Autoimmune polyglandular syndrome type 1 (APS-1) is a rare and complex recessively inherited disorder of immune-cell dysfunction with multiple autoimmunities. It presents as a group of symptoms including potentially life-threatening endocrine gland and gastrointestinal dysfunctions. Autoimmune disorders occur when antibodies and immune cells are launched by the body against one or several antigens of its own tissues. APS-1 is caused by changes (mutations) in the autoimmune regulator (AIRE) gene. HLA-DR/DQ genes also play a role in predisposing to which of the component autoimmune disease the patient actually develops. APS-I is a very rare disorder that tends to cluster in certain homogenous populations, including certain groups of Finns, Iranian Jews, and Sardinians. However, it can be found in numerous populations and among multiple ethnic groups. In the US, APS-1 probably affects as few as 1 in every 2-3 million newborns.