Special Issue

Autoimmune Regulator (AIRE) Gene in the Pathogenesis of Autoimmune Diseases

Message from the Guest Editor

Studying rare diseases with a Mendelian mode of inheritance such as Autoimmune polyglandular syndrome 1 (APS-1) could be a powerful tool to dissect the molecular mechanism underlying the pathogenesis of associated common autoimmune diseases. These include Addison's disease, hypoparathyroidism, autoimmune thyroid disease, type 1 diabetes (T1D) autoimmune hepatitis, vitiligo and alopecia areata. APS 1 is caused by mutations in the autoimmune regulator (AIRE) gene. AIRE regulates promiscuous expression of tissue-restricted antigens (TRA) in the thymus and plays an important role in central tolerance mechanisms. As the expression of TRA genes in the thymus is under the control of AIRE and the autoimmune attack is targeting peripheral tissues expressing these TRA, the therapy of autoimmunity diseases should be directed toward the thymus rather than the peripheral tissues. In this special edition, we elaborate on this and shed light on the role of AIRE in the pathogenesis of autoimmune diseases associated with APS 1.

Guest Editor

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