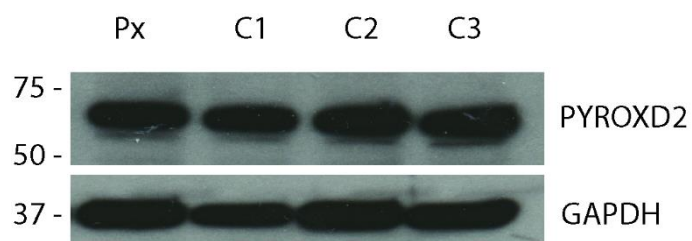


Attachment: Full western blots from data in article

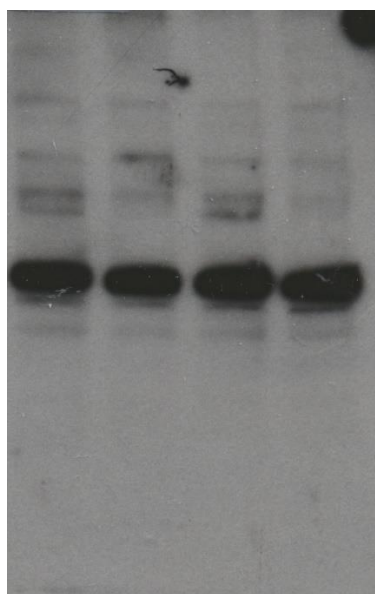
Article: *Biallelic variants in PYROXD2 cause a severe infantile metabolic disorder affecting mitochondrial function*



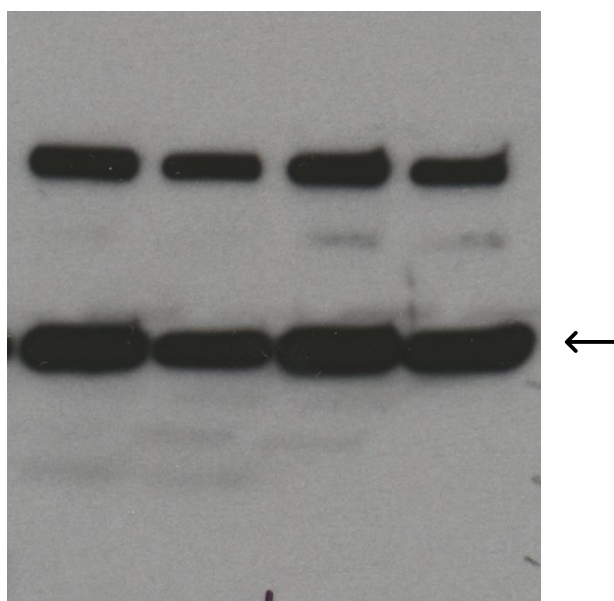
**Supplemental Figure 3. Western blot of PYROXD2 in patient fibroblasts.** Western blot of PYROXD2 in patient fibroblasts (Px) and three controls (C1 - C3) showing normal levels of PYROXD2 protein.

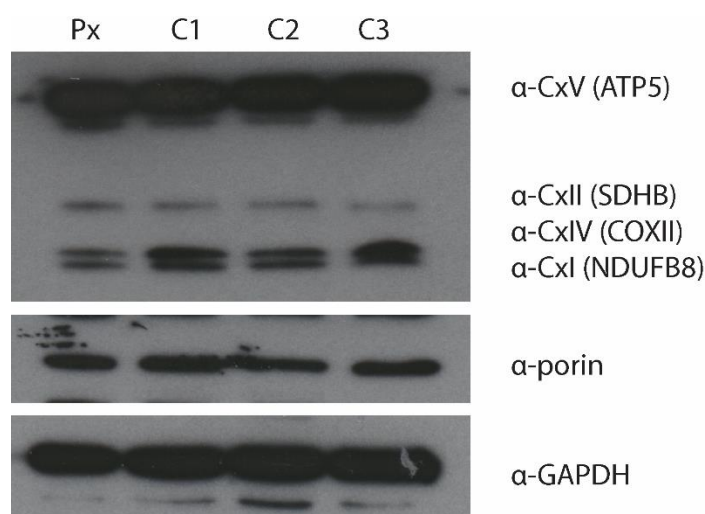
Full Western blots below

Anti-PYROXD2



Anti-GAPDH (indicated with arrow)





**Supplemental Figure 4: Western blot of OXPHOS complex subunits in patient fibroblasts.** Western blot of a representative subunit of each OXPHOS complex including; cytochrome c oxidase subunit 2 [COX2], cytochrome b-c1 complex subunit 2 [UQCRC2], succinate dehydrogenase [ubiquinone] flavoprotein subunit B [SDHB], NADH dehydrogenase [ubiquinone] 1 beta subcomplex subunit 8 [NDUFB8] and ATP synthase subunit alpha [ATP5A] in patient fibroblasts (Px) and three controls (C1 - C3) showing normal levels of OXPHOS subunit proteins.

#### Full Western blots

Anti-OXPHOS (CxV, CxII, CxIV and CxI)

Anti-Porin (after anti-OXPHOS probing). Porin indicated with an arrow

Anti-GAPDH

