

Product datasheet for AM31773PU-N

OriGene Technologies, Inc.

9620 Medical Center Drive, Ste 200 Rockville, MD 20850, US Phone: +1-888-267-4436 https://www.origene.com techsupport@origene.com EU: info-de@origene.com CN: techsupport@origene.cn

Stefin B (CSTB) Mouse Monoclonal Antibody [Clone ID: M1-C1]

Product data:

Product Type: Primary Antibodies

Clone Name: M1-C1

Applications: ELISA, IHC, WB

Recommended Dilution: ELISA.

Immunohistochemistry on Paraffin Sections: 5 µg/ml.

Western Blot.

Reactivity: Human
Host: Mouse
Isotype: IgG1

Clonality: Monoclonal

Immunogen: CSTB antibody was raised against cSTB (AAH03370.1, 1 a.a. ~ 98 a.a) full length recombinant

protein with GST tag. MW of the GST tag alone is 26 KDa.

Specificity: This antibody reacts to CSTB / Stefin B.

Formulation: PBS, pH 7.2

State: Purified

State: Liquid purified Ig fraction

Concentration: lot specific

Purification: Affinity chromatography on Protein A

Conjugation: Unconjugated

Storage: Store the antibody undiluted at 2-8°C for one month or (in aliquots) at -20°C for longer.

Avoid repeated freezing and thawing.

Stability: Shelf life: one year from despatch.

Gene Name: cystatin B

Database Link: Entrez Gene 1476 Human

P04080



Background:

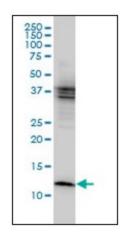
This is an intracellular thiol proteinase inhibitor. Tightly binding reversible inhibitor of cathepsins L, H and B.

Defects in CSTB are the cause of progressive myoclonic epilepsy type 1 (EPM1) [MIM:254800]. EPM1 is an autosomal recessive disorder characterized by severe, stimulus-sensitive myoclonus and tonic-clonic seizures. The onset, occurring between 6 and 13 years of age, is characterized by convulsions. Myoclonus begins 1 to 5 years later. The twitchings occur predominantly in the proximal muscles of the extremities and are bilaterally symmetrical, although asynchronous. At first small, they become late in the clinical course so violent that the victim is thrown to the floor. Mental deterioration and eventually dementia develop.

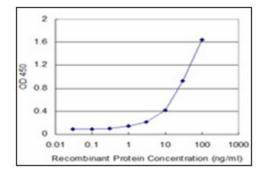
Synonyms:

Stefin-B, CPI-B, CSTB, CST6, STFB

Product images:

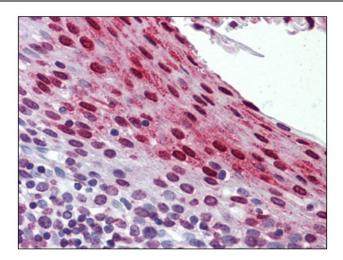


Western Blot analysis of CSTB expression in MCF-7 cell lysate.



Detection limit for recombinant GST tagged CSTB is approximately 1ng/ml as a capture antibody.





Human Tonsil: Formalin-Fixed, Paraffin-Embedded (FFPE)