

Acknowledgements

We thank the patients and their families who took part in this study; K.-D. Gerbitz, J.-J. Martin, M. de Visser, B. Mousson, L. D. Notarangelo, J. Villard and S. Zierz for access to their patient samples; E. Lamantea, B. Garavaglia, M. Rimoldi, A. Antonelli, F. Minoletti, F. Carrara and C. Gellera for their help and contribution to this work. This work was supported in part by a grant (to F.T.) from Telethon-Italia to the project "Molecular Analysis of Carnitine Palmitoyltransferase Deficiency." P.C. is a postdoctoral fellow supported by Telethon-Italia.

- McGarry, J.D. *et al.* Regulation of ketogenesis and the renaissance of carnitine palmitoyltransferase. *Diabetes/Metabol. Rev.* **5**, 271–284 (1989).
- Layzer, R.B. How muscles use fuel. *New Engl. J. Med.* **324**, 411–412 (1991).
- Felig, P. & Wahren, J. Fuel homeostasis in exercise. *New Engl. J. Med.* **293**, 1078–1084 (1975).
- DiDonato, S. Disorders of lipid metabolism affecting skeletal muscle. In *Myology* 2nd edn (eds Engel, A.G. & Franzini-Armstrong, C.) (McGraw-Hill, New York, in the press).
- Bremer, J. & Osmundsen, H. Fatty acid oxidation and its regulation. In *Fatty Acid Metabolism and Its Regulation* (ed. Numa, S.) 113–154 (Elsevier, New York, 1984).
- Bieber, L.L. Carnitine. *A. Rev. Biochem.* **57**, 261–283 (1988).
- Woeltje, K.F. *et al.* Inter-tissue and inter-species characteristics of the mitochondrial carnitine palmitoyltransferase enzyme system. *J. Biol. Chem.* **265**, 10714–10719 (1990).
- McKusick, V.A. *Mendelian Inheritance in Man* 10th edn (Johns Hopkins University Press, Baltimore, 1992).
- DiMauro, S. & Papadimitriou, A. Carnitine palmitoyltransferase deficiency. In *Myology* 1st edn (eds Engel, A.G. & Banker, B.Q.) 1697–1708 (McGraw-Hill, New York, 1986).
- DiMauro, S. & Melis-DiMauro, P. Muscle carnitine palmitoyltransferase deficiency and myoglobinuria. *Science* **182**, 929–931 (1973).
- Demaugre, F. *et al.* Hepatic and muscular presentations of carnitine palmitoyltransferase deficiency: two distinct entities. *Pediatr. Res.* **24**, 308–311 (1988).
- DiDonato, S. *et al.* Heterogeneity of carnitine-palmitoyltransferase deficiency. *J. Neurol. Sci.* **50**, 207–215 (1981).
- Demaugre, F. *et al.* Infantile form of carnitine palmitoyltransferase II deficiency with hepatomuscular symptoms and sudden death. Physiopathological approach to carnitine palmitoyltransferase II deficiencies. *J. Clin. Invest.* **87**, 859–864 (1991).
- Zinn, A.B. & Hoppel, C.L. An unusual form of carnitine palmitoyltransferase B (CPT-B) deficiency associated with neonatal cardiomyopathy and renal dysorganogenesis. *Am. J. Hum. Genet.* **49**, A109 (1991).
- Witt, D.R. *et al.* Carnitine palmitoyl transferase-type 2 deficiency: two new cases and successful prenatal diagnosis. *Am. J. Hum. Genet.* **49**, A109 (1991).
- Hug, G., Bove, K.E. & Soukup, S. Lethal neonatal multiorgan deficiency of carnitine palmitoyltransferase II. *New Engl. J. Med.* **325**, 1862–1864 (1991).
- Taroni, F. *et al.* Biochemical and molecular studies of carnitine palmitoyltransferase II deficiency with hepatocardiomyopathic presentation. In *New Developments in Fatty Acid Oxidation* (eds Coates, P.M. & Tanaka, K.) 521–531 (Wiley-Liss, New York, 1992).
- Finocchiaro, G. *et al.* cDNA cloning, sequence analysis and chromosomal localization of human carnitine palmitoyltransferase. *Proc. Natl. Acad. Sci. U.S.A.* **88**, 661–665 (correction 10981) (1991).
- Minoletti, F. *et al.* Localization of the human gene for carnitine palmitoyltransferase to 1p13-p11 by non-radioactive *in situ* hybridization. *Genomics* **13**, 1372–1374 (1992).
- Taroni, F. *et al.* Molecular characterization of inherited carnitine palmitoyltransferase II deficiency. *Proc. Natl. Acad. Sci. U.S.A.* **89**, 8429–8433 (1992).
- Gellera, C. *et al.* Molecular study of lethal neonatal carnitine palmitoyltransferase II (CPT II) deficiency. *Am. J. Hum. Genet.* **51**, A168 (1992).
- Bonnefont, J.P., Cepanec, C., Munnich, A., Saudubray, J.P. & Demaugre, F. Infantile form of CPT II deficiency: identification of a missense mutation in the CPT II gene. *Am. J. Hum. Genet.* **51**, A165 (1992).
- Verderio, E. *et al.* Two novel sequence polymorphisms of the human carnitine palmitoyltransferase II (CPT1) gene. *Hum. molec. Genet.* **2**, 334 (1993).
- Singh, R. *et al.* A case of carnitine palmitoyltransferase II deficiency in human skeletal muscle. *FEBS Lett.* **241**, 126–130 (1988).
- Silver, J. Inverse polymerase chain reaction. In *PCR: A Practical Approach* (eds McPherson, M.J., Quirke, P. & Taylor, G.R.) 137–146 (Oxford University Press, New York, 1991).
- Tonin, P., Lewis, P., Servidei, S. & DiMauro, S. Metabolic causes of myoglobinuria. *Ann. Neurol.* **27**, 181–185 (1990).
- Creighton, T.E. *Proteins: Structures and Molecular Properties* 2nd edn 108–114 (Freeman, New York, 1993).
- Woeltje, K.F. *et al.* Cloning, sequencing, and expression of a cDNA encoding rat liver mitochondrial carnitine palmitoyltransferase II. *J. Biol. Chem.* **265**, 10720–10725 (1990).
- Rechsteiner, M., Rogers, S. & Rote, K. Protein structure and intracellular stability. *Trends biochem. Sci.* **12**, 390–394 (1987).
- Isaya, G. *et al.* Mitochondrial import and processing of mutant human ornithine transcarbamylase precursors in cultured cells. *Molec. cell. Biol.* **8**, 5150–5158 (1988).
- Zierz, S. & Engel, A.G. Regulatory properties of a mutant carnitine palmitoyltransferase in human skeletal muscle. *Eur. J. Biochem.* **149**, 207–214 (1985).
- Kerner, J. & Bieber, L.L. Isolation of a malonyl-CoA-sensitive CPT/β-oxidation enzyme complex from heart mitochondria. *Biochemistry* **29**, 4326–4334 (1990).
- DiDonato, S. *et al.* Muscle carnitine palmitoyltransferase deficiency: a case with enzyme deficiency in cultured fibroblasts. *A. Neurol.* **4**, 465–467 (1978).
- Mongini, T. *et al.* Myoglobinuria and carnitine palmitoyl transferase deficiency in father and son. *J. Neurol.* **238**, 323–324 (1991).
- Chu, G., Hayakawa, H. & Berg, P. Electroporation for the efficient transfection of mammalian cells with DNA. *Nucl. Acids Res.* **15**, 1311–1326 (1987).
- Taroni, F. & Rosenberg, L.E. The precursor of the biotin-binding subunit of mammalian propionyl-CoA carboxylase can be translocated into mitochondria as apo- or holoprotein. *J. Biol. Chem.* **266**, 13267–13271 (1991).
- DiDonato, S. *et al.* Normalization of short-chain acylcoenzyme A dehydrogenase after riboflavin treatment in a girl with multiple acylcoenzyme A dehydrogenase-deficient myopathy. *Ann. Neurol.* **25**, 479–484 (1989).

correction

Transgenic mice containing a human heavy chain immunoglobulin gene fragment cloned in a yeast artificial chromosome

Ted K. Choi, Paul W. Hollenbach, Barbara E. Pearson, Roanna M. Ueda, Gregory N. Weddell, Carole G. Kurahara, Clive S. Woodhouse, Robert M. Kay & Jeanne F. Loring
Nature Genetics **4**, 117–123 (1993)

The final reference that was cited in the text was not included in the reference list. The reference list should have ended as follows:

42. Strauss, W.M. *et al.* Germ-line transmission of a yeast artificial chromosome spanning the murine Col1A1 (α1(I) collagen) locus. *Science* **259**, 1904–1907 (1993).

Fig. 3 A comparison of the temporal pattern of X inactivation as indicated by the changes in the proportion of β-gal expressing cells in 6 different tissue lineages of the post-implantation mouse embryo. The data at each time point between 7.5 and 11.5 d.p.c. represent the means of the percentage for the X^p and X^m embryos shown in Table 1. Brain includes all segment of the cephalic neural tube and somite also includes the presomitic mesoderm. The 7.5 d.p.c. value for the heart and cranial mesenchyme (CranialM) is that of the embryonic mesoderm and for the brain is the value for the embryonic ectoderm of the egg cylinder. The brain and somite completed X inactivation by 8.5 d.p.c., but the notochord, heart, gut, and cranial mesenchyme completed inactivation later at 11.5 d.p.c. The heart mesoderm remained around the 65% level at 10.5 d.p.c., suggesting that at least 30% of the cells still had two active X chromosomes at this stage. The notochord differed from all other tissues by maintaining a high proportion of β-gal expressing cells and only began to inactivate the X chromosome after 9.5 d.p.c. X inactivation in all somatic lineages was completed by 11.5 d.p.c.

X-chromosome inactivation occurs at different times in different tissues of the post-implantation mouse embryo

Seong-Seng Tan, Elizabeth A. Williams & Patrick P.L. Tam
Nature Genetics **3**, 170–174 (1993)

The revised version of Fig. 3, showing expression to 11.5 d.p.c., should have been published in the article. The legend to the Fig. remains unchanged.

