

# AGL (C-18): sc-130115

## BACKGROUND

AGL (amylo-1,6-glucosidase, 4- $\alpha$ -glucotransferase), also known as GDE (glycogen debranching enzyme), is a 1,532 amino acid protein that exists as three alternatively spliced isoforms which are expressed in kidney, liver, heart and muscle in an isoform-specific manner. Exhibiting multifunctional enzyme capabilities, AGL contains two catalytic active sites, one of which acts as an 4- $\alpha$ -glucotransferase and the other of which acts as an amylo-1,6-glucosidase during glycogen degradation. Defects in the gene encoding AGL are the cause of glycogen storage disease type 3 (GSD3), also known as Forbes disease. GSD3 is a metabolic disorder that is characterized by the presence of abnormal glycogen due to a lack of AGL activity. Symptoms of GSD3 generally include hypoglycemia, variable myopathy, hepatomegaly and short stature.

## REFERENCES

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## CHROMOSOMAL LOCATION

Genetic locus: AGL (human) mapping to 1p21.2.

## SOURCE

AGL (C-18) is a purified rabbit polyclonal antibody raised against a peptide mapping near the C-terminus of AGL of human origin.

## PRODUCT

Each vial contains 100  $\mu$ g IgG in 1.0 ml PBS with < 0.1% sodium azide and 0.1% gelatin.

## APPLICATIONS

AGL (C-18) is recommended for detection of AGL of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2  $\mu$ g per 100-500  $\mu$ g of total protein (1 ml of cell lysate)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

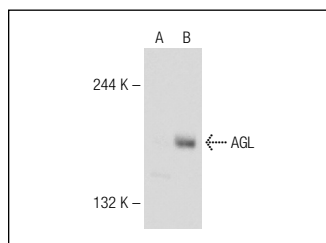
Suitable for use as control antibody for AGL siRNA (h): sc-88368, AGL shRNA Plasmid (h): sc-88368-SH and AGL shRNA (h) Lentiviral Particles: sc-88368-V.

Molecular Weight of AGL: 160 kDa.

## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml).

## DATA



AGL (C-18): sc-130115. Western blot analysis of AGL expression in non-transfected: sc-117752 (A) and mouse AGL transfected: sc-178264 (B) 293T whole cell lysates.

## STORAGE

Store at 4° C, \*\*DO NOT FREEZE\*\*. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

## RESEARCH USE

For research use only, not for use in diagnostic procedures.