomic, and energy metabolism systems.¹² The disease is characterized by the hallmark symptom post-exertional malaise (PEM) in which even small amounts of cognitive and physical exertion can exacerbate symptoms and cause a loss of function that can last for days, weeks or sometimes months. ME/CFS is also characterized by cognitive issues, orthostatic intolerance, unrefreshing sleep, joint and muscle pain, headaches, sensitivity to noise and light, and other symptoms. The NAM report recommended the name “Systemic exertion intolerance disease” and established a definition which requires PEM in recognition of this hallmark characteristic. Currently, HHS agencies, including the US Centers for Disease Control and Prevention and the National Institutes of Health use the term “ME/CFS.” This proposal refers to the disease by that name

References

¹ The CFS definitions used in the US include the 1988 Holmes definition, the 1994 Fukuda definition, and the 2005 Reeves definition, which is based on Fukuda. The Oxford CFS definition, commonly used in the UK, requires only fatigue and no other symptoms. While not used in US research, the findings of these studies have been incorporated into US clinical guidance.

- Holmes G, Kaplan J, Gantz N, Komaroff A, Schonberger L, Straus S, Jones J, Dubois R, Cunningham-Rundles C, Pahwa S, Tosato G, Zegans L, Purtilo D, Brown N, Schooley R, Brus I. “Chronic Fatigue Syndrome: A Working Case Definition.” *Annals of Internal Medicine*. March 1, 1988; 108(3): 387-389. PMID: 2829679. <http://dx.doi.org/10.7326/0003-4819-108-3-387>
- Fukuda K, Straus SE, Hickie I, Sharpe MC, Dobbins JG, Komaroff A and the International Chronic Fatigue Syndrome Study Group. “The chronic fatigue syndrome: a comprehensive approach to its definition and study.” *Ann Intern Med* 1994; 121(12): 953-9. PMID: 7978722. <http://dx.doi.org/10.7326/0003-4819-121-12-199412150-00009>
- Reeves W, Wagner D, Nisenbaum R, Jones J, Gurbaxani B, Solomon L, Papanicolaou D, Unger E, Vernon S, Heim C. “Chronic Fatigue Syndrome – A clinically empirical approach to its definition and study.” *BMC Medicine* December 2005; 3:19. PMID: 16356178. <http://dx.doi.org/10.1186/1741-7015-3-19>

² National Academy of Medicine. Beyond Myalgic Encephalomyelitis/Chronic Fatigue Syndrome: Redefining an Illness. *The National Academies Press*; May 2015. <https://www.nap.edu/catalog/19012/beyond-myalgic-encephalomyelitischronic-fatigue-syndrome-redefining-an-illness> Page 74, 75, 77

³ Evidence of neurological impairment submitted to WHO in March 2017 in a proposal for the G93.3 terms in ICD-11.

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- Evidence Supporting a Classification of ME and CFS in the Neurological Chapter 2017. <http://bit.ly/2mwLvYE>
 - Komaroff AL, Cho TA. Role of infection and neurologic dysfunction in chronic fatigue syndrome. *Semin Neurol* July 2011. <http://dx.doi.org/10.1055/s-0031-1287654>
 - Komaroff A. Presentation for U.S. disease organization *Solve ME/CFS Initiative*. November 2016. <https://youtu.be/ENrdrGjOp-c?t=555> Minute 14:23, 48:46. Presentation slides: <http://solvecfs.org/wp-content/uploads/2016/11/SolveME-CFS-Webinar-Dr-Komaroff.pdf>
- ⁴ Fourth Meeting of the JLMMS Task Force, Queensland, Australia, 11-14 July 2016
http://www.who.int/entity/classifications/icd/revision/2016.07.11-14_iSummaryMeetingReportQueensland.pdf
- ⁵ Personal correspondence between UK advocate Suzy Chapman and Dr. Robert Jakob of the World Health Organization, March 17, 2017
- ⁶ Proposal for Chronic Fatigue Syndrome. September 14, 2011. National Center for Health Statistics
<https://www.cdc.gov/nchs/data/icd/topicpacketforsept2011a.pdf> “The meeting proposal states, “In ICD-10-CM chronic fatigue syndrome NOS (that is not specified as being due to a past viral infection) was added to ICD-10-CM in Chapter 18 at R53.82, Chronic fatigue, unspecified.”
- ⁷ Ibid. Page 10. The meeting proposal states, “ICD-10-CM retained code G93.3 to allow the differentiation of cases of fatigue syndrome where the physician has determined the cause as being due to a past viral infection from cases where the physician has not established a post viral link.”
- ⁸ National Academy of Medicine. Beyond Myalgic Encephalomyelitis/Chronic Fatigue Syndrome: Redefining an Illness. *The National Academies Press*; May 2015. <https://www.nap.edu/catalog/19012/beyond-myalgic-encephalomyelitischronic-fatigue-syndrome-redefining-an-illness>. Page 60. Also see the following for an overview of the definitions used for myalgic encephalomyelitis and chronic fatigue syndrome. <http://bit.ly/2niiKfu>
- ⁹ International Association of CFS/ME Chronic fatigue syndrome/myalgic encephalomyelitis Primer for Clinical Practitioners Primer. 2014. http://iacfsme.org/portals/0/pdf/Primer_Post_2014_conference.pdf
- ¹⁰ National Academy of Medicine. Beyond Myalgic Encephalomyelitis/Chronic Fatigue Syndrome: Redefining an Illness. *The National Academies Press*; May 2015. <https://www.nap.edu/catalog/19012/beyond-myalgic-encephalomyelitischronic-fatigue-syndrome-redefining-an-illness> Page 209
- ¹¹ Hvidberg MF, Brinth LS, Olesen AV, Petersen KD, Ehlers L. 2015. The Health-Related Quality of Life for Patients with Myalgic Encephalomyelitis / Chronic Fatigue Syndrome (ME/CFS). *PLOS ONE* 10(7): e0132421.
<http://journals.plos.org/plosone/article?id=10.1371/journal.pone.0132421>
- Also see
- Nacul L, Lacerda E, Campion P, Pheby D, Drachler M, Leite J, Poland F, Howe A, Fayyaz S, Molokhia M. “The functional status and well being of people with myalgic encephalomyelitis/chronic fatigue syndrome and their carers” *BMC Public Health* May 2011. <https://bmcpublichealth.biomedcentral.com/articles/10.1186/1471-2458-11-402>
- ¹² National Academy of Medicine. Beyond Myalgic Encephalomyelitis/Chronic Fatigue Syndrome: Redefining an Illness. *The National Academies Press*; May 2015. <https://www.nap.edu/catalog/19012/beyond-myalgic-encephalomyelitischronic-fatigue-syndrome-redefining-an-illness> Quote on page 209. See the full report and the following reference for a discussion of the nature of the multi-system impairments
- Komaroff A. 12th International IACFS/ME Conference Emerging Science & Clinical Care. October 2016. <http://iacfsme.org/Conferences/2016-Fort-Lauderdale/Agenda/Dr-Komaroff-s-2016-Summary-Slides/IACFSME-Komaroff-Slides.asp>