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## anti-TIMM8A/DDP antibody (AA 31-97) (AbBy Fluor® 680)



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| Quantity:            | 100 μL  |
|----------------------|---|
| Target:              | TIMM8A/DDP (TIMM8A)   |
| Binding Specificity: | AA 31-97  |
| Reactivity:          | Human   |
| Host:                | Rabbit  |
| Clonality:           | Polyclonal  |
| Conjugate:           | This TIMM8A/DDP antibody is conjugated to AbBy Fluor® 680   |
| Application:         | Immunofluorescence (Cultured Cells) (IF (cc)), Immunofluorescence (Paraffin-embedded Sections) (IF (p)) |

#### **Product Details**

| Immunogen:            | KLH conjugated synthetic peptide derived from human TIMM8A |
|-----------------------|--|
| Isotype:              | IgG  |
| Predicted Reactivity: | Human,Mouse,Rat,Cow,Sheep,Pig                              |
| Purification:         | Purified by Protein A.                                     |

#### **Target Details**

| Target:           | TIMM8A/DDP (TIMM8A)  |
|-------------------|--|
| Alternative Name: | TIMM8A (TIMM8A Products)   |
| Background:       | Synonyms: DDP 1, DDP, DDP1, Deafness dystonia protein 1, Deafness/dystonia peptide, DFN 1, |

DFN1, MGC12262, Mitochondrial import inner membrane translocase subunit Tim8 A, MTS, TIM 8A, TIM8A, TIMM 8A, Translocase of inner mitochondrial membrane 8 homolog A, X linked deafness dystonia protein, TIM8A\_HUMAN.

Background: The majority of mitochondrial-directed proteins are encoded by the nuclear genome and are transported to the mitochondria via regulated processes involving the mitochondrial Tom and Tim proteins (1). The mitochondrial Tim protein family is comprised of a large group of evolutionarily conserved proteins that are found in most eukaryotes (1,2). Import of nuclear-encoded precursor proteins into and across the mitochondrial inner membrane is mediated by two distinct complexes, the Tim23 complex and the Tim22 complex, which differ in their substrate specificity (1). Defects in Tim proteins are implicated in several neuro-degenerative diseases, suggesting important roles for Tim proteins in development and health (3,4). Tim8A and Tim8B, which map to human chromosomes Xq22.1 and 11q23.1-q23.2, respectively, are conserved proteins of the mitochondrial intermembrane space, which are organized in hetero-oligomeric complex with Tim13 (5,6,7). Tim8A is highly expressed in fetal and adult brain (5). Tim8A is mutated in deafness dystonia syndrome, a novel type of disease that causes severe neurological defects, thought to be caused by a defective mitochondrial protein transport system (5,8).

#### **Application Details**

| Application Notes: | IF(IHC-P) 1:50-200    |
|--------------------|-----------------------|
|                    | IF(IHC-F) 1:50-200    |
|                    | IF(ICC) 1:50-200      |
| Restrictions:      | For Research Use only |

#### Handling

| Format:            | Liquid   |
|--------------------|--|
| Concentration:     | 1 μg/μL  |
| Buffer:            | Aqueous buffered solution containing 0.01M TBS (pH 7.4) with 1 % BSA, 0.03 % Proclin300 and 50 % Glycerol.         |
| Preservative:      | ProClin  |
| Precaution of Use: | This product contains ProClin: a POISONOUS AND HAZARDOUS SUBSTANCE, which should be handled by trained staff only. |
| Storage:           | -20 °C   |

### Handling

| Storage Comment: | Store at -20°C. Aliquot into multiple vials to avoid repeated freeze-thaw cycles. |
|------------------|---|
| Expiry Date:     | 12 months   |