

Additional file 3: Additional Structural Information

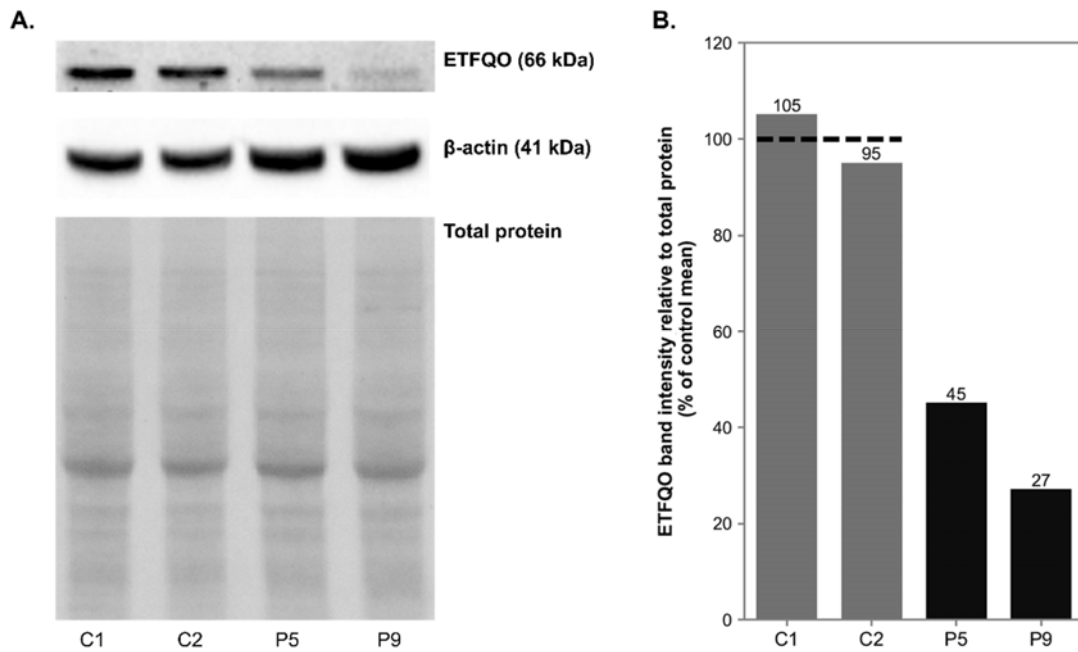


Figure S1.a–b Structural instability of ETFQO in skin fibroblasts of P5 and P9

(a) Whole cell lysates, prepared from primary skin fibroblasts of P5 (c.[1448C>T];c.[1448C>T]) and P9 (c.[1067G>A];c.[1448C>T]) as well as two healthy controls, C1 and C2 (matched in age, gender, and ethnicity to P9 and P5, respectively), were separated on a 10% SDS-PAGE gel and immunoblotted with primary antibodies against ETFQO (reported weight of the monomeric protein: 64 kDa) and β -actin (42 kDa). (b) Band volume intensities were normalised to the total protein content of each corresponding lane and quantified as a percentage, relative to the mean (100%) of both controls. Abbreviations: ETFQO: electron transfer flavoprotein-ubiquinone oxidoreductase.