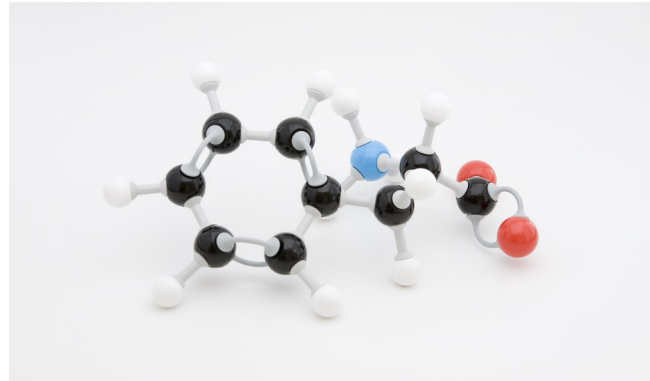


Phenylketonuria (PKU) is an autosomal recessive disorder which results in a defect in the enzyme phenylalanine hydroxylase which is responsible for the conversion of the amino acid phenylalanine to tyrosine. If not detected and treated in infancy, it can result in severe psychomotor retardation, seizures, heightened deep tendon reflexes, and abnormal gait and posture.

PKU is controlled by limiting the intake of phenylalanine from normal food sources such as milk, which, if controlled, can often prevent the development of any of the aforementioned symptoms. Phenylalanine is also present in non-food sources such as aspartame. Any foods containing phenylalanine must be marked as such so they can be more easily avoided by individuals with the disorder. The dietary restriction should be continued at least until adolescence for males. A longer regimen of dietary restriction is recommended for females.

Phenylalanine resides in the liver, and there are instances in which a liver transplant in a young patient treated for liver failure also resulted in a cure for his phenylketonuria. Some MRI studies have demonstrated abnormal neural myelination in patients, even those who exhibited no symptoms of the disease.



*Phenylalanine molecule*

The anesthetic plan for the patient with PKU should be aimed at avoiding any drugs that may promote seizure activity. Large doses of local anesthetics can lower the seizure threshold in these patients. In some cases, very high doses of opioids have been implicated in seizures in patients with epilepsy. Methohexital and meperidine may also increase the likelihood of epileptiform activity in these patients and may need to be avoided. Because patients with PKU are also more susceptible to vitamin B12 deficiency due to strict dietary restriction. As a result, it may be prudent to avoid the use of nitrous oxide. Some studies indicate an increased sensitivity to narcotics in patients with PKU, although the efficacy of some drugs such as nondepolarizing muscle relaxants may be reduced in patients who are already on anti-seizure medications.

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