

Gene Section

Mini Review

CIITA (MHC class II transactivator)

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Identity

Other names: MHC2TA (MHC class II transactivator); C2TA

HGNC (Hugo): CIITA

Location: 16p13

DNA/RNA

Transcription

6.7 kb mRNA; coding sequence: 3392 bp.

Protein

Description

1130 amino acids.

Function

Non-DNA-binding transcriptional coactivator; highly regulated; IFN-gamma-inducible; transactivator of the major histocompatibility complex (MHC) class II genes, herein the name; DM and Ii (molecules associated to MHC class II) are also regulated by MHC2TA; MHC2TA also transactivates MHC class I; lymphoid cells of MHC2TA-null mice do not express MHC class II.

Implicated in

t(3;16)(q27;p13)/ BCL6

Disease

Non-Hodgkin lymphoma.

Prognosis

Yet unknown (only 1 case available with certainty).

Hybrid/Mutated gene

5' MHC2TA - 3' BCL6; MHC2TA fuses with the second exon of BCL6.

Abnormal protein

No fusion protein, but promoter exchange.

Oncogenesis

BCL6 is a transcription repressor; it is supposed that substitution of the promoter of BCL6 may be responsible for BCL6 deregulation.

Bare lymphocyte syndrome (BLS)

Note

At least five complementation groups.

Disease

Severe immunodeficiency due to failure to express MHC class II genes.

References

Steimle V, Otten LA, Zufferey M, Mach B. Complementation cloning of an MHC class II transactivator mutated in hereditary MHC class II deficiency (or bare lymphocyte syndrome). *Cell*. 1993 Oct 8;75(1):135-46

Wright KL, Chin KC, Linhoff M, Skinner C, Brown JA, Boss JM, Stark GR, Ting JP. CIITA stimulation of transcription factor binding to major histocompatibility complex class II and associated promoters in vivo. *Proc Natl Acad Sci U S A*. 1998 May 26;95(11):6267-72

Fontes JD, Kanazawa S, Nekrep N, Peterlin BM. The class II transactivator CIITA is a transcriptional integrator. *Microbes Infect*. 1999 Sep;1(11):863-9

Yoshida S, Kaneita Y, Aoki Y, Seto M, Mori S, Moriyama M. Identification of heterologous translocation partner genes fused to the BCL6 gene in diffuse large B-cell lymphomas: 5'-RACE and LA - PCR analyses of biopsy samples. *Oncogene*. 1999 Dec 23;18(56):7994-9

Girdlestone J. Synergistic induction of HLA class I expression by RelA and CIITA. *Blood*. 2000 Jun 15;95(12):3804-8

Masternak K, Muhlethaler-Mottet A, Villard J, Zufferey M, Steimle V, Reith W. CIITA is a transcriptional coactivator that is recruited to MHC class II promoters by multiple synergistic interactions with an enhanceosome complex. *Genes Dev*. 2000 May 1;14(9):1156-66

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