

SUPPLEMENTARY MATERIAL

Fig. S1. Absence of association of plasma oxysterols with plasma cholesterol or anti-oxidant capacity.

Fig. S2. Free cholesterol levels and LDL-stimulated cholesterol esterification in skin fibroblasts from NPC1 subjects.

Table S1. Plasma oxysterols in NPC1 subjects.

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Table S1**Plasma oxysterols in NPC1 subjects**

Oxysterol	ng/ml \pm SEM
7α-HC	63.4 \pm 11.5
7β-HC	214.6 \pm 31.8
4β-HC	125.5 \pm 46.2
3β,5α,6β-triol	358.0 \pm 164.0
27-HC	77.7 \pm 34.2
24(S)-HC	63.1 \pm 23.0
20-S	31.4 \pm 7.4
25-HC	7.3 \pm 2.0
7α,27-HC	6.8 \pm 1.3
7β,27-HC	3.9 \pm 0.9
6-KC	22.2 \pm 7.3
7-KC	804.1 \pm 148.5

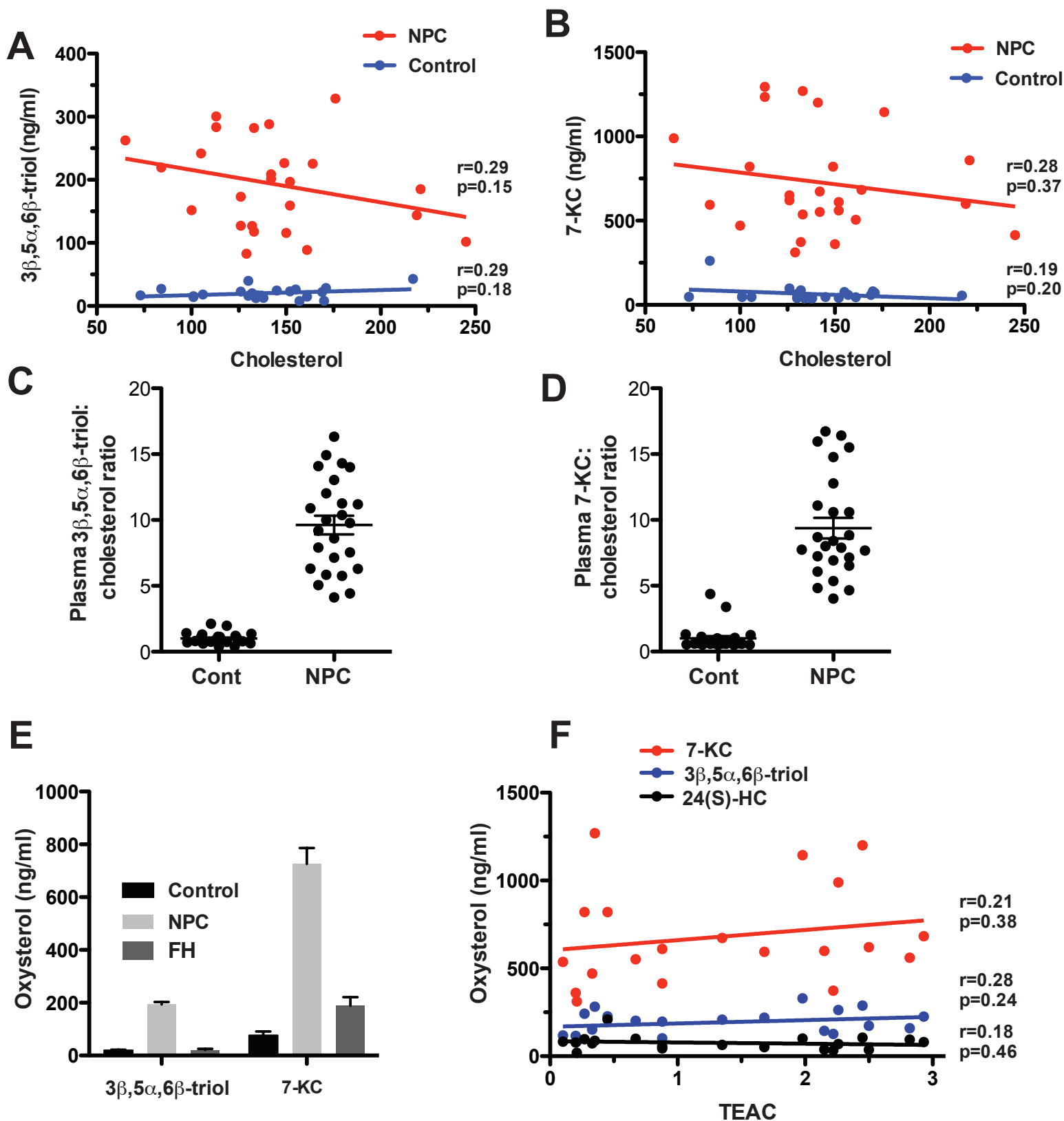


Fig. S1. Absence of association of plasma oxysterols with plasma cholesterol or anti-oxidant capacity. (A) Plasma 3 β ,5 α ,6 β -triol and (B) 7-KC levels in control and NPC1 subjects correlated with plasma total cholesterol levels. (C) Plasma 3 β ,5 α ,6 β -triol and (D) 7-KC levels in control and NPC1 subjects normalized to plasma total cholesterol levels. (E) Comparison of plasma 3 β ,5 α ,6 β -triol and 7-KC levels in control, NPC1 and Familial Hypercholesterolemia subjects. (F) Correlation of plasma 3 β ,5 α ,6 β -triol, 7-KC and 24(S)-HC levels with Trolox equivalent antioxidant capacity (TEAC) of plasma in NPC1 subjects.

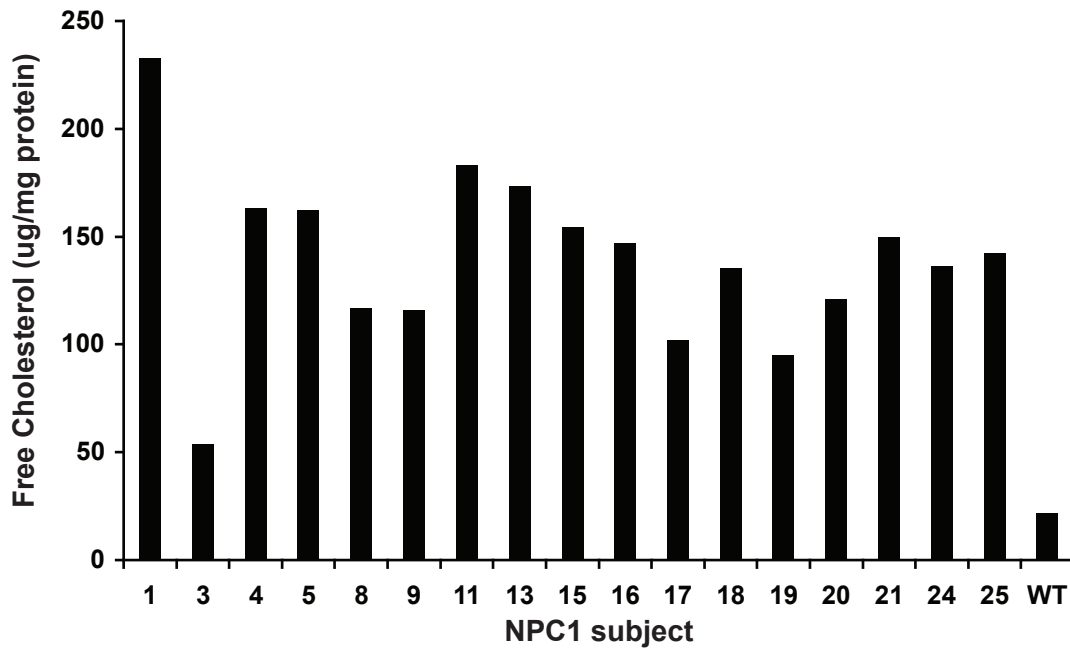
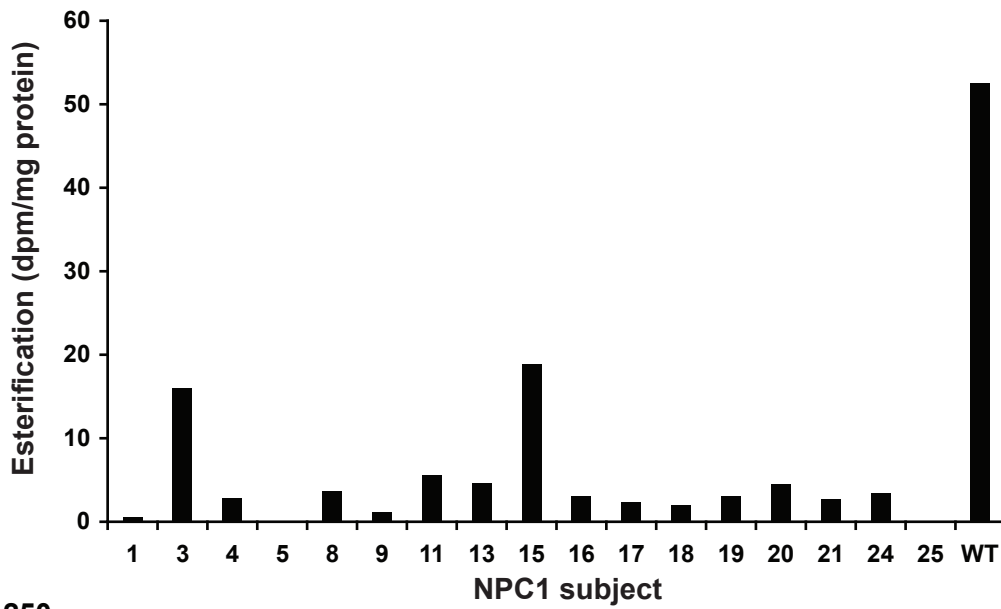
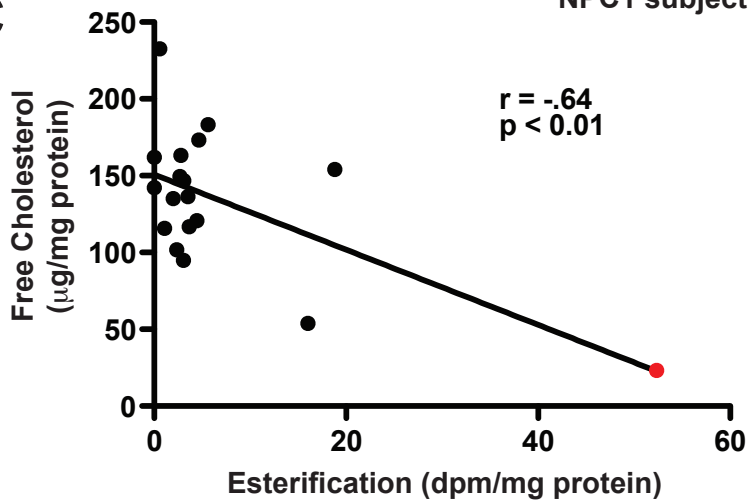
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Fig. S2. Free cholesterol levels and LDL-stimulated cholesterol esterification in skin fibroblasts from NPC1 subjects. (A) Free cholesterol levels in skin fibroblasts cultured for 48h under lipoprotein-deficient conditions. (B) Rates of LDL-stimulated cholesterol esterification in skin fibroblasts from NPC1 subjects. (C) Correlation of rates of LDL-stimulated cholesterol esterification with free cholesterol accumulation in fibroblasts obtained from NPC1 subjects. Red symbol indicates WT fibroblasts. $p < 0.01$.