

# ZNF341 Gene

Subjects: **Genetics & Heredity**

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Zinc finger protein 341

genes

## 1. Normal Function

The *ZNF341* gene provides instructions for making a transcription factor, which is a protein that attaches (binds) to specific regions of DNA and helps control the activity of particular genes. The *ZNF341* protein is thought to regulate the activity of the *STAT1* and *STAT3* genes, controlling production of the *STAT1* and *STAT3* proteins, respectively. Both proteins are involved in the immune system. They control pathways in cells that help fight foreign invaders such as viruses, bacteria, and fungi. The *STAT3* protein, in particular, transmits signals for the maturation of immune system cells, especially T cells and B cells. *STAT3* is also involved in normal development and maintenance of bones and other tissues.

Researchers suspect that the *ZNF341* protein controls the activity of other genes, although they have not been identified.

## 2. Health Conditions Related to Genetic Changes

### 2.1. Autosomal dominant hyper-IgE syndrome

At least five mutations in the *ZNF341* gene have been found to cause a condition similar to autosomal dominant hyper-IgE syndrome (AD-HIES), which is a disorder of the immune system characterized by recurrent skin and lung infections and abnormally high levels of an immune system protein called immunoglobulin E (IgE) in the blood. AD-HIES is usually caused by *STAT3* gene mutations and follows an autosomal dominant pattern of inheritance, which means one altered copy of the gene is sufficient to cause the disorder. In contrast, the condition caused by *ZNF341* gene mutations follows an autosomal recessive pattern of inheritance, which means both copies of the gene must be altered for immune system problems to develop.

The *ZNF341* gene mutations that cause an AD-HIES-like condition result in production of an abnormally short *ZNF341* protein or production of no protein. With little or no *ZNF341* protein, production of the *STAT1* and *STAT3* proteins is impaired. A shortage of functional *STAT3* blocks the maturation of T cells (specifically a subset known as Th17 cells) and other immune cells. The resulting immune system abnormalities make people with AD-HIES highly

susceptible to infections, particularly bacterial and fungal infections of the lungs and skin. A shortage of STAT1 protein is not thought to contribute to immune system problems in affected individuals.

## 3. Other Names for This Gene

- ZNF341 gene

## References

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