

Supplemental Information

Long-term correction of ornithine transcarbamylase deficiency in Spf-Ash mice with a translationally optimized AAV vector

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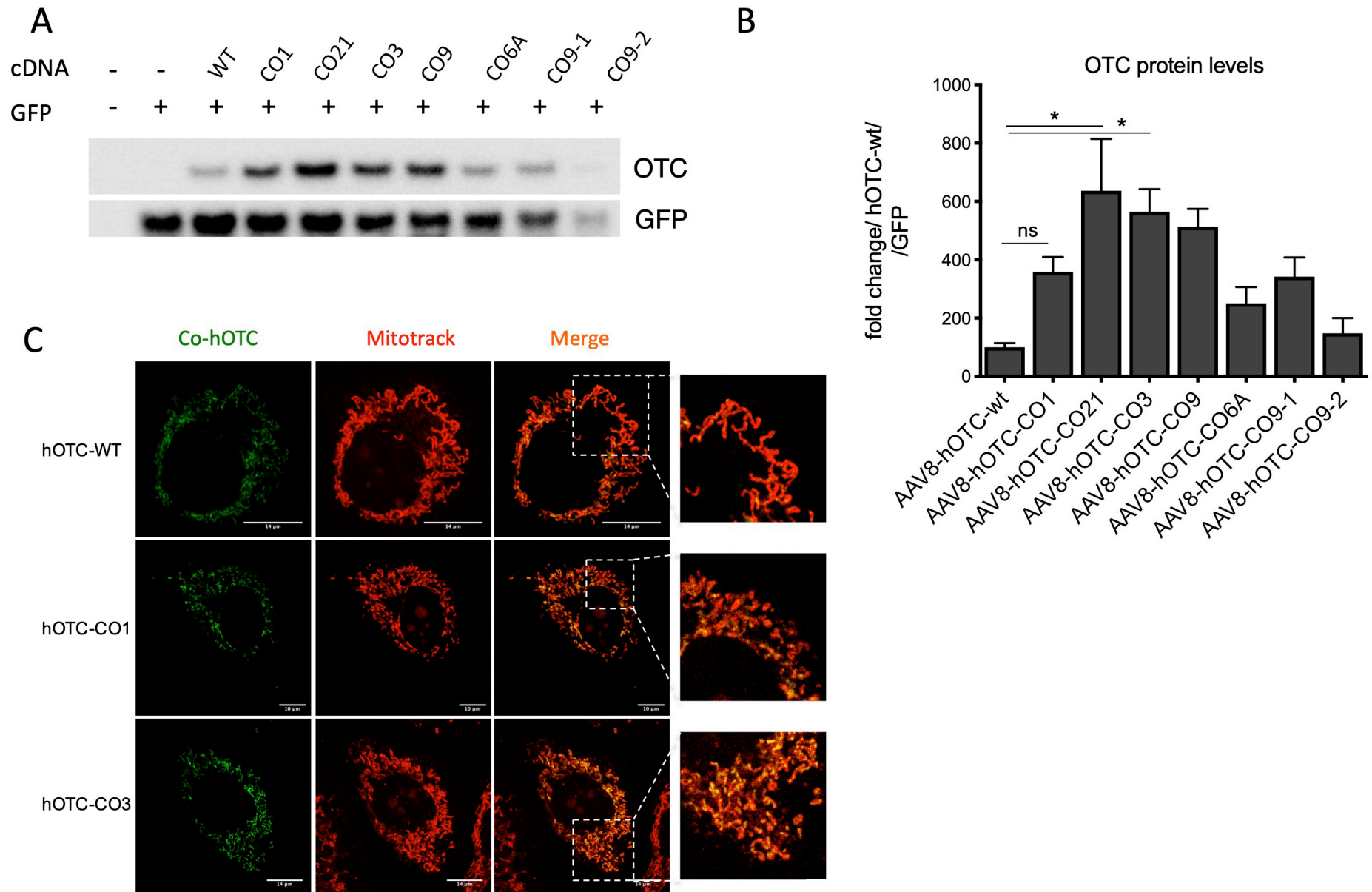


Figure S1. Codon-optimized hOTC variants tested in Huh-7 human liver cells. **A)** Representative Western blot analysis of OTC protein levels in cell lysates (15 μ g protein per lane) following co-transfection of pSMD2-hOTC-CO constructs and GFP plasmid. The control lane contains cell lysate from non-transfected cells, while the empty lane contains lysate from cells transfected with empty AAV8 plasmid (pSMD2) together with GFP plasmid. **B)** Densitometric quantification of OTC protein from the experiment shown in Panel A. GFP was used as transfection control. Values are expressed as fold-change respect to the AAV8-hOTC-wt. Data are shown as mean \pm SEM and statistical analyses was performed by one-way ANOVA with Turkey's Multiple comparison test ($n=2$, * $P < 0,05$). **C)** Sub-cellular localization studies of hOTC in Huh-7 human liver cells. Huh-7 cells were transfected with plasmids encoding the indicated hOTC-CO variants. Mitochondria (red, MITO-TRACK) and hOTC (green) were detected with a confocal microscope. On the right, a magnified picture of the indicated areas is shown. The scale bar corresponds to 14 μ m.

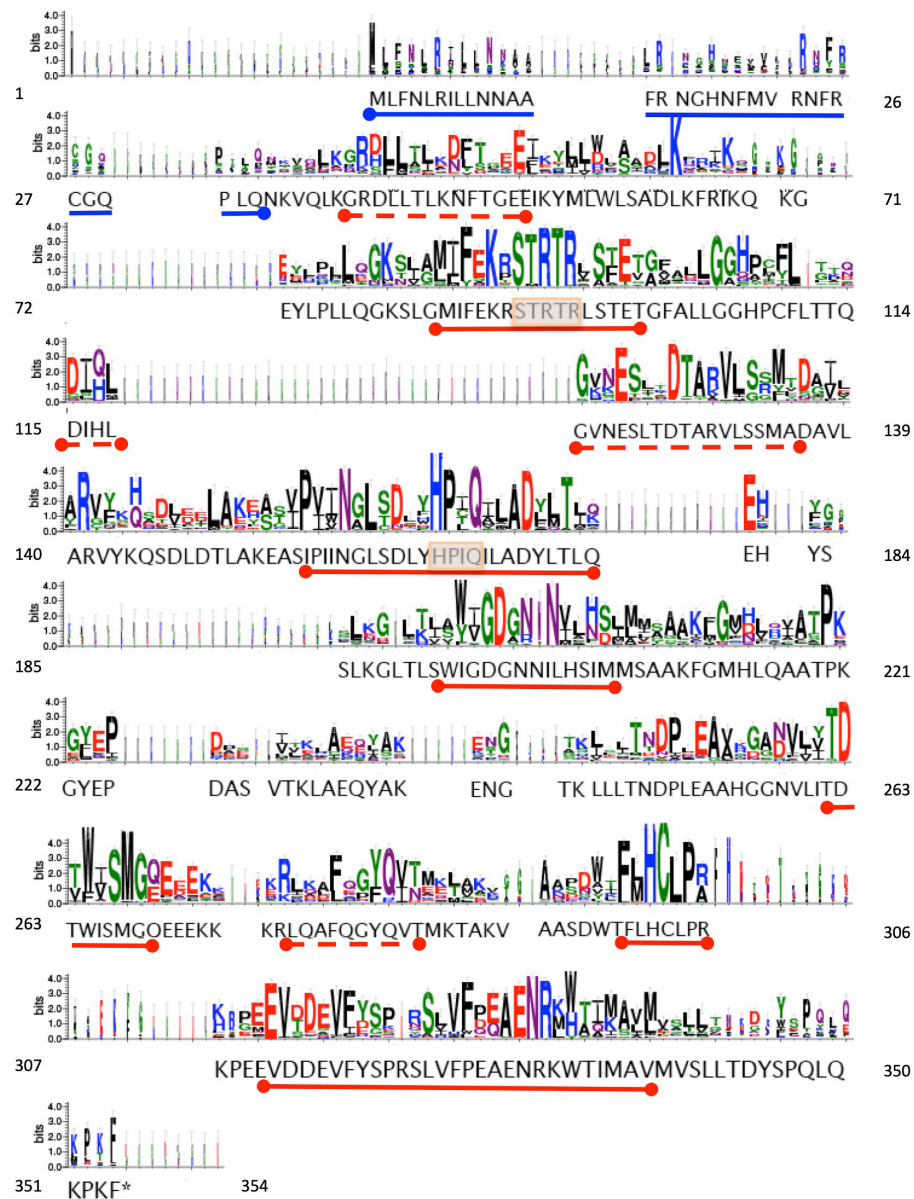


Figure S2. Logo representation of the alignment of 142 OTCases. The size of the letters indicates the degree of sequence conservation and most of these have catalytically or structurally important roles. The human full-length OTC primary sequence is shown below the logo. The signal peptide and mitochondrial localization sequence are indicated by a blue line below the human sequence. Very conserved regions are indicated by a red continuous line, while the dashed line indicate conserved regions. The active sites are indicated by an orange rectangle (90-94 and 160-171: Ornithine and carbamoyl phosphate binding site, respectively). The numbers at the side (from 1 to 354) correspond to the human OTC amino acid sequence. Gaps in the human sequence are caused by insertions in the OTC cDNAs in the different species, resulting in different-length sequences. Table S1 contains the list of species used in the representation.

5'

ATGCTGTTCAACCTGCGAATCCTGCTGAACAACGCCGCTTTTCGGAACGGGCACAAC
TTTATGGTGAGGAAC TTTCGCTGCGGACAGCCCCTCCAGAATAAGGTCCAGCTGAAG
GGCAGGGACCTGCTGACCCTGAAAAATTTACAGGGGAGGAAATCAAATACATGCTG
TGGCTGAGCGCCGATCTGAAGTTCAGAATCAAGCAGAAGGGCGAGTACCTGCCTCTG
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GACATTCACCTGGGAGTGAACGAGTCCCTGACCGACACTGCTCGCGTCCTGAGCTCT
ATGGCCGACGCCGTGCTGGCTCGGGTGTACAAACAGTCCGACCTGGATAACCCTGGCC
AAGGAAGCTTCCATCCCCATCATCAACGGCCTGAGCGACCTGTACCACCCCATCCAG
ATCCTGGCCGACTACCTGACCCTGCAGGAGCACTACAGCAGCCTGAAGGGCCTGACC
CTGAGCTGGATCGGCGACGGCAACAATATCCTGCACTCTATTATGATGTCTGCCGCC
AAGTTTGGAATGCACCTGCAGGCTGCTACCCCTAAAGGCTACGAACCCGATGCCTCT
GTGACAAAGCTGGCTGAACAGTACGCCAAAGAGAACGGCACAAGCTGCTGCTGACC
AACGACCCTCTGGAGGCCGCTCACGGAGGCAACGTGCTGATCACCGATACCTGGATT
AGTATGGGACAGGAGGAAGAGAAGAAGCGGCTGCAGGCCTTCCAGGGCTACCAG
GTCACCATGAAAACCGCTAAGGTGGCCGCCAGCGATTGGACCTTCTGCACTGCCTG
CCCAGAAAGCCCGAAGAGGTGGACGACGAGGTCTTCTACTCTCCAGAAAGCCTGGTG
TTTCCCGAAGCTGAGAATAGGAAGTGGACAATTATGGCAGTGATGGTGTCCCTGCTG
ACTGATTATTCTCCTCAACTGCAGAAACCTAAATTTTGA

3'

Figure S3. Nucleotide sequence of the codon optimized hOTC CO21 ORF. The 5' and 3' orientation of the sequence are indicated.

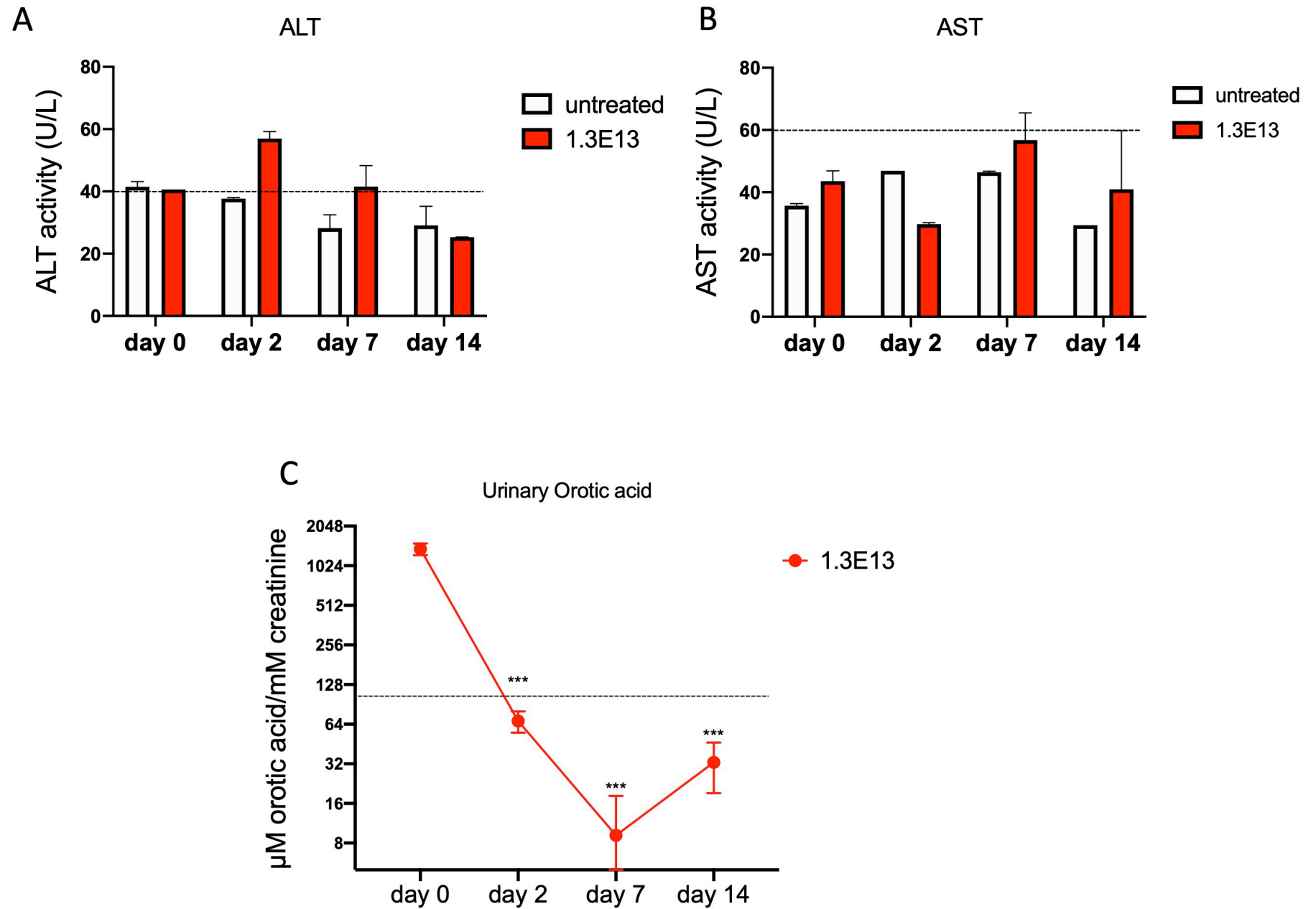


Figure S5. High dose treatment. Analysis of liver transaminases in plasma. Juvenile P30 OTC^{SpfAsh} mice were i.v. injected with 1.3E13 vg/kg of the or AAV8-hOTC-CO21Δice were i.v. inj2). Blood samples were taken at 2, 7 and 14 days after vector administration. AST **(A)**, ALT **(B)** liver transaminases, and urinary orotic acid **(C)** were determined. Data are shown as mean ± SEM and statistical analyses was performed by two-way ANOVA with mixed effect analysis for Panels A and B, and with one-way ANOVA with Bonferroni's multiple comparison test (***)P <0,0001).

Table S1. List of 142 species from which the ornithine transcarbamylase (ornithine carbamoyltransferase, OTCase) primary sequence were obtained. These sequences were then aligned to generate the logo representation of Figure S2.

Entry	Entry name	Status	Protein names	Gene names	Organism	Length
P18186	OTC_BACSU	reviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	argF BSU11250	Bacillus subtilis (strain 168)	319
O19072	OTC_PIG	reviewed	Ornithine carbamoyltransferase, mitochondrial (EC 2.1.3.3) [OrnithineOTC]	argF NMBH4476_0658 NMH_2186	Sus scrofa (Pig)	328
Q05M07	OTC_BORAP	reviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	arcB BAPK0_0897 BaPKo_0870	Borrelia afzelii (strain PK0)	328
EGN013	OTC_NEIMH	reviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	argF NMBH4476_0658 NMH_2186	Neisseria meningitidis serogroup B / serotype 15 (strain H44/76)	331
P11803	OTC_EMENI	reviewed	Ornithine carbamoyltransferase, mitochondrial (EC 2.1.3.3) [OrnithineargB AN4409]	argB AN4409	Microascus nidulans (strain FGSC A4 / ATCC 38163 / CBS 112.46 / NRRL 194 / M139) (Aspergillus nidulans)	359
OT55601	OTC_OTCSE	reviewed	Ornithine carbamoyltransferase, catabolic [OTCase] (EC 2.1.3.3)	arcB SA2434	Staphylococcus aureus (strain Mu50) / ATCC 700699	359
P31317	OTC_SCHPO	reviewed	Ornithine carbamoyltransferase, mitochondrial (EC 2.1.3.3) [OrnithineargS3 SPAC4G5.10]	argS3 SPAC4G5.10	Schizosaccharomyces pombe (strain 972 / ATCC 24843) (Fission yeast)	327
P11066	OTC_ASPNG	reviewed	Ornithine carbamoyltransferase, mitochondrial (EC 2.1.3.3) [OrnithineargB]	argB	Aspergillus niger	370
B1KX03	OTC_CLOMB	reviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	arcB CLK_1978	Clostridium botulinum (strain Loch Maree / Type A3)	333
P11725	OTC_MOUSE	reviewed	Ornithine carbamoyltransferase, mitochondrial (EC 2.1.3.3) [OrnithineOtc]	OTC	Mus musculus (Mouse)	354
P0CL21	OTC_COCIM	reviewed	Ornithine carbamoyltransferase, mitochondrial (EC 2.1.3.3) [OrnithineCMG_04084]	CMG_04084	Coccidioides immitis (strain RS) (Valley fever fungus)	349
P31226	OTC_LITCT	reviewed	Ornithine carbamoyltransferase, mitochondrial (EC 2.1.3.3) [Ornithine transcarbamylase] [OTCase]	OTC	Lithobates catesbeianus (American bullfrog) (Rana catesbeiana)	350
Q8CWM2	OTC2_STAES	reviewed	Ornithine carbamoyltransferase 2, catabolic [OTCase 2] (EC 2.1.3.3)	arcB2 SE_2216	Staphylococcus epidermidis (strain ATCC 12228)	335
Q9YH19	OTC_CHICK	reviewed	Ornithine carbamoyltransferase, mitochondrial (EC 2.1.3.3) [OrnithineOTC]	OTC	Gallus gallus (Chicken)	354
O50039	OTC_ARATH	reviewed	Ornithine carbamoyltransferase, chloroplastic (EC 2.1.3.3) [Ornithine OTC]	OTC	Arabidopsis thaliana (Mouse-ear cress)	375
A6Q668	OTC_STAAE	reviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	argF NWMN_1078	Staphylococcus aureus (strain Newman)	333
Q1JF21	OTC_STRDP	reviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	arcB MGAS10270_Spy1288	Streptococcus pyogenes serotype M2 (strain MGAS10270)	337
Q8G598	OTC2_LACHI	reviewed	Ornithine carbamoyltransferase, catabolic [OTCase] (EC 2.1.3.3)	arcB	Lactobacillus hilgardii	343
Q04391	OTC_ECOLU	reviewed	Ornithine carbamoyltransferase subunit I [OTCase-1] (EC 2.1.3.3)	argI b4254 JW4211	Escherichia coli (strain K12)	334
Q3J4K5	OTC_RHOSA	reviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	argF RHOSA_05910 RSP_2009	Rhodobacter sphaeroides (strain ATCC 17023 / 2.4.1 / NCIB 8253 / DSM 158)	308
Q8ET05	OTC_OCEIH	reviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	argF OB0460	Oceanobacillus ihayensis (strain DSM 14371 / CIP 107618 / JCM 11309 / KCTC 3954 / HT8831)	322
Q8C4U1	OTC1_STAES	reviewed	Ornithine carbamoyltransferase 1, catabolic [OTCase 1] (EC 2.1.3.3)	arcB1 SE_0103	Staphylococcus epidermidis (strain ATCC 12228)	332
P14995	OTC_PACTA	reviewed	Ornithine carbamoyltransferase, mitochondrial (EC 2.1.3.3) [OrnithineOTC]	OTC	Pachysolen tannophilus (Yeast)	347
OT55603	OTC_OTCSE	reviewed	Ornithine carbamoyltransferase 1, catabolic [OTCase 1] (EC 2.1.3.3)	arcB1 gbs2085	Streptococcus agalactiae serotype III (strain NEM316)	325
P00481	OTC_RAT	reviewed	Ornithine carbamoyltransferase, mitochondrial (EC 2.1.3.3) [OrnithineOtc]	OTC	Rattus norvegicus (Rat)	354
P65605	OTC2_STRA3	reviewed	Ornithine carbamoyltransferase 2, catabolic [OTCase 2] (EC 2.1.3.3)	arcB2 gbs2124	Streptococcus agalactiae serotype III (strain NEM316)	337
R8A010	OTC_SHEEP	reviewed	Ornithine carbamoyltransferase, mitochondrial (EC 2.1.3.3) [OrnithineOTC]	OTC	Ovis aries (Sheep)	355
BSXMC1	OTC_STR2P	reviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	arcB Spy49_1195C	Streptococcus pyogenes serotype M49 (strain NZ131)	337
Q48296	OTC_HALS5	reviewed	Ornithine carbamoyltransferase, catabolic [cOTCase] (EC 2.1.3.3)	argF VNG_6315G	Halobacterium salinarum (strain ATCC 700922 / JCM 11081 / NRC-1) (Halobacterium halobium)	295
OT5C2P2	OTC_OTCSE	reviewed	Ornithine carbamoyltransferase 1, catabolic [OTCase 1] (EC 2.1.3.3)	arcB1 SAG2216	Streptococcus agalactiae serotype V (strain ATCC BAA-611 / 2603 V/R)	335
Q9PNU6	OTCA_CAMIE	reviewed	Ornithine carbamoyltransferase, anabolic [OTCase] (EC 2.1.3.3)	argF G09942	Campylobacter jejuni subsp. jejuni serotype O:2 (strain ATCC 700819 / NCTC 11168)	306
Q43814	OTC_PEA	reviewed	Ornithine carbamoyltransferase, chloroplastic (EC 2.1.3.3) [Ornithine IARGF]	IARGF	Pisum sativum (Garden pea)	375
Q20247	OTC1A_PESH5	reviewed	Ornithine carbamoyltransferase 1, anabolic [OTCase 1] (EC 2.1.3.3) [OArgF]	OTC	Pseudomonas savastanoi pv. phaseolicola (Pseudomonas syringae pv. phaseolicola)	306
AAUV03	OTC_STR5Y	reviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	arcB SSUO5_0626	Streptococcus suis (strain 05ZVH33)	337
P00480	OTC_HUMAN	reviewed	Ornithine carbamoyltransferase, mitochondrial (EC 2.1.3.3) [OrnithineOTC]	OTC	Homo sapiens (Human)	354
OT55604	OTC_OTCSE	reviewed	Ornithine carbamoyltransferase 1, catabolic [OTCase 1] (EC 2.1.3.3)	arcB1 SAG2216	Streptococcus agalactiae serotype V (strain ATCC BAA-611 / 2603 V/R)	335
Q9N1U7	OTC_BOVIN	reviewed	Ornithine carbamoyltransferase, mitochondrial (EC 2.1.3.3) [OrnithineOTC]	OTC	Bos taurus (Bovine)	354
Q2YPH2	OTC_BRAU2	reviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	argB BAB1_0332	Bruella abortus (strain 2308)	312
Q972M1	OTC_SACS2	reviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	argF SS00871	Saccharolobus solfataricus (strain ATCC 35092 / DSM 1617 / JCM 11322 / F2) [Sulfolobus solfataricus]	307
P44770	OTC_HAEIN	reviewed	Ornithine carbamoyltransferase, catabolic [OTCase] (EC 2.1.3.3)	OTC	Haemophilus influenzae (strain ATCC 51907 / DSM 11121 / KW20 / Rd)	334
P68746	OTC2_PSEF5	reviewed	Ornithine carbamoyltransferase 2, phenol/oxalin-insensitive [OTCase argK]	argK	Pseudomonas syringae pv. actinidiae	327
OT57805	OTC_TKRAH	reviewed	Ornithine carbamoyltransferase, mitochondrial (EC 2.1.3.3) [Ornithine transcarbamylase] [OTCase]	OTC	Trametes hirsuta (White-rot fungus) (Coriolus hirsutus)	375
P68747	OTC2A_PESH5	reviewed	Ornithine carbamoyltransferase 2, anabolic [OTCase 2] (EC 2.1.3.3) [OArgF]	OTC	Pseudomonas savastanoi pv. phaseolicola (Pseudomonas syringae pv. phaseolicola)	327
Q1J4R4	OTC_STRPP	reviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	arcB MGAS2096_Spy1292	Streptococcus pyogenes serotype M12 (strain MGAS2096)	337
P09690	OTC2_ECOLU	reviewed	Ornithine carbamoyltransferase subunit F [OTCase-2] (EC 2.1.3.3)	argF b0273 JW0266	Escherichia coli (strain K12)	334
P65606	OTC2_STRA5	reviewed	Ornithine carbamoyltransferase 2, catabolic [OTCase 2] (EC 2.1.3.3)	arcB2 SAG2165	Streptococcus agalactiae serotype V (strain ATCC BAA-611 / 2603 V/R)	337
PC2CF3	OTC_ASPFN	reviewed	Ornithine carbamoyltransferase, mitochondrial (EC 2.1.3.3) [OrnithineargI arg-1 AT6G_05492]	arg-1 AT6G_05492	Aspergillus terreus (strain NIH 2624 / FGSC A1156)	361
P11724	OTC_PSEAE	reviewed	Ornithine carbamoyltransferase, anabolic [OTCase] (EC 2.1.3.3)	argF AR3537	Arthropis sinensis (Chinese alligator)	305
Q51742	OTC_PYRFO	reviewed	Ornithine carbamoyltransferase, anabolic [OTCase] (EC 2.1.3.3)	argF P0504	Pyrococcus furiosus (strain ATCC 43587 / DSM 3638 / JCM 8422 / Vc1)	315
AA24L2X5J4	AA02L2X5J4_9BACT	unreviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	OTC	Candidatus Gastranaerophilus sp. (ex Termes propinquus)	310
DBLH5	DBLH5_ECTS1	unreviewed	Ornithine transcarbamylase (EC 2.1.3.3)	OTC	Ectocarpus siliculosus (Brown alga) (Conferva siliculosa)	320
AA367YH05	AA0367YH05_9ASCO	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Candida viswanathii	343
AA04X1J355	AA04X1J355_PIG	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Sus scrofa (Pig)	354
AA024LQ222	AA024LQ222_TURTR	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Tursiops truncatus (Atlantic bottlenose dolphin) (Delphinus truncatus)	354
AA0340WR1A1	AA0340WR1A1_LIPVE	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Lipotes vexillifer (Yangtze river dolphin)	354
AA02Y9FRK9	AA02Y9FRK9_PHYMC	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Physeter macrocephalus (Sperm whale) (Physeter catodon)	354
AA02Y9GIU7	AA02Y9GIU7_NEOSC	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Neomonachus schauinslandi (Hawaiian monk seal) (Monachus schauinslandi)	354
AA01428FN0	AA01428FN0_9GAMM	unreviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	OTC	Endozoichomonas montiporae CL-33	338
AA0284SF3F	AA0284SF3F_STYPI	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Slyophilora pistillata (Smooth cauliflower coral)	376
AA01179S18	AA01179S18_ALLSI	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Trachymys sinensis (Chinese slider turtle)	354
Q9IAU8	Q9IAU8_TRASE	unreviewed	Ornithine transcarbamylase	OTC	Trachymys scripta elegans (Red-eared slider turtle)	354
WSUIC6	WSUIC6_ICTPU	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Ictalurus punctatus (Channel catfish) (Silurus punctatus)	352
AA01Y2FGM4	AA01Y2FGM4_PROLT	unreviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	OTC	Protomyces lactucaedebilis	353
AA021ZFB17	AA021ZFB17_9EURO	unreviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	OTC	Aspergillus candidus	365
M9ME20	M9ME20_PSEAF	unreviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	OTC	Pseudozyma acetivorans (strain T-34) (Yeast) (Candida antarctica)	382
AA024JUSV56	AA024JUSV56_LFEVW	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Trachymys sinensis (Chinese slider turtle)	354
AA03Q7SV11	AA03Q7SV11_VUIUV	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Vulpes vulpes (red fox) (Canis vulpes)	354
H2VP80	H2VP80_PONAB	unreviewed	Ornithine carbamoyltransferase	OTC	Pongo abelii (Sumatran orangutan) (Pongo pygmaeus abelii)	354
F1MNG5	F1MNG5_BOVIN	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Bos taurus (Bovine)	354
H2QV67	H2QV67_PANTR	unreviewed	Ornithine carbamoyltransferase	OTC	Pan troglodytes (Chimpanzee)	354
AA0152Z6E8	AA0152Z6E8_ERIEU	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Ermacrus europaeus (Western European hedgehog)	354
AA02Y9M4V18	AA02Y9M4V18_DELE1	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Delphinus phocaenoides (Beluga whale)	354
AA0480E1W6	AA0480E1W6_PIG	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Sus scrofa (Pig)	354
ISTWB5	ISTWB5_ASP03	unreviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	A03042_05393	Aspergillus oryzae (strain 3.042) (Yellow koji mold)	372
AA01C7M8Y7	AA01C7M8Y7_GRIFR	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Grifola frondosa (Maitake) (Polyporus frondosus)	342
G3AUC8	G3AUC8_SPAAP	unreviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	OTC	Spathospora passalidarum (strain NRRL Y-27907 / 11-Y1)	331
AA024ZHN29	AA024ZHN29_3HYPO	unreviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	OTC	Trichoderma parviseptum	353
AA0112VQ21	AA0112VQ21_9ASCO	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Aspergillus ruber (strain DSM 1968)	354
AA013TCN33	AA013TCN33_3HYPO	unreviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	OTC	Trichoderma guizhouense	352
AA028Y8289	AA028Y8289_HUMAN	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Homo sapiens (Human)	130
AA03L6FRW5	AA03L6FRW5_MAZE	unreviewed	Ornithine carbamoyltransferase, chloroplastic	OTC	Zea mays (Maize)	72
AA02G9100	AA02G9100_9LAMI	unreviewed	Ornithine carbamoyltransferase [OTCase] (EC 2.1.3.3)	OTC	Handroanthus impetiginosus	261
AA016H1D8	AA016H1D8_NICAT	unreviewed	Ornithine carbamoyltransferase, chloroplastic	OTC	Nicotiana attenuata (Coyote tobacco)	376
F8H550	F8H550_STRES	unreviewed	Ornithine carbamoyltransferase, catabolic [OTCase] (EC 2.1.3.3)	OTC	Streptococcus salivarius (strain 57.1)	354
F1NUG7	F1NUG7_CHICK	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Gallus gallus (Chicken)	354
Q9IAV1	Q9IAV1_9CHON	unreviewed	Ornithine transcarbamylase	OTC	Raja sp.	353
AA0178W4A4	AA0178W4A4_ARATH	unreviewed	OTC	AX17_At1g69770	Arabidopsis thaliana (Mouse-ear cress)	375
F1RXQ9	F1RXQ9_PIG	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Sus scrofa (Pig)	336
AA0347N1C8	AA0347N1C8_CALLUR	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Callorhynchus (Northern fur seal)	354
AA0341D9D3	AA0341D9D3_9EURO	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Neophocaena asiatorientalis asiatorientalis (Yangtze finless porpoise) (Neophocaena phocaenoides subsp. asiatorientalis)	343
AA02U3VW86	AA02U3VW86_ODORO	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Odobenus rosmarus divergens (Pacific walrus)	354
AA03Q7XZ17	AA03Q7XZ17_URSAR	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Ursus arctos horribilis	354
AA038329Y7	AA038329Y7_BALAS	unreviewed	Ornithine carbamoyltransferase, mitochondrial isoform X2	OTC	Balaenoptera acutorostrata scammonii (North Pacific minke whale) (Balaenoptera davidsoni)	299
AA038329Y1	AA038329Y1_BALAS	unreviewed	Ornithine carbamoyltransferase, mitochondrial isoform X1	OTC	Balaenoptera acutorostrata scammonii (North Pacific minke whale) (Balaenoptera davidsoni)	354
AA0384C5C2	AA0384C5C2_URSMA	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Ursus martimus (Polar bear) (Thalictos martimus)	354
Y8NCN3	Y8NCN3_PHYMC	unreviewed	Ornithine carbamoyltransferase, mitochondrial (Fragment)	OTC	Ophiophagus hannah (King cobra) (Naja hannah)	354
Q9IAV0	Q9IAV0_SCUN	unreviewed	Ornithine transcarbamylase	OTC	Sceloporus undulatus (Eastern fence lizard) (Stellio undulatus)	356
AA01V4C89	AA01V4C89_PATFA	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Patagonia fasciata monilis	335
Q9IAU7	Q9IAU7_PYTRG	unreviewed	Ornithine transcarbamylase (Fragment)	OTC	Python regius (Ball python) (Boa regia)	163
Q5K711	Q5K711_CHICK	unreviewed	Ornithine transcarbamylase (Fragment)	OTC	Gallus gallus (Chicken)	18
Q95959	Q95959_MYCSP	unreviewed	Ornithine transcarbamylase [EC 2.1.3.3] (Fragment)	OTC	Mycoplasma sp.	226
Q8R1A8	Q8R1A8_MOUSE	unreviewed	Ornithine carbamoyltransferase, mitochondrial (Otc protein)	OTC	Ch. (Mouse)	354
AA03Q0D8F7	AA03Q0D8F7_MESAU	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Mesocricetus auratus (Golden hamster)	251
AA03L7H3E8	AA03L7H3E8_CRIGR	unreviewed	OTC	CGPICR_014091	Cricetus griseus (Chinese hamster) (Cricetus barabensis griseus)	235
Q5M897	Q5M897_RAT	unreviewed	Otc protein	OTC	Rattus norvegicus (Rat)	350
AA0153G0L8	AA0153G0L8_DIPOR	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Dipodomys ordii (Ord's kangaroo rat)	355
G58M029	G58M029_HETGA	unreviewed	Ornithine carbamoyltransferase, mitochondrial	OTC	Heteropogon glaber (Naked mole rat)	354
A1D528	A1D528_LACBR	unreviewed	Ornithine transcarbamylase (Fragment)	OTC	Lactobacillus buchneri	59
BPV30	BPV30_LACBR	unreviewed	Ornithine transcarbamylase (Fragment)	OTC	Lactobacillus brevis	267
A1D525	A1D525_LACBR	unreviewed	Ornithine transcarbamylase (Fragment)	OTC	Lactobacillus brevis	60
A1D522	A1D522_PEPDE	unreviewed	Ornithine transcarbamylase (Fragment)	OTC	Pediococcus pentoseus	55
AA075HZ36						