

Why Immune Responses Fail (and how to correct them)

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Primary immunodeficiency diseases (PID) are caused by pathogenic gene mutations leading to defects in the development or functions of immune cells. Patients with this group of diseases are susceptible not only to frequent and severe infections but also to autoimmunity and certain types of cancers. Since these diseases began to be characterized at the cellular and molecular levels in the latter half of the 20th century, researches on PID has been proved to be critical leads for important immunology discoveries. We have been investigating the role of reactive oxygen species (ROS) produced by innate immune cells through studying the immune defects in patients with PID and animal models of these human PID in mice with the mutations corresponding to human patients. The results from our studies revealed novel immune-regulatory and effector mechanisms critical for the pathogenesis in the PID patients and general populations. These novel mechanisms often lead to more effective treatment for patients with immunological diseases. Moreover, we are trying to develop novel treatments based on the molecular insights to correct the ineffective immune responses.