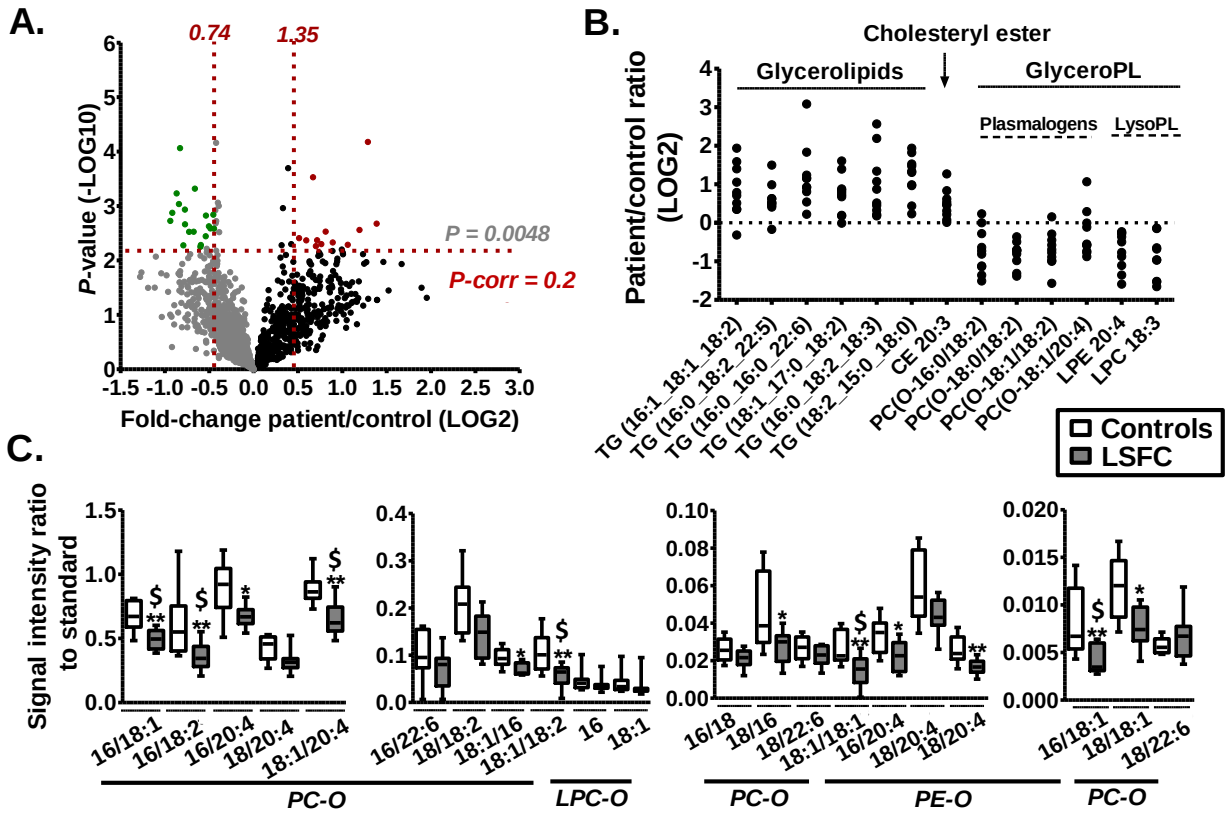
SUPPLEMENTAL INFORMATION

SUPPLEMENTAL FIGURES

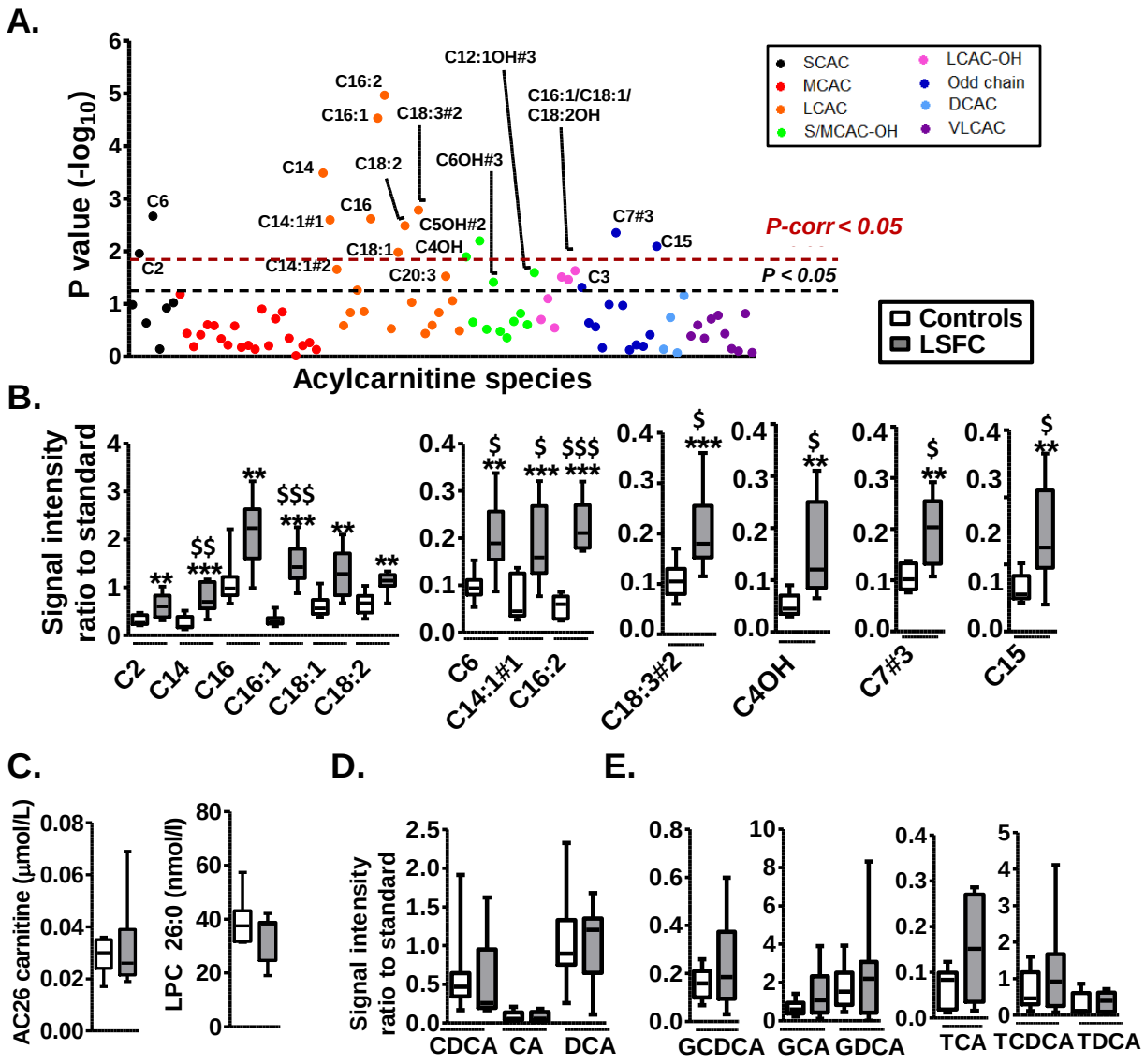
Figure S1. Untargeted (A-B) and targeted (C) lipidomics unveil major plasma lipid homeostasis, including lower circulating plasmalogen levels, in LSFC patients (related to Figure 1).



(A) Volcano plot from LC-QTOF-based untargeted lipidomics of plasma from LSFC and control subjects (n=9/group; after a "nutrient intake challenge") depicting the 1,478 features obtained following MS data processing. Using a corrected *P*-value threshold of 0.2 (corresponding to an uncorrected *P*-value (*P*-corr) of 0.0048; horizontal red dotted line) and a fold-change (FC) >1.35 or <0.74 (vertical red dotted lines), 31 features significantly discriminated LSFC patients from controls, of which 11 were increased (red

dots) and 20 decreased (green dots). **(B)** Dot plot of 13 selected lipids significantly discriminating LSFC patients from controls and identified by MS/MS using LC-QTOF. Each dot represents a log2-transformed patient/matched control signal intensity ratio (n=9) for the indicated lipid (sub)classes with their acyl side chain(s): (i) 2 lysophospholipids (LPC and LPE); (ii) 6 glycerolipids: triacylglycerol (TG) (iii) 1 cholesteryl ester (CE); (iv) 4 phosphatidylcholine plasmalogens (PC-*O*). **(C)** Box plots of LC-QQQ-based lipidomic analysis of plasmalogens, 21 of which were detected in plasma from LSFC patients (grey; n=9) and controls (white; n=9): 2 LPC-*O*, 15 PC-*O* and 4 phosphatidylethanolamine (PE) plasmalogens (PE-*O*). Statistics using paired Student *t*-test: **P* <0.05, ***P* <0.01 before and $^{\$}$*P* <0.05 after Benjamini Hochberg correction.

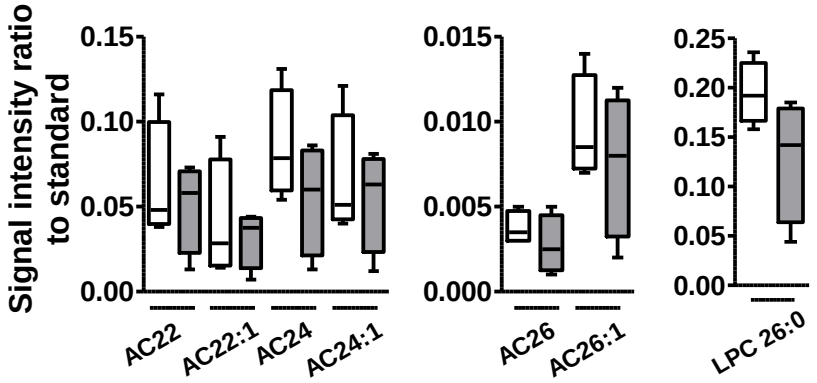
Figure S2. Targeted profiling of acylcarnitines (ACs) and lysophosphatidylcholine (LPC) 26:0 (A-C) and unconjugated/conjugated bile acids (BAs; D-E) reveals additional metabolic perturbations in plasma from LSFC patients (related to Figure 2).



A-B: LC-QQQ-based profiling of 91 ACs from plasma of fasted LSFC patients (black or grey; n=9) and controls (white; n=9). **(A)** Dot plot of *P*-values obtained using paired Student *t*-test analysis for the various AC species: short chain (SCAC; black), medium

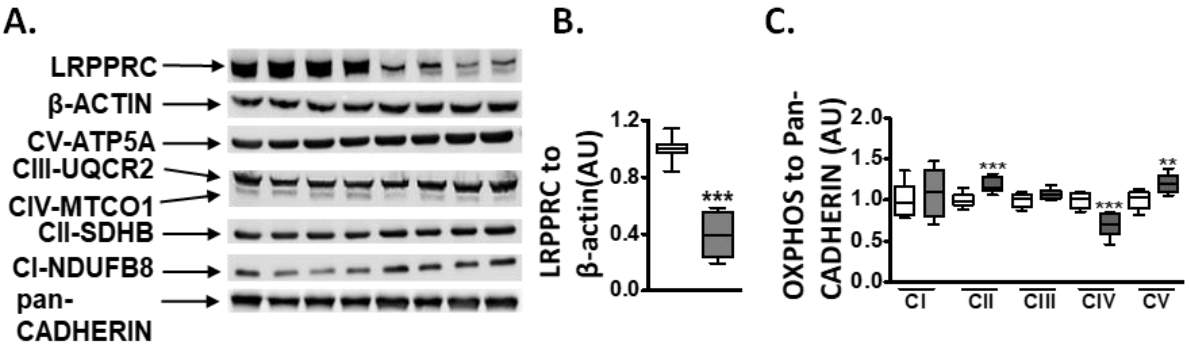
chain (MCAC; red), long chain (LCAC; orange), hydroxylated short/medium chain (S/MCAC-OH; green), hydroxylated long chain (LCAC-OH; pink), odd chain (dark blue), dicarboxylic (DCAC; light blue) and very long chain (VLCAC; purple). Significantly elevated ACs are above the dotted line (black $P < 0.05$, red P-corr < 0.05). **(B)** Box plots of selected significantly (according to P-corr) elevated AC species in fasted LSFC patients (grey) and controls (white). (C) Box plots of quantitative values of very long chain AC, AC26 and the corresponding lysophosphatidylcholine (LPC) 26:0 **(D-E):** Box plots from LC-QQQ-based profiling from plasma of post-smoothie LSFC patients (grey; n=4-9) and controls (white; n=6-9) for (D) unconjugated BA, (E) glyco-conjugated BAs and (F) tauro-conjugated BAs. Unequal distribution is ascribed to values below the limit of detection. Statistics using paired Student *t*-test: $^{*}P < 0.05$, $^{**}P < 0.01$, $^{***}P < 0.001$ before and $^{\$}P < 0.05$, $^{\$\$}P < 0.01$, $^{\$\$\$}P < 0.001$ after Benjamini-Hochberg correction.

Figure S3. Targeted profiling of plasma very long chain acylcarnitines (AC) and lysophosphaditylcholine (LPC) 26:0 do not reveal any significant changes between H-*Lrpprc*$^{-/-}$ mice *vs.* controls (related to Figure 3).



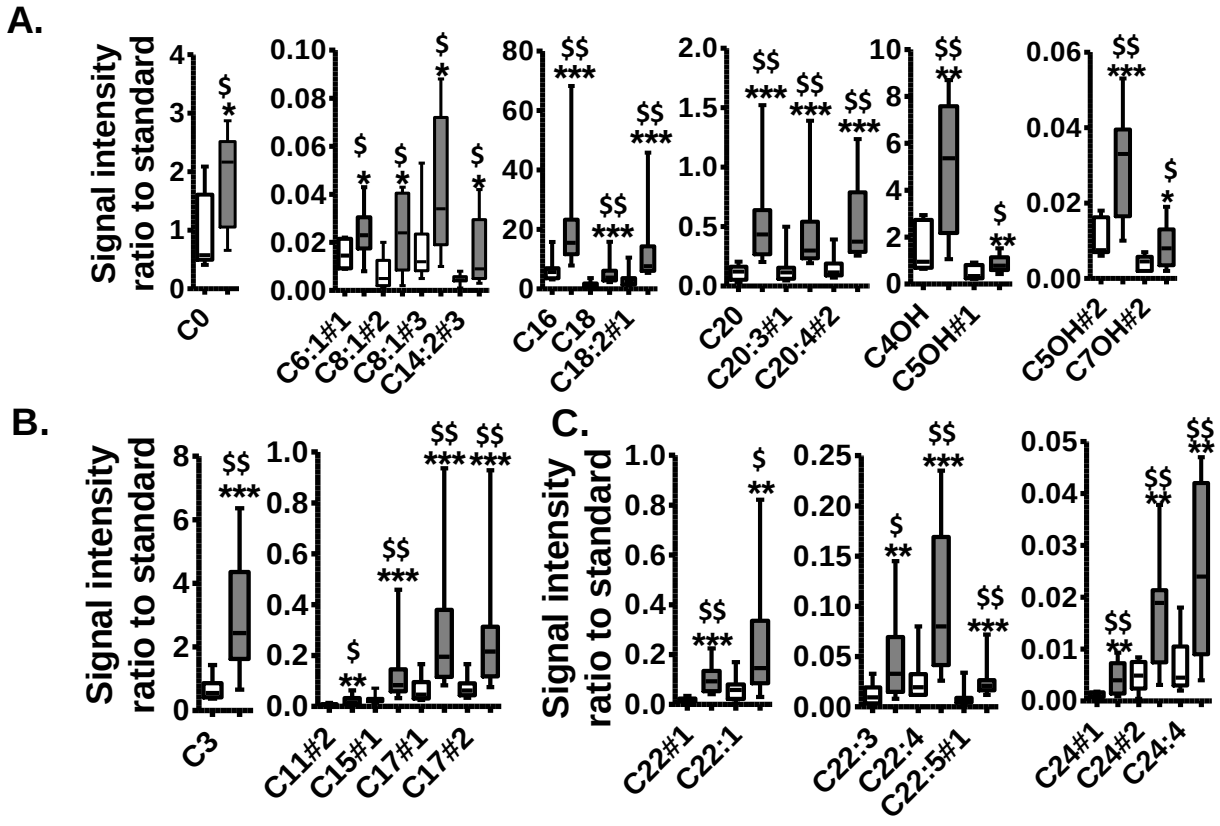
Box plots of very long chain-ACs and LPC 26:0 from LC-QQQ-based profiling of mouse plasma (n=4/group, controls in white and H-*Lrpprc*$^{-/-}$ in grey).

Figure S4. Immunoblot analysis of LRPPRC and OXPHOS complexes subunits in livers from H-*Lrpprc*$^{-/-}$ mice.



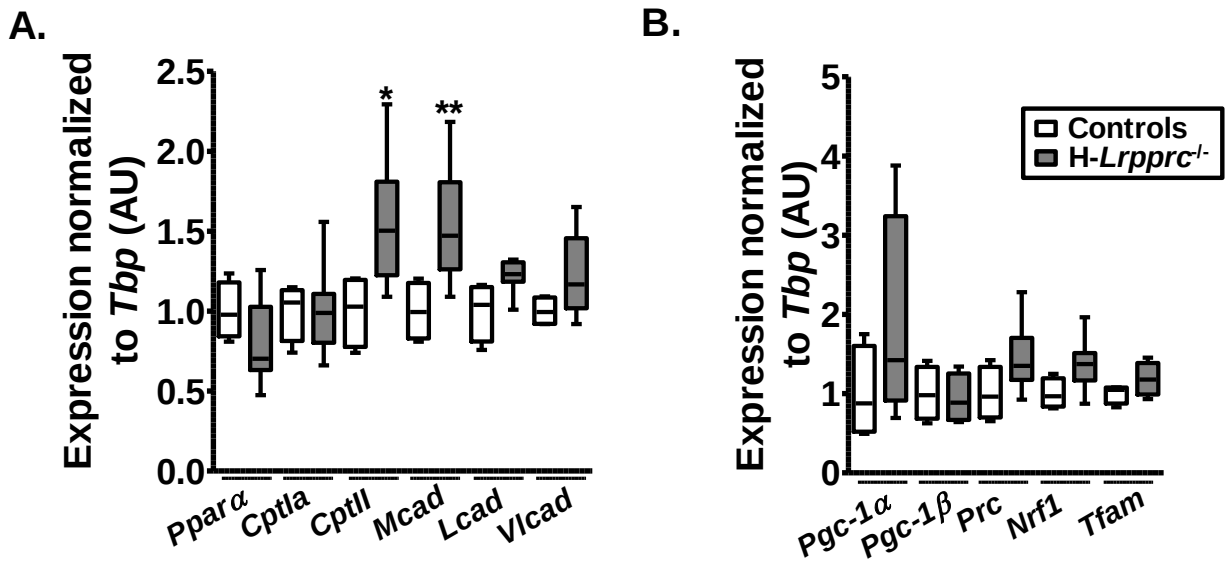
.A) Representative images and quantitation (box plots; controls in white, n=8; H-*Lrpprc*$^{-/-}$ in grey, n=8) of **B)** LRPPRC, β-actin – for LRPPRC normalization – and **C)** OXPHOS complexes subunits (CV-ATP5A, CIII-UQCR2, CIV-MTCO1, CII-SDHB, CI-NDUFB8) as well as pan-cadherin – for OXPHOS normalization – immunoreactivity. ***P* <0.01, ****P* <0.001 using unpaired two-tailed Student *t*-test analysis.

Figure S5. Targeted profiling of liver acylcarnitines (ACs) reveal disturbances in mitochondrial and peroxisomal fatty acid metabolism (related to Figure 5).



(A-B) Box plots of LC-QQQ-based profiling of SCACs, MCACs and LCACs, including hydroxylated and non-hydroxylated ACs **(B)** or odd chain and VLC-ACs **(C)** in livers from fed H-*Lrpprc*$^{-/-}$ mice (n=13; grey) and their littermate controls (n=8; white). Statistics using two-tailed unpaired Student *t*-test: **P* <0.05; ***P* <0.01, ****P* <0.001 before and $^{\$}$p < 0.05, $^{\$\$}$p < 0.01 after Benjamini-Hochberg correction.

Figure S6. Livers from H-*Lrpprc*$^{-/-}$ mice display significant changes for molecular markers of mitochondrial fatty acid oxidation albeit not for mitochondrial biogenesis (related to Figure 6).



Levels of transcripts were assessed in whole liver extracts from H-*Lrpprc*$^{-/-}$ mice (grey, n=13) and controls (white, n=8). (**A-B)** Box plots depicting levels of mRNA, normalized to *Tbp*, are shown for markers of (**A**) mitochondrial biogenesis (*Pgc-1α, Pgc-1β, Prc, Nrf1, tfam*) and (**B**) fatty acid oxidation (*Pparα, CptIa, CptII, Mcad, Lcad, Vlcad*). **P* <0.05, ** *P* <0.01 using two-tailed unpaired Student *t*-test.

SUPPLEMENTAL TABLES

Table S1: Related to Figure 1. List of lipids identified by MS/MS that significantly discriminated plasma from LSFC patients *vs.* controls by untargeted lipidomic analysis using a corrected *P*-value threshold of 0.2 (corresponding to an uncorrected *P*-value of 0.0034) and a fold-change (FC) > 1.35 or < 0.74.

MS/MS ID	FC	*Uncorrected P*-value	Corrected *P*-value
	Glycerophospholipids		
PC-plasmalogens			
LPC (O-18:1)	0.64	1.5E-03	0.168
PE (O-16:0/18:2)	0.60	1.2E-03	0.177
PE (O-18:1/18:2)	0.61	1.9E-03	0.188
PE (O-18:1/20:4)	0.69	3.4E-03	0.195
	Acylcarnitines		
AC16:1	6.03	2.4E-05	5.0E-03
AC18:1	1.63	1.3E-03	0.177
	Glycerolipids		
TG (18:2_16:1_18:2)	3.27	2.7E-03	0.188
TG (18:0_18:2_20:4)	2.42	3.1E-03	0.195
TG (18:1_16:1_22:4)	1.82	3.4E-03	0.195
TG (16:0_16:0_22:6)	2.56	1.0E-03	0.177
TG (16:0_18:1_22:6)	1.85	2.9E-03	0.195
TG (16:0_18:2_22:6)	2.33	2.9E-03	0.149
DG (18:2/18:2)	2.42	3.4E-03	0.195
	Cholesteryl ester		
CE (20:3)	1.72	2.6E-03	0.188

Abbreviations: AC, acylcarnitine; CE, cholesteryl ester; DG, diacylglycerol; FC, fold-change; LPC (O-), lysophosphatidylcholine plasmalogen; ID, identification; PC (O-), phosphatidylcholine plasmalogen; PE (O-), phosphatidylethanolamine plasmalogen; TG, triacylglycerol.

Table S2: Related to Figure 3. List of lipids identified by MS/MS that significantly discriminated plasma from H-*Lrpprc*$^{-/-}$ mice *vs.* their littermate counterparts by untargeted lipidomic analysis using a corrected *P*-value threshold of 0.05 (corresponding to an uncorrected *P*-value of 0.01) and a FC >2 or <0.5.

MS/MS ID	FC	*Uncorrected P*-value	Corrected *P*-value
	Glycerophospholipids		
PC-enriched in 22:6			
LPC (22:6)	0.49	4.8E-03	2.9E-02
PC (22:6_20:4)	0.30	1.0E-06	2.2E-04
PC (20:0_22:6)	0.29	5.4E-04	9.0E-03
PC (18:2_22:6)	0.47	5.0E-04	9.0E-03
PE-plasmalogens-enriched in 22:6			
PE (O-16:0/22:6)	0.44	1.0E-05	8.9E-04
PE (O-18:0/22:6)	0.49	4.8E-05	2.0E-03
PC-enriched in 22:5			
LPC (22:5)	2.64	2.0E-03	1.6E-02
PC (18:0_22:5)	2.05	8.0E-04	1.1E-02
PC (18:1_22:5)	3.28	8.6E-05	3.0E-03
Other PC			
PC (16:0_20:5)	0.28	2.2E-08	2.8E-05
PC (18:0_16:0)	0.46	7.0E-03	3.7E-02
PC (20:0_20:4)	0.38	5.4E-03	3.2E-02
PC (20:2_18:2)	2.03	1.8E-03	1.7E-02
	Acylcarnitines		
AC18:0	2.04	8.5E-04	1.2E-02
	Glycerolipids		
TG (18:1_18:1_16:0)	0.49	1.2E-02	1.9E-02
TG (20:5_18:2_16:0)	0.45	2.0E-03	4.9E-02
TG (22:6_16:0_16:1)	0.40	3.0E-03	2.0E-02
TG (18:2_16:1_22:6)	0.25	1.5E-04	4.0E-03
TG (18:2_20:3_18:2)	2.72	1.0E-03	1.0E-02
TG (22:5_18:2_16:0)	2.27	5.0E-04	9.0E-03
TG (22:5_18:1_18:1)	2.57	1.0E-03	1.0E-02
TG (22:5_18:2_18:1)	5.46	7.9E-05	3.0E-03
TG (20:3_18:2_20:4)	2.43	3.5E-05	2.0E-03
TG (22:5_18:2_18:2)	5.24	3.1E-05	2.0E-03
TG (20:4_18:2_18:2)	2.59	2.2E-04	5.3E-03
TG (20:4_18:1_22:5)	5.95	7.4E-06	6.9E-04
TG (22:6_18:0_20:4)	2.11	2.0E-03	2.0E-02
TG (20:4_18:2_22:5)	2.04	7.0E-03	3.5E-02
TG (22:5_18:2_22:6)	2.94	3.3E-06	4.3E-04
	Cholesteryl ester		
CE (16:1)	0.47	7.0E-03	4.0E-02
CE (18:0)	0.37	7.0E-03	4.0E-02
CE (20:3)	0.40	2.0E-03	2.0E-02
CE (20:5)	0.25	8.5E-07	2.2E-04
CE (22:4)	0.17	2.0E-03	2.0E-02
CE (22:6)	0.40	2.0E-03	2.0E-02

Abbreviations: AC, acylcarnitine; CE, cholesteryl ester; FC, fold-change; LPC, lysophosphatidylcholine; ID, identification; PC, phosphatidylcholine; PE (O-), phosphatidylethanolamine plasmalogen; TG, triacylglycerol.

Table S3: Related to Figure 4. List of lipids identified by MS/MS that significantly discriminated liver from H-*Lrpprc*$^{-/-}$ mice *vs.* their littermate counterparts by untargeted lipidomic analysis using a corrected *P*-value threshold of 0.05 (corresponding to an uncorrected *P*-value of 0.015) and a FC >2.5 or <0.4.

MS/MS ID	FC	*P*-value	Corrected *P*-value
	Glycerophospholipids		
PG/PS			
PG (18:2/18:2)	3.12	2.0E-08	2.0E-06
PG (18:2_20:4)	3.32	7.0E-07	9.0E-06
PS (22:5_18:0)	3.73	9.0E-10	3.0E-07
PS (20:4_16:0)	0.30	1.0E-07	5.0E-06
PC			
PC (16:0_18:3)	0.37	6.7E-03	2.5E-02
PC (16:0_20:5)	0.34	3.0E-07	9.0E-06
PC (22:6_20:4)	0.35	1.0E-07	5.0E-06
PC (18:0_20:2)	2.61	7.0E-07	3.0E-04
PC (17:0_22:5)	7.37	5.0E-06	8.0E-05
PC (18:0_22:5)	2.91	1.0E-07	5.0E-06
PC (18:1_22:5)	3.83	2.0E-05	2.0E-04
PC (18:2_22:5)	3.26	5.4E-03	2.2E-02
PC (20:2_22:5)	2.94	1.5E-02	4.7E-02
PE			
PE (16:0_20:2)	3.06	2.0E-05	3.0E-04
PE (16:0_22:6)	3.44	5.2E-03	2.1E-02
PE (18:0_20:2)	4.27	5.0E-07	1.0E-05
PE (18:0_22:5)	5.44	8.0E-10	3.0E-07
PE (18:0_22:6)	2.94	2.0E-06	4.0E-05
PE (18:1_18:1)	2.97	1.0E-04	1.3E-03
PE (18:1_20:2)	3.22	1.5E-03	8.2E-03
PE (18:1_22:5)	5.85	6.0E-08	4.0E-06
PE (20:2_22:6)	2.59	3.0E-04	2.6E-03
	Acylcarnitines		
AC16	2.96	2.0E-05	2.0E-04
AC16:1	3.57	5.0E-05	6.0E-04
AC18	3.48	2.0E-06	4.0E-05
AC18:2	3.50	1.0E-05	1.0E-04
	Glycerolipids		
TG (18:1_16:1_16:0)	0.26	1.5E-03	8.2E-03
TG (18:1_17:1_16:0)	0.17	2.9E-03	1.3E-02
TG (18:2_18:3_18:2)	0.32	3.5E-03	1.6E-02
TG (18:1_18:1_22:6)	0.21	1.6E-02	4.8E-02
TG (18:2_18:1_22:6)	0.35	2.9E-03	1.3E-02
TG (18:4_18:2_16:0)	0.16	8.0E-04	5.0E-03
TG (14:2_18:2_16:0)	5.00	6.4E-03	2.5E-02
TG (18:2_12:0_18:2)	3.46	6.0E-03	2.4E-02
TG (18:2_14:2_18:2)	3.98	4.5E-03	1.9E-02
TG (16:0_14:0_22:6)	4.44	2.5E-03	1.2E-02
TG (18:2_18:2_20:2)	3.62	4.0E-03	3.3E-03

TG (22:5_18:1_18:2)	3.92	2.6E-03	1.3E-02
TG (22: 6_16:0_22:5)	3.60	6.5E-03	2.5E-02
TG (18:2_22:5_18:2)	2.76	1.8E-03	9.3E-03
TG (22:5_20:3_18:2)	2.77	5.0E-04	3.4E-03

Abbreviations: AC, acylcarnitine; FC, fold-change; ID, identification; PC, phosphatidylcholine; PE, phosphatidylethanolamine; PG, phosphatidylglycerol; PS, phosphatidylserine; TG, triacylglycerol.

Table S4. Characteristics of targeted LC-QQQ method for semi-quantitative analysis of plasmalogens.

Analyte	Precursor ion	Quantifier	Qualifier
PC(14:0/14:0)(Standard)	722.5	227.2	662.5
PC(O-16:0/18:0)	792.6	283.3	732.6
PC(O-16:0/18:1)	790.6	281.2	221.2
PC(O-16:0/18:2)	788.6	279.2	728.6
PC(O-16:0/20:4)	812.6	303.2	752.6
PC(O-16:0/22:6)	836.6	327.2	776.6
PC(O-18:0/16:0)	792.6	255.2	732.4
PC(O-18:0/18:1)	818.6	281.2	758.6
PC(O-18:0/18:2)	816.6	279.2	756.6
PC(O-18:0/20:4)	840.6	303.2	780.6
PC(O-18:0/22:6)	864.6	327.2	804.6
PC(p-18:0/16:0)	790.6	255.2	730.6
PC(p-18:0/18:1)	816.6	281.2	756.6
PC(p-18:0/18:2)	814.6	279.2	754.6
PC(p-18:0/20:4)	838.6	303.2	778.6
PC(p-18:0/22:6)	866.6	327.2	806.6
PC(p-16:0/18:1)	788.6	281.2	728.6
PE(O-16:0/18:1)	702.5	281.2	
PE(O-16:0/20:4)	724.5	303.2	
PE(O-18:0/20:4)	752.6	303.2	
PE(P-18:0/20:4)	750.5	303.2	
LPC(O-18:1)	552.4	492.4	
LPC(O-16:0)	526.4	466.4	

Abbreviations: LPC, lysophosphatidylcholine plasmalogen; PC, phosphatidylcholine; PE, phosphatidylethanolamine.

Table S5. Characteristic of targeted LC-QQQ method for semi-quantitative analysis of very long chain acylcarnitines and lysophosphatidylcholine.

Analyte	Internal standard	RT (min)	MRM transition (m/z)	CV inter-day (%) on human plasma
Docosanoyl carnitine (C22:0 Car)	Hexacosanoyl DL-carnitine-d_9	33.17	484.4->85^{Quant} 484.4->60^{Qual}	13.1
Tetracosanoyl carnitine (C24:0 Car)	Hexacosanoyl DL-carnitine-d_9	33.85	512.5->85^{Quant} 512.5->60^{Qual}	13.2
Hexacosanoyl carnitine (C26:0 Car)	Hexacosanoyl DL-carnitine-d_9	34.45	540.5->85^{Quant} 540.5->60^{Qual}	14.4
Docosenoyl carnitine (C22:1 Car)	Hexacosanoyl DL-carnitine-d_9	32.48	482.4->85^{Quant} 482.4->60^{Qual}	11.3
Tetracosenoyl carnitine (C24:1 Car)	Hexacosanoyl DL-carnitine-d_9	33.24	510.5->85^{Quant} 510.5->144^{Qual}	13.6
Hexacosenoyl carnitine (C26:1 Car)	Hexacosanoyl DL-carnitine-d_9	33.88	538.5->85^{Quant} 538.5->60^{Qual}	12.6
Hexacosanoyl-hydroxy-glycerophosphocholine (C26:0 LPC)	1-Hexacosanoyl-d_4-2-hydroxy-sn-glycero-3-phosphocholine	35.06	636.5->104.1^{Quant} 636.5->618.3^{Qual}	15

Table S6. Characteristics of targeted LC-QQQ method for semi-quantitative analysis of bile acids.

Analytes	Internal standard	Quantifier	Qualifier (MRM transition)
Cholic acid (CA)	d_4-CA	407.3	407.3->325.2^{Qual}
			407.3->289.3^{Qual}
			407.3->232.8^{Qual}
Taurocholic acid (TCA)	d_4-CA	514.3	514.3->124.^{Qual}
			514.3->106.9^{Qual}
			514.3->79.9^{Qual}
Glycocholic acid (GCA)	d_4-GCA	464.3	464.3->262.2^{Qual}
			464.3->74.1^{Qual}
Taurochenodeoxycholic acid (TCDCA)	d_4-CA	498.3*	498.3->124.0^{Qual}
			498.3->106.9^{Qual}
			498.3->80.1^{Qual}
Taurodeoxycholic acid (TDCA)	d_4-CA	498.3*	498.3->124.0^{Qual}
			498.3->106.9^{Qual}
			498.3->80.1^{Qual}
Glycochenodeoxycholic acid (GCDCA)	d_4-GCDCA	452.3*	452.3->390.5^{Qual}
			452.3->73.9^{Qual}
Glycodeoxycholic acid (GDCA)	d_4-GDCA	452.3*	452.3->390.5^{Qual}
			452.3->73.9^{Qual}
Chenodeoxycholic acid (CDCA)	d_4-CDCA	391.3	391.3->69.2^{Qual}
Deoxycholic acid (DCA)	d_4-DCA	391.3	391.3->355.4^{Qual}
			391.3->345.2^{Qual}
			391.3->327.3^{Qual}
			391.3->311.3^{Qual}
			391.3->69.2^{Qual}
d_4-CA		411.3	411.3->346.9^{Qual}
			411.3->327.7^{Qual}
			411.3->290.2^{Qual}
d_4-GCA		468.3	468.3->406.3^{Qual}
			468.3->74.2^{Qual}
d_4-GCDCA		452.3	452.3->452.4^{Qual}
			452.3->452.5^{Qual}
d_4-GDCA		448.3	448.3->386.0^{Qual}
			448.3->73.9^{Qual}

d_4-CDCA	395.3	395.3->72.0^{Qual}
d_4-DCA	395.3	395.3->359.2^{Qual}
		395.3->349.3^{Qual}
		395.3->330.4^{Qual}
		395.3->72.2^{Qual}

*Differentiated through their retention time.

Table S7. Targeted LC-QQQ method validation for semi-quantitative analysis of bile acids in plasma.

Analyte	Internal standard for normalization	LOQ (pmole)	CV of LC-MS system (%)	CV intra-day of entire workflow (%)	CV inter-day of entire workflow (%)
Cholic acid (CA)	d4-CA	3.0	5.0	6.1	11.7
Taurocholic acid (TCA)	d4-CA	1.0	4.4	8.5	11.3
Glycocholic acid (GCA)	d4-GCA	1.0	1.4	3.4	5.9
Taurochenodeoxycholic acid (TCDCA)	d4-CA	1.0	2.9	8.8	11.1
Taurodeoxycholic acid (TDCA)	d4-CA	1.0	4.3	10.6	8.1
Glycochenodeoxycholic acid (GCDCA)	d4-GCDCA	3.0	2.5	9.6	10.4
Glycodeoxycholic acid (GDCA)	d4-GDCA	10.0	1.3	2.4	23.9
Chenodeoxycholic acid (CDCA)	d4-CDCA	10.0	3.4	9.0	21.5
Deoxycholic acid (DCA)	d4-DCA	3.0	3.8	2.8	7.7

Abbreviations: CV, coefficient of variation; LOQ, limit of quantification.

Table S8: List of murine oligonucleotides used for RT-qPCR.

Gene	Accession Number	Primer (sense)	Primer (anti-sense)
Mitochondrial biogenesis and lipid metabolism genes			
Pgc-1α	NM_008904	TGGATGAAGACGGATTGC	TGGTTCTGAGTGCTAAGAC
Pgc-1β	NM_133249.2	TGGAAAGCCCCTGTGAGAGT	TTGTATGGAGGTGTGGTGGG
Prc	NM_001081214	CCAGAACTGGCCAACGTG	ATAACTGGTGGGGAGGGGTA
Nrf1	NM_010938	TTACTCTGCTGTGGCTGATGG	CCTCTGATGCTTGCGTCGTCT
Tfam	NM_009360.4	GAAAGCACAAATCAAGAGGAG	CTGCTTTTCATCATGAGACAG
Pppara	NM_011144	CAACATGAACAAGGTCAAGGC	GGCAGCAGTGGAAGAATCG
Cpt1a	NM_013495.2	TCCTTCCCATTTGACACCTT	GAAGAGCCGAGTCATGGAAG
Cpt2	NM_009949	TGCTCCGAGGCGTTTGTCAGGG	GAGACATTGCAGCCTATCCAGT
Mcad	NM_007382	TTGACGGAACAGCAGAAAG	CCATACGCCAACTCTTCG
Lcad	NM_017381	ATGCCCTATATTGCGAATTACG	CCTTGCTTCCATTGAGAATCC
Vlcad	NM_017366	GGCTCTCCAAGGCTGTATG	ACCACTGCGACTTAACTCTG
Peroxisomal biogenesis and lipid metabolism genes			
Pex11α	NM_011068	TCAAGAGGCTGGAGACCAGT	CGGTTGAGGTTGGCTAATGT
Pex11β	NM_011069	CTATGGGCTGGAAAGTCTGG	CTCATAAGCATCACGGCTCA
Pex11γ	NM_026951.2	GTGTCAGCCCAGTTCAATCA	GGCGATATGCTCACAAGGAT
Pex14	NM_019781.2	GAGATTGACCTGGCTTTCCA	AATGCAATTCCTGCCATGAT
Pex3	NM_019961.3	CTCGGCGACAGTACCATTTT	TGCTGCATTAAGGCCTCTCT
Pex16	NM_145122.2	CTCGTCATTGCTCTCATCCA	CTGTGCCTGAGTTTCCCTGT
Pex19	NM_023041.3	TGTACCCATCCCTGAAGGAG	TGTGCTGCTGCTGGTACTTC
Pex1	NM_001293806.1	GCTCAAGGAATCCACACGTT	TCCAAGTCAGGGAAATTGCT
Pex6	NM_145488	GACGGAGTGGAGATTCTGGA	CCGATCACAAACACATCCTG
Pex10	NM_001042407.1	ACCTGGCCAAGAGACTAGCA	GATGCAAGAGGGAGATCAGC
Pex12	NM_134025.3	GCTCAGGACATGCAAGCTATC	CCTCCCACAGCTTTCTTCAG
Pex26	NM_028730.6	ATGGAGAGAAGCCCTGTCCT	GGGCTCTTTCATTTTGCTGT
Pxmp4	NM_021534.3	GCTCCTGCTGTTTGGAGAGA	CACTTGAGCGCAGGGATATAG
Abcd1	NM_007435.2	TAGCTGTCCCCATCATCACA	CGCTCACTAACCAGCTCCTC
Pex7	NM_008822.2	TTAGGGGCCATGAGAGTGTC	TGCTGGAATCACAATCCGTA
Acox1	NM_015729	CCTCCAATCATGCGATAGTCC	TTGTCCATCTTCAGGTAGCC
Acox2	NM_053115.2	ATCTTGGCATGTTGGTGACA	TTCCAGGATTTTTGCCTCTG
Ehhadh	NM_023737.3	TGATCTGTGGAGCAAATGACA	CCACCACTGGCTTCTGGTAT
Pthio	NM_130864.3	AATATCTCTTCCCGCCTGCT	CGCAAAGTCATCCTGCTTCT
Mfp2	NM_008292.4	CCATTGAAGGCAGGAAGAAC	TAGGCCACCATTTTCCTCAC
Agps	NM_172666.3	AAAGGAGAGGATAAGAAGGG	CATCATAAGTCTGTGCACC
Gnpat	NM_010322.3	TTGGCCTGATAACAAACTTC	TTCATTTTCAGAACCACCTG
Far1	NM_027379.4	GGTATTCTGGAGTTAATAGACC	GTTAGATTTACATTGGGCCA
Housekeeping genes			
Polr2A	NM_009089	CAACATGCTGACAGATATGACC	TGATGATCTTCTTCTTGTTGTCTG
Tbp	NM_013684	GGCCTCTCAGAAGCATCACTA	GCCAAGCCCTGAGCATAA
Rpl32	NM_172086	GCTGCTGATGTGCAACAAA	GGGATTGGTGACTCTGATGG
Rplp0	NM_007475	GCGACCTGGAAGTCCAACTA	TTGTCTGCTCCCACAATGAA
Hprt1	NM_013556	CCAGCGTCGTGATTAGCG	AGCAAGTCTTTCAGTCCTGTC
Ppia	NM_008907	CCGATGACGAGCCCTTGG	GCCGCCAGTGCCATTATG
Ywhaz	NM_011740	AGACGGAAGGTGCTGAGAAA	GAAGCATTGGGGATCAAGAA

Abbreviations: *Pgc-1α*, peroxisome proliferator-activated receptor gamma coactivator 1-alpha; *Pgc1-β*, peroxisome proliferator-activated receptor gamma coactivator 1-beta; *Prc*,

peroxisome proliferator-activated receptor gamma coactivator-related 1; *Nrf1*, nuclear respiratory factor 1; *Tfam*, mitochondrial transcription factor 1; *Pparα*, peroxisome proliferator-activated receptor alpha; *Cpt*, carnitine palmitoyl transferase; *Mcad*, medium-chain acyl-CoA dehydrogenase; *Lcad*, long-chain acyl-CoA dehydrogenase; *Vlcad*, very long-chain acyl-CoA dehydrogenase; *Pex*, peroxine; *Abcd1*, ATP binding cassette subfamily D1; *Acox*, acyl-CoA oxidase; *Ehhadh*, enoyl-CoA hydratase and 3-hydroxyacyl CoA dehydrogenase; *Pthio*, peroxisomal 3-oxoacyl-CoA thiolase; *Mfp2*, multifunctional protein 2; *Agps*, alkylglycerone phosphate synthase; *Gnpat*, glyceronephosphate O-acyltransferase; *Far1*, fatty acyl CoA reductase 1; *Polr2a*, RNA polymerase II subunit A; *Tbp*, tata binding protein; *Rpl*, ribosomal protein l; *Rplp*, ribosomal protein lateral stalk subunit p0; *Hprt1*, hypoxanthine phosphoribosyltransferase 1; *Ppia*, peptidylprolyl isomerase A; *Ywhaz*, tyrosine 3-monooxygenase/tryptophan 5-monooxygenase activation protein.

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