Technical Data Sheet

Purified Mouse Anti-Ufd1L

Product Information

 $\begin{tabular}{llll} \textbf{Material Number:} & \textbf{611642} \\ \textbf{Size:} & 50~\mu g \\ \textbf{Concentration:} & 250~\mu g/ml \\ \textbf{Clone:} & 19/Ufd1L \\ \end{tabular}$

Immunogen: Mouse Ufd1L aa. 120-241

 Isotype:
 Mouse IgG1

 Reactivity:
 QC Testing: Mouse

Tested in Development: Human, Rat

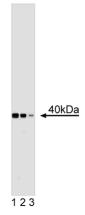
Target MW: 40 kDa

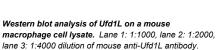
Storage Buffer: Aqueous buffered solution containing BSA, glycerol, and ≤0.09% sodium

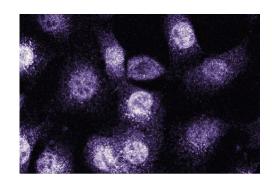
azide.

Description

Microdeletions of the chromosomal region 22q11.2 are associated with defects in cardiac and craniofacial development that are referred to as the CATCH 22 phenotype and include DiGeorge syndrome, the velo-cardio-facial syndrome, and the Opitz GBBB syndrome. Deleted in CATCH 22 patients, UFD1L encodes the human homolog of the yeast ubiquitin fusion degradation 1 protein (UFD1p), an essential component of the ubiquitin-dependent proteolytic pathway. The murine homolog (Ufd11) is expressed in the eyes and in the inner ear primordia during embryogenesis. In rat liver cytosol, Ufd11 binds to the nuclear transport protein Np14 and this binary complex competes with p47 for binding to p97, which inhibits Golgi membrane fusion. The tertiary complex Ufd11/Np14/p97 may also be involved in mitotic ubiquitin-dependent processes. Both Ufd11 and Np14 are found in the nucleus, while p97 is found in the cytoplasm and nucleus. In humans, Ufd1L is primarily expressed in adult heart, placenta, skeletal muscle, and pancreas, and fetal liver and kidney. Thus, Ufd1L is thought to be involved in cell functions that are critical for normal cell development in the cardiac and craniofacial regions.







Immunofluorescence staining of NIH/3T3 cells (Mouse embryo fibroblast cells; ATCC CRL-1658).

Preparation and Storage

The monoclonal antibody was purified from tissue culture supernatant or ascites by affinity chromatography. Store undiluted at -20°C.

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Application Notes

Application

Western blot	Routinely Tested
Immunofluorescence	Tested During Development

Recommended Assay Procedure:

Western blot: Please refer to http://www.bdbiosciences.com/pharmingen/protocols/Western_Blotting.shtml

Suggested Companion Products

Catalog Number	Name	Size	Clone	
611479	Mouse Macrophage Cell Lysate	500 μg	(none)	
554002	HRP Goat Anti-Mouse Ig	1.0 ml	(none)	
554001	FITC Goat Anti-Mouse Ig	0.5 mg	Polyclonal	

Product Notices

- 1. Since applications vary, each investigator should titrate the reagent to obtain optimal results.
- 2. Please refer to www.bdbiosciences.com/pharmingen/protocols for technical protocols.
- Caution: Sodium azide yields highly toxic hydrazoic acid under acidic conditions. Dilute azide compounds in running water before discarding to avoid accumulation of potentially explosive deposits in plumbing.
- 4. Source of all serum proteins is from USDA inspected abattoirs located in the United States.

References

Meyer HH, Shorter JG, Seemann J, Pappin D, Warren G. A complex of mammalian ufd1 and npl4 links the AAA-ATPase, p97, to ubiquitin and nuclear transport pathways. *EMBO J.* 2000; 19(10):2181-2192.(Biology)

Nagahama M, Suzuki M, Hamada Y, et al. SVIP is a novel VCP/p97-interacting protein whose expression causes cell vacuolation. *Mol Biol Cell.* 2003; 14(1):262-273.(Biology: Western blot)

Pizzuti A, Novelli G, Ratti A. UFD1L, a developmentally expressed ubiquitination gene, is deleted in CATCH 22 syndrome. *Hum Mol Genet.* 1997; 6(2):259-265. (Biology)

Yamagishi H, Garg V, Matsuoka R, Thomas T, Srivastava D. A molecular pathway revealing a genetic basis for human cardiac and craniofacial defects. *Science*. 1999; 283(5405):1158-1161.(Biology)

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