Product datasheet

Anti-CBS antibody ab135626

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Overview

<table>
<thead>
<tr>
<th>Product name</th>
<th>Anti-CBS antibody</th>
</tr>
</thead>
<tbody>
<tr>
<td>Description</td>
<td>Rabbit polyclonal to CBS</td>
</tr>
<tr>
<td>Host species</td>
<td>Rabbit</td>
</tr>
<tr>
<td>Tested applications</td>
<td>Suitable for: WB, IHC-P, Flow Cyt, ICC/IF</td>
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<tr>
<td>Species reactivity</td>
<td>Reacts with: Mouse, Human</td>
</tr>
<tr>
<td>Immunogen</td>
<td>Synthetic peptide conjugated to KLH, corresponding to a region within internal sequence amino acids 308-338 of Human CBS</td>
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<tr>
<td>Positive control</td>
<td>Mouse kidney tissue lysates, Human brain tissue and 293 cells.</td>
</tr>
</tbody>
</table>

Properties

<table>
<thead>
<tr>
<th>Form</th>
<th>Liquid</th>
</tr>
</thead>
</table>
| Storage buffer   | Preservative: 0.09% Sodium azide  
                              Constituent: 99% PBS |
| Purity           | Immunogen affinity purified |
| Purification notes| ab135625 is purified through a protein A column, followed by peptide affinity purification. |
| Clonality        | Polyclonal |
| Isotype          | IgG |

Applications

Our Abpromise guarantee covers the use of ab135626 in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

<table>
<thead>
<tr>
<th>Application</th>
<th>Notes</th>
</tr>
</thead>
<tbody>
<tr>
<td>WB</td>
<td>1/100 - 1/500. Predicted molecular weight: 61 kDa.</td>
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</tbody>
</table>
**Function**

Only known pyridoxal phosphate-dependent enzyme that contains heme. Important regulator of hydrogen sulfide, especially in the brain, utilizing cysteine instead of serine to catalyze the formation of hydrogen sulfide. Hydrogen sulfide is a gastratransmitter with signaling and cytoprotective effects such as acting as a neuromodulator in the brain to protect neurons against hypoxic injury.

**Tissue specificity**

In the adult strongly expressed in liver and pancreas, some expression in heart and brain, weak expression in lung and kidney. In the fetus, expressed in brain, liver and kidney.

**Pathway**

Amino-acid biosynthesis; L-cysteine biosynthesis; L-cysteine from L-homocysteine and L-serine: step 1/2.

**Involvement in disease**

Defects in CBS are the cause of cystathionine beta-synthase deficiency (CBSD) [MIM:236200]. CBSD is an enzymatic deficiency resulting in altered sulfur metabolism and homocystinuria. The clinical features of untreated homocystinuria due to CBS deficiency include myopia, ectopia lentis, mental retardation, skeletal anomalies resembling Marfan syndrome, and thromboembolic events. Light skin and hair can also be present. Biochemical features include increased urinary homocysteine and methionine.

**Sequence similarities**

Belongs to the cysteine synthase/cystathionine beta-synthase family. Contains 1 CBS domain.

**Cellular localization**

Cytoplasm. Nucleus.

**Images**

Anti-CBS antibody (ab135626) at 1/100 dilution + Mouse kidney tissue lysate at 35 µg

*Predicted band size: 61 kDa*
**Western blot** - Anti-CBS antibody (ab135626)

**All lanes**: Anti-CBS antibody (ab135626) at 1/100 dilution

**Lane 1**: nontransfected 293 cells lysate

**Lane 2**: lysate of 293 cells transiently transfected with the CBS gene

Lysates/proteins at 2 µg per lane.

**Predicted band size**: 61 kDa

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**Immunohistochemistry** (Formalin/PFA-fixed paraffin-embedded sections) - Anti-CBS antibody (ab135626)

Immunohistochemical analysis of Formalin-fixed, Paraffin-embedded Human brain tissue labelling CBS with ab135626 at 1/50 dilution followed by peroxidase-conjugated secondary antibody and DAB staining.

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**Flow Cytometry** - Anti-CBS antibody (ab135626)

Flow cytometric analysis of ab135626 staining CBS at 1/10 in 293 cells (bottom histogram) compared to a negative control cell (top histogram). FITC-conjugated goat-anti-rabbit secondary antibodies were used for the analysis.
Confocal immunofluorescent analysis of CBS using ab135626 at 1/10 dilution in 293 cell followed by Alexa Fluor 488 conjugated goat anti-rabbit IgG (green). Actin filaments have been labeled with Alexa Fluor 555 phalloidin (red). DAPI was used to stain the cell nuclear (blue).

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