

# VITAMIN B<sub>6</sub> METABOLISM IN HEALTH AND DISEASE

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The biologically active form of vitamin B<sub>6</sub>, pyridoxal 5'-phosphate (PLP), is a cofactor in over 200 enzyme activities involved in many metabolic pathways, including neurotransmitter synthesis and degradation. In humans, PLP is recycled from food and degraded PLP-dependent enzymes in a salvage pathway requiring the action of pyridoxal kinase, pyridoxine 5'-phosphate oxidase and phosphatases. Once PLP is made, it is targeted to the dozens different apoenzymes that need it as cofactor. The regulation of the salvage pathway and the mechanism of targeting of PLP to the apoenzymes are poorly understood. Severe neurological disorders, such as convulsions, epileptic encephalopathy and axonal polyneuropathy, result from inborn errors in proteins involved in vitamin B<sub>6</sub> metabolism. This seminar will present the latest achievements in this field, focusing on the molecular basis of the above-mentioned neurological diseases and on the mechanisms of PLP homeostasis.