LYME BORRELIOSIS AS PRESENTED IN THE ICD11

For decades, the World Health Organization (WHO) has recognized Lyme borreliosis 'as a disease of consequence'. Recognition of the potential morbidity and mortality of Lyme borreliosis has increased in the WHO's eleventh revision of the International Classification of Diseases Codes (ICD11). ICD11 now recognize fifteen complications from Lyme borreliosis [1] whereas the ICD10 recognized three complications from the disease [2].

ICD10 Codes: Lyme borreliosis	ICD11 Codes: Lyme borreliosis
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
	1C1G.2 Congenital Lyme borreliosis
	1C1GY Other specified Lyme borreliosis
	6D85.Y Dementia due to other specified diseases classified elsewhere Dementia due to Lyme Disease
	9C20.1 Infectious panuveitis Infectious panuveitis in Lyme disease
	9B66.1 Infectious intermediate Chorioditis Infectious intermediate uveitis in Lyme disease
	8A45.0Y Other Specified white matter disorders due to infections Central Nervous System demyelination due to Lyme borreliosis

In ICD11, six of the 15 codes for Lyme specify complications of central nervous system: 1C1G.10 Lyme Neuroborreliosis; 1C1G.12 Ophthalmic Lyme borreliosis; 6D85.Y Dementia due to Lyme Disease; 9C20.1 Infectious panuveitis in Lyme disease; 9B66.1 Infectious intermediate uveitis in Lyme disease; and 8A45.0Y Central Nervous System demyelination due to Lyme borreliosis.

Furthermore, five of the codes identify complications documented as life threatening: 1C1G.10 Lyme Neuroborreliosis; 1C1G.11Lyme Carditis; 1C1G.2 Congenital Lyme borreliosis; 6D85.Y Dementia due to Lyme Disease; and 8A45.0Y Central Nervous System demyelination due to Lyme borreliosis.

In the ICD11, there is instruction on how add associated conditions to the primary codes, such as 1C1G Lyme borreliosis,

“Coding Note: Use additional code if desired, to identify any associated condition. Use additional code, if desired, to identify any sequelae. The extension code 'Cause of late effect' is used in addition to both codes to show the relationship between the causative condition and the resulting sequelae.”

Postcoordination: Add detail to Lyme borreliosis; Use additional code, if desired, examples are:

1D00.0 Bacterial encephalitis
1D01.0 Bacterial meningitis
1D01.0Y Other specified bacterial meningitis
1D01.0Z Bacterial meningitis, unspecified
1D02.0 Bacterial myelitis
BC42.1 Infectious myocarditis
8B88.0 Bell palsy
9A10.0 Infections of the lacrimal gland
9C20.2 Purulent endophthalmitis”

However, there are many associated conditions that can be added to show Lyme borreliosis sequalea and promote recognition for diagnosis and effective treatment as as well epidemiological purposes. The next table gives two examples; there are many more.

Postcoordination	Supporting references
'Meningo-vascular borreliosis and neuroborreliosis — with cerebral infarcts and/or intracranial aneurysm' … … can be a sequalea added under … **8A45.0Y Other specified white matter disorders due to infections**	J, Gahn G, von Kummer R, Reichmann H. Cerebral vasculitis with multiple infarcts caused by lyme disease. *Cerebrovasc Dis*. 2004;17:79–81. Schmitt AB, Kuker W, Nacimiento W. [Neuroborreliosis with extensive cerebral vasculitis and multiple cerebral infarcts]. [in German]. *Nervenarzt*. 1999;70:167–71. Shadick NA, Phillips CB, Logigian EL, et al. The long-term clinical outcomes of Lyme disease. A population-based retrospective cohort study. *Ann Intern Med*. 1994;121:560–7. Sparsa L, Blanc F, Lauer V, Cretin B, Marescaux C, Wolff V. Recurrent ischemic strokes revealing Lyme meningovascularitis. *Rev Neurol (Paris)* 2009;165:273–7. Topakian R, Stieglbauer K, Aichner FT. Unexplained cerebral vasculitis and stroke: keep Lyme neuroborreliosis in mind. *Lancet Neurol*. 2007;6:756–7. Uldry PA, Regli F, Bogousslavsky J. Cerebral angiopathy and recurrent strokes following Borrelia burgdorferi infection. *J Neurol Neurosurg Psychiat*. 1987;50:1703–4. Van Snick S, Duprez TP, Kabamba B, Van De Wyngaert FA, Sindic CJ. Acute ischaemic pontine stroke revealing lyme neuroborreliosis in a young adult. *Acta Neurol Belg*. 2008;108:103–6 Veenendaal-Hilbers JA, Perquin WV, Hoogland PH, Doornbos L. Basal meningovasculitis and occlusion of the basilar artery in two cases of Borrelia burgdorferi infection. *Neurology*. 1988;38:1317–9. Wittwer B, Pelletier S, Ducrocq X, Maillard L, Mione G, Richard S. Cerebrovascular Events in Lyme Neuroborreliosis. *J Stroke Cerebrovasc Dis*. 2015 Jul;24(7):1671-8. doi: 10.1016/j.jstrokecerebrovasdis.2015.03.056. Zajkowska J, Garkowski A, Moniuszko A et al. Vasculitis and stroke due to Lyme neuroborreliosis – a review. *Infectious Diseases*. 2014;47(1):1-6. doi:10.3109/00365548.2014.961544. Zhang Y, Lafontant G, Bonner FJ., Jr Lyme neuroborreliosis mimics stroke: a case report. *Arch phys Med Rehab*. 2000;81:519–21.

FB30 Infectious Myositis Bacterial myositis	Brtkova J, Jirickova P, Kapla J, Dedic K, Pliskova L. Borrelia arthritis and chronic myositis accompanied by typical chronic dermatitis. JBR-BTR. 2008;91(3):88-9. Carvounis PE, Mehta AP, Geist CE. Orbital myositis associated with Borrelia burgdorferi (Lyme disease) infection. Ophthalmology. 2004;111(5):1023-8. DOI: 10.1016/j.ophtha.2003.08.032. Holak H, Holak N, Huzarska M, Holak S. Tick inoculation in an eyelid region: report on five cases with one complication of the orbital myositis associated with Lyme borreliosis. Klin Oczna. 2006;108(4-6):220-4. Holmgren AR, Matteson EL. Lyme myositis. Arthritis Rheum. 2006;54(8):2697- 700. Sauer A, Speeg-Schatz C, Hansmann Y. Two cases of orbital myositis as a rare feature of lyme borreliosis. Case Rep Infect Dis. 2011;2011:372470. doi: 10.1155/2011/372470. Waton J, Pinault AL, Pouaha J, Truchetet F. [Lyme disease could mimic dermatomyositis]. [Article in French] Rev Med Interne. 2007 May;28(5):343-5.

SIGNIFICANCE OF THE CHANGES BETWEEN ICD10 AND ICD11

The changes between ICD10 and ICD11 are significant on many levels as unlike the disease described in ICD10, the disease presented in ICD11 is not '*simple to diagnose, treat and cure*'.

Changes in ICD11 reveal this infection to have a number of potentially fatal outcomes and demonstrates its affinity for 'immune privileged sites' and the central nervous system. Furthermore, many of the complications validated in the ICD11 codes are the result of undiagnosed later stage infection. Later stage complications indicate:

- Common Lyme diagnostic technologies are unreliable
    - Some pathogenic strains belonging to the *B. burgdorferi* sl (Lyme borreliosis) complex have a worldwide distribution, yet they are rarely tested for or considered in diagnoses [3]
    - Standard recommended diagnostic tests have an approximate accuracy of 50 percent for males and 40 percent accuracy rate for females [4] [5] [6]

- Globally promoted standard treatment show treatment failure for 20 percent in the acute phase of the disease and approximately 36 percent of those infected develop long-term illness [7] [8] [9]

- Healthcare professionals need updated training to perform clinical diagnosis, make use of treatment options that have met internationally accepted standards and treat the disease according to patient response [7]

- The current case definition for epidemiological purposes may only capture a fraction of the cases of this 'notifiable disease'

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Endnotes

[1] https://icd.who.int/browse11/l-m/en#/ICD11 Codes for Lyme borreliosis
[2] http://apps.who.int/classifications/icd10/browse/2010/en
[3] Perronne C (2014) Lyme and associated tick-borne diseases: Global challenges of Lyme disease. Front. Cell. Infect. Microbiol. 4:74
[4] Cook M, Puri B. Commercial test kits for detection of Lyme borreliosis: A meta-analysis of test accuracy. International Journal of General Medicine. 2016; Volume 9:427–440. doi:10.2147/ijgm.s122313
[5] Schwarzwalder A1, Schneider MF, Lydecker A, Aucott JN Sex differences in the clinical and serologic presentation of early Lyme disease: Results from a retrospective review. Gend Med. 2010 Aug;7(4):320-9. doi: 10.1016/ j.genm.2010.08.002.
[6] Stricker RB, Johnson L. Let's tackle the testing. Letter. 2007;335(7628):1008. doi:10.1136/bmj.39394. 676227.BE. http://dx.doi.org/10.1136/bmj.39394.676227.BE. Accessed February 19, 2017
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[8] Aucott JN, Rebman AW, Crowder LA, Kortte KB. Post-treatment Lyme disease syndrome symptomatology and the impact on lifefunctioning: is there something here? Qual Life Res. 2013 Feb; 22(1): 75–84. Published online 2012 Feb 1. doi: 10.1007/s11136-012-0126-6 PMCID: PMC3548099
[9] Many State Actors and medical societies base their Lyme guidelines on the 2006 Infectious Diseases Society of America Lyme Guidelines that are elaborated as a case study in conflicts of interest, medical and scientific bias and other poor practices in the (US) Institutional of Medicine's (IOM) 2011 Clinical Practice Guidelines We Can Trust (page 56, BOX 3-1). Standards Committee, Institute of Medicine, Board on Health Care Services. Clinical practice guidelines we can trust. Graham R, Mancher M, Miller Wolman D, Greenfield S, Steinberg E, eds. Washington, DC: National Academies Press; June 16, 2011