

Supplementary Fig. 1. Discordance of successive repeat PDC testing in fibroblasts (A and B) and skeletal muscle (C and D). Plot of minimum vs maximum fibroblast PDC activity noted as % control mean (%mean) for the *PDHA1*-Only (A and C) and All-CIDEM (B and D) groups. "Low" PDC cutoff values (<52% and <38% for fibroblast and skeletal muscle assays, respectively and correspond to <3rd %ile of control range) noted as green lines along with the area of discordance (green box). See Materials and Methods Section 2.2 for control mean and range. The red and blue asterisks (C and D) represent values off the plot scale for the respective subjects; one female subject was considered discordant but was off scale. Y-axis (Max %mean) and X-axis (Min %mean) represent maximum and minimum PDC activities for a subject as % control mean, respectively.