

## Update for patients and practitioners: The ICD-11 and Lyme borreliosis

Jenna Luché-Thayer.  
August 13, 2023

ICD-10 codes for Lyme borreliosis were limited. The ICD-11 recognizes many complications from Lyme borreliosis infection, including complications that can disable and cause death. *(See the table below for the new Lyme ICD-11 codes.)*

ICD10	<i>ICD-11 is far more comprehensive, allowing almost all complications from Lyme to be represented and hopefully result in faster diagnosis, covered care and extended treatment and disability support as needed.</i>	
A69.2 Lyme Disease	1C1G	Lyme borreliosis
M01.2 Arthritis due to Lyme	1C1G.0	Early cutaneous Lyme borreliosis
G01 Meningitis due to Lyme	1C1G.1	Disseminated Lyme borreliosis
G63.0 Polyneuropathy due to Lyme	1C1G.10	Lyme Neuroborreliosis
	1C1G.11	Lyme Carditis
	1C1G.12	Ophthalmic Lyme borreliosis
	1C1G.13	Lyme arthritis
	1C1G.14	Late cutaneous Lyme borreliosis
	1C1G.1Y	Other specified disseminated Lyme borreliosis
	1C1G.1Z	Disseminated Lyme borreliosis, unspecified
	1C1GY	Other specified Lyme borreliosis
	6D85.Y	Dementia due to Lyme Disease
	9C20.1	Infectious panuveitis in Lyme disease
	9B66.1	Infectious intermediate uveitis in Lyme disease
	8A45.0Y	Other Specified white matter disorders due to infections; Central Nervous System demyelination due to Lyme borreliosis
	"KA6Y Other specified infections of the fetus or newborn" can be coordinated with "XN13C Borrelia Burgdorferi" to represent Congenital Lyme.	

These updated ICD-11 codes were developed over a multiyear multi-stakeholder scientific review process and represent consensus among 194 World Health Organization (WHO) member states, including Poland. WHO added 15 new medical diagnostic codes for Lyme disease (aka borreliosis) complications, effective on January 1, 2022.

These codes benefit patients by validating many complications that were previously unrepresented. The codes can also better serve researchers and policy makers attempting to track, understand and address the depth and breadth of health harms caused by this infection.

- For decades, the CDC's Lyme case definition emphasizes arthritis as the signifying complication. In contrast, of the fifteen new codes, six describe infection in the central nervous system.
- The ICD-11 demonstrates and confirms Lyme has affinity for 'immune privileged sites' such as the central nervous system.
- Five of the fifteen codes identify complications documented as life threatening: Lyme Neuroborreliosis, Lyme Carditis, Congenital Lyme borreliosis, Dementia due to Lyme Disease, and Central Nervous System demyelination due to Lyme borreliosis.
- ICD-11 codes for Lyme now describe a disease which may cause severe and potentially fatal central nervous system complications and is passed from pregnant mother to fetus.
- Many of the new codes describe late stage and systemic complications. These codes demonstrate the infection is not easy to diagnose and underscore the unreliability of the recommended serology diagnostics — meaning they do not capture infection.
- The multiple codes for late stage and systemic complications demonstrate widespread treatment failure following the standard short-term antibiotics.
- The need for multiple codes for late stage and systemic complications underscore the infection is not easy to treat nor cure.
- Post Treatment Lyme disease syndrome (PTLDS) is a vague term used to describe 'patients who have symptoms like fatigue, pain and joint and muscle aches after the treatment of Lyme disease'.

PTLDS does not capture the many complications of this infection and is not recognized in ICD-11, nor in the previous versions of the ICD.

Furthermore, the theoretical basis for this syndrome is a psychosomatic condition which has been repudiated by the American Psychiatric Association and deleted from their guidance.

### **Congenital Lyme is representable in ICD-11**

Post-coordination is being used extensively in ICD-11. Correspondence from WHO's Director-General for Metrics and Measurement (Dr Lubna Abdulrahman Alansari) and WHO's ICD-11 Medical and Scientific Advisory Committee to Jenna Luche-Thayer stated Congenital Lyme is represented via ICD-11 with post

coordination of codes. WHO stated that appropriate post-coordination options would be incorporated explicitly into ICD for neonatal infections where the organism is not specified. For example: "KA6Y Other specified infections of the fetus or newborn" can be coordinated with "XN13C *Borrelia Burgdorferi*"

KA6Y can also be post coordinated with *other pathogenic Borrelia infections* that cause congenital complications. For example: XN5R4 *Borrelia recurrentes* (relapsing fever borreliosis) also has congenital complications. See "Tickborne Relapsing Fever in a Mother and Newborn Child — Colorado, 2011 Weekly" [https://www.cdc.gov/mmwr/preview/mmwrhtml/mm6110a3.htm?s\\_cid=mm6110a3\\_w](https://www.cdc.gov/mmwr/preview/mmwrhtml/mm6110a3.htm?s_cid=mm6110a3_w)

Other post coordination codes are options. For example: KA61 Other bacterial infections of the fetus or newborn; KA65 Neonatal infections of certain specified sites and/or KA00 Fetus or newborn affected by maternal infectious diseases.

Please share this information regarding the new codes with your practitioners. The information regarding these codes is not, nor should be considered, or used as a substitute for, medical advice, diagnosis, or treatment. Advice on the testing, treatment or care of an individual patient should be obtained through consultation with a physician who has examined that patient or is familiar with that patient's medical history.