

Two types of hyperprolinemia are recognized by physicians and clinical researchers. Each represents an inherited inborn error of metabolism involving the amino acid, proline. Hyperprolinemia Type II is characterized by an abnormally high level of the amino acid proline in the blood. Fevers associated with seizures are common and mild mental retardation may be present. Hyperprolinemia Type II is a very rare disorder that is present at birth. It affects males and females in equal numbers. HP-II is recognized by elevated blood proline and elevated P-5-C levels in the urine. (Normal blood proline levels are about 450 units whereas elevated blood proline levels in subjects with HP-II reach 1900-2000 units.)