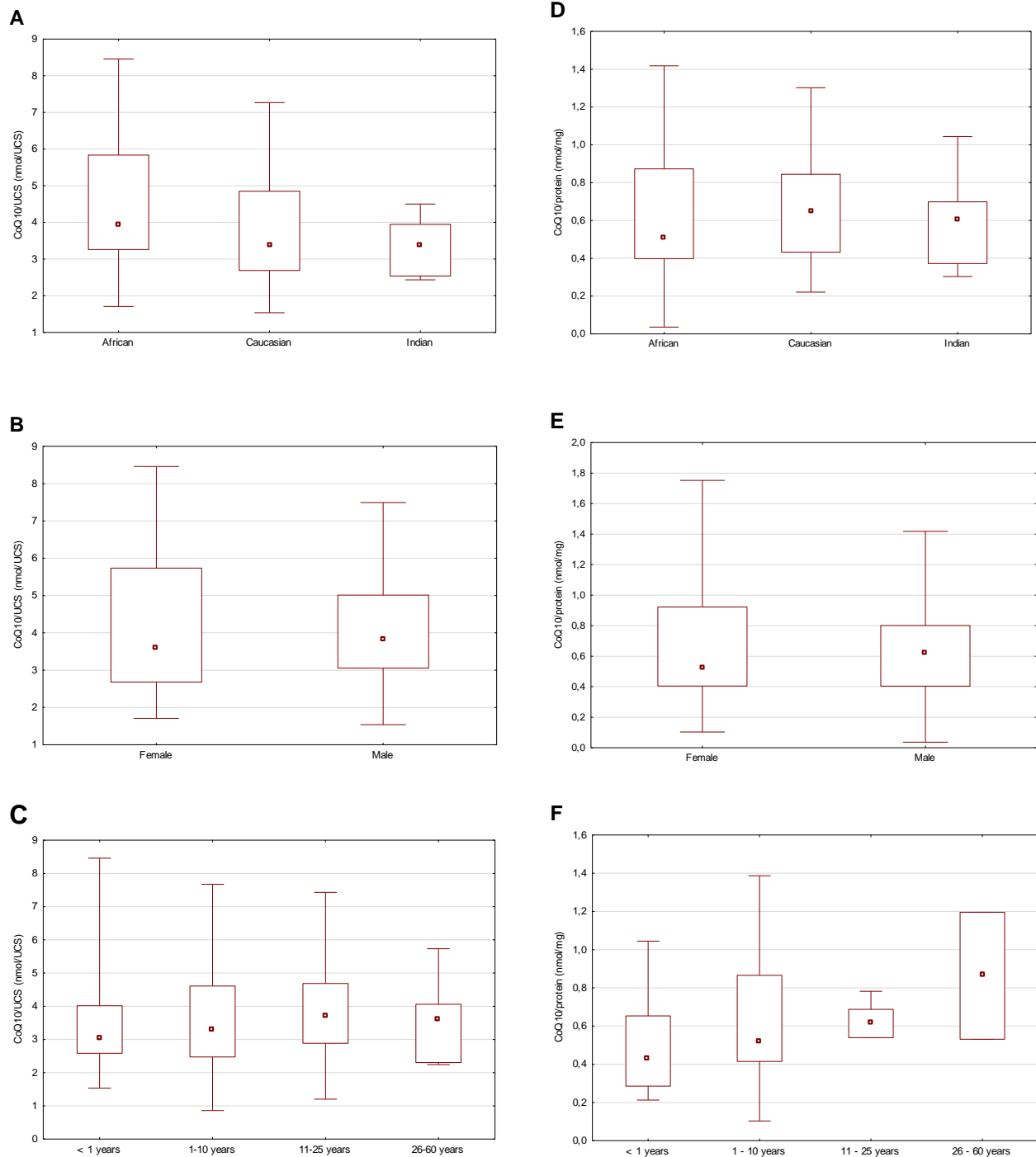# The dilemma of diagnosing coenzyme Q_{10} deficiency in muscle

Online supplementary document

Figure S1. The effect of ethnicity (A, D), gender (B, E) and age (C, F) on muscle CoQ$_{10}$ when expressed against CS activity (A, B, C) and protein content (D, E, F) in the clinically referred controls (CRC). CRC samples (n = 80) were divided into ethnic groups [Black African (n = 38), Caucasian (n = 35) and Indian (n = 7)], gender groups (39 female and 41 male) and the following age groups: < 1 years (n = 16), 1 - 10 years (n = 57), 11 - 25 years (n = 5) and 26 - 60 years (n = 2). Box plots show the median, 25th and 75th percentiles as well as the range. Muscle CoQ$_{10}$/UCS were not significantly influenced by ethnicity, gender or age. When expressing the CoQ$_{10}$ against protein content (nmol/mg protein), no significant effects were detected either.
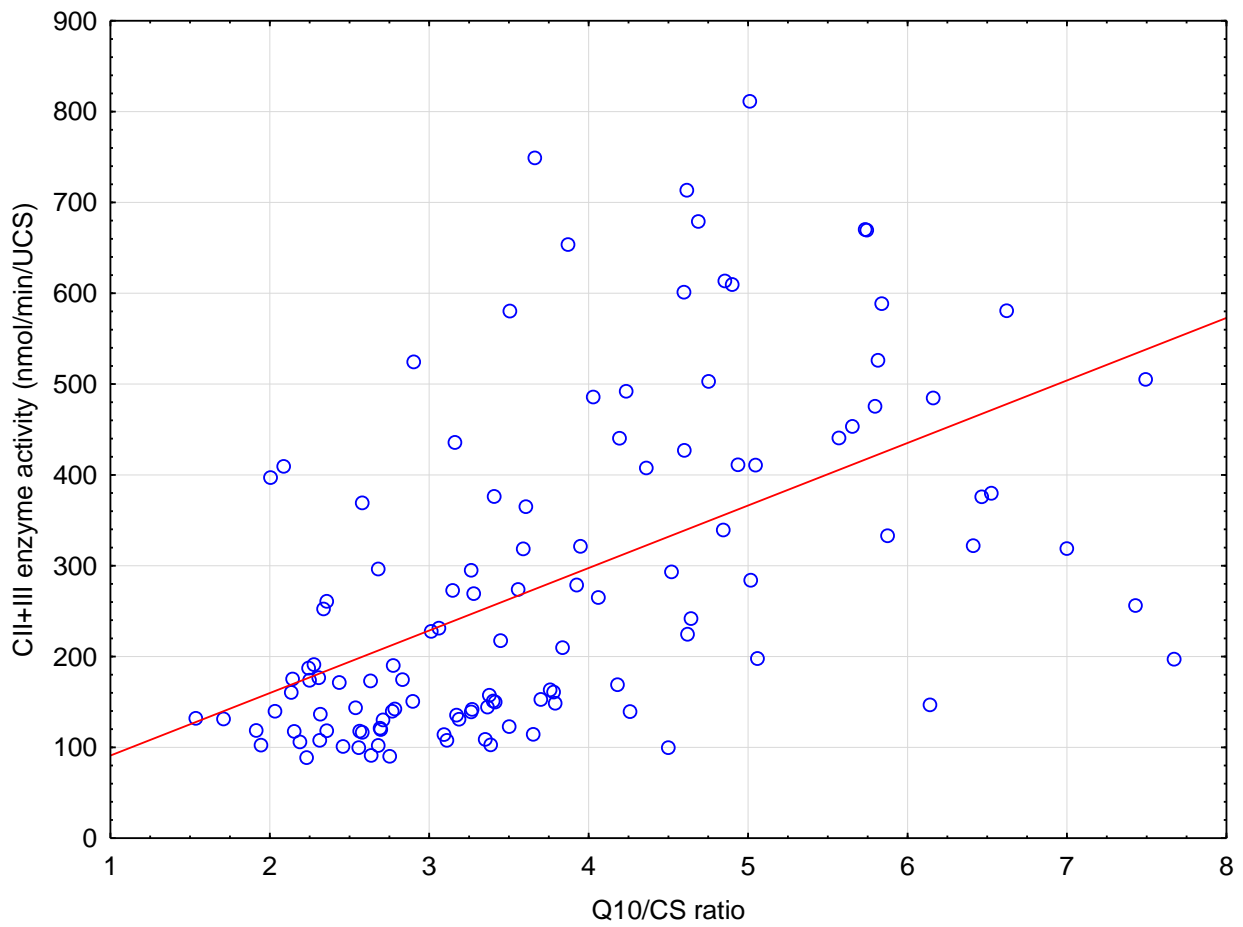
Figure S2. Correlation of muscle CoQ$_{10}$ concentrations with complex II+III RC enzyme activity. Data from the CRC and Other RCD groups was used after removal of outliers using Tukey's method, resulting in a total of 119 samples. Muscle CoQ$_{10}$ showed a strong correlation with CII+III enzyme activity (Spearman's correlation = 0.62). The correlation was also significant (P < 0.001).
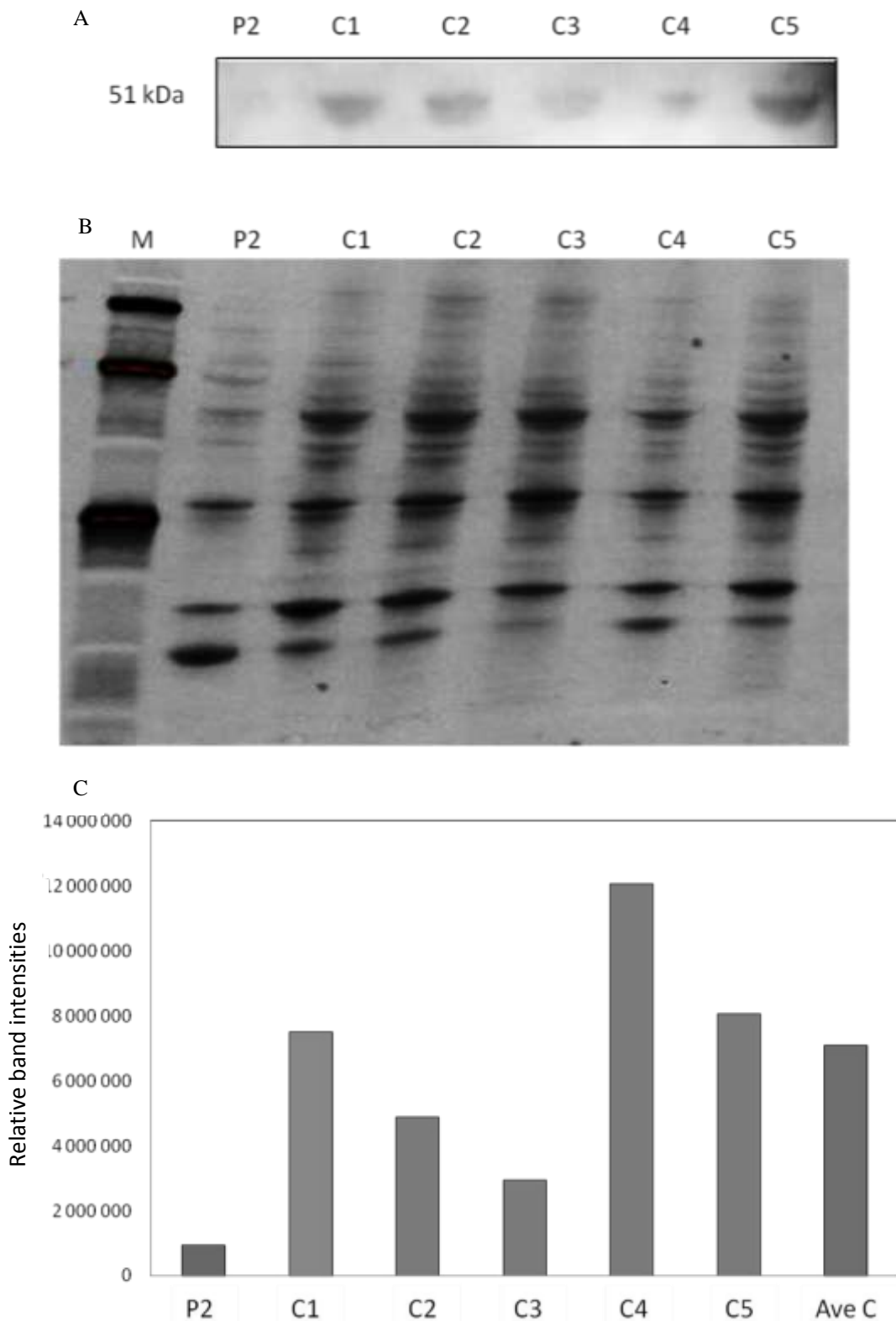
Figure S3. Immunoblotting analysis of COQ6 expression in P2 and patient controls. 600 x g supernatant protein (25 µg) from muscle tissue, separated using SDS-PAGE and transferred using western blotting, muscle tissue was subjected to immunoblot analysis using monoclonal rabbit anti-COQ6 antibodies and goat-anti-rabbit horse radish conjugated polyclonal antibodies. (A) The COQ6 protein was specifically detected as a band at ~51 kDa of the molecular weight marker. (B) Stain free blot used for total protein normalization of P2 and patient controls. C) The relative band intensities of which the expression level of P2 was notably lower from that of the average- and relative band intensities of the individual controls. P: patient; C: control; kDa: kilodalton; M: protein molecular weight marker; Ave: average