SEN1 encodes a Pol II transcriptional termination factor that is also involved in the maturation of noncoding RNAs. Based on sequence similarity to and functional characterization of the S. pombe ortholog, S. cerevisiae Sen1p is presumed to be a 5'-3' ATP-dependent RNA/DNA helicase, although this activity has not been directly shown in S. cerevisiae. Sen1p localizes to the nucleus in a granular pattern. SEN1 is an essential gene but loss of function mutations in Sen1p cause altered accumulation of tRNA and ribosomal RNA precursors, mislocalization of core snoRNPs, and 3'-extension and transcriptional read-through of some snoRNAs, snRNAs, and short protein-encoding mRNAs. Mutations in the human homolog Senataxin, result in the progressive neurological diseases Ataxia-Oculomotor Apraxia 2and juvenile Amyotrophic Lateral Sclerosis 4.