

Abstract: The authors have used a 3.6-kb DNA fragment containing the *hprt* gene to construct a transgenic mouse model of human *hprt* deficiency. The resulting mice, which express human *hprt* in all tissues, are fertile and healthy. The authors have also used this model to study the effects of various *hprt* mutations on the development of the immune system. The results show that mice with a mutation in the *hprt* gene that encodes the active site of the enzyme develop a severe combined immunodeficiency (SCID) that is similar to the one seen in humans with the same mutation.

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