BPL1 encodes biotin-apoprotein ligase, an enzyme that modifies proteins by covalently attaching biotin. Bpl1p is essential; among its substrates is Acc1p, acetyl-CoA carboxylase, which acts in fatty acid biosynthesis. Bpl1p has strong sequence similarities to biotin--apoprotein ligases from E. coli and human; BPL1 can complement a deletion of E. coli birA. Mutations in the human biotin--apoprotein ligase are associated with ataxia and carboxylase deficiency.