# CERTIFICATE OF ANALYSIS – TECHNICAL DATA SHEET

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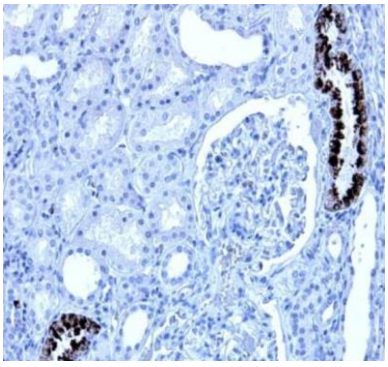
|---|---|---|---|
| **Product name** | Alpha-1-antitrypsin, Human, clone 2C1 | | |
| **Catalog number** | HM2289 | | |
| **Lot number** | - | **Expiry date** | - |
| **Volume** | 1 ml | **Amount** | 100 µg |
| **Formulation** | 0.2 µm filtered in PBS+0.1%BSA+0.02%NaN3 | **Concentration** | 100 µg/ml |
| **Host Species** | Mouse IgG1 | **Conjugate** | None |
| **Endotoxin** | N.A. | **Purification** | Protein G |
| **Storage** | 4°C | | |

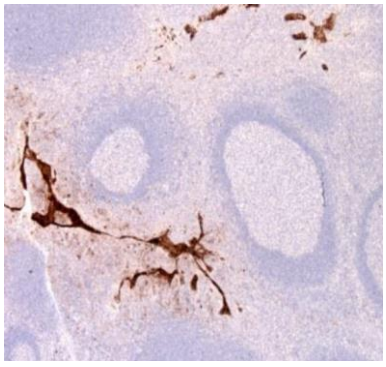
## Application notes

| | IHC-F | IHC-P | IF | FC | FS | IA | IP | W |
|---|---|---|---|---|---|---|---|---|
| Reference # | | 1,3,6 | 7,9 | | | 1-3,5,6,10 | 1,5 | 1,2,4-6, 8,10 |
| Yes | | ● | ● | | | ● | ● | ● |
| No | | | | | | | | |
| N.D. | ● | | | ● | ● | | | |

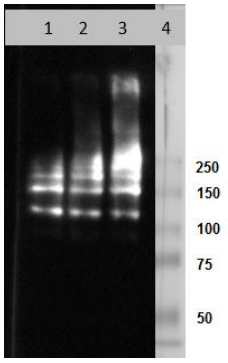
*N.D.= Not Determined; IHC = Immuno histochemistry; F = Frozen sections; P = Paraffin sections; IF = Immuno Fluorescence; FC = Flow Cytometry; FS = Functional Studies; IA = Immuno Assays; IP = Immuno Precipitation; W = Western blot*



IHC: sections of human kidney. HM2289 was used in a concentration of 2 µg/ml.



IHC: sections of human tonsil. HM2289 was used in a concentration of 2 µg/ml.




W: Western blot with Za1AT polymer lysate (20 µg) and HM2289 in 1, 2 and 4 µg/ml (respectively lane 1, 2 and 3).

Dilutions to be used depend on detection system applied. It is recommended that users test the reagent and determine their own optimal dilutions. The typical starting working dilution is 1:50.

- W: a non-denaturating gel was used. The bands obtained were ~110, 165, 220 and 275 kDa.
- IA: HM2289 can be used as capture and detection antibody.
- Positive control: Human tonsil and kidney; Negative control: For Western blot, any irrelevant protein can be used.

## General Information

**Description**

The monoclonal antibody clone 2C1 recognizes polymeric forms of human alpha-1-antitrypsin. Alpha-1-antitrypsin is the most abundant circulating protease inhibitor. Serpinopathies are conformational diseases characterized by the polymerization and intracellular retention of members of the serine protease inhibitor or serpin superfamily of proteins.1 The best known is a1-antitrypsin deficiency, with the most common severe deficiency allele being the Z mutation (Glu342Lys). The severe Z deficiency allele (Glu342Lys) causes the protein to undergo a conformational transition and form ordered polymers that are retained within hepatocytes. This causes neonatal hepatitis, cirrhosis, and hepatocellular carcinoma. Clone 2C1 recognizes polymers formed by Z α1-antitrypsin in vivo. It also recognizes polymers formed by the Siiyama (Ser53Phe) and Brescia (Gly225Arg) mutants, and the novel His334Asp shutter domain mutant of a1-antitrypsin that is associated with prolonged neonatal jaundice in a 6-week-old boy. These data show that Z and shutter domain mutants form polymers with a shared epitope.

**Immunogen**

Z a1-antitrypsin polymers prepared from a1-antitrypsin purified from the plasma of PI*Z homozygotes.

**Aliases** Alpha-1 protease inhibitor, Alpha-1-antiproteinase, Serpin A1

**Gene** Gene name: SERPINA1 Entrez Gene ID: 5265 Uniprot: P01009

**References**


1. Miranda, E et al; A novel monoclonal antibody to characterize pathogenic polymers in liver disease associated with alpha1-antitrypsin deficiency. Hepatology 2010, *52*: 1078.
2. Ekeowa, U et al; Defining the mechanism of polymerization in the serpinopathies. PNAS 2010, *107*: 17146.
3. Morris, H et al; ANCA-associated vasculitis is linked to carriage of the Z allele of α1 antitrypsin and its polymers. Ann Rheum Dis 2011, *70*: 1851.
4. Yamasaki, M et al; Molecular basis of α1-antitrypsin deficiency revealed by the structure of a domain-swapped trimer. EMBO reports 2011, *12*: 1011.
5. Ordonez, A et al; A single-chain variable fragment intrabody prevents intracellular polymerization of Z a1-antitrypsin while allowing its antiproteinase activity. FASEB Journal 2015, *29*: 2667
6. Tan, L et al; Characterising the association of latency with α1-antitrypsin polymerisation using a novel monoclonal antibody. Int J Bio & Cell Biol 2015, *58*: 81
7. Dickens, J et al; The endoplasmic reticulum remains functionally connected by vesicular transport after its fragmentation in cells expressing Z-a1-antitrypsin. FASEB Journal 2016, *30*: 4083
8. Haq, I et al; Deficiency Mutations of Alpha-1 Antitrypsin. Am J Respir Cell Mol Biol 2016, *1*: 71
9. Khodayari, N et al; Erdj3 Has an Essential Role for Z Variant Alpha-1-Antitrypsin Degradation. J Cell Bio 2017, *118*: 3090
10. Miranda, E et al; The pathological Trento variant of alpha-1-antitrypsin (E75V) shows nonclassical behaviour during polymerization. FEBS Journal 2017, *284*: 2110


**Storage&stability** Product should be stored at 4°C. Under recommended storage conditions, product is stable for at least one year.

**Precautions** For research use only. Not for use in or on humans or animals or for diagnostics. It is the responsibility of the user to comply with all local/state and federal rules in the use of this product. Hycult Biotech is not responsible for any patent infringements that might result from the use or derivation of this product.

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We hereby certify that the above-stated information is correct and that this product has been successfully tested by the Quality Control Department. This product was released for sale according to the existing specifications. This document has been produced electronically and is valid without a signature.

| Approved by Manager of QC | Date |
|---|---|
| Robbert Zwinkels | 09/04/2018 |

Do you have any questions or comments regarding this product? Please contact us via support@hycultbiotech.com.