

Product name:	ERCC1(1B10)Mouse Monoclonal Antibody
Cat number:	MABN10578
Conjugate:	Unconjugated
Size:	100μL
Clone:	Monoclonal
Concentration:	1mg/ml
Host:	Mouse
Isotype:	IgG
Immunogen:	Synthetic Peptide of ERCC1
Reactivity:	Human
Applications:	WB 1:500-1:2000,IHC 1:100-1:300,ICC/IF 1:50-1:200
Molecular Weight:	36kDa
Purification:	Affinity purification
Form:	Liquid
Buffer:	PBS, pH 7.4, containing 0.5%BSA, 0.02% New type preservative N as Preservative and 50% Glycerol.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

Background:

The product of this gene functions in the nucleotide excision repair pathway, and is required for the repair of DNA lesions such as those induced by UV light or formed by electrophilic compounds including cisplatin. The encoded protein forms a heterodimer with the XPF endonuclease (also known as ERCC4), and the heterodimeric endonuclease catalyzes the 5' incision in the process of excising the DNA lesion. The heterodimeric endonuclease is also involved in recombinational DNA repair and in the repair of inter-strand crosslinks. Mutations in this gene result in cerebrooculofacioskeletal syndrome, and polymorphisms that alter expression of this gene may play a role in carcinogenesis. Multiple transcript variants encoding different isoforms have been found for this gene. The last exon of this gene overlaps with the CD3e molecule, epsilon associated protein gene. Defects in ERCC1 are the cause of cerebro-oculo-facio-skeletal syndrome type 4 (COFS4) [MIM:610758]. COFS is a degenerative autosomal recessive disorder of prenatal onset affecting the brain, eye and spinal cord. After birth, it leads to brain atrophy, hypoplasia of the corpus callosum, hypotonia, cataracts, microcornea, optic atrophy, progressive joint contractures and growth failure. Facial dysmorphism is a constant feature. Abnormalities of the skull, eyes, limbs, heart and kidney also occur. **function:**Structure-specific DNA repair endonuclease responsible for the 5'-incision during DNA repair. **similarity:**Belongs to the ERCC1/RAD10/SWI10 family. **subunit:**Heterodimer composed of ERCC1 and XPF/ERCC4.