GCV3 encodes the H-protein of the glycine cleavage system or glycine decarboxylase complex. GDC is a multienzyme complex that catalyzes the reversible oxidative cleavage of glycine into CO2 and NH3 and connects the metabolism of one, two and three-carbon compounds. The other subunits of the GDC complex are Gcv1p, a tetrahydrofolate transferase; Gcv2p, a glycine dehydrogenase; and Lpd1p, a lipoamide dehydrogenase. Null mutations in GCV3 prevent utilization of glycine as a nitrogen source.Gcv3p also has a role in protein lipoylation. The gcv3 null mutant displays a complete absence of protein lipoylation. Gcv3p is itself lipoylated, and this modification is required for the lipoylation of other proteins: blocking Gcv3p lipoylation by mutation of the target residue also prevents all other proteins from being lipoylated. However, this role is unrelated to the role of Gcv3p in glycine cleavage, since mutation of the gcv1 or gcv2 genes encoding other subunits of that complex has no effect on lipoylation. Protein lipoylation also requires Lip2p, Lip5p, and Aim22p, which may function together in a complex with Gcv3p.GCV3 is up-regulated by glycine and repressed by the metabolic products that require one-carbon units for their synthesis. In addition, regulation of GCV3 by the general amino acid control system is mediated by the promoter elements GCRE1, GCRE2, GCRE3, and a TATA box.H-proteins are highly conserved from bacteria to mammals. Mutations in any of the glycine cleavage system genes can result in Glycine encephalopathy or Nonketotic hyperglycinemia.