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# **LRBA**

Cat.No. 524 003; Polyclonal rabbit antibody, 50 µg specific antibody (lyophilized)

# **Data Sheet**

Reconstitution/ Storage	50 μg specific antibody, lyophilized. Affinity purified with the immunogen. Albumin and azide were added for stabilization. For <b>reconstitution</b> add 50 μl H <sub>2</sub> O to get a 1mg/ml solution in PBS. Then aliquot and store at -20°C to -80°C until use. Antibodies should be stored at +4°C when still lyophilized. Do not freeze! For detailed information, see back of the data sheet.
Applications	WB: 1: 1000 up to 1: 5000 (AP staining) IP: not tested yet ICC: 1: 500 up to 1: 2000 IHC: 1: 2000 IHC: 1: 1000
Immunogen	Recombinant protein corresponding to residues near the central region of mouse LRBA (UniProt Id: Q9ESE1)
Reactivity	Reacts with: mouse (Q9ESE1). Other species not tested yet.
Specificity	K.O. validated PubMed: <u>28814779</u>
Remarks	<b>WB</b> : To avoid protein aggregation, do not heat samples for SDS-PAGE. Due to the large size of this protein, we recommend NuPAGE 3-8% Tris-Acetate gels for SDS-PAGE.

TO BE USED IN VITRO / FOR RESEARCH ONLY NOT TOXIC, NOT HAZARDOUS, NOT INFECTIOUS, NOT CONTAGIOUS

#### **Background**

LRBA (Lipopolysaccharide-Responsive and Beige-Like Anchor Protein) is a member of the BEACH-protein family. It is a large cytosolic protein comprising several functional domains, including the Plekstrin Homology (PH)-like domain, BEACH domain, and multiple WD40 repeats (1).

LRBA is essential for the proper functioning of immune cells, particularly B cells and T cells. It has been shown to regulate the recycling of the immune receptor CTLA-4, which is crucial for T cell activation and downregulation (2, 3). LRBA deficiency manifests as immunodeficiency, autoimmune diseases, and various hematological disorders, like reduced levels of B cells, impaired immunoglobulin production and more (4).

Dysfunctional LRBA is also connected to several CNS disorders like sensorineural hearing loss (5), cerebral granulomatous lesions, nerve demyelination and atrophy (6).

LRBA is widely expressed in multiple tissues including lymphoid organs, the gastrointestinal tract, kidney, the pancreas, and neuronal cells (7).

#### Selected References for 524 003

The BEACH Protein LRBA Promotes the Localization of the Heterotrimeric G-protein Golf to Olfactory Cilia. Kurtenbach S, Gießl A, Strömberg S, Kremers J, Atorf J, Rasche S, Neuhaus EM, Hervé D, Brandstätter JH, Asan E, Hatt H, et al. Scientific reports (2017) 71: 8409. WB, ICC, IHC; KO verified; tested species: mouse

LRBA, a BEACH protein mutated in human immune deficiency, is widely expressed in epithelia, exocrine and endocrine glands, and neurons.

Roussa E, Juda P, Laue M, Mai-Kolerus O, Meyerhof W, Sjöblom M, Nikolovska K, Seidler U, Kilimann MW Scientific reports (2024) 141: 10678. . IHC; KO verified; tested species: mouse

#### **Selected General References**

The BEACH is hot: a LYST of emerging roles for BEACH-domain containing proteins in human disease. Cullinane AR et al. Traffic (2013) PubMed:23521701

LRBA, a BEACH protein mutated in human immune deficiency, is widely expressed in epithelia, exocrine and endocrine glands, and neurons.

Roussa E et al. Sci Rep (2024) PubMed:38724551

Central nervous system manifestations of LRBA deficiency: case report of two siblings and literature review. Mangodt TC et al. BMC Pediatr (2023) PubMed:37443020

Murine LRBA deficiency causes CTLA-4 deficiency in Tregs without progression to immune dysregulation. Burnett DL et al. Immunol Cell Biol (2017) PubMed:28611475

The BEACH protein LRBA is required for hair bundle maintenance in cochlear hair cells and for hearing. Vogl C et al. EMBO Rep (2017) PubMed:28893864

Spectrum of Phenotypes Associated with Mutations in LRBA. Alkhairy OK et al. J Clin Immunol (2016) PubMed:26707784

AUTOIMMUNE DISEASE. Patients with LRBA deficiency show CTLA4 loss and immune dysregulation responsive to abatacept therapy.

Lo B et al. Science (2015) PubMed:26206937

Access the online factsheet including applicable protocols at <a href="https://sysy.com/product/524003">https://sysy.com/product/524003</a> or scan the QR-code.



# FAQ - How should I store my antibody?

# **Shipping Conditions**

 All our antibodies and control proteins / peptides are shipped lyophilized (vacuum freezedried) and are stable in this form without loss of quality at ambient temperatures for several weeks.

### Storage of Sealed Vials after Delivery

- Unlabeled and biotin-labeled antibodies and control proteins should be stored at 4°C before reconstitution. They must not be stored in the freezer when still lyophilized!
   Temperatures below zero may cause loss of performance.
- Fluorescence-labeled antibodies should be reconstituted immediately upon receipt. Long term storage (several months) may lead to aggregation.
- **Control peptides** should be kept at -20°C before reconstitution.

# Long Term Storage after Reconstitution (General Considerations)

- The storage freezer must not be of the frost-free variety ("no-frost freezer"). This cycle
  between freezing and thawing (to reduce frost-build-up), which is exactly what should be
  avoided. For the same reason, antibody vials should be placed in an area of the freezer that
  has minimal temperature fluctuations, for instance towards the back rather than on a door
  shelf.
- Aliquot the antibody and store frozen (-20°C to -80°C). Avoid very small aliquots (below 20 µl)
  and use the smallest storage vial or tube possible. The smaller the aliquot, the more the stock
  concentration is affected by evaporation and adsorption of the antibody to the surface of the
  storage vial or tube. Adsorption of the antibody to the surface leads to a substantial loss of
  activity.
- The addition of glycerol to a final concentration of 50% lowers the freezing point of your stock and keeps your antibody at -20°C in liquid state. This efficiently avoids freeze and thaw cycles.

# **Product Specific Hints for Storage**

#### Control proteins / peptides

• Store at -20°C to -80°C.

#### **Monoclonal Antibodies**

- Ascites and hybridoma supernatant should be stored at -20°C up to -80°C. Prolonged storage at 4°C is not recommended! Unlike serum, ascites may contain proteases that will degrade the antibodies.
- **Purified IgG** should be stored at -20°C up to -80°C. Adding a carrier protein like BSA will increase long term stability. Many of our antibodies already contain carrier proteins. Please refer to the data-sheet for detailed information.

#### **Polyclonal Antibodies**

- Crude antisera: With anti-microbials added, they may be stored at 4°C. However, frozen storage (-20°C up to -80°C) is preferable.
- Affinity purified antibodies: Less robust than antisera. Storage at -20°C up to -80°C is
  recommended. Adding a carrier protein like BSA will increase long term stability. Most of our
  antibodies already contain carrier proteins. Please refer to the data-sheet for detailed
  information.

#### Fluorescence-labeled Antibodies

• Store as a liquid with 1:1 (v/v) glycerol at -20°C. Protect these antibodies from light exposure.

# Avoid repeated freeze-thaw cycles for all antibodies!

# FAQ - How should I reconstitute my antibody?

#### Reconstitution

- All our purified antibodies are lyophilized from PBS. To reconstitute the antibody in PBS, add
  the amount of deionized water given in the respective datasheet. If higher volumes are
  preferred, add water as mentioned above and then the desired amount of PBS and a
  stabilizing carrier protein (e.g. BSA) to a final concentration of 2%. Some of our antibodies
  already contain albumin. Take this into account when adding more carrier protein.
   For complete reconstitution, carefully remove the lid. After adding water, briefly vortex the
  solution. You can spin down the liquid by placing the vial into a 50 ml centrifugation tube filled
  with paper.
- If desired, add small amounts of azide or thimerosal to prevent microbial growth. This is especially recommended if you want to keep an aliquot a 4°C.
- After reconstitution of fluorescence-labeled antibodies, add 1:1 (v/v) glycerol to a final
  concentration of 50%. This lowers the freezing point of your stock and keeps your antibody in
  liquid state at -20°C.
- Glycerol may also be added to unlabeled primary antibodies. It is a suitable way to avoid freezethaw cycles.
- Please refer to our tips and hints for subsequent storage of reconstituted antibodies and control peptides and proteins.