Fig S1. Variants in VLCAD-deficient patients and their conservative analysis. 
(a) Genotypes of VLCAD deficiency pedigrees; (b) Variants distribution in ACADVL; (c) Conservative analysis of six novel missense variants in species (Homo sapiens, NP_000009.1; Pan troglodytes, XP_016786900.1; Bos taurus, NP_776919.1; Ratus norvegicus, NP_037023.1; Mus musculus, NP_059062.1; Danio rerio, NP_997776.1)
Fig S2. Expression levels of HA-tagged VLCAD in cells. (a) transient transfected cells and (b) the stable cells. β-Actin is used as a loading control on the same blot.
Fig S3. FAO capacity for utilization of exogenous palmitate. Oxygen-consumption rate (OCR) was monitored by sequential injections of oligomycin, FCCP, antimycin A and rotenone (downward arrows) in the presence of bovine serum albumin alone (BSA) or conjugated to palmitate (Palm-BSA).
Fig. S4 Structural predictions in wild-type and mutant residues. The structure of the mutant was shown in sky blue with the WT in tan or green. The black and red dashed represent the electrostatic interactions of mutant and WT, respectively. The occupancy of the hydrogen bonds was shown in red.