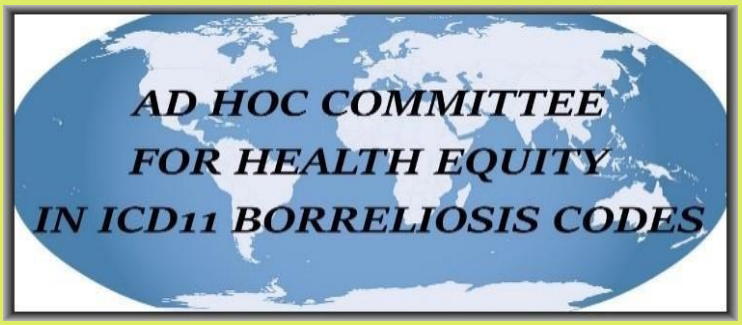

UPDATING ICD11 Borreliosis Diagnostic Codes

Edition One
March 29, 2017

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Abstract

This paper analyzes how, on a global scale, existing ICD10 diagnostic codes are preventing proper diagnosis and treatment of borrelioses including Lyme borreliosis and relapsing fever borreliosis. Across the globe, clinicians, scientists, researchers and patients examined and compiled references from peer-reviewed scientific literature to demonstrate the wide-ranging manifestations experienced by patients afflicted with these diseases.

Caused by the bite of ticks, in the United States (US) alone there an estimated 380,000 new annual Lyme borreliosis cases - more cases than breast cancer and more than six times the number of new HIV/AIDS cases.[1] Nevertheless, the few ICD10 diagnostic codes related to the borreliosis illness known Lyme disease cover a fraction of the conditions Lyme disease may cause. On the other hand, several highly unusual and rare conditions currently have their own diagnostic codes including:

W61.62XD	Struck by duck
W55.1	Bitten by a cow
W61.33	Pecked by a chicken
V91.07	Burn due to water skis on fire
V95.40	Unspecified spacecraft accident injuring occupant
R46.1	Bizarre personal appearance

Without accurate and appropriate diagnostic codes, physicians are impeded in their ability to properly care for their patients, leaving those patients invisible and marginalized within the medical system and to those guiding public policy. This results in great personal hardship, pain, disability and expense. This unnecessarily burdens health care systems, governments, families and society as a whole. With accurate diagnostic codes in place, robust data can guide medical and public health research, health policy, track mortality and save health care dollars.

WHO recognizes Lyme disease to be a 'disease of consequence'. WHO has met and consulted with other patient groups, scientists and medical professionals and non-governmental stakeholders from other 'diseases of consequence' during the ICD11 revision process.

We therefore recommend WHO engage with the global borreliosis stakeholder community who recognize the wide-ranging manifestations experienced by borreliosis patients. We also advise WHO use the peer-reviewed scientific literature that recognizes the wide-ranging manifestations experienced by patients as the basis for updating and modernizing the ICD11 diagnostic codes for Lyme borrelioses and relapsing fever borrelioses.

[1] Source is the CDC. (2013) International Conference on Lyme Borreliosis and other Tick-borne Diseases. 18 Aug 2013

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Excerpts from the report now follow: Recommendations and Tables of Conditions that have been added to the ICD11 revision by the Ad Hoc Committee via report to WHO, the ICD Team and to the digital Beta Platform of the ICD11 codes.

I. Recommendations for ICD Updates for Borreliosis

Currently, Lyme borreliosis and relapsing fever borreliosis are interpreted as two unrelated diseases. However, these borrelioses are, in fact, closely related and share similar features. Therefore, given the extensive genetic heterogeneity of the *Borrelia* family, the non-specific and multisystemic nature of symptoms that largely dominate both types of borreliosis, the ICD borreliosis codes should be more integrated and facilitate better characterization of these diseases and their wide range of manifestations.

Poor coding may contribute to misdiagnosis. For example, when the Lyme disease conditions of arthritis and other arthropathies, hearing and vision failure, and neurodegenerative diseases such as dementia and Alzheimer's disease are attributed to aging rather than Lyme disease, the older Lyme disease patient loses the opportunity to reverse and reduce these conditions when treated with antibiotics.

ICD modernization for borreliosis diseases will recognize and integrate advances from scientific research to more accurately reflect the worldwide distribution and range of human illnesses that result from these of diseases. It is recommended that:

1. Revised ICD codes should include a more complete list of the various stages and manifestations of Lyme borreliosis and relapsing fever. For example, ICD codes should reflect that:
   - The immune, endocrine and reticulo-endothelial systems, the skin, the gastrointestinal tract and various organs can all be involved.
   - *Borrelia* infection is found in organic brain syndromes and diverse neurologic manifestations such as seizure disorder, encephalopathy, and a diverse range of neuropsychiatric symptoms.
   - Immunosuppression due to *Borrelial* infection is recognized in the scientific literature, as are *Borrelia*-triggered autoimmune and neuro-autoimmune phenomena.

2. Codes for borreliosis associated with cases of sudden death due to unrecognized Lyme carditis, chronic congestive cardiomyopathies, and chronic and progressive encephalomyelitis including with fatal outcomes, require specific delineation in ICD coding.

3. Model borreliosis codes to reflect the scope and variability of infection, as is done with syphilis coding to include latent, serovariability and seronegative infections. Codes to specify these forms of syphilitic infection exist but are missing for Lyme disease.
   - Just as with syphilis, Lyme borreliosis codes should reflect latency (asymptomatic infection) in early and late stages as well as the serovariability during continuous active infection.

4. Congenital Lyme borreliosis requires clear articulation in the codes.

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5. Tick-borne pathogens have been reported in a wide range domestic animals and wildlife reservoirs. There is the need to codify borreliosis - Lyme, Lyme-like and relapsing fever - as zoonoses to guide diagnosis, treatment and prophylaxis, including provisions for recreation and occupational exposures.

6. Many patients with Lyme borreliosis and other tick-borne diseases develop varying degrees of disability, which is sometimes severe. The clinical and laboratory findings that support the recognition of disability from Lyme borreliosis and other tick-borne diseases require attention and articulation in ICD11.

7. Codes should be revised to focus on borreliosis diseases rather than vector sources and revised to accurately reflect multiple vectors of transmission for Pathogens transmitted by body lice or soft-bodied ticks are typically diagnosed as the cause of relapsing fever borreliosis.
   - The ICD10 codes for relapsing fever borreliosis are separate from those that represent Lyme disease. However, these code distinctions are based on the type of louse or tick that transmit the bacteria causing the disease, and not on the clinical presentation of the disease itself which may resemble Lyme disease' or relapsing fever or have symptoms from both.
   - There are relapsing fever borreliosis that include transmission from lice, and hard and soft-bodied ticks.
   - Furthermore, there are indications of hybrid genospecies among the disease causing borreliosis.

8. A single tick bite can result in transmission a multitude of bacterial, viral or protozoal agents. *For this reason, it is vital for the ICD codes to include a code for tick bite, as it does spider bite and insect bite.* In this way, should certain symptoms linked to tickborne illnesses manifest, medical records will link back to the tick bite incident and assist with arriving at the proper diagnosis.

9. A number of un-coded conditions that may be caused by borreliosis should be properly noted under those conditions; as is found under the ICD10 code G01 "Meningitis in bacterial diseases classified elsewhere".

10. There are other tick-borne diseases, while not addressed within the scope of this paper, that deserve more articulation of their manifestations and be updated in the ICD11 by WHO.

ICD10

G01 Meningitis in bacterial diseases classified elsewhere

- meningitis (in):
- gonococcal (A54.81)
- leptospirosis (A27.81)
- listeriosis (A32.11)
- **Lyme disease (A69.21)**
- meningococcal (A39.0)
- neurosyphilis (A52.13)
- tuberculosis (A17.0)

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The following recommendations are related to the broader mandates of WHO

11. The lack of support for borreliosis research in sub Saharan Africa is reflected in under-reporting and skewed global prevalence rates. There should be effort made to improve surveillance and study of these diseases in this region.

12. There should be a concerted effort to make provisions for inclusion of Borrelia in differential diagnoses for malaria, dengue, lassa and other fevers of unknown etiology in sub Saharan Africa. This would ensure patients with clinical borreliosis are not excluded from treatment due to missed diagnoses.

13. Governmental travel guidance should integrate accurate health warnings for travelers in the countries and regions under the wide global range of borreliosis.

14. Questions on travel history should be integrated into the medical and clinical reviews of patients.

15. Studies beyond the acute presentation of borreliosis are required to investigate the biological basis for the clinical disease variability observed in humans. For example, there is significant variation in disease presentation, which is likely due to a combination of various factors influencing pathogen-host interactions, including the virulence of the infecting *Borrelia* genospecies, the age, the genetic predisposition, and the immune status of the host.

16. There is the ongoing need to find promising treatments from antibiotic combinations and non-antibiotic treatments and to increase understanding regarding the transmission of borreliosis infection by other biting insects, blood transfusions, organ transplants and possible sexual transmission.

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Tables of LB Conditions

The Ad Hoc Committee submitted each manifestation noted in the following tables.

Some definitions of Terminology

- ✓ The secondary stage results from the dissemination of Borrelia during the early latent stage.
- ✓ The late stage is considered chronic if the duration is more than six months.
- ✓ ** (double red asterisks) indicates possibility of fatality from the condition

TABLE 1 Congenital Lyme

Congenital Lyme disease *Borrelia burgdorferi* can potentially infect the fetus and cause adverse fetal outcomes ****	**References** Bale JF, Murph JR. Congenital infections and the nervous system. *Pediatric Clinics of North America*. 1992;39(4):669–690. doi:10.1016/s0031-3955(16)38370-5. [PubMed] Brzostek T. [Human granulocytic ehrlichiosis co-incident with Lyme borreliosis in pregnant woman--a case study] [in Polish] *Przegl Epidemiol*. 2004;58(2):289–94. [PubMed] Gardner T. Lyme disease. In: Remington JS, Klein JO, eds. *Infectious Diseases of the Fetus and Newborn*. 5th ed. Philadelphia: Saunders; 1995:447–528chap 11. Gardner T. Lyme disease. In: Remington JS, Klein JO. *Infectious diseases of the fetus and newborn infant*. 4th ed. Philadelphia: W B Saunders Co; December 13, 1994. Goldenberg RL, Thompson C. The infectious origins of stillbirth. *American Journal of Obstetrics and Gynecology*. 2003;189(3):861–873. doi:10.1067/s0002-9378(03)00470-8. [PubMed] Gustafson JM, Burgess EC, Wachal MD, Steinberg H. Intrauterine transmission of Borrelia burgdorferi in dogs. *American Journal of Veterinary Research*. 1993;54(6):882–890. [PubMed]

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Congenital Lyme disease **	**References** MacDonald AB, Benach JL, Burgdorfer W. Stillbirth following maternal Lyme disease. *N Y State J Med*. 1987;11:615–616. [PubMed] MacDonald AB. Gestational Lyme borreliosis. Implications for the fetus. *Rheum Dis Clin North Am*. 1989;15(4):657–677. [PubMed] Macdonald AB. Human fetal borreliosis, toxemia of pregnancy, and fetal death. *Zentralblatt für Bakteriologie, Mikrobiologie und Hygiene. Series A: Medical Microbiology, Infectious Diseases, Virology, Parasitology*. 1986;263(1-2):189–200. doi:10.1016/s0176-6724(86)80122-5. [PubMed] Maraspin V, Cimperman J, Lotric-Furlan S, Pleterski-Rigler D, Strle F. Erythema migrans in pregnancy. *Wiener klinische Wochenschrift*. 2000;111:933–40. [PubMed] Markowitz LE, Steere AC, Benach JL, Slade JD, Broome CV. Lyme disease during pregnancy. *JAMA: The Journal of the American Medical Association*. 1986;255(24):3394. doi:10.1001/jama.1986.03370240064038. PubMed Schlesinger PA, Duray PH, Burke BA, Steere AC, Stillman MT. Maternal-fetal transmission of the Lyme disease Spirochete, Borrelia burgdorferi. *Annals of Internal Medicine*. 1985;103(1):67. doi:10.7326/0003-4819-103-1-67. [PubMed] Silver RM, Yang L, Daynes RA, Branch WD, Salafia CM, Weis JJ. Fetal outcome in Murine Lyme disease. *Infection and Immunity*. 1995;63(1):66–72. [PubMed] Strobino BA, Williams CL, Abid S, Ghalson R, Spierling P. Lyme disease and pregnancy outcome: A prospective s of two thousand prenatal patients. *American Journal of Obstetrics and Gynecology*. 1993;169(2):367–374. doi:10.1016/0002-9378(93)90088-z. [PubMed] Weber K, Bratzke H-J, Neubert U, Wilske B, Duray PH. Borrelia burgdorferi in a newborn despite oral penicillin for Lyme borreliosis during pregnancy. *The Pediatric Infectious Disease Journal*. 1988;7(4):286–288. doi:10.1097/00006454-198804000-00010. [PubMed]

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TABLE 2 Primary Infection & Erythema migrans

Primary infection - Erythema migrans	**References** Christova I, Komitova R. Clinical and epidemiological features of Lyme borreliosis in Bulgaria. Wien Klin Wochenschr. 2004;116(1-2):42-6. [PubMed] Hercogová J, Tománková M, Barták P. Contributions to the treatment of dermatologic manifestations of Lyme borreliosis. Cutis. 1992 Jun;49(6):409-11. [PubMed] Lipsker D, Hansmann Y, Limbach F, Clerc C, Tranchant C, Grunenberger F, Caro-Sampara F, Attali P, Frey M, Kubina M, Piémont Y, Sibilia J, Jaulhac B; GEBLY Study Group. Study Group for Lyme Borreliosis. Disease expression of Lyme borreliosis in northeastern France. Eur J Clin Microbiol Infect Dis. 2001;20(4):225-30. [PubMed] Melski JW, Reed KD, Mitchell PD, Barth GD. Primary and secondary erythema migrans in central Wisconsin. Arch Dermatol. 1993;129(6):709-16. [PubMed] Schmid GP. Epidemiology and clinical similarities of human spirochetal diseases. Rev Infect Dis. 1989;11Suppl 6:S1460-9. Review type. [PubMed]
Primary Infection, seronegative Early Lyme disease 'seronegative' is not a 'separate condition' - it is an aspect of the disease recognizing patients may test negative when infected.	Coyle PK, Deng Z, Schutzer SE, Belman AL, Benach J, Krupp LB, Luft B. Detection of Borrelia burgdorferi antigens in cerebrospinal fluid. Neurology 1993;43:1093-1097. [PubMed] Coyle PK, Schutzer SE, Deng Z, et al. Detection of Borrelia burgdorferi-specific antigen in antibody-negative cerebrospinal fluid in neurologic Lyme disease. Neurology. 1995;45(11):2010-2015. [PubMed] Dattwyler RJ, Volkman DJ, Luft BJ, Halperin JJ, Thomas J, Golightly MG. Seronegative Lyme Disease. Dissociation of T- and B-Lymphocyte Responses to Borrelia burgdorferi. N Engl J Med 1988;319:1441-6. [PubMed] Holak H1, 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. [PubMed] Karma A, Seppälä I, Mikkilä H, Kaakkola S, Viljanen M, Tarkkanen A. Diagnosis and clinical characteristics of ocular Lyme borreliosis. Am J Ophthalmol. 1995;119(2):127-35. [PubMed] Lawrence C, Lipton RB, Lowy FD, Coyle PK. Seronegative Chronic Relapsing Neuroborreliosis. Eur Neurol 1995;35:113-117. [PubMed]

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TABLE 3 Persistent Infection in Secondary & Late Stages

Persistent infection	**References** Feng J., Wang T., Shi W., Zhang S., Sullivan D., Auwaerter P.G., Zhang Y. Identification of novel activity against Borrelia burgdorferi persisters using an FDA approved drug library. Emerg. Microbes Infect. 2014;3:e49. doi: 10.1038/emi.2014.53. [PMC free article] [PubMed] [Cross Ref] Oksi J., Marjamaki M., Nikoskelainen J., Viljanen M.K. *Borrelia burgdorferi* detected by culture and PCR in clinical relapse of disseminated Lyme borreliosis. Ann. Med. 1999;31:225–232. doi: 10.3109/07853899909115982. [PubMed] [Cross Ref] Preac-Mursic V, Pfister HW, Spiegel H, Burk R, Wilske B, Reinhardt S, Bohmer R. First Isolation of Borrelia burgdorferi from an Iris Biopsy. J Clin Neuro-ophthalmol 1993;13:155-161. [PubMed] Preac-Mursic V, Weber K, Pfister HW, Wilske B, Gross B, Baumann A, Prokop J. Survival of Borrelia burgdorferi in Antibiotically Treated Patients with Lyme borreliosis. Infection 1989;17:355-359. [PubMed] Schmidli J., Hunziker T., Moesli P., Schaad U.B. Cultivation of *Borrelia burgdorferi* from joint fluid three months after treatment of facial palsy due to Lyme borreliosis. J. Infect. Dis. 1988;158:905–906. doi: 10.1093/infdis/158.4.905. [PubMed] [Cross Ref]

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TABLE 4 Secondary and Late infection - Cutaneous

Borrelial lymphocytoma (BL)	**References** Arnež M, Ružić-Sabljić E. Borrelial Lymphocytoma in Children. Pediatr Infect Dis J. 2015;34(12):1319-22. [PubMed] Colli C, Leinweber B, Müllegger R, Chott A, Kerl H, Cerroni L. Borrelia burgdorferi-associated lymphocytoma cutis: clinicopathologic, immunophenotypic, and molecular study of 106 cases. J Cutan Pathol. 2004;31(3):232-40. [PubMed] Glatz M, Resinger A, Semmelweis K, Ambros-Rudolph CM, Müllegger RR. Clinical spectrum of skin manifestations of Lyme borreliosis in 204 children in Austria. Acta Derm Venereol. 2015;95(5):565-71. [PubMed] Gordillo-Pérez G, Torres J, Solórzano-Santos F, de Martino S, Lipsker D, Velázquez E, Ramon G, Onofre M, Jaulhac B.Borrelia burgdorferi infection and cutaneous Lyme disease, Mexico. Emerg Infect Dis. 2007;13(10):1556-8. [PubMed] Krbkova L, Stanek G. Therapy of Lyme borreliosis in children. Infection. 1996;24(2):170-3. [PubMed] Lenormand C, Jaulhac B, De Martino S, Barthel C, Lipsker D. Species of Borrelia burgdorferi complex that cause borrelial lymphocytoma in France. Br J Dermatol. 2009;161(1):174-6. [PubMed] Maraspin V, Cimperman J, Lotric-Furlan S, Ruzić-Sabljić E, Jurca T, Picken RN, Strle F. Solitary borrelial lymphocytoma in adult patients. Wien Klin Wochenschr. 2002;114(13-14):515-23. [PubMed] Maraspin V, Nahtigal Klevišar M, Ružić-Sabljić E, Lusa L, Strle F. Borrelial Lymphocytoma in Adult Patients. Clin Infect Dis. 2016;63(7):914-21. [PubMed] Müllegger RR, Means TK, Shin JJ, Lee M, Jones KL, Glickstein LJ, Luster AD, Steere AC. Chemokine signatures in the skin disorders of Lyme borreliosis in Europe: predominance of CXCL9 and CXCL10 in erythema migrans and acrodermatitis and CXCL13 in lymphocytoma. Infect Immun. 2007;75(9):4621-8. [PubMed] Strle F, Maraspin V, Pleterski-Rigler D, Lotric-Furlan S, Ruzić-Sabljić E, Jurca T, Cimperman J. Treatment of borrelial lymphocytoma. Infection. 1996;24(1):80-4. [PubMed]

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Granuloma annulare, morphea, localized scleroderma, lichen sclerosus & atrophicus	Aberer E, Schmidt BL, Breier F, Kinaciyan T, Luger A. Amplification of DNA of Borrelia burgdorferi in urine samples of patients with granuloma annulare and lichen sclerosus et atrophicus. 1999;135(2):210-2. [PubMed] Asbrink E, Brehmer-Andersson E, Hovmark A. Acrodermatitis chronica atrophicans--a spirochetosis. Clinical and histopathological picture based on 32 patients; course and relationship to erythema chronicum migrans Afzelius. Am J Dermatopathol. 1986;8(3):209-19. [PubMed] Buechner SA, Winkelmann RK, Lautenschlager S, Gilli L, Rufli T. Localized scleroderma associated with Borrelia burgdorferi infection. Clinical, histologic, and immunohistochemical observations. J Am Acad Dermatol. 1993;29(2 Pt 1):190-6. [PubMed] Eisendle K, Grabner T, Zelger B. Morphoea: a manifestation of infection with Borrelia species? Br J Dermatol. 2007;157(6):1189-98. [PubMed] Kaya G, Berset M, Prins C, Chavaz P, Saurat JH. Chronic borreliosis presenting with morphea- and lichen sclerosus et atrophicus-like cutaneous lesions. a case report. Dermatology. 2001;202(4):373-5. [PubMed] Malane MS, Grant-Kels JM, Feder HM Jr, Luger SW. Diagnosis of Lyme disease based on dermatologic manifestations. Ann Intern Med. 1991;114(6):490-8. [PubMed]

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TABLE 5 Secondary and late Lyme meningitis, oculopathy, iridocyclitis, iritis, uveitis

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TABLE 6 Secondary & Late Lyme nephritis, hepatitis, lymphadenopathy, myositis & other

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TABLE 8 Late NB - neuritis or neuropathy, meningovascular, NB with cerebral infarcts, Lyme parkinsonism, Lyme encephalitis

Symptomatic Late Lyme neuroborreliosis **	**References** Burakgazi AZ. Lyme disease –induced polyradiculopathy mimicking amyotrophic lateral sclerosis. *Inter J of Neuroscience*. 2014;124(11):859–862. doi:10.3109/00207454.2013.879582. [PubMed] Christova I, Komitova R. Clinical and epidemiological features of Lyme borreliosis in Bulgaria. *Wien Klin Wochenschr*. 2004;116(1-2):42-6. [PubMed] Lawrence C, Lipton RB, Lowy FD, Coyle PK. Seronegative Chronic Relapsing Neuroborreliosis. *Eur Neurol* 1995;35:113-117. [PubMed] Marconi RT, Hohenberger S, Jauris-Heipke S, Schulte-Spechtel U, LaVoie CP, Rößler D, Wilske B. Genetic Analysis of Borrelia garinii OspA Serotype 4 Strains Associated with Neuroborreliosis: Evidence for Extensive Genetic Homogeneity. *J Clin Microbiol*. 1999;37(12): 3965–3970. [PubMed Central] Nafeev AA, Klimova LV. [Clinical manifestations of neuroborreliosis in the Volga region]. [Article in Russian]. *Ter Arkh*. 2010;82(11):68-70. [PubMed]
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TABLE 10 Late atrophic form of Lyme meningoencephalitis with dementia & subacute presenile dementia & Neuropsychiatric manifestations

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TABLE 11 Late Lyme Bone & Joint & Musculoskeletal

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TABLE 12 Late - oculopathy & liver & kidney & respiratory

Late Lyme oculopathy	**References** Drenckhahn A, Spors B, Knierim E. Acute isolated partial oculomotor nerve palsy due to Lyme neuroborreliosis in a 5 year old girl. *European Journal of Paediatric Neurology*. 2016;20(6):977-979. doi:10.1016/j.ejpn.2016.05.022. [PubMed] Karma A, Seppälä I, Mikkilä H, Kaakkola S, Viljanen M, Tarkkanen A. Diagnosis and clinical characteristics of ocular Lyme borreliosis. *Am J Ophthalmol*. 1995;119(2):127-135. [PubMed] Golubić D, Vinković T, Turk D, Hranilović J, Slugan I. [Ocular manifestations of Lyme borreliosis in northwest Croatia]. [in Croatian]. *Lijec Vjesn*. 2004;126(5-6):124-128. [PubMed] Mikkilä HO, Seppälä IJ, Viljanen MK, Peltomaa MP, Karma A. The expanding clinical spectrum of ocular lyme borreliosis. *Ophthalmology*. 2000;107(3):581-587. [PubMed]
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Late Lyme disease of kidney & ureter ****	**References** Finnian R. Mc Causland, Sophie Niedermaier, Vanesa Bijol, Helmut G. Rennke, Mary E. Choi, John P. Forman; Lyme disease-associated glomerulonephritis. *Nephrol Dial Transplant* 2011; 26 (9): 3054-3056. doi: 10.1093/ndt/gfr335 [PubMed] Florens N, Lemoine S, Guebre-Egziabher F, et al. Chronic Lyme borreliosis associated with minimal change glomerular disease: a case report. *BMC Nephrol*. 2017;18(1). doi:10.1186/s12882-017-0462-4. [PubMed] Kelly B, Finnegan P, Cormican M, et al. . Lyme disease and glomerulonephritis, *Ir Med J*, 1999, vol. 92 pg. 372 [PubMed] Papineni P, Doherty T, Pickett T, et al. . Membranous glomerulonephritis secondary to *Borrelia burgdorferi* infection presenting as nephrotic syndrome, *NDT Plus*, 2010, vol. 3 (pg. 105-106) [PubMed] Google ScholarPubMed Rawal B, Rovner L, Thakar C, et al. MPGN and nephrotic syndrome secondary to Lyme disease, *Am J Kidney Dis (abstract)*, 2008, vol. 51 pg. B83 Google ScholarCrossRef
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TABLE 13 Latent Lyme disease, unspecified

Latent Lyme disease, unspecified	References
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March 29, 2017

30 YEARS AND COUNTING

V International Conference
on Lyme Borreliosis

PROGRAM AND ABSTRACTS

Arlington, Virginia, U.S.A.
May 30-June 2, 1992
Hyatt Regency Crystal City

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CULTURE-CONFIRMED TREATMENT FAILURE OF CEFOTAXIME AND MINOCYCLINE IN A CASE OF LYME MENINGOENCEPHALOMYELITIS IN THE UNITED STATES.

Kenneth B. Liegner, Carl E. Rosenkilde, Grant L. Campbell*, Thomas J. Quan, and David T. Dennis, Armonk, NY, USA, Mount Kisco, NY, USA, and Centers for Disease Control, Fort Collins, CO, USA.

In 1987, a 37-year-old woman living in Westchester County, NY, developed spastic paraparesis, bilateral Babinski reflexes, and cranial nerve and bulbar dysfunction characterized by dysphagia, dysphonia, diplopia, absent gag reflex, and dysfunction of bowel and bladder control. CSF contained 19 WBC/mm^3 (86% lymphs). A test for antibodies to *Borrelia burgdorferi* (*Bb*) in serum was negative. No etiology was established despite an extensive workup. Symptoms and signs reportedly worsened gradually from 1988 to present. There was a past history of splenectomy for idiopathic thrombocytopenic purpura diagnosed in 1975. In 1989, the right frontal region and right basal ganglia were abnormal on brain MRI. In January 1990, CSF contained 6 WBC/mm^3 (93% lymphs), but no oligoclonal bands or myelin basic protein. Paired CSF and serum tests for antibodies to *Bb*, and PCR for *Bb*-specific oligonucleotides in CSF, were negative. An empiric 21-day course of cefotaxime (3 g/12 hr i.v.) was given in January, 1990 with no clear clinical benefit. Following treatment, CSF contained 9 WBC/mm^3 (93% lymphs). Four months of minocycline (200 mg/day p.o.) begun in November, 1990 also yielded no clear clinical benefit. In December, 1990 a T-cell stimulation test with *Bb* antigens was strongly positive. In December, 1991 CSF contained 6 WBC/mm^3 (89% lymphs) and elevated IgG. Paired serum and CSF samples were strongly positive for antibodies to *Bb*, with a CSF-to-serum index of 1.04. Culture of this CSF specimen in BSK-II yielded a strain of *Bb*. Culture-confirmed treatment failures have been previously reported for three Lyme neuroborreliosis cases in Europe. The present case apparently is the first of this type to be reported from the United States.

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