The COX10 gene encodes the mitochondrial enzyme hemeA:farnesyltransferase, also known as heme O synthase or protoheme IX farnesyltransferase. It catalyzes the transfer of a farnesyl group to protoheme IX, forming heme O. Heme O is then converted to heme A, probably by the action of a monooxygenase composed of Cox15p, Yah1p, and Arh1p. Two heme A molecules generated by this pathway are incorporated into the Cox1p subunit of cytochrome c oxidase, which is the terminal member of the mitochondrial electron transport chain involved in cellular respiration.Because the heme A cofactor is essential to cytochrome c oxidase and Cox10p is required for its synthesis, cox10 mutations lead to a deficiency in respiratory growth. Cytochrome c oxidase subunits are present in the cox10 mutant but are not assembled into a functional enzyme.Homologs of Cox10p are found in various organisms from prokaryotes to humans. The human homolog functionally complements the yeast cox10 mutation. Mutations in the human homolog lead to cytochrome c oxidase deficiency, which may manifest itself as tubulopathy, leukodystrophy, Leigh syndrome, or infantile hypertrophic cardiomyopathy.