

## PHENYLALANINE AMMONIUM LYASE (PAL) FOR PHENYLKETONURIA (PKU)

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Phenylketonuria is an inherent disorder of metabolism, which is a result of a deficiency or defect in the phenylalanine hydroxylating system that metabolizes the essential amino acids phenylalanine to tyrosine affecting the biosynthesis of the neurotransmitters dopamine, noradrenaline, and adrenaline. If phenylalanine accumulates high concentrations in blood, mental retardation, microcephaly, spasticity and tremors occur. PKU patients are treated immediately after birth through a low-Phe diet. PAL is a nonmammalian enzyme and very stable under a wide temperature range. PAL requires no cofactors for degrading phenylalanine and products the harmless metabolite trans-cinnamic acid converted in the liver to benzoic acid, which is then excreted via the urine mainly as hippurate. PAL is widely distributed in plants and some fungi and yeasts and produced from *E.coli*. In fungal cells, PAL has a catabolic role while in plants, it is a key biosynthetic enzyme in stress responses such as including pathogenic attack, tissue wounding and UV irradiation and low temperature. PAL is purified from *Rhodotorula glutinis* yeast. Red spring wheat, winter wheat, fall rye, green lentil, romano bean, soybean, black turtle bean, pinto bean etc. have high PAL activity. The pH optima of PAL are close to the average pH of the small intestine, however enzyme must be protected. To preserve of PAL, PAL (from *R. glutinis*) with artificial cells is immobilized, coated gelatin capsules, entrapped with silk fibroin and adenocarcinoma cell line Caco-2. PAL enzyme production is also become a viable option with recombinant technology.

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