

## [KD Validated] Anti-HTT Rabbit mAb

Purified Recombinant Rabbit Monoclonal Antibody

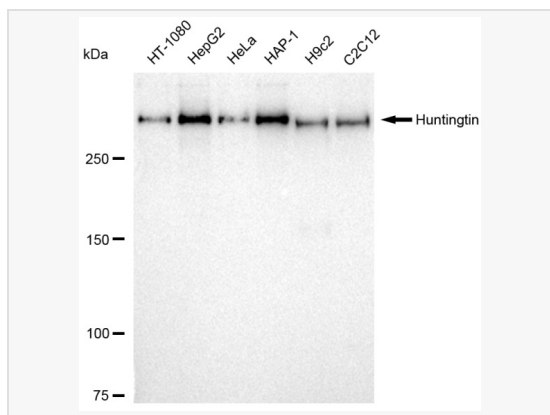
Catalog # R021114

### Product Information

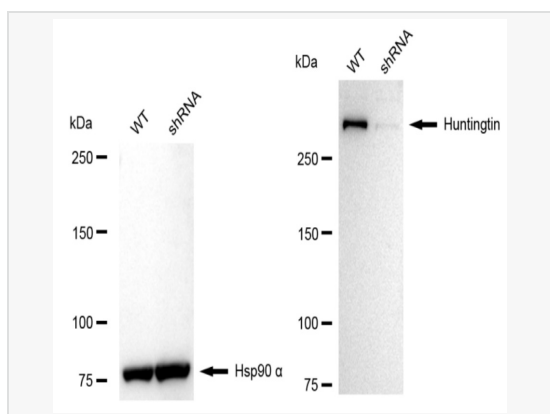
|             |   |
|-------------|---|
| Application | WB, FC, IF (Cell)/ICC, IHC-P/IF (Tissue-P)  |
| Reactivity  | Human, Mouse, Rat   |
| Dilution    | WB 1:1,000~1:5,000; FC 1:200~1:2,000; IF 1:100~1:1,000; IHC-P 1:100~1:200   |
| Host        | Rabbit  |
| Clonality   | Monoclonal  |
| Clone No.   | 24D84P30  |
| Isotype     | IgG   |
| Label       | Unconjugated  |
| Immunogen   | A synthesized peptide derived from human Huntingtin   |
| Format      | Affinity purified monoclonal antibody supplied in PBS with 0.02% sodium azide and 50% glycerol, pH 7.3.                         |
| Storage     | Shipped on wet ice. Store at -20°C. Stable for 12 months from date of receipt. Aliquoting is unnecessary for -20°C storage.     |
| Precautions | [KD Validated] Anti-HTT Rabbit mAb [24D84P30] is for research use only and not for use in diagnostic or therapeutic procedures. |

### Protein Information

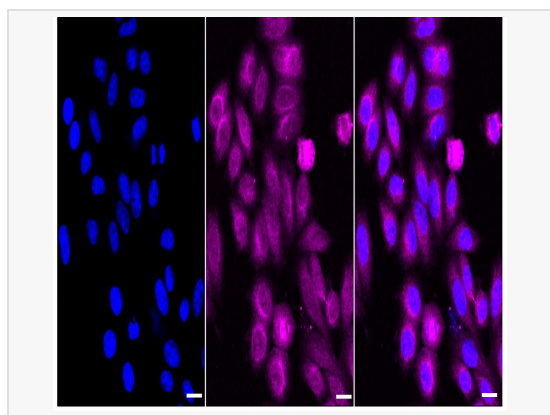
|                   |  |
|-------------------|--|
| Synonyms          | HTT; Huntingtin; IT15; HD; Huntington Disease Protein; Huntingtin (Huntington Disease); HD Protein; LOMARS.  |
| Calculated MW     | Calculated MW: 348 kDa, Observed MW: 348 kDa   |
| Uniprot ID        | P42858   |
| Gene ID           | 3064   |
| Background        | Huntingtin is a disease gene linked to Huntington's disease, a neurodegenerative disorder characterized by loss of striatal neurons. This is thought to be caused by an expanded, unstable trinucleotide repeat in the huntingtin gene, which translates as a polyglutamine repeat in the protein product. A fairly broad range of trinucleotide repeats (9-35) has been identified in normal controls, and repeat numbers in excess of 40 have been described as pathological. The huntingtin locus is large, spanning 180 kb and consisting of 67 exons. The huntingtin gene is widely expressed and is required for normal development. It is expressed as 2 alternatively polyadenylated forms displaying different relative abundance in various fetal and adult tissues. The larger transcript is approximately 13.7 kb and is expressed predominantly in adult and fetal brain whereas the smaller transcript of approximately 10.3 kb is more widely expressed. The genetic defect leading to Huntington's disease may not necessarily eliminate transcription, but may confer a new property on the mRNA or alter the function of the protein. One candidate is the huntingtin-associated protein-1, highly expressed in brain, which has increased affinity for huntingtin protein with expanded polyglutamine repeats. This gene contains an upstream open reading frame in the 5' UTR that inhibits expression of the huntingtin gene product through translational repression. [provided by RefSeq, Jul 2016] |
| Cellular Location | Cytoplasm. Nucleus. The mutant Huntingtin protein colocalizes with AKAP81 in the nuclear matrix of Huntington's disease  |



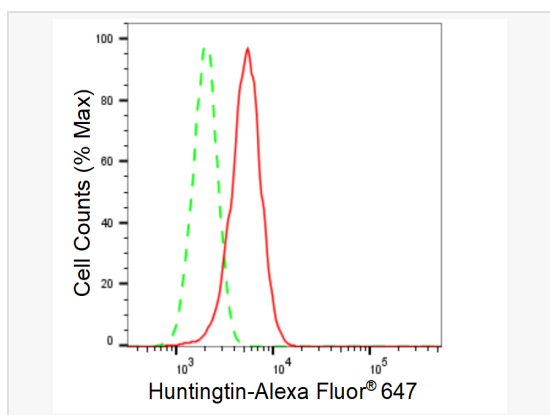
Western blotting analysis using Huntingtin antibody (R021114). Total cell lysates (30  $\mu$ g) from various cell lines were loaded and separated by SDS-PAGE. The blot was incubated with Huntingtin antibody (R021114, 1:5,000) and HRP-conjugated goat anti-rabbit secondary antibody (1:20,000) respectively. Image was developed using ECL Substrate Kit.



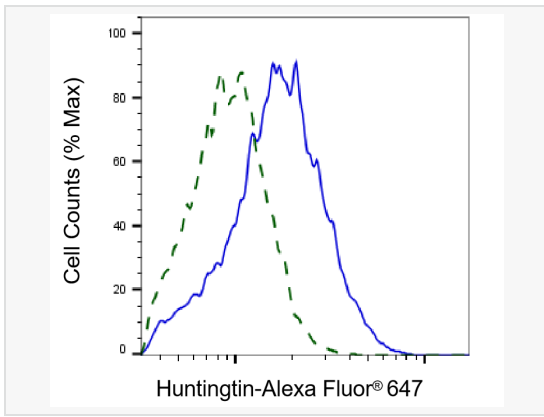
Western blotting analysis using Huntingtin antibody (R021114). Huntingtin expression in wild type (WT) and Huntingtin shRNA knockdown (KD) HeLa cells with 20  $\mu$ g of total cell lysates. Hsp90  $\alpha$  serves as a loading control. The blot was incubated with Huntingtin antibody (R021114, 1:5,000) and HRP-conjugated goat anti-rabbit secondary antibody (1:20,000) respectively. Image was developed using ECL Substrate Kit.



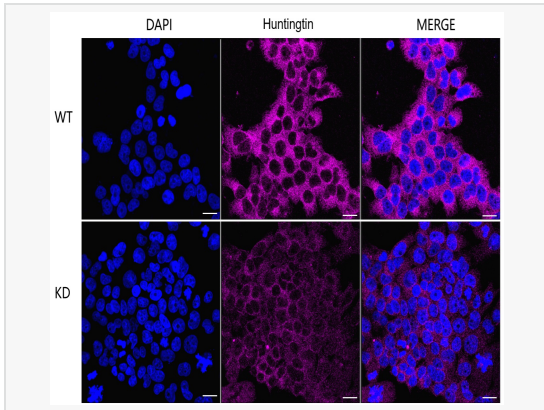
Immunocytochemical staining of HepG2 cells with Huntingtin antibody (R021114, 1:1,000). Nuclei were stained blue with DAPI; Huntingtin was stained magenta with Alexa Fluor<sup>®</sup> 647. Images were taken using Leica stellaris 5. Protein abundance based on laser Intensity and smart gain: Medium. Scale bar: 20  $\mu$ m.



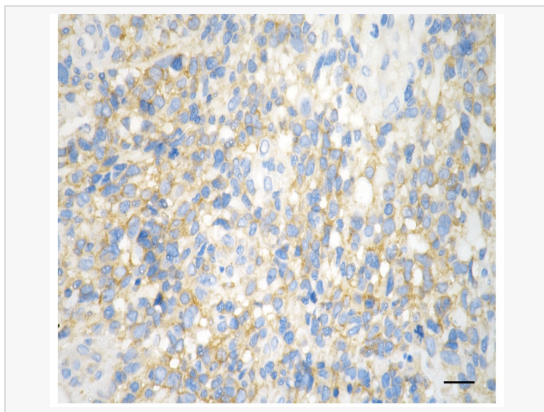
Flow cytometric analysis of Huntingtin expression in HepG2 cells using Huntingtin antibody (R021114, 1:2,000). Green, isotype control; red, Huntingtin.



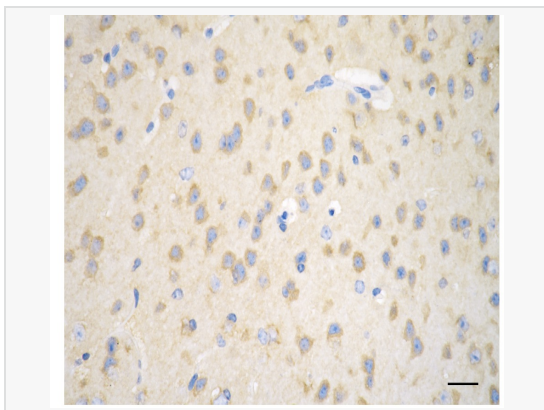
Validation of Huntingtin knockdown using flow cytometry. Wild-type(WT, Blue) and knockdown(KD, Green) HeLa cells were stained with Huntingtin antibody (R021114, 1:2,000) and analyzed using BD flow cytometer.



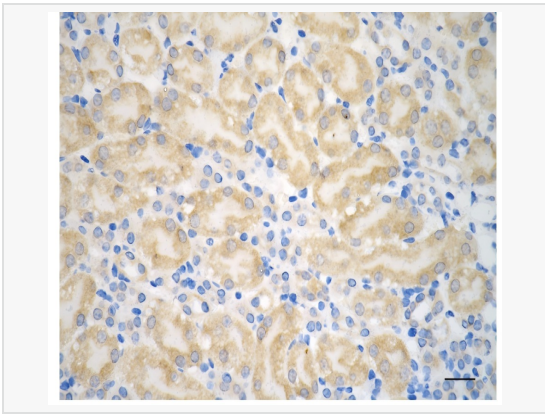
Immunocytochemical staining of HeLa cells using Huntingtin antibody (R021114, 1:1,000), Top panel: wild-type (WT); Bottom panel: Huntingtin shRNA knockdown (KD). Nuclei were stained blue with DAPI; Huntingtin was stained magenta with Alexa Fluor® 647. Scale bar, 20  $\mu$ m. Permeabilization: Triton.



Immunohistochemistry was performed on paraffin-embedded human glioblastoma using huntingtin antibody (R021114, 1:200). Antigen retrieval was done in sodium citrate buffer (pH 6.0). DAB was used for detection, with hematoxylin counterstaining. Images were acquired using a Nikon Ci-L Plus microscope (40 $\times$  objective). Scale bar: 25  $\mu$ m.



Immunohistochemistry was performed on paraffin-embedded mouse brain using huntingtin antibody (R021114, 1:200). Antigen retrieval was done in sodium citrate buffer (pH 6.0). DAB was used for detection, with hematoxylin counterstaining. Images were acquired using a Nikon Ci-L Plus microscope (40 $\times$  objective). Scale bar: 25  $\mu$ m.



Immunohistochemistry was performed on paraffin-embedded mouse kidney using huntingtin antibody (R021114, 1:200). Antigen retrieval was done in sodium citrate buffer (pH 6.0). DAB was used for detection, with hematoxylin counterstaining. Images were acquired using a Nikon Ci-L Plus microscope (40× objective). Scale bar: 25  $\mu$ m.