Cockayne Syndrome (CS) is a rare, autosomal recessive, neurodegenerative disorder. CS Type II, the congenital form of the disease, is caused by up to 61 different mutations in the ERCC6 gene, which codes for the Cockayne Syndrome B protein (CSB) [1]. CSB is a major factor in successful transcription-coupled nucleotide excision repair (TC-NER). When RNA polymerase II is halted by a DNA lesion during transcription, CSB helps recognize the damaged DNA and recruits essential NER factors that ultimately assemble into the TC-NER complex and fix the DNA so that RNA polymerase can continue transcription [2]. One might think of CSB as a trailblazer; it works slightly upstream of RNA polymerase II, gathers information on the integrity of the DNA, and sends help if it detects a problem. Because many DNA lesions are the result of UV radiation, it makes sense that dysfunction CSB protein leads to an increased sensitivity to light, a key symptom in all types of CS. However, CS is a complex multisystem disorder with many other symptoms where the link between molecular defect and phenotype is not as clear. CS is a progeroid disorder, meaning that part of it’s general phenotype includes premature aging. It is hypothesized that the buildup of oxidative damage in cells and the inability to process all of the damage is a major factor that leads to aging. Interestingly, CSB is thought to play a role in processing this type of DNA damage also, but its exact function is not clear [3]. In order to further understand the complex phenotype of CS and how defective CSB results in thprogeroid symptoms, *the role of CSB in repairing oxidative damage must be further understood.*

**[1]** Laugel, V., Dalloz, C., Durand, M. et al. 2010. Mutation update for the *CSB*/*ERCC6* and *CSA*/*ERCC8* genes involved in Cockayne syndrome. *Hum. Mutat*., 31: 113–126. [doi:10.1002/humu.21154](http://onlinelibrary.wiley.com.ezproxy.library.wisc.edu/wol1/doi/10.1002/humu.21154/abstract)

**[2]** Vermeulen, W., Fousteri, M. 2013. Mammalian transcription-coupled excision repair. *Cold Spring Harb. Perspect. Biol*. 5: a012625.

**[3]** Stevnsner, T., Muftuoglu, M., Aamann, M. D., & Bohr, V. A. 2008. The role of Cockayne Syndrome group B (CSB) protein in base excision repair and aging. *Mechanisms of Ageing and Development*, *129*(7-8), 441–448. http://doi.org/10.1016/j.mad.2008.04.009