

BTHS patient	Mutation
TAZ001	c.153C>G
TAZ002	c.239-1G>A
TAZ006	c.170G>T
TAZ013	c.110-1G>C

Table S1. *The four TAZ mutations examined in this study.* Patients were diagnosed with Barth Syndrome (BTHS) based on (ML)CL analysis (Houtkooper et al., 2009). Patient 1 has nonsense mutation, patient 6 has a missense mutation, patients 2 and 13 have a mutation in splice acceptor sites of exon 3 and exon 2 respectively.

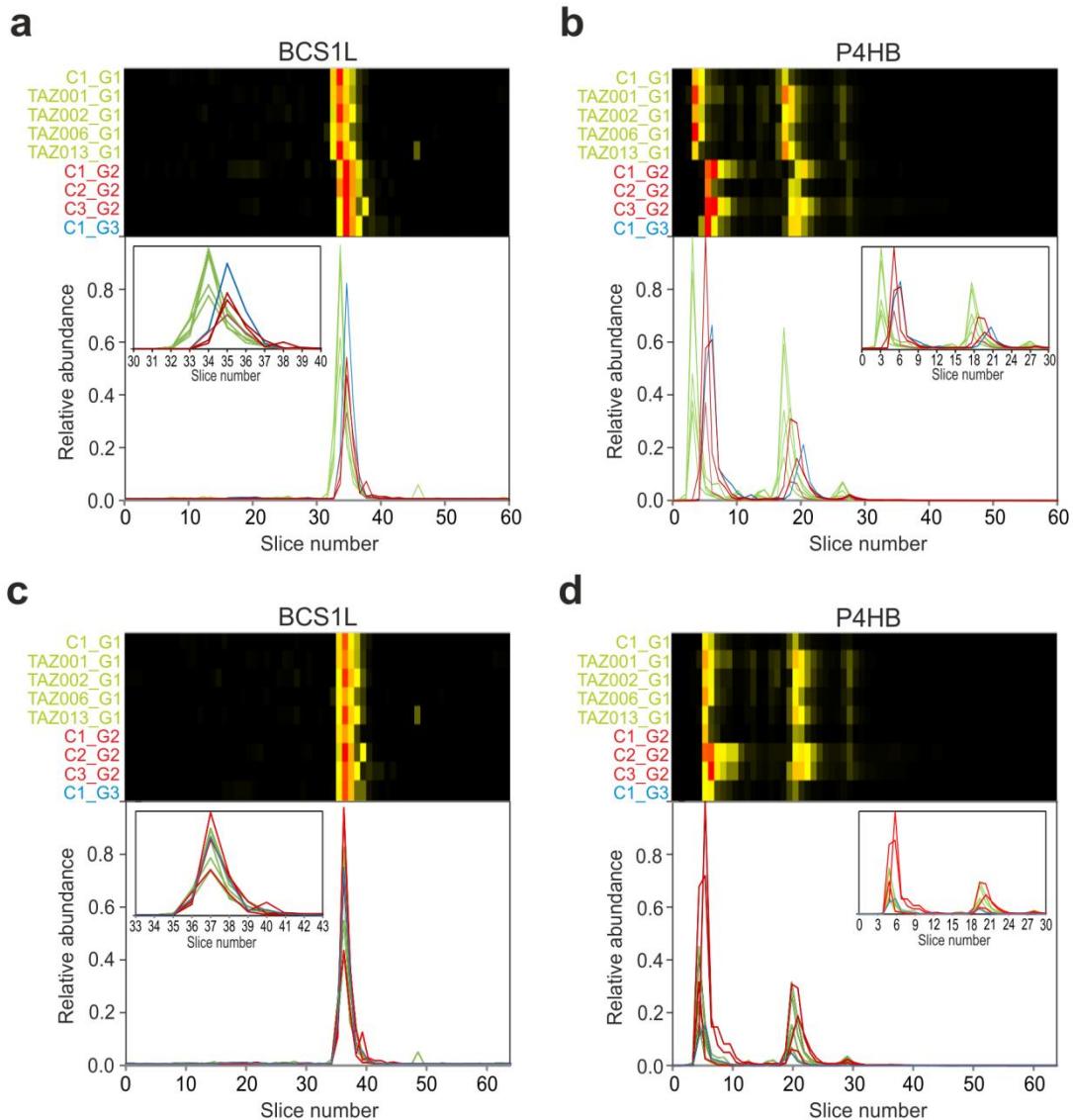


Figure S1. Migration patterns of the P4HB and BCS1L proteins before (a, b) and after (c, d) COPAL alignment. The color code of the gels is the same as in Figure 2. Slices inserted in the complexomes, based on the migration profiles of all mitochondrial proteins, correctly align the peaks of two individual proteins. The insets zoom into the region of the aligned peaks.

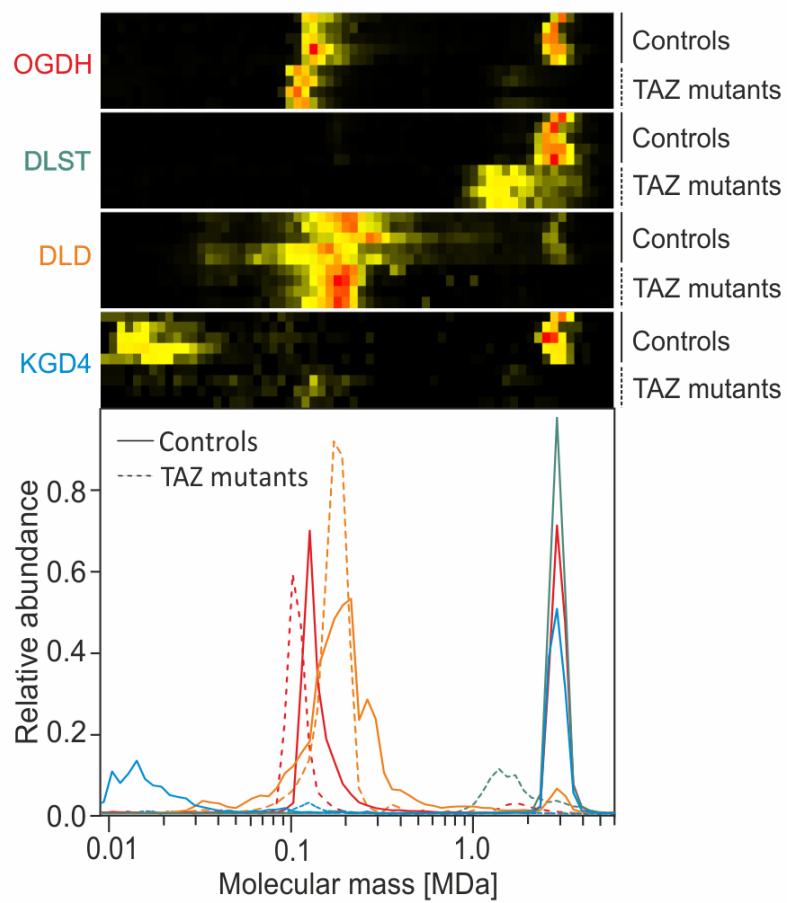


Figure S2. 2-oxoglutarate dehydrogenase complex is disassembled in TAZ mutation fibroblasts.

The 2-oxoglutarate dehydrogenase complex, consisting of dihydrolipoyllysine-succinyltransferase (DLST), dihydrolipoyl-dehydrogenase (DLD), oxoglutarate dehydrogenase (OGDH) and KGD4 (MRPS36) (Heublein et al., 2014), is markedly affected by the mutations. The 3 MDa complex was essentially absent in mitochondria from the BTHS fibroblasts. Instead small amounts of two complexes at ~2.0 MDa containing DLST and OGDH and at ~1.5 MDa containing DLST were observed. While DLST was only detectable in these two large complexes, a similar large reservoir of trimeric DLD (~150 kDa) and monomeric OGDH (~110 kDa) was present in both control and BTHS mitochondria.

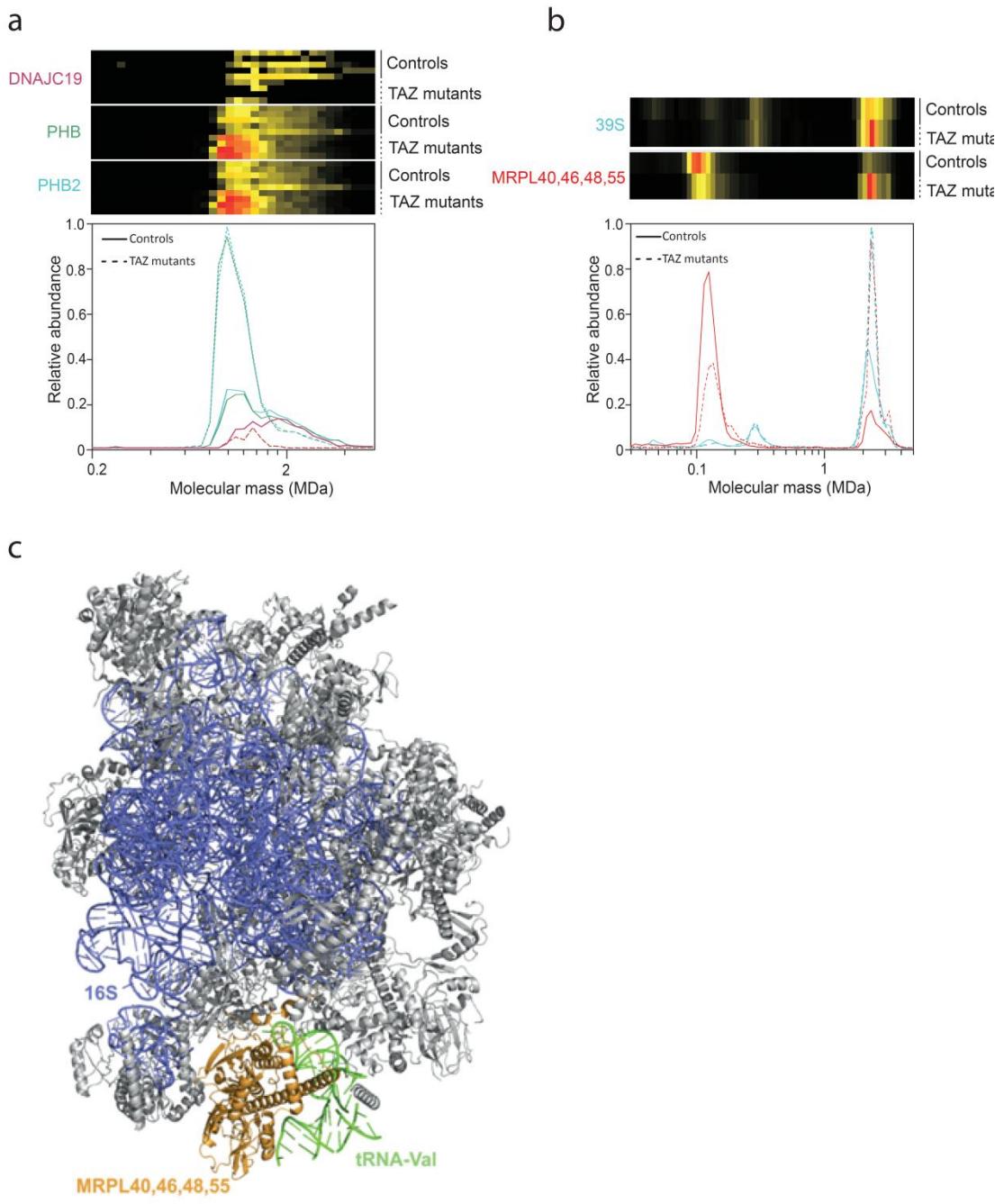


Figure S3. Significant effects of the TAZ mutation on prohibitin and the 39S subunit of the mitochondrial ribosome

a) The prohibitins (PHB, PHB2) forming a complex of ~1.2 MDa exhibited a ~4-fold increase in abundance while a shoulder in the range from 1.8 - 4.0 MDa was slightly reduced. This corresponded to marked decrease in the abundance of DNAJC19 in the same mass range. Prohibitin is known to take part in CL remodeling in association with DNAJC19 (Richter-Dennerlein et al., 2014).

b) The average of all detected proteins of the large 39S subunit of the mitochondrial ribosome from all samples exhibited a higher and narrower peak at ~ 2.5 MDa in the BTHS profiles as compared to the controls. An explanation for this shift of the 39S subunit is that the tRNA binding subcomplex of MRPL40, 46, 48, 55 has shifted from being mainly a separate subcomplex in the controls to being part of the 39S subunit in the BTHS mitochondria. c) Structure of the 39S subunit (Amunts et al., 2015) in which the tRNA binding proteins MRPL40, 46, 48, 55 have been highlighted. Abundance profiles of the (sub)complexes have been rescaled to include them in the same panel.

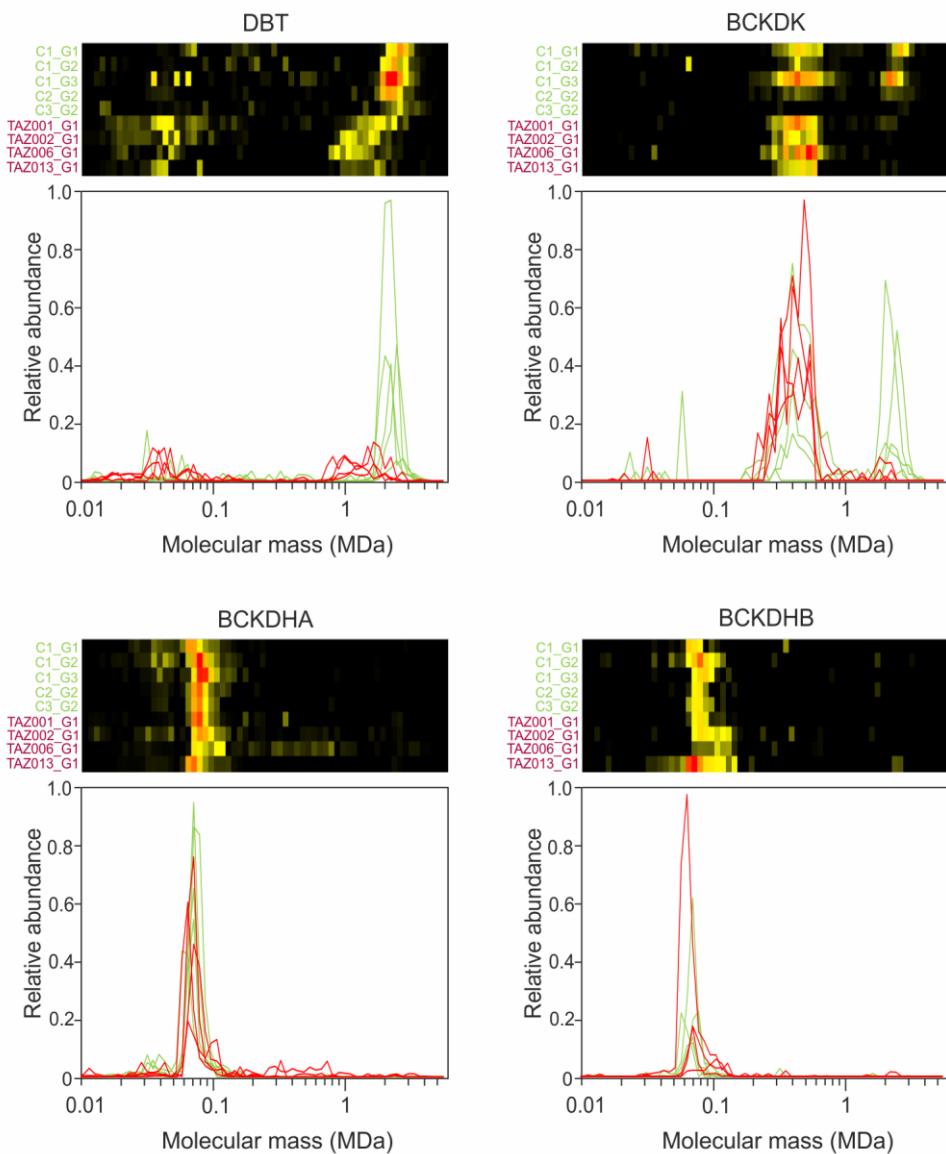


Figure S4 Large variation in the presence of the branched-chain amino acid dehydrogenase

The branched chain dehydrogenase that was found to be disassembled (loss of the peak at ~3 MDa) in BTHS mitochondria compared to one control Chatzispyrou *et al.* (submitted) was not detected as significantly changed in the automated analysis using COPAL and five controls. Closer inspection of the results showed that none of the individual components obtained a high Hausdorff score (Table S3) because there was a large variation in the presence of the ~3 MDa (DBT and BCKDK, upper panels) assembled complex among the controls and because the dominant peak remained at the same mass (BCKDHA, BCKDHB).

Supplementary methods. The CORUM database

Manual inspection of the human mitochondrial protein complex list in CORUM revealed a few known complexes that were absent from “human” complexes in the database but that were present in other mammalian species. These were added to the list used: 2-oxoglutarate dehydrogenase complex, cytochrome *bc_I*-complex, F_IF_O-ATP synthase - IF1 (inhibitor protein) complex, respiratory chain supercomplex (complex I, III, IV) and succinate dehydrogenase (complex II). Furthermore, we added MIC12/QIL1 to the MICOS complex, MTX3 to the MIB complex (Huynen et al., 2016) and we adapted the mitochondrial ribosome subunits based on recent literature (Amunts et al., 2015; Greber et al., 2015).

References supplementary material

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Table S4) Mitochondrial proteins sorted by the Hausdorff effect size of a mutation in the taffazin gene on their abundance profile. See main text for details.

geneID	Symbol	Hausdorff effect size
37693993	NME3	5,698269487
42476281	VDAC2	5,545355702
48255924	NDUFV3	5,04861963
4507879	VDAC1	4,984911691
4505773	PHB	4,92687829
154354966	IMMT	4,59386202
8923390	CHCHD3	4,491541016
22202629	AIFM1	4,444893553
221307584	PHB2	4,441212599
157388993	SYNJ2BP	4,181426523
13129148	APOO	4,079813398
61743952	AGPAT5	4,018977422
40807491	ACSL1	3,898963304
13027606	MRPL34	3,794394658
25188179	VDAC3	3,765905721
19718741	OSBPL1A	3,610092226
222136639	MTHFD1	3,565611237
28872734	MRPL43	3,520692071
224831243	OPA1	3,462036089
62244044	OCIAD2	3,440127521
156105685	ABCB8	3,409320726
19923748	DLST	3,321998369
21361497	ACAD9	3,301940088
7661806	MRPL15	3,295692041
21265070	MRPL2	3,276890602
7657347	MTCH2	3,046349164
154354962	IMMT	2,948902832
188497754	HK1	2,877320611
323276668	MINOS1	2,871036321
48526509	TIMM50	2,866989413
259013537	DTYMK	2,793277466
49169828	TOMM5	2,790566535
156104864	ACOX3	2,65437332
285002233	GPD2	2,626824726
281604151	PGAM5	2,623024178
186928854	MRPS31	2,590513314
31563366	MRPL55	2,589550244
4758714	MGST3	2,568657415
14165270	MRPL13	2,533693851
45387955	C19orf70	2,531946514
5729937	MTX2	2,525987211
31652226	MRPL21	2,511714286
38505218	ACAD11	2,511145377
32698876	CMC1	2,501802923
333470683	MRPL45	2,498901258
225543166	SAMM50	2,463099397
117168279	MGARP	2,425518702
4757732	AIFM1	2,416757658
7705704	GSTK1	2,305584622
149193319	GRSF1	2,301245964
13027604	MRPS34	2,297998013
4506001	PPOX	2,292038833

21265080	MRPL18	2,279140122
27886582	MRPL39	2,229405238
169636418	MRPL38	2,192239238
56699456	ISCU	2,165575258
7524346	AK2	2,158191979
38016911	STOM	2,139745602
268370293	VARS2	2,117446823
4502987	COX7A1	2,087716194
22547134	MRPL37	2,079076282
5174723	TOMM40	2,068577487
194097323	ECHS1	2,0671733
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223468571	MRPL23	2,029085422
13376717	OPA3	2,024908878
96975135	RAB24	2,021678568
6912482	LETM1	2,020883856
4557303	ALDH3A2	2,011397857
261862352	SHMT2	2,009578668
7661910	EMC2	2,007228771
51243059	MCAT	1,984742024
300192933	AFG3L2	1,9801032
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160420328	ISCA2	1,962979025
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4758484	GSTO1	1,945309237
38569423	ACLY	1,943222556
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282396088	COA7	1,927573911
13376617	PTGES2	1,914375448
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4557032	LDHB	1,894732444
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29789409	NDUFAF2	1,865014914
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4501853	ACAA1	1,843849565
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59710109	TIMMD1	1,834680831
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22547138	MRPL4	1,821260386
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51558774	ATP5S	1,707309359
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117606353	MSRB2	1,704470627
7705851	CHCHD2	1,697770514
116063536	COQ5	1,696085238
62988355	CCDC58	1,695975727
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4505143	ME1	1,149908986
226371731	HSDL1	1,147485494
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4758788	NDUFS3	1,14128779
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148491091	SLC25A24	1,135104939
13654274	FAHD1	1,133804328
155030240	TRNT1	1,129672692
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