

DAFTAR PUSTAKA

1. Catalina MD, Owen KA, Labonte AC, Grammer AC, Lipsky PE. The pathogenesis of systemic lupus erythematosus: Harnessing big data to understand the molecular basis of lupus. *J Autoimmun.* 2020;110:1-20.
2. Barber MRW, Drenkard C, Falasinnu T, Hoi A, Mak A, Kow NY, et al. Global epidemiology of systemic lupus erythematosus. *Nat Rev Rheumatol.* 2021;17(9):515-32.
3. Tanaka Y, O'Neill S, Li M, Tsai I, Yang Y. Systemic lupus erythematosus: Targeted literature review of the epidemiology, current treatment, and disease burden in the Asia Pacific region. *Arthritis Care & Research.* 2022;74(2):187-98.
4. Lee YH, Choi SJ, Ji JD, Song GG. Overall and cause-specific mortality in systemic lupus erythematosus: an updated meta-analysis. *Lupus.* 2016;25(7):727-34.
5. Hamijoyo L, Candrianita S, Rahmadi AR, Dewi S, Darmawan G, Suryajaya BS, et al. The clinical characteristics of systemic lupus erythematosus patients in Indonesia: a cohort registry from an Indonesia-based tertiary referral hospital. *Lupus.* 2019;28(13):1604-9.
6. Pusdatin. Situasi lupus di Indonesia. *Pusat Data dan Informasi Kementerian Kesehatan RI.* 2017:1-8.
7. Sawaf M, Dumortier H, Monneaux F. Follicular helper T cells in systemic lupus erythematosus: Why should they be considered as interesting therapeutic targets? *J Immunol Res.* 2016;2016:1-13.
8. Bertsias G, Cervera R, Boumpas DT. Systemic lupus erythematosus: Pathogenesis and clinical features. *EULAR textbook of rheumatic disease.* 2012:476-505.
9. Paredes JL, Fernandez-Ruiz R, Niewold TB. T cells in systemic lupus erythematosus. *Rheum Dis Clin North Am.* 2021;47(3):379-93.
10. Fan X, Lin C, Han J, Jiang X, Zhu J, Jin T. Follicular helper CD4+ T cells in human neuroautoimmune diseases and their animal models. *Mediators Inflamm.* 2015:1-11.

11. Abbas AK, Lichtman AH, Pillai S. Humoral immune responses. In: *Basic Immunology Functions and Disorders of the Immune System*. 6th ed. Elsevier; 2020:137-57.
12. Rodríguez-Rodríguez N, Rosetti F, Crispín JC. T cells. In: *Systemic Lupus Erythematosus*. Elsevier; 2016:113-9.
13. Zhang X, Lindwall E, Gauthier C, Lyman J, Spencer N, Alarakhia A, et al. Circulating CXCR5+CD4+helper T cells in systemic lupus erythematosus patients share phenotypic properties with germinal center follicular helper T cells and promote antibody production. *Lupus*. 2015;24(9):1-9.
14. Liu D, Yan J, Sun J, Liu B, Ma W, Li Y, et al. BCL6 controls contact-dependent help delivery during follicular T-B cell interactions. *Immunity*. 2021;54(10):2245-55
15. Nurieva RI, Chung Y, Martinez GJ, Yang X, Tanaka S, Matskevitch T, et al. Bcl6 mediates the development of T follicular helper cells. *Science*. 2009;325(5943):1001-5.
16. Zhang X, Ing S, Fraser A, Chen M, Khan O, Zakem J, et al. Follicular helper T cells: new insights into mechanisms of autoimmune diseases. *Ochsner J*. 2013;13(1):131-9.
17. Choi JY, Ho JH en, Pasoto SG, Bunin V, Kim S, Carrasco S, et al. Circulating follicular helper-like T cells in systemic lupus erythematosus: Association with disease activity: circulating Tfh-like cells in SLE. *Arthritis Rheumatol*. 2015;67(4):988-99.
18. Huang X, Wu H, Qiu H, Yang H, Deng Y, Zhao M, et al. The expression of BCL6 in circulating follicular helper-like T cells positively correlates with the disease activity in systemic lupus erythematosus. *Clin Immunol*. 2016;173:161-70.
19. Gensous N, Schmitt N, Richez C, Ueno H, Blanco P. T follicular helper cells, interleukin-21 and systemic lupus erythematosus. *Rheumatology*. 2016;56(4):1-8.
20. Terrier B, Costedoat-Chalumeau N, Garrido M, Geri G, Rosenzweig M, Musset L, et al. Interleukin 21 correlates with T cell and B cell subset

- alterations in systemic lupus erythematosus. *J Rheumatol*. 2012;39(9):1819-28.
21. Ding S, Rao Y, Lu Q. Are BCL6 and EZH2 novel therapeutic targets for systemic lupus erythematosus? *Cell Mol Immunol*. 2022;19(7):863-5.
 22. Perhimpunan Reumatologi Indonesia. Diagnosis dan pengelolaan lupus eritematosus sistemik. *Perhimpunan Reumatologi Indonesia*. 2019:1-129.
 23. Gergianaki I, Bortoluzzi A, Bertias G. Update on the epidemiology, risk factors, and disease outcomes of systemic lupus erythematosus. *Best Pract Res Clin Rheumatol*. 2018;32(2):188-205.
 24. Khanna S, Pal H, Pandey RM, Handa R. The relationship between disease activity and quality of life in systemic lupus erythematosus. *Rheumatology*. 2004;43(12):1536-40.
 25. Izmirly PM, Parton H, Wang L, McCune W, Lim S, Drenkard C, et al. Prevalence of systemic lupus erythematosus in the United States: Estimates from a meta-analysis of the centers for disease control and prevention national lupus registries. *Arthritis Rheumatol*. 2021;73(6):991-6.
 26. Kaul A, Gordon C, Crow MK, Touma Z, Urowitz M, van Vollenhoven R, et al. Systemic lupus erythematosus. *Nat Rev Dis Primers*. 2016;2(1):1-21.
 27. Suarjana IN. Imunopatogenesis lupus eritematosus sistemik. In: *Buku Ajar Ilmu Penyakit Dalam*. 6th ed. Interna Publishing; 2014:3331-45.
 28. Hahn BH. Systemic lupus erythematosus. In: *Harrison's Principles of Internal Medicine*. 20th ed. Mc Graw Hill; 2018:2515-26.
 29. Tsokos GC, Lo MS, Reis PC, Sullivan KE. New insights into the immunopathogenesis of systemic lupus erythematosus. *Nat Rev Rheumatol*. 2016;12(12):716-30.
 30. Munoz L, van Bavel C, Franz S, Berden J, Herrmann M, van der Vlag J. Apoptosis in the pathogenesis of systemic lupus erythematosus. *Lupus*. 2008;17(5):371-5.
 31. Suárez-Fueyo A, Crispín JC, Tsokos GC. T cells. In: *Dubois' Lupus Erythematosus and Related Syndromes*. Elsevier; 2019:116-24.
 32. Kim SJ, Lee K, Diamond B. Follicular helper T cells in systemic lupus erythematosus. *Front Immunol*. 2018;9:1-8.

33. Feng X, Wang D, Chen J, Lu L, Hua B, Li X, et al. Inhibition of aberrant circulating Tfh cell proportions by corticosteroids in patients with systemic lupus erythematosus. Zissel G, ed. *PLoS One*. 2012;7(12):e51982.
34. Crotty S. Follicular helper CD4 T cells (Tfh). *Annu Rev Immunol*. 2011;29(1):621-63.
35. Ding S, Zhang Q, Luo S, Gao L, Huang J, Lu J, et al. BCL-6 suppresses miR-142-3p/5p expression in SLE CD4+ T cells by modulating histone methylation and acetylation of the miR-142 promoter. *Cell Mol Immunol*. 2020;17(5):474-82.
36. Choi J, Crotty S. Bcl6-mediated transcriptional regulation of follicular helper T cells (Tfh). *Trends Immunol*. 2021;42(4):336-49.
37. Long D, Chen Y, Wu H, Zhao M, Lu Q. Clinical significance and immunobiology of IL-21 in autoimmunity. *J Autoimmun*. 2019;99:1-14.
38. Leonard WJ, Wan CK. IL-21 signaling in immunity. *F1000Res*. 2016;5:1-10.
39. Rozo C, Chinenov Y, Maharaj RK, Gupta S, Leuenberger L, Kirou K, et al. Targeting the RhoA-ROCK pathway to reverse T-cell dysfunction in SLE. *Ann Rheum Dis*. 2017;76(4):740-7.
40. Alsén S, Cervin J, Deng Y, Deng Y, Szeponik L, Wenzel U, et al. Antigen-presenting B cells program the efferent lymph T helper cell response. *Front Immunol*. 2022;13:813203.
41. Shen C, Xue X, Zhang X, Wu L, Duan X, Su C. Dexamethasone reduces autoantibody levels in MRL/lpr mice by inhibiting Tfh cell responses. *J Cell Mol Med*. 2021;25(17):8329-37.
42. Cardenas MG, Oswald E, Yu W, Xue F, MacKerell AD, Melnick AM. The expanding role of the BCL6 oncoprotein as a cancer therapeutic target. *Clin Cancer Res*. 2017;23(4):885-893.
43. McLachlan T, Matthews WC, Jackson ER, Staudt D, Douglas A, Findlay I, et al. B-cell lymphoma 6 (BCL6): from master regulator of humoral immunity to oncogenic driver in pediatric cancers. *Mol Cancer Res*. 2022;20(12):1711-23.
44. Eto D, Lao C, DiToro D, Barnett B, Escobar T, Kageyama R, et al. IL-21 and IL-6 are critical for different aspects of B cell immunity and redundantly

- induce optimal follicular helper CD4 T cell (Tfh) differentiation. Poh LNF, ed. *PLoS One*. 2011;6(3):1-11.
45. Choi JY, Seth A, Kashgarian M, Terrillon S, Fung E, Huang L, et al. Disruption of pathogenic cellular networks by IL-21 blockade leads to disease amelioration in murine lupus. *J Immunol*. 2017;198(7):2578-88.
 46. McPhee CG, Bubier JA, Sproule TJ, Park G, Steinbuck M, Schott W, et al. IL-21 Is a double-edged sword in the systemic lupus erythematosus–like disease of BXSB.Yaa mice. *J Immunol*. 2013;191(9):4581-8.
 47. Fröhlich C, Ehrhardt J, Krüger D, Trojnaraska D, Zygmunt M, Muzzio DO. Pregnancy status alters IL-21-mediated effects on murine B lymphocytes. *Reproduction*. 2020;159(3):351-9.
 48. Iannello A, Tremblay C, Routy JP, Boulassel MR, Toma E, Ahmad A. Decreased levels of circulating IL-21 in HIV-Infected AIDS patients: correlation with CD4+T-cell counts. *Viral Immunol*. 2008;21(3):385-8.
 49. Asao H. Interleukin-21 in viral infections. *IJMS*. 2021;22(17):1-15.
 50. Costa RT, Araújo ORD, Brunialti MKC, Assunção M, Azevedo L, Freitas F, et al. T helper type cytokines in sepsis: time-shared variance and correlation with organ dysfunction and hospital mortality. *Braz J Infect Dis*. 2019;23(2):79-85.
 51. Miao T, Pu Y, Zhou B, Chen P, Wang Y, Song Y, et al. Association between polymorphisms in IL21 gene and risk for sepsis. *Biomarkers*. 2017;22(1):14-8.
 52. Fava A, Petri M. Systemic lupus erythematosus: Diagnosis and clinical management. *J Autoimmun*. 2019;96:1-13.
 53. Kasjmir YI, Handono K, Wijaya LK, Hamijoyo L, Albar Z, Kalim H, et al. Diagnosis dan pengelolaan lupus eritematosus sistemik. In: *Buku Ajar Ilmu Penyakit Dalam*. Vol III. 6th ed. Interna Publishing; 2014:3360-77.
 54. Galarza-Maldonado C, Kourilovitch MR, Molineros JE, Cardiel M, Zurita L, Soroka N, et al. The administration of low doses of rituximab followed by hydroxychloroquine, prednisone and low doses of mycophenolate mofetil is an effective therapy in Latin American patients with active systemic lupus erythematosus. *Autoimmun Rev*. 2010;10(2):108-11.

55. Ibañez D, Gladman DD, Touma Z, Nikpour M, Urowitz MB. Optimal frequency of visits for patients with systemic lupus erythematosus to measure disease activity over time. *J Rheumatol*. 2011;38(1):60-3.
56. Nakou M, Papadimitraki ED, Fanouriakis A, Bertias GK, Choulaki C, Sidiropoulos P, et al. Interleukin-21 is increased in active systemic lupus erythematosus patients and contributes to the generation of plasma B cells. *Clin Exp Rheumatol*. 2013;31(2):172-9.
57. Kurniawan RE. Perbedaan kadar glikoprotein P serum dan jumlah limfosit antara pasien lupus eritematosus sistemik aktif dan tidak aktif yang mendapat terapi metilprednisolon. *Tesis Bagian Ilmu Penyakit Dalam FK UNAND*. 2019:45-6. (*Unpublished*)
58. Dhistiarnie PA. Pengaruh pemberian kortikosteroid pulse dose terhadap jumlah sel limfosit T CD4+, kadar sel T regulator, interleukin-2 dan nilai mex sledai pada pasien lupus eritematosus sistemik. *Tesis Bagian Ilmu Penyakit Dalam FK UNAND*. 2018:47-8. (*Unpublished*)
59. Bae EH, Lim SY, Han KD, Jung JH, Choi HS, Kim HY, et al. Trend of prevalence and incidence of systemic lupus erythematosus in South Korea, 2005 to 2015: a nationwide population-based study. *Korean J Intern Med*. 2020;35(3):652-61.
60. Jakes RW, Bae S, Louthrenoo W, Mok C, Navarra SV, Kwon N. Systematic review of the epidemiology of systemic lupus erythematosus in the Asia-Pacific region: Prevalence, incidence, clinical features, and mortality. *Arthritis Care Res*. 2012;64(2):159-68.
61. Kim JW, Kim HA, Suh CH, Jung JY. Sex hormones affect the pathogenesis and clinical characteristics of systemic lupus erythematosus. *Front Med*. 2022;9:1-15.
62. Petri M. Sex hormones and systemic lupus erythematosus. *Lupus*. 2008;17(5):412-5.
63. Cramer DW, Xu H. Predicting age at menopause. *Maturitas*. 1996;23(3):319-26.

64. Nusbaum JS, Mirza I, Shum J, Freilich R, Cohen R, Pillinger M, et al. Sex differences in systemic lupus erythematosus. *Mayo Clin Proc.* 2020;95(2):384-94.
65. Rider V, Abdou NI. Gender differences in autoimmunity: molecular basis for estrogen effects in systemic lupus erythematosus. *International Immunopharmacology.* 2001;1(6):1009-24.
66. Rees F, Doherty M, Grainge MJ, Lanyon P, Zhang W. The worldwide incidence and prevalence of systemic lupus erythematosus: a systematic review of epidemiological studies. *Rheumatology.* 2017;56(11):1945-61.
67. Susilo DH, Wachid Achadiono DN, Paramaiswari A. Correlation between MEX-SLEDAI and mean platelet volume in systemic lupus erythematosus patients. *Ina J Rheum.* 2021;12(2):293-301.
68. Sixto LB, Isenberg D. Systemic lupus erythematosus: causes and manifestations. *Trends Urol & Men's Health.* 2020;11(1):26-9.
69. Cooper EE, Pisano CE, Shapiro SC. Cutaneous manifestations of lupus: systemic lupus erythematosus and beyond. Rothschild BM, ed. *Int J Rheumatol.* 2021;2021:1-19.
70. Gasparotto M, Gatto M, Binda V, Doria A, Moroni G. Lupus nephritis: clinical presentations and outcomes in the 21st century. *Rheumatology.* 2020;59(Suppl5):v39-v51.
71. Santacruz JC, Mantilla MJ, Rueda I, Pulido S, Rodriguez-Salas G, Londono J. A practical perspective of the hematologic manifestations of systemic lupus erythematosus. *Cureus.* 2022;14(3):1-12.
72. Voulgarelis M, Kokori S, Ioannidis J, Tzioufas A, Kyriaki D, Moutsopoulos H. Anaemia in systemic lupus erythematosus: aetiological profile and the role of erythropoietin. *Ann Rheum Dis.* 2000;59(3):217-22.
73. Liang Y, Leng RX, Pan HF, Ye DQ. The prevalence and risk factors for serositis in patients with systemic lupus erythematosus: a cross-sectional study. *Rheumatol Int.* 2017;37(2):305-11.
74. Zhao J, Bai W, Zhu P, Zhang X, Liu S, Wu L, et al. Chinese SLE treatment and research group (CSTAR) registry VII: prevalence and clinical significance

- of serositis in Chinese patients with systemic lupus erythematosus. *Lupus*. 2016;25(6):652-657.
75. Sarwar S, Mohamed AS, Rogers S, Sarmast ST, Kataria S, Mohamed KH, et al. Neuropsychiatric systemic lupus erythematosus: A 2021 update on diagnosis, management, and current challenges. *Cureus*. 2021;13(9):1-11.
 76. Zhang Y, Han H, Chu L. Neuropsychiatric lupus erythematosus: future directions and challenges; a systematic review and survey. *Clinics*. 2020;75:1-7.
 77. Bubier JA, Sproule TJ, Foreman O, Spolski R, Shaffer D, Morse H, et al. A critical role for IL-21 receptor signaling in the pathogenesis of systemic lupus erythematosus in BXSB-Yaa mice. *Proc Natl Acad Sci U S A*. 2009;106(5):1518-23.
 78. Shater H, Fawzy M, Farid A, El-Amir A, Fouad S, Madbouly N. The potential use of serum interleukin-21 as biomarker for lupus nephritis activity compared to cytokines of the tumor necrosis factor (TNF) family. *Lupus*. 2022;31(1):55-64.
 79. Zhou S, Li Q, Zhou S, Zhao M, Lu L, Wu H, et al. A novel humanized cutaneous lupus erythematosus mouse model mediated by IL-21-induced age-associated B cells. *J Jaut*. 2021;123:1-10.
 80. Fanouriakis A, Tziolos N, Bertias G, Boumpas DT. Update on the diagnosis and management of systemic lupus erythematosus. *Ann Rheum Dis*. 2021;80(1):14-25.
 81. Porta S, Danza A, Arias Saavedra M, Carlomagno A, Goizueta M, Vivero F, et al. Glucocorticoids in systemic lupus erythematosus. Ten questions and some issues. *JCM*. 2020;9(9):1-13.
 82. Fanouriakis A, Kostopoulou M, Alunno A, Aringer M, Bajema I, Boleris J, et al. 2019 update of the EULAR recommendations for the management of systemic lupus erythematosus. *Ann Rheum Dis*. 2019;78(6):736-45.
 83. Kim S, Boehme L, Nel L, Casian A, Sangle S, Nova LE, et al. Defective STAT5 activation and aberrant expression of BCL6 in naive CD4 T cells enhances follicular Th cell-like differentiation in patients with granulomatosis with polyangiitis. *J Immunol*. 2022;208(4):807-18.

84. Ricard L, Jachiet V, Malard F, Ye Y, Stocker N, Senet P, et al. Circulating follicular helper T cells are increased in systemic sclerosis and promote plasmablast differentiation through the IL-21 pathway which can be inhibited by ruxolitinib. *Ann Rheum Dis*. 2019;78(4):539-50.
85. Lan Y, Luo B, Wang JL, Jiang YW, Wei YS. The association of interleukin-21 polymorphisms with interleukin-21 serum levels and risk of systemic lupus erythematosus. *Gene*. 2014;538(1):94-8.
86. Pan HF, Wu GC, Fan YG, Leng RX, Peng H, Zhou M, et al. Decreased serum level of IL-21 in new-onset systemic lupus erythematosus patients. *Rheumatol Int*. 2013;33(9):2337-42.
87. Kang K, Kim HO, Kwok SK, Ju J, Park K, Sun D, et al. Impact of interleukin-21 in the pathogenesis of primary Sjogren's syndrome: increased serum levels of interleukin-21 and its expression in the labial salivary glands. *Arthritis Res Ther*. 2011;13(5):1-10.
88. Ma CS, Deenick EK, Batten M, Tangye SG. The origins, function, and regulation of T follicular helper cells. *JEM*. 2012;209(7):1241-53.
89. Wang L, Zhao P, Ma L, Shan Y, Jiang Z, Wang J, et al. Increased interleukin 21 and follicular helper T-like cells and reduced interleukin 10+ B cells in patients with new-onset systemic lupus erythematosus. *J Rheumatol*. 2014;41(9):1781-92.
90. Zickert A, Amoudruz P, Sundström Y, Rönnelid J, Malmström V, Gunnarsson I. IL-17 and IL-23 in lupus nephritis - association to histopathology and response to treatment. *BMC Immunol*. 2015;16(1):7.
91. Cain DW, Cidlowski JA. Immune regulation by glucocorticoids. *Nat Rev Immunol*. 2017;17(4):233-47.
92. Nazir A, Zaki N, Mansour A, Farrag N, Agamy E. Plasma interleukin-21 and CXCL13 in primary immune thrombocytopenic patients receiving corticosteroids as a first line therapy. *wjpmr*. 2017;3(8):405-10.
93. Li Y, Rauniyar VK, Yin WF, Hu B, Ouyang S, Xiao B, et al. Serum IL-21 levels decrease with glucocorticoid treatment in myasthenia gravis. *Neuro Sci*. 2014;35(1):29-34.

94. Ren HM, Lukacher AE, Rahman ZSM, Olsen NJ. New developments implicating IL-21 in autoimmune disease. *J Autoimmun.* 2021;122:1-30.
95. Caprioli F, Sarra M, Caruso R, Stolfi C, Fina D, Sica G, et al. Autocrine regulation of IL-21 production in human T lymphocytes. *J Immunol.* 2008;180(3):1800-7.
96. Simpson N, Gatenby PA, Wilson A, Malik S, Fulcher D, Tangye S, et al. Expansion of circulating T cells resembling follicular helper T cells is a fixed phenotype that identifies a subset of severe systemic lupus erythematosus. *Arthritis Rheum.* 2010;62(1):234-44.
97. Jin H, Carrio R, Yu A, Malek TR. Distinct activation signals determine whether IL-21 induces B cell costimulation, growth arrest, or bim-dependent apoptosis. *J Immunol.* 2004;173(1):657-65.
98. Good KL, Bryant VL, Tangye SG. Kinetics of human B cell behavior and amplification of proliferative responses following stimulation with IL-21. *J Immunol.* 2006;177(8):5236-47.
99. Singh RR. IL-4 and many roads to lupuslike autoimmunity. *Clin Immunol.* 2003;108(2):73-9.

