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Title: Identification of KCNRG, a bronchial autoantigen, in a children with IPEX syndrome

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Body: IPEX syndrome is a rare disorder of immune regulation caused by mutations in the FOXP3 gene, which is required for the suppressive function of naturally arising CD4 + CD25 + regulatory T cells. It is associated with the presence of common autoantibodies associated with autoimmune disorders. We report an infant, who manifested at first week after birth a type 1 diabetes mellitus and eczema. IPEX syndrome was confirmed by proved V408M mutation in the FOXP3 gene. At one month old he developped wheezing, cough, respiratory distress without infection. Despite treatment with salbutamol and budesonide nebulized, he was still symptomatic. High-resolution tomography scan showed hyperinflation, trapping, and ground-glass opacities. Analysis of bronchoalveolar lavage showed 1 350 000 cells/ml, with 65% alveolar macrophages without infection. Autoantibodies to KCNRG were present in serum at one month and still persist at nine months. Treatment with mycophenolate mofetil was started because of combination of severe diarrhea, persistent respiratory symptoms with an optimal nebulized corticotherapy. One month after the begining of immunosuppresive therapy the corticotherapy was reduced. KCNRG, is a potassium channel regulating protein expressed in bronchial epithelial cells. The presence of this antibody was first decribed in the polyendocrine syndrome type 1. The presence of this antibody in an other autoimmune syndrome confirms the fact that KCNRG is a major bronchial autoantigen.The recognition of pulmonary autoimmunity, and its distinction from asthma is important because the autoimmune bronchiolitis in this case respond well to immunosuppression.