A) ATP-production in mitochondria isolated from muscle using glutamate + succinate (Glu+Succ), N, N, N', N'-tetramethyl-p-fenylendiamin + ascorbate (TMPD+Asc), pyruvate + malate (Pyr+Mal), palmitoyl-L-carnitine + malate (PalCar+Mal) and succinate + rotenone (Succ+Rot) as substrates. B) Respiratory chain enzyme activities in mitochondria isolated from muscle. C) Respiratory chain enzyme activities in mitochondria isolated from fibroblasts, n=3. Δ) Activities outside the control range ±2SD are considered pathological (Fig A-B). *) p<0.01 using Student’s t-test (Fig C).