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Nota di contenuto	1. Cytokines in cytokine storm syndromes -- 2. History of HLH -- 3. The history of macrophage activation syndrome in autoimmune diseases -- 4. Clinical features of cytokine storm syndromes -- 5. Laboratory features and pathology of cytokine storm syndromes -- 6. Criteria for cytokine storm syndromes -- 7. Familial HLH genetics -- 8. Secondary HLH genetics -- 9. Genetics of macrophage activation syndrome in systemic juvenile idiopathic arthritis -- 10. CD8+ T cell biology in cytokine storm syndromes -- 11. Immunology of cytokine storm syndromes: Natural killer cells -- 12. Myeloid cells in the immunopathogenesis of cytokine storm syndrome -- 13. Cytokine storm -- 14. Primary immunodeficiencies -- 15. Infectious triggers of cytokine storm syndromes: Herpes virus family (non-EBV) -- 16. CSS associated with Epstein Barr Virus -- 17. Hemorrhagic Fever and other viruses -- 18. Cytokine storm syndrome as a manifestation of primary HIV infection -- 19. Bacteria-associated cytokine storm syndrome -- 20. Zoonotic bacterial infections triggering cytokine storm syndrome -- 21. Parasitic and fungal triggers -- 22. COVID-19 pneumonia and

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Sommario/riassunto

Cytokine Storm Syndromes, including HLH and MAS, are frequently fatal disorders, particularly if not recognized early and treated during presentation. The genetics of Cytokine Storm Syndromes are being defined with many of the risk alleles giving rise to mutations in the perforin-mediated cytolytic pathway used by CD8 cytotoxic T cells and natural killer cells. These are being studied using murine models. Up to 10% of the general population may carry risk alleles for developing Cytokine Storm Syndromes, and Cytokine Storm Syndromes are being increasingly recognized around the world in pediatric and adult hospitals. A variety of infectious, rheumatic, and oncologic triggers are commonly associated with Cytokine Storm Syndromes, but understanding this disorder is critical for all researchers and physicians to ensure timely and appropriate therapy. This second edition addresses all aspects of the disorder, from genetics, pathophysiology, and ongoing research, to clinical presentations, risk factors and treatment. New-to-this-edition features include dedicated chapters on severe COVID-19 CSS, and post-COVID CSS of multi-system inflammatory syndrome in children (MIS-C). In addition, novel topics including CSS associated with Castleman disease, pregnancy, therapeutics, transplantation, and cardiac bypass, as well as treatment with JAK inhibitors are addressed. .
