COX20 encodes a specific chaperone that facilitates proteolytic processing of the mitochondrial gene product Cox2p and its assembly into the mitochondrial inner membrane enzyme cytochrome c oxidase. Cox20p is required for N-terminal processing of Cox2p by the mitochondrial inner membrane peptidase, but is not required for processing of other inner membrane peptidase substratesPhysical interaction between Cox20p, which is an integral protein of the mitochondrial inner membrane, and Cox2p can be detected both before and after Cox2p N-terminal processing. The cox20 null mutant contains unprocessed Cox2p that is not assembled into cytochrome c oxidase, causing a respiratory growth defect due to lack of cytochrome c oxidase activity.