Autosomal dominant hyper IgE syndrome (AD-HIES) is a rare multisystem primary immunodeficiency disorder. Symptoms often become apparent early during infancy or childhood. The disorder is characterized by repeated bacterial infections of the skin and lungs (pneumonia), skeletal abnormalities, and characteristic facial features. The first symptom is often the development of a dry, red flaky skin rash (eczema) at birth or early during infancy. Researchers have discovered that mutations in the STAT3 gene cause AD-HIES in over 60% of the patients. Most cases of AD-HIES occur as the result of a new mutation in this gene. There are two main forms of hyper IgE syndrome - one inherited in an autosomal dominant pattern and one in an autosomal recessive pattern. Both involve defects of the immune system and elevated levels of immunoglobulin E (hyper IgE) in the blood. For years, researchers considered them different expressions of the same disorder, but now researchers consider them similar, yet distinct disorders.