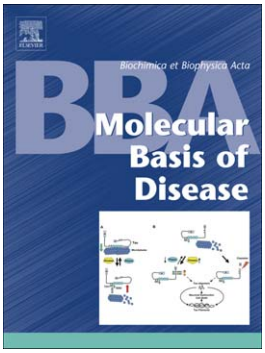

Bioenergetics dysfunction, mitochondrial permeability transition pore opening and lipid peroxidation induced by hydrogen sulfide as relevant pathomechanisms underlying the neurological dysfunction characteristic of ethylmalonic encephalopathy

Gabriela Miranda Fernandez Cardoso, Julia Tauana Pletsch, Belisa Parmeggiani, Mateus Grings, Nícolas Manzke Glanzel, Larissa Daniele Bobermin, Alexandre Umpierrez Amaral, Moacir Wajner, Guilhian Leipnitz

PII: S0925-4439(17)30202-8
DOI: doi:10.1016/j.bbadis.2017.06.007
Reference: BBADIS 64790

To appear in: *BBA - Molecular Basis of Disease*

Received date: 26 January 2017
Revised date: 16 May 2017
Accepted date: 10 June 2017

Please cite this article as: Gabriela Miranda Fernandez Cardoso, Julia Tauana Pletsch, Belisa Parmeggiani, Mateus Grings, Nícolas Manzke Glanzel, Larissa Daniele Bobermin, Alexandre Umpierrez Amaral, Moacir Wajner, Guilhian Leipnitz, Bioenergetics dysfunction, mitochondrial permeability transition pore opening and lipid peroxidation induced by hydrogen sulfide as relevant pathomechanisms underlying the neurological dysfunction characteristic of ethylmalonic encephalopathy, *BBA - Molecular Basis of Disease* (2017), doi:10.1016/j.bbadis.2017.06.007

Bioenergetics dysfunction, mitochondrial permeability transition pore opening and lipid peroxidation induced by hydrogen sulfide as relevant pathomechanisms underlying the neurological dysfunction characteristic of ethylmalonic encephalopathy

Gabriela Miranda Fernandez Cardoso[a,e], Julia Tauana Pletsch[a,e], Belisa Parmeggiani[a], Mateus Grings[a], Nícolas Manzke Glanzel[a], Larissa Daniele Bobermin[a], Alexandre Umpierrez Amaral[a,d], Moacir Wajner[a,b,c], Guilhian Leipnitz[a,b,*]

[a]Programa de Pós-Graduação em Ciências Biológicas: Bioquímica, Universidade Federal do Rio Grande do Sul, Rua Ramiro Barcelos, 2600-Anexo, CEP 90035-003, Porto Alegre, RS, Brazil

[b]Departamento de Bioquímica, Instituto de Ciências Básicas da Saúde, Universidade Federal do Rio Grande do Sul, Rua Ramiro Barcelos, 2600-Anexo, CEP 90035-003, Porto Alegre, RS, Brazil

[c]Serviço de Genética Médica, Hospital de Clínicas de Porto Alegre, Rua Ramiro Barcelos, 2350, CEP 90035-903, Porto Alegre, RS, Brazil

[d]Departamento de Ciências Biológicas, Universidade Regional Integrada do Alto Uruguai e das Missões, Avenida Sete de Setembro, 1621, CEP 99709-910, Erechim, RS, Brazil.

[e]Co-first authors. Both authors have contributed equally to this work.

*Corresponding Author: Guilhian Leipnitz, Departamento de Bioquímica, Instituto de Ciências Básicas da Saúde, Universidade Federal de Rio Grande do Sul, Ramiro Barcelos Street, 2600 – Anexo, CEP 90035-003, Porto Alegre, RS – Brazil. Phone: +55 51 3308-5551, fax: +55 51 3308-5540, e-mail: guilhian@ufrgs.br