MSH1 is one of six MutS homologs in S. cerevisiae involved in mismatch repair but it is the only member of the family to function in the repair and maintenance of mitochondrial DNA. MSH1 is required for mitochondrial functioning and maintenance of mitochondrial morphology. Msh1p has DNA-dependent ATPase activity and preferentially 3684>binds DNA substrates containing nucleotide mismatches and unpaired nucleotides. Although MSH1 expression is increased during meiosis, there is no evidence that MSH1 is involved in meiotic recombination.Msh1p shares 29% sequence identity with the bacterial MutS. The apparent lack of a MSH1 homolog in humans or other eukaryotes has led to the proposal that the high mutation rate of human mtDNA is due to limited mismatch repair.