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Anti-Human/Mouse BCS1L Monoclonal Antibody

单克隆抗体

产品货号: MA01874

第三版

免疫原:Fusion protein of BCS1L

亚型:IgG1

克隆:59

描述:Human BCS1-like (BCS1L),a mitochondrial inner-membrane protein,is a chaperone necessary for the assembly of mitochondrial respiratory chain complex III. Recently studies indicated that the mutations in this protein may cause the GRACILE (growth retardation,aminoaciduria,cholestasis,iron overload,lactacidosis,and early death) syndrome,a recessively inherited lethal disease characterized by fetal growth retardation,lactic acidosis,aminoaciduria,cholestasis,and abnormalities in iron metabolism.

配方:

浓度:None

保存: +4 ° C 保存 (1-2 周). 长期保存在-20 ° C或者-70 ° C. 避免反复冻融.

应用:ELISA, WB, ICC/IF