

Two types of hyperprolinemia are recognized by physicians and clinical researchers. Each represents an inherited inborn error of metabolism involving the amino acid, proline. Proline is abundant in nature and readily found in a variety of foods. Hyperprolinemia Type I is the result of proline oxidase deficiency and is characterized by an abnormally high proline blood level. Levels of the amino acids hydroxyproline and glycine in the blood are also higher than normal. Some clinicians believe that kidney abnormalities may be associated with HP-I, although this is disputed. The signs and symptoms of HP-I, other than the high levels of amino acids in the blood and urine, are vague. Often, this disorder is described as benign. Hyperprolinemia Type I is a very rare disorder that is present at birth. It affects males and females in equal numbers. HP-I is recognized by elevated blood proline levels. (The normal level is approximately 450 units, but people with HP-1 may have levels of 1900 to 2000 units.. Often, the diagnosis is made by exclusion. After failure to arrive at a diagnosis by other means, a blood proline level is ordered. The result confirms the diagnosis.