

# Ku70 shown to be critical regulator of DNA damage in Huntington's disease

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Ku70, a component of the DNA repair complex, is shown to be a new critical player in the DNA damage-linked pathologies of Huntington's disease (HD), according to a study in the May 3 issue of the *Journal of Cell Biology*.

DNA repair defends against naturally occurring or disease-related DNA damage during the long lifespan of neurons. Impairments to this process underlie "polyQ" diseases, a major group of hereditary neurodegenerative disorders that includes HD. Understanding the multiple pathogenic pathways that lead to such [DNA repair](#) dysfunction is key for the development of new therapies.

In this study, Hitoshi Okazawa and colleagues report that expression of mutant huntingtin (Htt)—the protein responsible for HD—in neurons causes double-strand breaks (DSBs) in genomic DNA and impairs DNA repair. The researchers identify Ku70 as a mediator of the DNA repair dysfunction in mutant Htt-expressing neurons—mutant Htt interacts with Ku70, impairing its function in nonhomologous end joining, which consequently increases DSB accumulation. Boosting Ku70 levels rescues mutant Htt-induced neurodegeneration in a [mouse model](#) of HD, suggesting that Ku70 is a critical regulator of [DNA damage](#) in HD pathology.

**More information:** Enokido, Y., et al. 2010. *J. Cell Biol.*  
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