

Online Supplemental Figure S1 for

Hypomorphic pathogenic variant in *SFTPB* leads to adult pulmonary fibrosis

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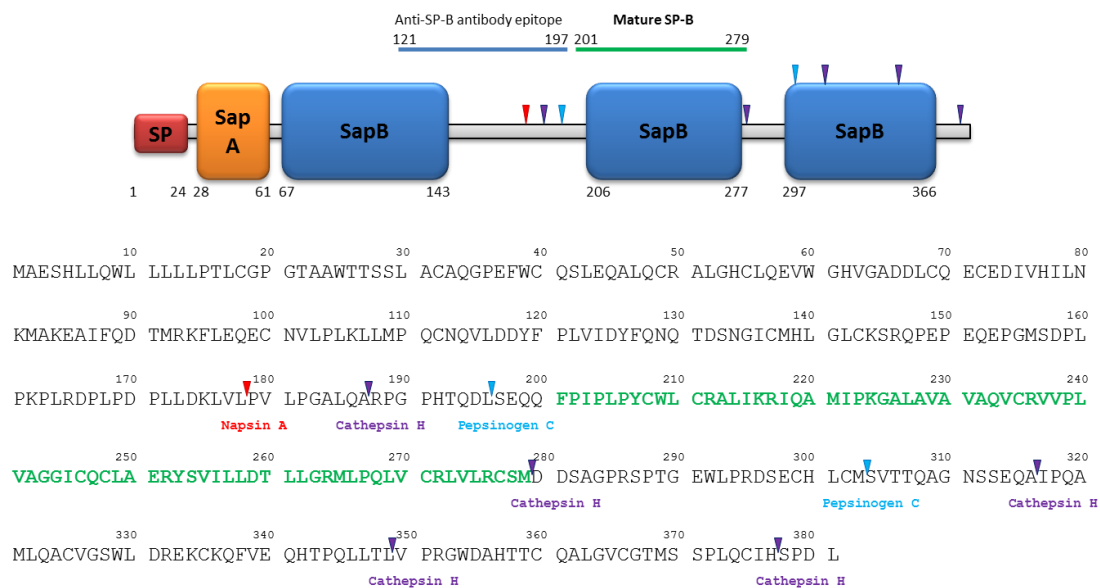


Figure S1: SP-B predicted domains and amino acids (aa) sequence

Domains were predicted by SMART from the SP-B protein sequence. Mature protein (amino acids 201-279) is represented in green (predictive domains and sequence). Epitope sequence for the anti-SP-B antibody (HPA062148) is represented by a blue line. Pro-SP-B cleavage sites by Napsin A,

Cathepsin H and Pepsinogen C ^(1,2) are represented by red, purple and blue triangles respectively. SapA and SapB: saposin A and B; SP: signal peptide.

⁽¹⁾ Ueno T, Linder S, Na CL, Rice WR, Johansson J, Weaver TE. Processing of pulmonary surfactant protein B by napsin and cathepsin H. *J Biol Chem.* 2004;279(16):16178-16184. doi:10.1074/jbc.M312029200

⁽²⁾ Brasch F, Ochs M, Kahne T, et al. Involvement of napsin A in the C- and N-terminal processing of surfactant protein B in type-II pneumocytes of the human lung. *J Biol Chem.* 2003;278(49):49006-49014. doi:10.1074/jbc.M306844200]]

Links

SMART: <http://smart.embl-heidelberg.de/>