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Adenine nucleotide translocase is involved in a mitochondrial coupling defect in MFN2-related Charcot–Marie–Tooth type 2A disease

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Titre	Adenine nucleotide translocase is involved in a mitochondrial coupling defect in MFN2-related Charcot–Marie–Tooth type 2A disease
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Auteur	Guillet, Virginie [1], Guegen, Naig [2], Verny, Christophe [3], Ferré, Marc [4], Homedan, Chadi [5], Loiseau, Dominique [6], Procaccio, Vincent [7], Amati-Bonneau, Patrizia [8], Bonneau, Dominique [9], Reynier, Pascal [10], Chevrollier, Arnaud [11]
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Résumé en anglais	Charcot–Marie–Tooth type 2A disease (CMT2A), a dominantly inherited peripheral neuropathy, is caused by mutations in MFN2, a mitochondrial fusion protein. Having previously demonstrated a mitochondrial coupling defect in CMT2A patients' fibroblasts, we here investigate mitochondrial oxygen consumption and the expression of adenine nucleotide translocase (ANT) and uncoupling proteins from eight other patients with the disease. The mitochondrial uncoupling was associated with a higher respiratory rate, essentially involving complex II proteins. Furthermore, a twofold increase in the expression of ANT led to the reduced efficiency of oxidative phosphorylation in CMT2A cells, suggesting that MFN2 plays a role in controlling ATP/ADP exchanges.
URL de la notice	http://okina.univ-angers.fr/publications/ua287 [20]
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