Fig. 1

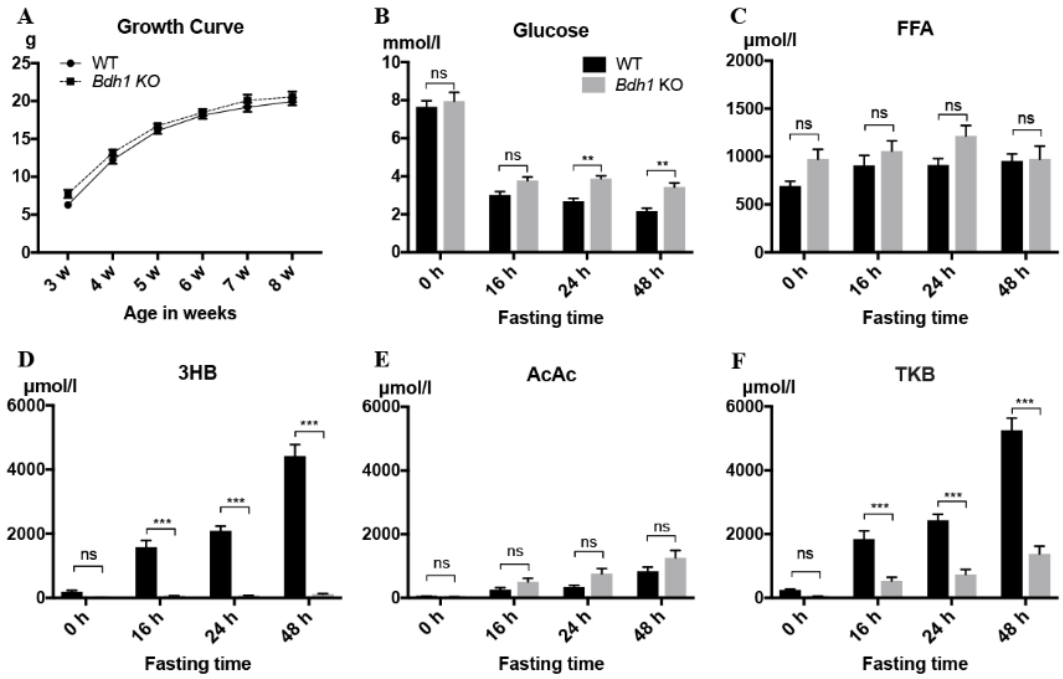
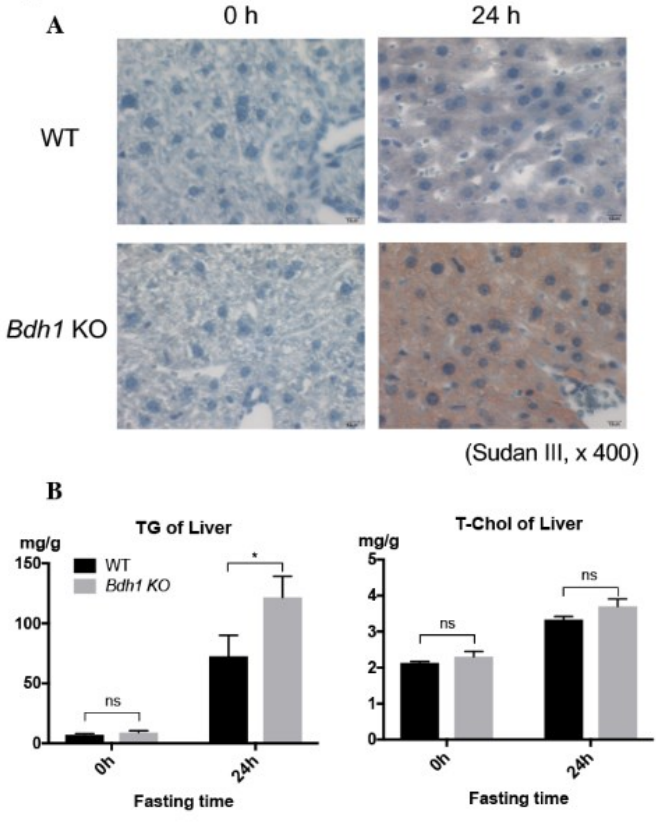
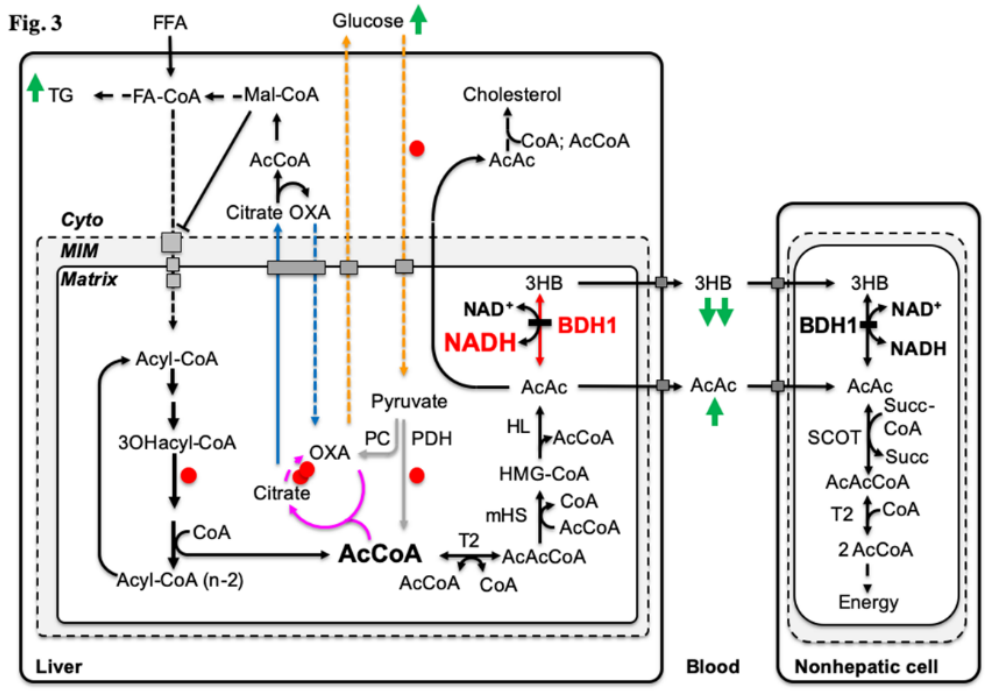
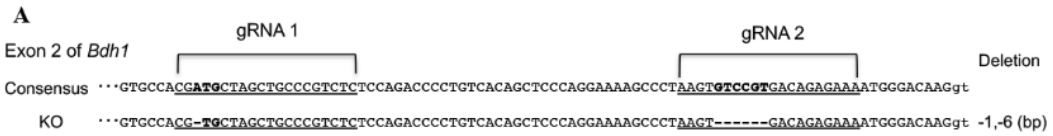

A
Growth Curve
g
WT
Bdh1 KO
3 w
4 w
5 w
6 w
7 w
8 w
Age in weeks
B
Glucose
mmol/l
WT
Bdh1 KO
ns
**
0 h
16 h
24 h
48 h
Fasting time
C
FFA
μmol/l
ns
Fasting time
D
3HB
μmol/l
***
Fasting time
E
AcAc
μmol/l
Fasting time
F
TKB
μmol/l
***
Fasting time

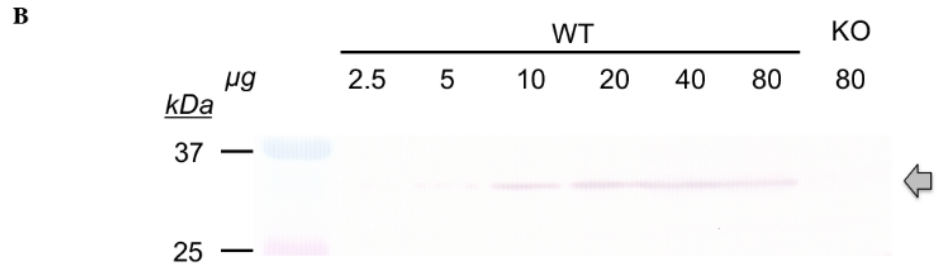
Fig. 2


A
0 h
24 h
WT
Bdh1 KO
(Sudan III, x 400)
B
TG of Liver
mg/g
WT
Bdh1 KO
*
ns
0h
24h
Fasting time
T-Chol of Liver
mg/g
ns
Fasting time



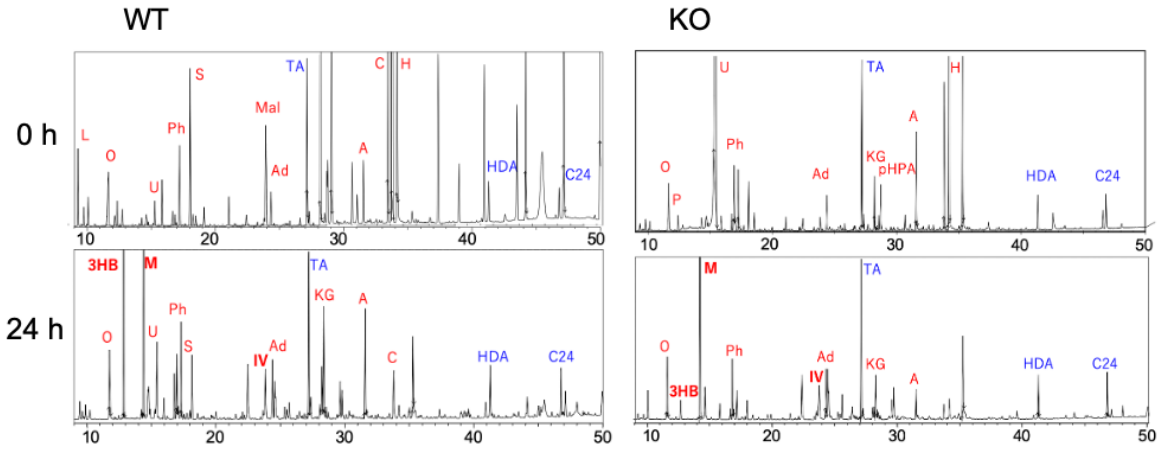
Supplemental Fig. 1





Supplemental Fig. 1A: Sequences of wild type and *Bdh1* KO mouse in exon2 of *Bdh1* in the region of the initiation methionine codon. The initiation ATG methionine codon is in red. The two gRNA target regions are underlined. KO had both a deletion of c.1A of the initiator methionine codon and c.58_63del (GTCCGT). Combined, these changes alter both reading frame and length.
B: Immunoblot analysis of Bdh1. Homogenized liver samples were measured for protein content and resolved on a 12% SDS-PAGE gel. On the left, WT samples, 2.5 to 80 μg, were applied as positive controls, and 80 μg of protein from KO was applied. A rabbit polyclonal antibody to BDH1 was used as first antibody and an alkaline phosphatase-conjugated polyclonal antibody against rabbit IgG was used as the second antibody. Bdh1 is detected as a 31 kDa signal (grey arrow) in lanes containing 5 μg or more of protein from WT mice. In contrast, no signal is detectable in *Bdh1* KO homogenate (80 μg).

Supplemental Fig. 2



Supplemental Fig. 2: Urinary organic acids analysis of WT and *Bdh1* KO mouse with/without fasting.

TA, HDA, C24 were added as internal standards. AcAc were not detected because of their instability. 3HB, malonate, isovalerylglycine were detected in 24 fasting of both KO and WT. The increase of 3HB is clearly higher in WT. L=lactate; O=oxalate; P=pyruvate; 3HB=3-OH-butyrate; M=malonate; U=urea; Ph=phosphate; S=succinate; IV=isovalerylglycine; Ad=adipate; TA=tropate (IS); KG=2-ketoglutarate; pHPA=p-OH-phenylacetate; A=aconitate; C=citrate; H=hippurate; HDA=heptadecanoate (IS); C24=tetracosane (IS)