PRODUCT INFORMATION

Carbamylated Alpha-1 Antitrypsin

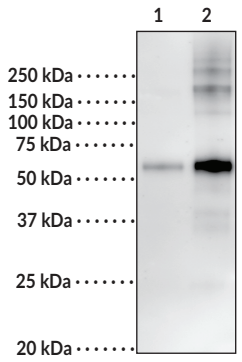
Item No. 21798

Overview and Properties

Synonyms: Carbamylated A1A, Carbamylated A1AT, Carbamylated AAT, Carbamylated α_1-Antitrypsin
Source: Human
Uniprot No.: P01009
Molecular Weight: 44.325 kDa
Storage: -80°C (as supplied)
Stability: ≥1 year
Purity: ***batch specific*** (≥90% estimated by SDS-PAGE)
Supplied in: Lyophilized from PBS, pH 7.4

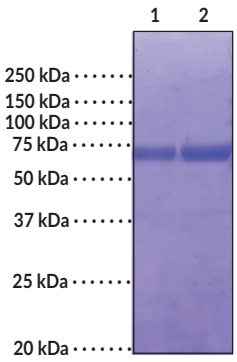
Information represents the product specifications. Batch specific analytical results are provided on each certificate of analysis.

Images



Lane 1: A1AT
Lane 2: Carbamylated A1AT

A1AT (Lane 1) and Carbamylated A1AT (Lane 2) were reacted with a biotin labeled probe reactive towards carbamylated lysines. After separation by SDS-PAGE, the proteins were blotted to nitrocellulose and detected using strepatavidin-HRP.



Lane 1: A1AT (2 µg)
Lane 2: A1AT (4 µg)

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Description

Alpha-1 antitrypsin is a serine protease inhibitor and member of the serpin superfamily.[1] It has a five-stranded A β-sheet and a mobile reactive center loop that acts as a pseudosubstrate for various proteases. Alpha-1 antitrypsin binds to a protease, undergoes proteolytic cleavage, and forms a covalent linkage between a carboxyl group in the reactive loop and the serine hydroxyl of the protease active site, effectively inactivating the enzyme which is then cleared from circulation. The primary targets of alpha-1 antitrypsin are neutrophil elastase and proteinase 3, however, it also inhibits trypsin, kallikreins 7 and 14, and matriptase.[2] Alpha-1 antitrypsin protects the lower respiratory tract from proteolytic destruction *via* inhibition of neutrophil elastase and reduced serum levels of alpha-1 antitrypsin have been linked to early-onset liver disease and emphysema.[3] Alpha-1 antitrypsin is an acute-phase protein that reduces production of inflammatory cytokines, inhibits apoptosis, blocks leukocyte degranulation and migration, as well as suppresses NF-κB nuclear translocation in monocytes. It delays disease onset in mouse models of inflammatory disease, including collagen-induced arthritis and experimental autoimmune encephalomyelitis (EAE). Alpha-1 antitrypsin is subject to post-translational modifications such as glycosylation and carbamylation *in vivo*. Carbamylated alpha-1 antitrypsin has been found in synovial fluid samples from rheumatoid arthritis patients and is predicted to act as an autoantigen.[4]

References

1. Elliott, P.R., Abrahams, J.P., and Lomas, D.A. Wild-type alpha 1-antitrypsin is in the canonical inhibitory conformation. *J. Mol. Biol.* **275(3)**, 419-425 (1998).
2. Janciauskiene, S.M., Bals, R., Koczulla, R., *et al*. The discovery of α1-antitrypsin and its role in health and disease. *Respir. Med.* **105(8)**, 1129-1139 (2011).
3. Ehlers, M.R. Immune-modulating effects of alpha-1 antitrypsin. *Biol. Chem.* **395(10)**, 1187-1193 (2014).
4. Verheul, M.K., Yee, A., Seaman, A., *et al*. Identification of carbamylated alpha 1 anti-trypsin (A1AT) as an antigenic target of anti-CarP antibodies in patients with rheumatoid arthritis. *J. Autoimmun.* **80**, 77-84 (2017).

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