

本公司提供的电子版说明书仅供参考，实验请以收到的纸质手册为准。

Recombinant Human ALDH4A1 Protein

产品货号: TP06954

第三版

别名:ALDH4;P5CD;P5CDh

**描述:**ALDH4A1 is a member of the aldehyde dehydrogenase family. Aldehyde dehydrogenase enzymes function in the metabolism of many molecules including certain fats (cholesterol and other fatty acids) and protein building blocks (amino acids). Additional aldehyde dehydrogenase enzymes detoxify external substances, such as alcohol and pollutants, and internal substances, such as toxins that are formed within cells. ALDH4A1 is expressed abundantly in liver followed by skeletal muscle, kidney, heart, brain, placenta, lung and pancreas. It is a mitochondrial matrix NAD-dependent dehydrogenase which catalyzes the second step of the proline degradation pathway, converting pyrroline-5-carboxylate to glutamate. Defects in ALDH4A1 are the cause of hyperprolinemia type 2 (HP-2). HP-2 is characterized by the accumulation of delta-1-pyrroline-5-carboxylate (P5C) and proline. The disorder may be causally related to neurologic manifestations, including seizures and mental retardation.

**配方:**PBS

**分子量:**59.2 kDa

**序列:**Lys 25-Gln 563

**纯度:**> 95% by HPLC

**浓度:**

**内毒素:**<1.0 EU per 1 ug of protein (determined by LAL method)

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