

# Accepted Manuscript

Severe disease and greater impairment of NF- $\kappa$ B activation in I $\kappa$ B $\alpha$  point mutants versus truncation mutants in autosomal dominant anhidrotic ectodermal dysplasia with immune deficiency

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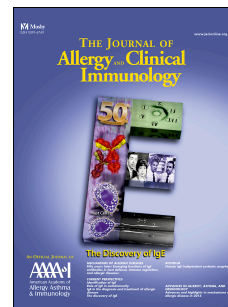
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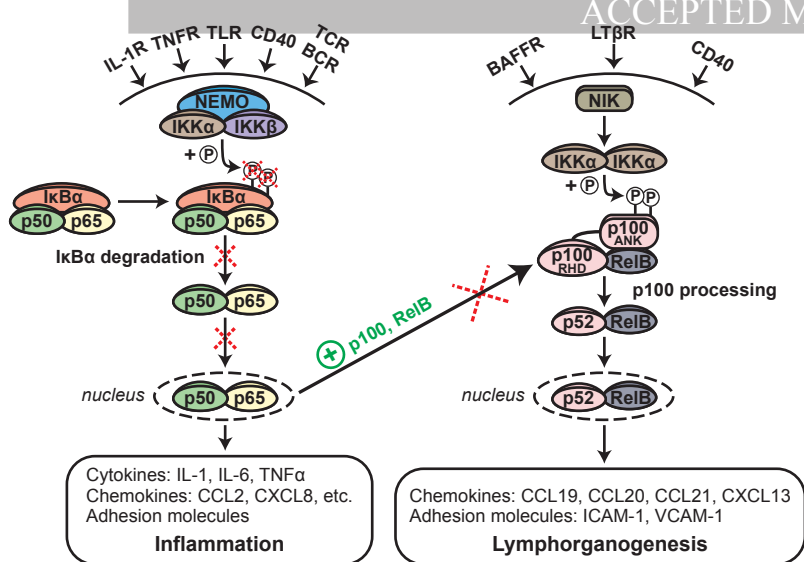
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Canonical NF- $\kappa$ B pathwayNon-canonical NF- $\kappa$ B pathway

ANK, Ankyrin repeat domain; BAFFR, B cell-activating factor receptor; BCR, B cell receptor  
 IL-1R, Interleukin-1 receptor; LT $\beta$ R, Lymphotoxin beta receptor; RHD, Rel homology domain  
 TCR, T cell receptor; TLR, Toll-like receptor; TNFR, Tumor necrosis factor receptor

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