Electronic Supplementary Material (ESI) for Metallomics

Mutation in the CPC motif-containing 6th transmembrane domain affects intracellular localization, trafficking and copper transport efficiency of ATP7A protein in *mosaic* mutant mice – an animal model of Menkes disease

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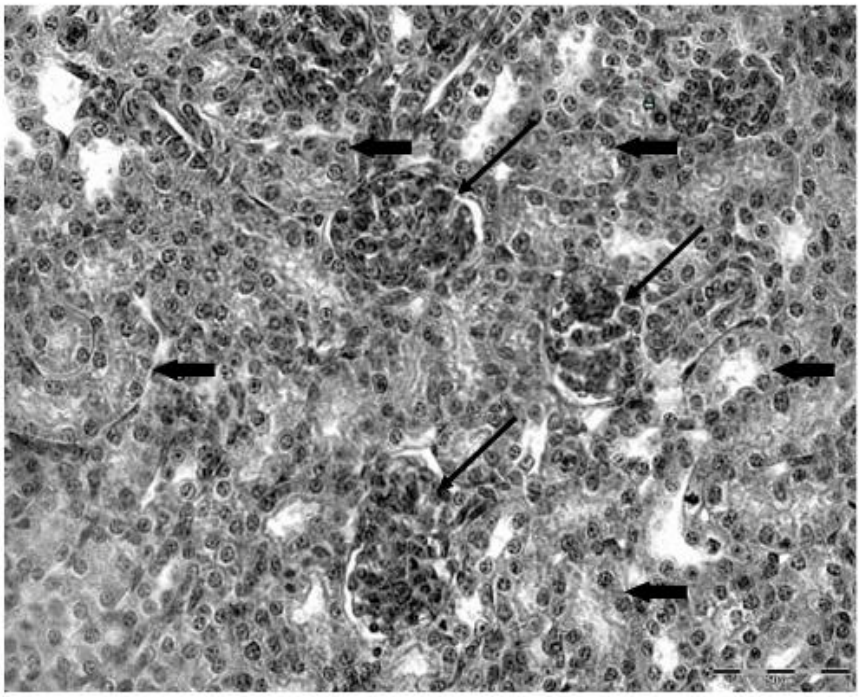

Supplementary Figure 1. Section through the renal cortex of the 14-day-old mouse, H&E stained. Renal cortex is composed from the renal tubules (wide arrows) and renal glomeruli (arrows). Corresponding region of the kidney is shown in the Figure 3.

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Mus musculus                  PETLREAIEDMGFDAALPADMKEPLVVIAQPSLE
Rattus norvegicus             PEPLREAIEDMGFDAVLPADMKEPLVVIAQPSLE
Cricetulus griseus            PETLREVIEDMGFDAVLP-DMSEPLVVIAQPSLE
Homo sapiens                  PETLRGAIEDMGFDATLS-DTNEPLVVIAQPSSE
Nomascus leucogenys           PETLRGAIEDMGFDATLS-DTNEPLVVIAQPSSE
Pongo abelii                  PETLRGAIEDMGFDATLS-DMNEPLVVIAQPSSE
Macaca mulatta                PETLRGAIEDMGFDATLS-DTNEPLVVIAQPSSE
Callithrix jacchus            PETLREAIEDMGFDATLS-DVNEPLVVIAQPSSE
Oryctolagus cuniculus         PETLREAIEDMGFDATLS-DMNEPLVVIAQPSSD
Bos taurus                    PETLREAIENMGFDASLS-DTNEPLVVIAQPSSE
Sus scrofa                    PETLREAIEDLGFDASLS-DTNEPLVVIAQSSSE
Canis lupus familiaris        PETLREAIEDMGFDAALS-DINEPLVIIGQTSSE
Equus caballus                PETLRKAIEDMGFDATLS-DTNEPLVVIAQPSSE
Loxodonta africana            PETLREAIENMGFDATLP-DVNEPSVLITQPSLE
Ailuropoda melanoleuca        PEALREAIEDMGFDAALS-DTNEPLVIIAQTSSE
Ornithorhynchus anatinus      PETLREVIEDMGFDASLS-EMMEPVVVITQPSLD
Monodelphis domestica         PETLREAIEDMGFDATLK----------DSDKET
Gallus gallus                 PEDLRSSIENMGFDASLPEKTELPVGITQPTSKE
Meleagris gallopavo           PEDLRSSIENMGFDASLPEKTELPVGVTQSTPKE
Anolis carolinensis           PDDLRNAIEDMGFDASLTGTPSPDPVVLITQPSQ
Oreochromis niloticus         PEELREAIEDMGFDAFLPETNSLLPSPHPLSSKS
Sparus aurata                 PEELREAIEDMGFDAFLPGTNSLLPEPDRSLSKS
Opsanus beta                  PEELREAIEDMGFDAFLPETNSLLPVSDSSLSKS
Tetraodon nigroviridis        PAELREAIEDMGFDAFLPATNSLLPEPACKRSNS
Danio rerio                   PEELRAAIEDMGFDAFLPETNSLVPSVVKSPSPS
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Supplementary Figure 2. Multiple alignment of the ATP7A protein sequence. Highlighted sequence represents an additional alanine (and surrounding amino acids) resulting from and additional CAG sequence in the *Atp7a* transcript, as published previously [34].

Supplementary Table 1. Primers used for mutation analysis of the *Atp7a* gene

Exon number	Primers sequence
Atp7a E1FP Atp7a E1RP	GTGGACCATTTCCCTAGTCG CCCCAGCTTAAAGTGGTGAC
Atp7a E2FP Atp7 E2RP	CATTTTCAACTGGAGGCATGT AGCCAACCTCCAGCCCTA
Atp7a E3FP Atp7a E3RP	GGAGGACTTGGTTCTTGATAATTG CATACTTGGGAAGAAAGCAGAAAG
Atp7a E4FP1 Atp7a E4RP1 Atp7a E4FP2 Atp7a E4RP2	TTGACTCTGAAGGGGACCAG AGAGCTGGCGGTACTTTCAA TCTTTAGAGAATAGGTCAGCCATTG AAAGGTGAAACTTCAGTGACTGTG
Atp7a E5FP Atp7a E5RP	GGTGTGGCTGTGACTTTTGA CAAAGCATGGCCATAAAACA
Atp7a E6FP Atp7a E6RP	GAAGGCACGTGAAGATGGTAA AAGTTTTCTGTGCAGGCCTCT
Atp7a E7FP Atp7a E7RP	AGGCAATCCTGTGCTACGAT CCAGATGTGATTCCAGAAGGT
Atp7a E8FP Atp7a E8RP	GGACAGCTTCATGCTTCCAT ACCTCCACAGTCTCCTGCAT
Atp7a E9FP Atp7a E9RP	AGCCTGAGTGAGATGGTTTCTC TCCCTCAGCCCTCACTTTTA
Atp7a E10FP Atp7a E10RP	TGAGGCTCGAGAACAGTGTTTA CATGAGCTCTGGGGGTACAT
Atp7a E11FP Atp7a E11RP	TGCAAAGGGGATCTGGTTTA CGAGGAGAAAACTCCCACAA
Atp7a E12FP Atp7a E12RP	TTTGGGCCAAGAATTTGTTC AGATTCCCACATTGCAGAGG
Atp7a E13FP Atp7a E13RP	TTGCAGTTTCCTCTGCAATG TTAACAAACCCCAGGAATGC
Atp7a E14FP Atp7a E14RP	AGTTGCATTCCTGGGGTTT AGCATTTCTCCAGCTCCTCA
Atp7a E15FP Atp7a E15RP	GGGTTGACCAGTGTTGCTTT GCCTAAGAACCACAGCCAAA
Atp7a E16FP Atp7a E16RP	TTGTGAAGCCAAAGTCCAGTT TTCAAAGACCAGCCCAGATT
Atp7a E17FP Atp7a E17RP	CCCAAAATTTGAAGTCCTGTCT ACTGTGGTCAGGATGCATTG

Atp7a E18FP Atp7a E18RP	GAACTTGCCTCATGAGATGGA TCTGCCTCTCTCAGGTTTGC
Atp7a E19FP Atp7a E19RP	TGAGTTAAACCAGTTCTCAACAGC GGTCCTTGTACCATATCATCCTG
Atp7a E20FP Atp7a E20RP	TGAGCTCAGCATTATTTGCAG CCACAGGACGTACCCACTCT
Atp7a E21FP Atp7a E21RP	AATTCCCAGTTTGTCCCACA CGGTAAGCCTGTTCTTCCAA
Atp7a E22FP Atp7a E22RP	ACAGCAGCACACATCTCCAC ACACACATGCACGTCAGACA
Atp7a E23FP1 Atp7a E23RP1 Atp7a E23FP2 Atp7a E23RP2	GTTTTGTAGCAGTAGCCACTTTTG GAGCATGAGATTGACATAAGATCC GGGAAGATGATGACACCACA AAAACCATTGCTCCCCAGT