

Update on the classification of PVFS, ME and CFS for ICD-11

ICD-11 Report One | November 2019

Q: What is the “ICD”?

The International Classification of Diseases (ICD) is the global standard diagnostic classification of diseases for use in epidemiology, health management and clinical practice. ICD is maintained and published by the World Health Organization (WHO).

It is used to classify and monitor causes of injury and mortality (death) and to monitor morbidity (illness and disease) trends for analysing data compiled from many types of health and other records, including death certificates and insurance.

More than 100 countries (WHO Member States) use the ICD system to report mortality data. ICD is increasingly used in clinical care and research to define diseases, study disease patterns, manage health care, monitor outcomes and inform policy makers on health related spending and resource allocation.

Q: What is the current revision of ICD?

The current revision of ICD is the 10th Revision [1]. The WHO has said that the final update for ICD-10 will be the 2019 release but Member States can continue to use ICD-10 until they have prepared their health systems for transition to ICD-11.

Q: How are the terms: postviral fatigue syndrome; myalgic encephalomyelitis; and chronic fatigue syndrome currently classified in ICD-10?

Image 1: ICD-10 Browser Version: 2016, Accessed November 01, 2019:

<https://icd.who.int/browse10/2016/en#/G93.3>

ICD-10 Version:2016

Search [Advanced Search]

ICD-10 Versions - La

G90-G99 Other disorders of the nervous system

- ▶ G90 Disorders of autonomic nervous system
- ▶ G91 Hydrocephalus
- ▶ G92 Toxic encephalopathy
- ▶ **G93 Other disorders of brain**
 - G93.0 Cerebral cysts
 - G93.1 Anoxic brain damage, not elsewhere classified
 - G93.2 Benign intracranial hypertension
 - G93.3 Postviral fatigue syndrome**
 - G93.4 Encephalopathy, unspecified
 - G93.5 Compression of brain
 - G93.6 Cerebral oedema
 - G93.7 Reye syndrome

G93.2 Benign intracranial hypertension
Excl.: hypertensive encephalopathy

G93.3 Postviral fatigue syndrome
Benign myalgic encephalomyelitis

G93.4 Encephalopathy, unspecified
Excl.: encephalopathy:

- alcoholic (G31.2)
- toxic (G92)

G93.5 Compression of brain
Compression | of brain (stem)
Herniation

Excl.: traumatic compression of brain
traumatic compression of brain

- focal (S06.3)

Image location: <https://dxrevisionwatch.files.wordpress.com/2019/10/image-1.png>

Postviral fatigue syndrome is the ICD-10 Concept Title term. It is classified in **Volume 1: Tabular List** in *Chapter VI: Diseases of the nervous system* (the neurological chapter) at code G93.3, under category block G90-G99 Other disorders of the nervous system > G93 Other disorders of brain.

Benign myalgic encephalomyelitis is the inclusion term under G93.3 Postviral fatigue syndrome.

There is an exclusion for “postviral fatigue syndrome (G93.3)” under F48.0 Neurasthenia and its inclusion, Fatigue syndrome. There is also an exclusion for “fatigue syndrome • postviral (G93.3)” under R53 Malaise and fatigue [2].

Chronic fatigue syndrome is not included in the Tabular List but is indexed to the G93.3 code in the **Alphabetical Index (Volume 3)**.

Q: Are all Member States using the same version of ICD-10?

No. Around 25 Member States have been licensed by the WHO to adapt the ICD-10 classification for the collection of morbidity (illness and disease) data. These are known as “national modifications” or “clinical modifications” and their category content and code structure varies from the WHO’s unmodified ICD-10.

Member States that have developed clinical modifications of ICD-10 include: Canada, Germany, the United States (also used by Belgium, Luxembourg and Spain), Australia (also used by Ireland and Slovenia) and Thailand.

Q: How do Canada, Germany and the United States classify the G93.3 terms?

Canada (ICD-10-CA): Postviral fatigue syndrome is the G93.3 Tabular List Concept Title. Benign myalgic encephalomyelitis; and Chronic fatigue syndrome are both inclusions under G93.3 in the Tabular List.

Germany (ICD-10-GM): Chronisches Müdigkeitssyndrom [Chronic fatigue syndrome] is the G93.3 Tabular List Concept Title. Chronisches Müdigkeitssyndrom bei Immundysfunktion; Myalgische Enzephalomyelitis; and Postvirales Müdigkeitssyndrom are all inclusions under G93.3 in the Tabular List.

United States (ICD-10-CM): Postviral fatigue syndrome is the G93.3 Tabular List Concept Title. Benign myalgic encephalomyelitis is the inclusion under G93.3 in the Tabular List.

But Chronic fatigue syndrome (as “chronic fatigue syndrome NOS”) is classified in the Tabular List in *Chapter 18: Symptoms, signs and abnormal clinical and laboratory findings, not elsewhere classified* as an inclusion under code R53.82 Chronic fatigue, unspecified. (NOS = Not Otherwise Specified.)

There is an Excludes1: postviral fatigue syndrome (G93.3) exclusion under the R53.82 code and a reciprocal Excludes1: chronic fatigue syndrome NOS (R53.82) exclusion under the G93.3 code.

Note: The United States’ ICD-10-CM (which has been adopted for use by Belgium, Luxembourg and parts of Spain) is the only modification of ICD-10 that classifies chronic fatigue syndrome (as “chronic fatigue syndrome NOS”) under the *Symptoms, signs* chapter.

2 PDF versions of ICD-10 (Fifth edition 2016) Volume 1: Tabular List and Volume 3: Alphabetical Index are now available as free downloads from the WHO’s IRIS document archive: <https://apps.who.int/iris/handle/10665/246208>

(For listing of PVFS and BME, see p376) ICD-10 Volume 1: Tabular List (9.952 Mb): <https://apps.who.int/iris/bitstream/handle/10665/246208/9789241549165-V1-eng.pdf>

(For indexing of CFS, see p604) ICD-10 Volume 3: Alphabetical Index (3.110 Mb): <https://apps.who.int/iris/bitstream/handle/10665/246208/9789241549165-V3-eng.pdf>

Q: How have the G93.3 terms been classified for ICD-11?

From 2008, the WHO began assembling external working groups (known as “Topic Advisory Groups” or “TAGs”). Topic Advisory Groups reported directly to the Revision Steering Group and were tasked with reviewing ICD-10 chapter content and structure, conducting literature reviews, making recommendations for revisions, proposing additions for ICD-11 and editing description texts.

The role of the TAGs and their sub working groups was advisory and final decisions rested with WHO classification experts and the Revision Steering Group; later with the Revision Task Force, and more recently with the Classifications and Statistics Advisory Committee (CSAC) and the Medical Scientific Advisory Committee (MSAC).

The TAG with responsibility for the G93.3 terms was **TAG Neurology**, chaired by Prof Raad Shakir.

A full account of the tortuous progression of the G93.3 terms through the nine year long Alpha and Beta drafting stages and the advocacy undertaken is beyond the scope of this update. But in summary:

- As the ICD-11 Beta draft stood in early 2013, TAG Neurology was recommending that Chronic fatigue syndrome should replace Postviral fatigue syndrome as Concept Title term, with Benign myalgic encephalomyelitis retained as the specified inclusion. Postviral fatigue syndrome was recommended to be relocated under a list of 14 historical and alternative Synonyms/Index terms.
- In early 2013, all three terms were unaccountably removed from view in the public version of the Beta drafting platform, making it very difficult to monitor, input or comment on proposals.
- In March 2017, the terms were restored to the public version of the Beta drafting platform under parent block: Other disorders of the nervous system, with Postviral fatigue syndrome as the Concept Title. Benign myalgic encephalomyelitis; and chronic fatigue syndrome were both specified as inclusion terms. I was informed by Dr Robert Jakob that, *“While the optimal place in the classification is still being identified, the entity has been put back to its original place in ICD.”*
- In March 2017, Chapman & Dimmock submitted a detailed proposal and rationale requesting, among other recommendations, that Postviral fatigue syndrome should be removed as the Concept Title term and that Myalgic encephalomyelitis; and Chronic fatigue syndrome should be elevated to Concept Titles and a unique code assigned to each term.
- In March 2017, Lily Chu MD submitted a proposal on behalf of the IACFS/ME, requesting that all three terms should be elevated to Concept Titles and a unique code assigned to each term.

Dr Tarun Dua is a medical officer working on the *Program for Neurological Diseases and Neuroscience, Management of Mental and Brain Disorders*, Department of Mental Health and Substance Abuse, World Health Organization, and had served as Lead WHO Secretariat and Managing Editor to TAG Neurology. TAG Neurology had ceased operating in October 2016 when a number of TAGs were stood down.

- In November 2017, Dr Dua submitted a proposal which she later clarified was being submitted on behalf of TAG Neurology and which she said, *“reiterates the TAG’s earlier conclusions.”*

Since the WHO had stated that decisions about potential changes to these terms were on hold pending completion of the literature review, it was unclear how this new proposal sat in relation to the review, which was understood to be still in progress. It was also unclear if this submission was supported by the WHO/ICD Revision or represented only the position of TAG Neurology and its former Managing Editor.

Dr Dua’s proposal recommended that “Myalgic encephalitis/Chronic Fatigue Syndrome (ME/CFS) [sic] should be removed from the *Diseases of the nervous system* chapter and reclassified in *Chapter 21: Symptoms, signs or clinical findings, not elsewhere classified*, as a child category under parent block: *Symptoms, signs or clinical findings of the musculoskeletal system*.

- **In November 2018, the WHO rejected Dr Dua's proposal.** In their brief rationale for this decision, the WHO stated that following completion of their literature review, they concluded that Postviral fatigue syndrome should remain the indexing target, that there was currently no evidence to suggest a better place to relocate these terms and that the terms would remain classified within the *Diseases of the nervous system* chapter.

The WHO further confirmed their decision was supported by the Classifications and Statistics Advisory Committee (CSAC) and the Medical Scientific Advisory Committee (MSAC). Discussion of this topic was also noted in a November 2018 presentation given by WHO's Dr Robert Jakob and referenced in the Summary Report of the October 2018 WHO-FIC Network Annual Meeting.

To recap: Postviral fatigue syndrome remains classified in the *Diseases of the nervous system* chapter (**Chapter 08 in ICD-11**). Postviral fatigue syndrome's new code is **8E49** and its parent block is **Other disorders of the nervous system**. (The ICD-10 parent class "Other disorders of brain" has been retired for ICD-11.)

Benign myalgic encephalomyelitis and **chronic fatigue syndrome** are both specified as inclusion terms within the ICD-11 Foundation and ICD-11's equivalent to the Tabular List, and assigned code **8E49**.

The **8E49** code maps directly to the ICD-10 **G93.3** code in the ICD-11 to ICD-10 Mapping Tables.

Image 2: ICD-11 for Mortality and Morbidity Statistics (Version: 04/2019), Accessed November 01, 2019:
<https://icd.who.int/browse11/l-m/en#/http%3a%2f%2fid.who.int%2fcd%2fentity%2f569175314>

ICD-11 for Mortality and Morbidity Statistics (Version : 04 / 2019)

Search [Advanced Search] Browse Coding Tool

- ▶ Disorders of consciousness
- ▼ **Other disorders of the nervous system**
 - ▶ 8E40 Disorders of the meninges excluding infection
 - ▶ 8E41 Pachymeningitis
 - ▶ 8E42 Superficial siderosis of the nervous system
 - ▶ 8E43 Pain disorders
 - ▶ 8E44 Post anoxic brain damage
 - ▶ 8E45 Locked-in syndrome
 - ▶ 8E46 Reye syndrome
 - ▶ 8E47 Encephalopathy, not elsewhere classified
 - ▶ 8E48 Encephalitis, not elsewhere classified
 - 8E49 Postviral fatigue syndrome**
 - ▶ 8E4A Paraneoplastic or autoimmune disorders of the nervous system
 - MH10 Brain death
 - 4B20.3 Neurosarcoidosis

Foundation Id : <http://id.who.int/icd/entity/569175314>

8E49 Postviral fatigue syndrome

All ancestors up to top

- 08 Diseases of the nervous system
- Other disorders of the nervous system
- 8E49 Postviral fatigue syndrome

Inclusions

- Benign myalgic encephalomyelitis
- chronic fatigue syndrome

Exclusions

- **Fatigue (MG22)**

Image location: <https://dxrevisionwatch.files.wordpress.com/2019/10/image-2.png>

The current version of the "Blue Browser" does not display the list of 14 Synonyms/Index terms but these are viewable in the "Orange Browser" (click on "Show all index terms [14]" to display the index term list):
<https://icd.who.int/dev11/l-m/en#/http://id.who.int/icd/entity/569175314>

The **Coding Tool** is the ICD-11 Index. All 14 index terms are searchable in the database and direct coders and clinicians to the 8E49 code: https://icd.who.int/ct11/icd11_mms/en/release

Exclusions under Fatigue: For ICD-11, the ICD-10 category R53 Malaise and fatigue has been revised to MG22 Fatigue. For ICD-10, there is an exclusion for the G93.3 code under R53 Malaise and fatigue but this exclusion had not been carried forward to the ICD-11 Alpha and Beta drafts.

In December 2014, I submitted proposals for exclusions for Postviral fatigue syndrome; (Benign) myalgic encephalomyelitis; and Chronic fatigue syndrome under MG22 Fatigue. These proposals for exclusions were approved and implemented and can be seen at the end of the Exclusions list at:

<https://icd.who.int/dev11/f/en#http%3a%2f%2fid.who.int%2fid%2fentity%2f1109546957>

In January 2018, I submitted a proposal and rationale for deletion of the ICD-10-CM term, “chronic fatigue, unspecified” from the 8E49 Index terms list. This was approved and implemented in March 2019.

Q: Which proposals have not been approved?

The proposals submitted by Chapman & Dimmock (March 2017) for removing PVFS as Concept Title and assigning two unique Concept Title codes for ME; and CFS; and by Lily Chu MD on behalf of IACFS/ME (March 2017) for assigning three unique Concept Title codes for PVFS; ME; and CFS were not approved.

The proposals submitted by Chapman & Dimmock (March 2017) and by Lily Chu MD on behalf of IACFS/ME (March 2017) for removing the word “Benign” from “Benign myalgic encephalomyelitis” were not approved. (Note: the terms “myalgic encephalomyelitis” and “ME - [myalgic encephalomyelitis]” are included under the Synonyms/Index terms list and are assigned the 8E49 code in the Coding Tool.)

The proposals submitted by Chapman (March 2017), Chapman & Dimmock (March 2017) and by Lily Chu MD on behalf of IACFS/ME (March 2017) for exclusions for the 8E49 terms under 6C20 Bodily distress disorder and for exclusion of 6C20 Bodily distress disorder under 8E49 were not approved.

A Table setting out the status of proposals for PVFS, BME, CFS, and BDD can be downloaded from:

ICD-11 Proposal Mechanism, status of processed proposals at 01.11.19 v4:

<https://dxrevisionwatch.files.wordpress.com/2019/10/outcome-of-icd-11-processed-proposals-v4.pdf>

Q: Are definitions for the 8E49 terms included in ICD-11?

In ICD-10, there are no description texts or definitions for any of the G93.3 terms. For ICD-10, only disorders coded in *Chapter V Mental and behavioural disorders* have description texts.

For ICD-11, the convention is that coded terms *in all chapters* will have description texts but the textual content for many of these “Description” fields has yet to be drafted. At the time of compiling this report, there is no “Description” or “Definition” text approved or proposed for 8E49 Postviral fatigue syndrome or either of its inclusion terms.

Q: When will Member States start using ICD-11?

The WHO published a stable version of ICD-11 in June 2018. This version was presented as an “advance preview” to enable Member States to start planning for implementation, prepare national translations and begin training health professionals. An updated version was released in April 2019.

On May 25, 2019, the 72nd World Health Assembly (WHA) voted unanimously to adopt ICD-11. WHA’s endorsement of the new edition won’t come into effect until **January 01, 2022, which is the earliest date from which Member States can begin using ICD-11 for reporting data.**

Note that there is no mandatory implementation date — Member States will transition from ICD-10 to ICD-11 at their own pace and according to their countries’ specific timelines, requirements and resources. Global adoption of ICD-11 is anticipated to be a prolonged process and during this period of transition, the WHO will be accepting data recorded using both ICD-10 and the new ICD-11 code sets.

No countries have announced implementation schedules yet. The WHO anticipates early implementers will take several years to evaluate the new edition, determine how they will use ICD-11, produce training and implementation materials, complete translations and prepare their health systems for migration.

Q: Where can I view ICD-11?

The “Blue Browser” for **ICD-11 for Mortality and Morbidity Statistics (Version: 04/2019)** can be accessed here: <https://icd.who.int/browse11/l-m/en>

The **ICD-11 Coding Tool** (the electronic index and search engine for coders and clinicians for looking up disease terms and their associated codes) is here: https://icd.who.int/ct11/icd11_mms/en/release

The ICD-11 “Blue Browser” does not currently display the index terms. These can be seen in the **Update and Maintenance Platform** (the “Orange Browser”) or searched for in the Coding Tool:

Update and Maintenance Platform Foundation Component: <https://icd.who.int/dev11/f/en/>

Update and Maintenance Platform MMS Linearization: <https://icd.who.int/dev11/l-m/en/>

Q: Has the content of ICD-11 been completed?

No. Many of the Description field texts have still to be completed. There remain hundreds of proposals in the ICD-11 Proposal Mechanism waiting to be processed. None of the specialty or derivative publications, for example, the *ICD-11 Clinical Descriptions and Diagnostic Guidelines for Mental and Behavioural Disorders (CDDG)* or the proposed ICD-11 PHC guideline for 27 mental disorders, have been released.

Update and revision cycle: Official releases of ICD-11 will be produced annually for international use in mortality and morbidity. Approved proposals for updates, additions and corrections will be incorporated into ICD-11 on an annual or five yearly cycle, according to their potential impact on data collection.

Information in this report is correct at November 01, 2019 and based on ICD-11 MMS Version: 04/2019.

My next ICD-11 update will be **Report Two: Update on ICD-11 and Bodily distress disorder.**

Please note that I have retired from active advocacy work around classifications and coding. It is crucial that advocates, patient groups and their professional allies inform themselves, become confident around the various classification and terminology systems and take responsibility, as stakeholders, for submitting timely responses and requests for changes, because I shall no longer be doing this work.

For a summary of the classification and terminology systems that are going to require close monitoring in the future and potential input from stakeholders see my Dx Revision Watch post:

PVFS, ME and CFS in classification and terminology systems: notes for the future:

<https://dxrevisionwatch.com/2019/06/30/pvfs-me-and-cfs-in-classification-and-terminology-systems-notes-for-the-future/>

Dedicated to the memory of Drs Maarten “Maartensz”, Amsterdam, Netherlands, who died suddenly in August 2019.

Suzy Chapman, Dx Revision Watch

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