HLA-DPB1 gene

major histocompatibility complex, class II, DP beta 1

Normal Function

The *HLA-DPB1* gene provides instructions for making a protein that plays a critical role in the immune system. The *HLA-DPB1* gene is part of a family of genes called the human leukocyte antigen (HLA) complex. The HLA complex helps the immune system distinguish the body's own proteins from proteins made by foreign invaders such as viruses and bacteria.

The HLA complex is the human version of the major histocompatibility complex (MHC), a gene family that occurs in many species. The *HLA-DPB1* gene belongs to a group of MHC genes called MHC class II. MHC class II genes provide instructions for making proteins that are present on the surface of certain immune system cells. These proteins attach to protein fragments (peptides) outside the cell. MHC class II proteins display these peptides to the immune system. If the immune system recognizes the peptides as foreign (such as viral or bacterial peptides), it triggers a response to attack the invading viruses or bacteria.

The protein produced from the *HLA-DPB1* gene attaches (binds) to the protein produced from another MHC class II gene, *HLA-DPA1*. Together, they form a functional protein complex called an antigen-binding DPαβ heterodimer. This complex displays foreign peptides to the immune system to trigger the body's immune response.

Each MHC class II gene has many possible variations, allowing the immune system to react to a wide range of foreign invaders. Researchers have identified hundreds of different versions (alleles) of the *HLA-DPB1* gene, each of which is given a particular number (such as *HLA-DPB1*03:01*).

Health Conditions Related to Genetic Changes

Granulomatosis with polyangiitis

At least one variant of the *HLA-DPB1* gene has been associated with granulomatosis with polyangiitis (GPA). This condition occurs when the immune system malfunctions and attacks the body's own tissues and organs (autoimmunity), causing inflammation that affects the lungs, airways, and kidneys. The associated variant, called *HLA-DPB1*0401*, has been found more frequently in people with GPA than in those who do not

have the condition; this variant is thought to increase the risk of developing GPA.

Because the *HLA-DPB1* gene is involved in the immune system, changes in it might be related to the autoimmune response and inflammation that damage the lungs, kidneys, and other organs. However, it is unclear what specific role the *HLA-DPB1* gene variant plays in development of this condition. It is likely that environmental factors trigger the condition in people who are genetically predisposed to it. Other genetic factors are also likely to be involved in GPA.

Juvenile idiopathic arthritis

MedlinePlus Genetics provides information about Juvenile idiopathic arthritis

Rheumatoid arthritis

MedlinePlus Genetics provides information about Rheumatoid arthritis

Other disorders

Variants of the *HLA-DPB1* gene are associated with immune reactions to beryllium, a metallic element that can be toxic. Beryllium exposure can occur in manufacturing plants and the nuclear and aerospace industries. About 2 to 10 percent of people exposed to beryllium develop beryllium sensitization or chronic beryllium disease. Sensitization is an immune reaction that occurs in response to beryllium exposure; sensitization can cause an increase in the number of certain immune system cells in the blood, but it may not lead to any symptoms. In some people, sensitization leads to chronic beryllium disease, which is a lung disease characterized by the formation of small masses of inflammatory cells (granulomas). The lungs can become scarred and stiff and lose their ability to function. Having variants of the *HLA-DPB1* gene that contain the protein building block (amino acid) glutamic acid at position 69 (written as E69) increases the risk of developing beryllium sensitization or chronic beryllium disease.

Other Names for This Gene

- beta1 domain MHC class II HLA DPB
- class II HLA beta chain
- DPB1
- DPB1_HUMAN
- HLA class II histocompatibility antigen, DP beta 1 chain
- HLA class II histocompatibility antigen, DP(W4) beta chain
- HLA DP14-beta chain
- HLA-DP
- HLA-DP histocompatibility type, beta-1 subunit
- HLA-DP1B
- HLA-DPB

- major histocompatibility complex class II antigen beta chain
- MHC class II antigen beta chain
- MHC class II antigen DP beta 1 chain
- MHC class II antigen DPB1
- MHC class II antigen DPbeta1
- MHC class II HLA-DP-beta-1
- MHC class II HLA-DRB1
- MHC HLA DPB1

Additional Information & Resources

Tests Listed in the Genetic Testing Registry

- Tests of HLA-DPB1 (https://www.ncbi.nlm.nih.gov/gtr/all/tests/?term=3115[geneid])

Scientific Articles on PubMed

- PubMed (https://pubmed.ncbi.nlm.nih.gov/?term=%28HLA-DPB1%5BTIAB%5D%29+AND+%28%28Genes%5BMH%5D%29+OR+%28Genetic+Phenomena%5BMH%5D%29%29+AND+english%5Bla%5D+AND+human%5Bmh%5D+AND+%22last+720+days%22%5Bdp%5D)

Catalog of Genes and Diseases from OMIM

- MAJOR HISTOCOMPATIBILITY COMPLEX, CLASS II, DP BETA-1; HLA-DPB1 (https://omim.org/entry/142858)

Gene and Variant Databases

- NCBI Gene (https://www.ncbi.nlm.nih.gov/gene/3115)
- ClinVar (https://www.ncbi.nlm.nih.gov/clinvar?term=HLA-DPB1[gene])

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Genomic Location

The *HLA-DPB1* gene is found on chromosome 6 (https://medlineplus.gov/genetics/chromosome/6/).

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