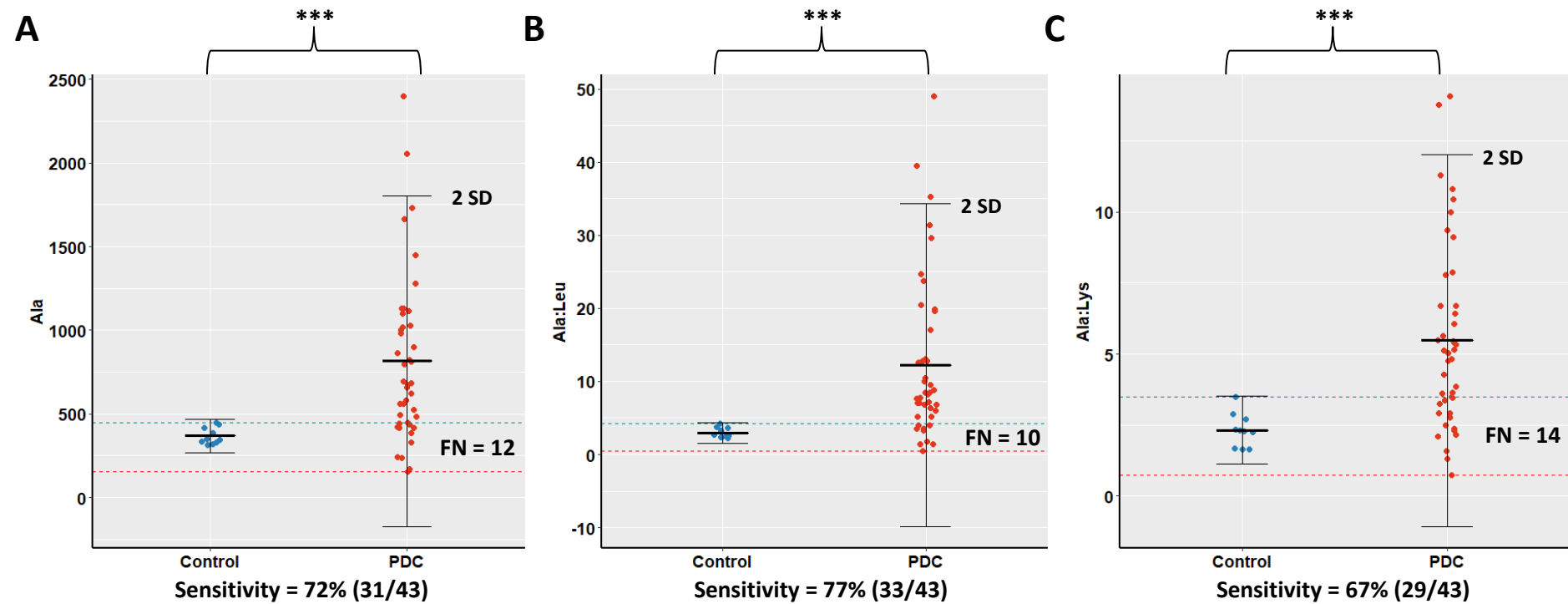


# Suppl. Fig. 1



**Supplementary Fig. 1.** Distribution of plasma Ala, Ala:Leu, and Ala:Lys from PDC deficient subjects due to *PDHA1* (20) or *PDHB* (2) mutations and normal controls. A) Ala ( $\mu\text{M}$ ), B) Ala:Leu, and C) Ala:Lys from PDC deficient subjects (red dots) and controls (blue dots). Mean and 2 standard deviation (SD) bars shown. Number of false negative (FN) between the maximum value for controls (blue dotted line) and minimum value for PDC deficient subjects (red dotted line) shown. Percentage difference of mean ( $\% \Delta$ ), for Ala, Ala:Leu and Ala: Lys metrics were 75%, 122% and 81% respectively. P values were obtained from two-sample t-test for un-pooled variances with one-tailed significance level of  $p < 0.05$ .  $***$ ,  $p < 0.0005$ .