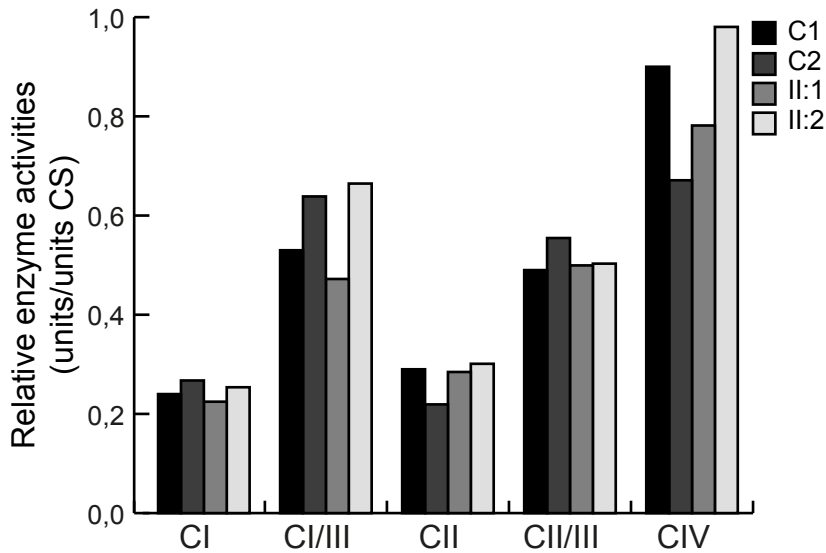


A)



**Figure S2.** Patient fibroblast fail to show mitochondrial enzymes defects when grown in galactose. (A) Relative enzyme activities of respiratory chain enzyme complex I (NADH coenzyme Q reductase), complex I/III (NADH cytochrome c reductase), complex II (succinate dehydrogenase), complexes II /III (succinate:cytochrome c reductase, SCR) and complex IV (cytochrome c oxidase) in isolated mitochondria of fibroblasts from subjects (II:1 and II:2) and non-related controls (C1 and C2) grown in galactose. Data are represented as mean  $\pm$  standard deviation (SD), n=1.

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