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Оглавление

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- L10.5 ,
- L10.8 (PAMS)/ (PNP) / IgA

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, 9,3 %.

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1. Joly P, Litrowski N. Pemphigus group (vulgaris, vegetans, foliaceus, herpetiformis, brasiliensis). *Clin Dermatol* 2011; 29: 432–436.
2. Kneisel A, Hertl M. Autoimmune bullous skin diseases. Part 1: clinical manifestations. *J Dtsch Dermatol Ges* 2011; 9: 844–856; quiz 857.
3. Kneisel A, Hertl M. Autoimmune bullous skin diseases. Part 2: diagnosis and therapy. *J Dtsch Dermatol Ges* 2011; 9: 927–947.
4. Amagai M, Klaus-Kovtun V, Stanley JR. Autoantibodies against a novel epithelial cadherin in pemphigus vulgaris, a disease of cell adhesion. *Cell* 1991; 67: 869–877.
5. M Amagai, T Hashimoto, N Shimizu, and T Nishikawa Absorption of pathogenic autoantibodies by the extracellular domain of pemphigus vulgaris antigen (Dsg3) produced by baculovirus. *J Clin Invest.* 1994 Jul; 94(1): 59–67.
6. Vincenzo Ruocco, Eleonora Ruocco, Ada Lo Schiavo, Giampiero Brunetti, Luigi Pio Guerrera, Ronni Wolf. Pemphigus: Etiology, pathogenesis, and inducing or triggering factors: Facts and controversies. *Clinics in Dermatology, Volume 31, Issue 4, Pages 374-381.*
7. Vincenzo Ruocco MD, Sarah Brenner MD and Eleonora Ruocco MD. Pemphigus and diet: does a link exist? *Int J Dermatol.* 2001 Mar;40(3):161-3.
8. Michael H, Cassian S. Pathogenesis, Clinical Manifestations and Diagnosis of Pemphigus. Available from: URL: [http:// www.uptodate.com/store](http://www.uptodate.com/store). Accessed July 7, 2013.
9. M. Hertl, H. Jedlickova, S. Karpati, B. Marinovic, S. Uzun, S. Yayli, D. Mimouni, L. Borradori, C. Feliciani, D. Ioannides, P. Joly, C. Kowalewski, G. Zambruno, D. Zillikens, M.F. Jonkman. Pemphigus. S2 Guideline for diagnosis and treatment – guided by the European Dermatology Forum (EDF) in cooperation with the European Academy of Dermatology and Venereology (EADV) *JEADV* 2015, 29, 405–414
10. Poot AM, Diercks GF, Kramer D et al. Laboratory diagnosis of paraneoplastic pemphigus. *Br J Dermatol* 2013; 169: 1016–1024.
11. Thompson AJ, Newman WG, Elliott RA, Roberts SA, Tricker K, Payne K. The cost-effectiveness of a pharmacogenetic test: a trial-based evaluation of TPMT genotyping for azathioprine. *Value Health* 2014; 17: 22–33.
12. Mentink LF, Mackenzie MW, Toth GG et al. Randomized controlled trial of adjuvant oral dexamethasone pulse therapy in pemphigus vulgaris: PEMPULS trial. *Arch Dermatol* 2006; 142: 570–576.
13. Toth GG, Westerlaken BO, Eilders M, Laseur M, Jonkman MF, Uges DR. Dexamethasone pharmacokinetics after high-dose oral therapy for pemphigus. *Ann Pharmacother* 2002; 36: 1108–1109.
14. Guslandi M. Steroid ulcers: any news? *World J Gastrointest Pharmacol Ther* 2013; 4: 39–40.
15. Martin LK, Werth VP, Villaneuva EV, Murrell DF. A systematic review of randomized controlled trials for pemphigus vulgaris and pemphigus foliaceus. *J Am Acad Dermatol* 2011; 64: 903–908.
16. Chams-Davatchi C, Mortazavizadeh A, Daneshpazhooh M et al. Randomized double blind trial of prednisolone and azathioprine, vs. prednisolone and placebo, in the treatment of pemphigus vulgaris. *J Eur Acad Dermatol Venereol* 2013; 27(10): 1285–1292.

17. Beissert S, Werfel T, Frieling U et al. A comparison of oral methylprednisolone plus azathioprine or mycophenolate mofetil for the treatment of pemphigus. *Arch Dermatol* 2006; 142: 1447–1454.
18. Beissert S, Mimouni D, Kanwar AJ, Solomons N, Kalia V, Anhalt GJ. Treating pemphigus vulgaris with prednisone and mycophenolate mofetil: a multicenter, randomized, placebo-controlled trial. *J Invest Dermatol* 2010; 130: 2041–2048.
19. Ioannides D, Apalla Z, Lazaridou E, Rigopoulos D. Evaluation of mycophenolate mofetil as a steroid-sparing agent in pemphigus: a randomized, prospective study. *J Eur Acad Dermatol Venereol* 2012; 26: 855–860.
20. Chams-Davatchi C, Esmaili N, Daneshpazhooch M et al. Randomized controlled open-label trial of four treatment regimens for pemphigus vulgaris. *J Am Acad Dermatol* 2007; 57: 622–628.
21. Badalamenti SA, Kerdel FA. Azathioprine. In: Wolverton SE, ed. *Comprehensive Dermatologic Drug Therapy*. 2nd ed. London: Elsevier, 2007: 183–195.
22. Meggitt SJ, Anstey AV, Mohd Mustapa MF, Reynolds NJ, Wakelin S. British Association of Dermatologists' guidelines for the safe and effective prescribing of azathioprine 2011. *Br J Dermatol* 2011; 165: 711–734.
23. Kasperkiewicz M, Shimanovich I, Meier M et al. Treatment of severe pemphigus with a combination of immunoabsorption, rituximab, pulsed dexamethasone and azathioprine/mycophenolate mofetil: a pilot study of 23 patients. *Br J Dermatol* 2012; 166: 154–160.
24. Ahmed AR, Spigelman Z, Cavacini LA, Posner MR. Treatment of pemphigus vulgaris with rituximab and intravenous immune globulin. *N Engl J Med* 2006; 355: 1772–1779.
25. Joly P, Mouquet H, Roujeau JC et al. A single cycle of rituximab for the treatment of severe pemphigus. *N Engl J Med* 2007; 357: 545–552.
26. Hertl M, Zillikens D, Borradori L et al. Recommendations for the use of rituximab (anti-CD20 antibody) in the treatment of autoimmune bullous skin diseases. *J Dtsch Dermatol Ges* 2008; 6: 366–373.
27. Amagai M, Ikeda S, Shimizu H et al. A randomized double-blind trial of intravenous immunoglobulin for pemphigus. *J Am Acad Dermatol* 2009; 60: 595–603.
28. Zillikens D, Derfler K, Eming R et al. Recommendations for the use of immunoapheresis in the treatment of autoimmune bullous diseases. *J Dtsch Dermatol Ges* 2007; 5: 881–887.
29. Behzad M, M€obbs C, Kneisel A et al. Combined treatment with immunoabsorption and rituximab leads to fast and prolonged clinical remission in difficult-to-treat pemphigus vulgaris. *Br J Dermatol* 2012; 166: 844–852.
30. Pasricha JS, Khaitan BK, Raman RS, Chandra M. Dexamethasonecyclophosphamide pulse therapy for pemphigus. *Int J Dermatol* 1995; 34: 875–882.
31. Nousari CH, Brodsky R, Anhalt GJ. Evaluating the role of immunoablative high-dose cyclophosphamide therapy in pemphigus vulgaris. *J Am Acad Dermatol* 2003; 49: 148–150.
32. Baum S, Greenberger S, Samuelov L et al. Methotrexate is an effective and safe adjuvant therapy for pemphigus vulgaris. *Eur J Dermatol* 2012; 22: 83–87.
33. Werth VP, Fivenson D, Pandya AG et al. Multicenter randomized, double-blind, placebo-controlled, clinical trial of dapsone as a glucocorticoid-sparing agent in maintenance-phase pemphigus vulgaris. *Arch Dermatol* 2008; 144: 25–32.

34. Iraji F, Asilian A, Siadat AH. Pimecrolimus 1% cream in the treatment of cutaneous lesions of pemphigus vulgaris: a double-blind, placebo-controlled clinical trial. *J Drugs Dermatol* 2010; 9: 684–686.
35. Cohen SN, Lim RP, Paul CJ, Abdullah A. Equal efficacy of topical tacrolimus and clobetasone butyrate in pemphigus foliaceus. *Int J Dermatol* 2006; 45: 1379.
36. Murrell DF, Dick S, Ahmed AR et al. Consensus statement on definitions of disease, end points, and therapeutic response for pemphigus. *J Am Acad Dermatol* 2008; 58: 1043–1046.