Cytochrome C Oxidase deficiency is a very rare inherited metabolic disorder characterized by deficiency of the enzyme cytochrome C oxidase (COX), or Complex IV, an essential enzyme that is active in the subcellular structures that help to regulate energy production (mitochondria). Deficiency of COX may be limited (localized) to the tissues of the skeletal muscles or may affect several tissues, such as the heart, kidney, liver, brain, and/or connective tissue (fibroblasts); in other cases, the COX deficiency may be generalized (systemic). Four distinct forms of Cytochrome C Oxidase deficiency have been identifed. The first form of this disorder is known as COX deficiency, benign infantile mitochondrial myopathy. Affected infants exhibit many of the same symptoms as those with the more severe infantile form of the disease; however, because the COX deficiency is limited (localized) to tissues of the skeletal muscles, they typically do not have heart or kidney dysfunction.