REFERENCES

Abad, M.F.C., Di Benedetto, G., Magalhaes, P.J., Filippin, L., Pozzan, T., 2004. Mitochondrial pH monitored by a new engineered green fluorescent protein mutant. J. Biol. Chem. 279, 11521–11529.

Abrahams, J.P., Leslie, A.G., Lutter, R., Walker, J.E., 1994. Structure at 2.8 Å resolution of F1-ATPase from bovine heart mitochondria. Nature 370, 621–628.

Abramson, J., Svensson, E., Byrne, B., Iwata, S., 2001. Structure of cytochrome c oxidase: a comparison of the bacterial and mitochondrial enzymes. Biochim. Biophys. Acta 1544, 1–9.

Abramson, J., Smirnova, I., Kasho, V., Verner, G., Kaback, H.R., Iwata, S., 2003. Structure and mechanism of the lactose permease of *Escherichia coli*. Science 301, 610–615.

Acin-Perez, R., Salazar, E., Kamenetsky, M., Buck, J., Levin, L.R., Manfredi, G., 2009. Cyclic AMP produced inside mitochondria regulates oxidative phosphorylation. Cell Metab. 9, 265–276.

Adachi, K., Nishizaka, T., Kinosita, K., 2012. Rotational catalysis by the ATPase. Comp. Biophys. 8, 266–288.

Affourtit, C., Brand, M.D., 2005. Stronger control of ATP/ADP by proton leak in pancreatic beta cell than skeletal muscle mitochondria. Biochem. J. 393, 151–159.

Ahn, B.H., Kim, H.S., Song, S., Lee, I.H., Liu, J., Vassilopoulos, A., et al., 2008. A role for the mitochondrial deacetylase Sirt3 in regulating energy homeostasis. Proc. Natl. Acad. Sci. USA 105, 14447–14452.

Ainscow, E.K., Rutter, G.A., 2002. Glucose-stimulated oscillations in free cytosolic ATP concentration imaged in single islet beta-cells: evidence for a Ca^{2+}-dependent mechanism. Diabetes 51 (**Suppl 1**), S162–S170.

Alano, C.C., Garnier, P., Ying, W., Higashi, Y., Kauppinen, T.M., Swanson, R.A., 2010. NAD^+ depletion is necessary and sufficient for poly(ADP-ribose) polymerase-1-mediated neuronal death. J. Neurosci. 30, 2967–2978.

Albury, M.S., Elliott, C., Moore, A.L., 2009. Towards a structural elucidation of the alternative oxidase in plants. Physiol. Plant. 137, 316–327.

Amara, C.E., Marcinek, D.J., Shankland, E.G., Schenkman, K.A., Arakaki, L.S., Conley, K.E., 2008. Mitochondrial function *in vivo*: spectroscopy provides window on cellular energetics. Methods 46, 312–318.

Amo, T., Sato, S., Saiki, S., Wolf, A.M., Toyomizu, M., Gautier, C.A., et al., 2011. Mitochondrial membrane potential decrease caused by loss of PINK1 is not due to proton leak, but to respiratory chain defects. Neurobiol. Dis. 41, 111–118.

Amunts, A., Drory, O., Nelson, N., 2007. The structure of a plant photosystem I supercomplex at 3.4 Å resolution. Nature 447, 58–63.

Amunts, A., Toporik, H., Borovikova, A., Nelson, N., 2010. Structure determination and improved model of plant photosystem I. J Biol. Chem. 285, 3478–3486.

Anderson, K.A., Hirschey, M.D., 2012. Mitochondrial protein acetylation regulates metabolism. Essays Biochem. 52, 23–35.

Andrews, R.M., Kubacka, I., Chinnery, P.F., Lightowlers, R.N., Turnbull, D.M., Howell, N., 1999. Reanalysis and revision of the Cambridge reference sequence for human mitochondrial DNA. Nat. Genet. 23, 147.

Anflous, K., Armstrong, D.D., Craigen, W.J., 2001. Altered mitochondrial sensitivity for ADP and maintenance of creatine-stimulated respiration in oxidative striated muscles from VDAC1-deficient mice. J. Biol. Chem. 276, 1954–1960.

Arco, A.D., Satrustegui, J., 2005. New mitochondrial carriers: an overview. Cell Mol. Life Sci. 62, 2204–2227.

Arnou, B., Nissen, P., 2012. Structure–function relationships in P-type ATPases. Compr. Biophys. 8, 10–34.

Azzu, V., Brand, M.D., 2009. The on–off switches of the mitochondrial uncoupling proteins. Trends Biochem. Sci. 35, 298–307.

Bai, P., Canto, C., 2012. The role of PARP-1 and PARP-2 enzymes in metabolic regulation and disease. Cell Metab. 16, 290–295.

Bai, X., Ma, D., Liu, A., Shen, X., Wang, Q.J., Liu, Y., et al., 2007. Rheb activates mTOR by antagonizing its endogenous inhibitor, FKBP38. Science 318, 977–980.

Baker, L.A., Watt, I.N., Runswick, M.J., Walker, J.E., Rubinstein, J.L., 2012. Arrangement of subunits in intact mammalian mitochondrial ATP synthase determined by cryo-EM. Proc. Natl. Acad. Sci. USA 109, 11675–11680.

Balaban, R.S., 2009a. Domestication of the cardiac mitochondrion for energy conversion. J Mol. Cell. Cardiol. 46, 832–841.

Balaban, R.S., 2009b. The role of Ca^{2+} signaling in the coordination of mitochondrial ATP production with cardiac work. Biochim. Biophys. Acta Bioenerg. 1787, 1334–1341.

Baltzer, C., Tiefenbock, S.K., Frei, C., 2010. Mitochondria in response to nutrients and nutrient-sensitive pathways. Mitochondrion 10, 589–597.

Baniulis, D., Yamashita, E., Zhang, H., Hasan, S.S., Cramer, W.A., 2008. Structure–function of the cytochrome b_6f complex. Photochem. Photobiol. 84, 1349–1358.

Baradaran, R., Berrisford, J.M., Minhas, G.S., Sazanov, L.A., 2013. Crystal structure of the entire respiratory complex I. Nature 494, 443–448.

Bartelt, A., Heeren, J., 2012. The holy grail of metabolic disease: brown adipose tissue. Curr. Opin. Lipidol. 23, 190–195.

Bason, J.V., Runswick, M.J., Fearnley, I.M., Walker, J.E., 2011. Binding of the inhibitor protein IF(1) to bovine F(1)-ATPase. J. Mol. Biol. 406, 443–453.

Baughman, J.M., Perocchi, F., Girgis, H.S., Plovanich, M., Belcher-Timme, C.A., Sancak, Y., et al., 2011. Integrative genomics identifies MCU as an essential component of the mitochondrial calcium uniporter. Nature 476, 341–345.

Baumgartner, H.K., Gerasimenko, J.V., Thorne, C., Ferdek, P., Pozzan, T., Tepikin, A.V., et al., 2009. Calcium elevation in mitochondria is the main Ca^{2+} requirement for mitochondrial permeability transition pore (mPTP) opening. J. Biol. Chem. 284, 20796–20803.

Ben-Shem, A., Frolow, F., Nelson, N., 2003. Crystal structure of plant photosystem I. Nature 426, 630–635.

Bentzinger, C.F., Romanino, K., Cloetta, D., Lin, S., Mascarenhas, J.B., Oliveri, F., et al., 2008. Skeletal muscle-specific ablation of raptor, but not of rictor, causes metabolic changes and results in muscle dystrophy. Cell Metab. 8, 411–424.

Berrisford, J.M., Sazanov, L.A., 2009. Structural basis for the mechanism of respiratory complex I. J. Biol. Chem. 284, 29773–29783.

Berry, R.M., Sowa, Y., 2012. The rotary bacterial flagellar motor. Compr. Biophys. 8, 50–71.

Biegel, E., Schmidt, S., Gonzalez, J.M., Muller, V., 2011. Biochemistry, evolution and physiological function of the Rnf complex, a novel ion-motive electron transport complex in prokaryotes. Cell Mol. Life Sci. 68, 613–634.

Birket, M.J., Orr, A.L., Gerencser, A.A., Madden, D.T., Vitelli, C., Swistowski, A., et al., 2011. A reduction in ATP demand and mitochondrial activity with neural differentiation of human embryonic stem cells. J. Cell. Sci. 124, 348–358.

Blagosklonny, M.V., 2011. Hormesis does not make sense except in the light of TOR-driven aging. Aging 3, 1051–1062.

Blomain, E.S., McMahon, S.B., 2012. Dynamic regulation of mitochondrial transcription as a mechanism of cellular adaptation. Biochim. Biophys. Acta 1819, 1075–1079.

Bowler, M.W., Montgomery, M.G., Leslie, A.G., Walker, J.E., 2007. Ground state structure of F_1-ATPase from bovine heart mitochondria at 1.9Å resolution. J. Biol. Chem. 282, 14238–14242.

Brand, M.D., 1997. Regulation analysis of energy metabolism. J. Exp. Biol. 200 (**Pt 2**), 193–202.

Brand, M.D., 2010. The sites and topology of mitochondrial superoxide production. Exp. Gerontol. 46, 466–472.

Brand, M.D., Nicholls, D.G., 2011. Assessing mitochondrial dysfunction in cells. Biochem. J. 435, 297–312.

Brand, M.D., Chien, L.F., Ainscow, E.K., Rolfe, D.F., Porter, R.K., 1994. The causes and functions of mitochondrial proton leak. Biochim. Biophys. Acta 1187, 132–139.

Brandes, R., Bers, D.M., 2002. Simultaneous measurements of mitochondrial NADH and Ca^{2+} during increased work in intact rat heart trabeculae. Biophys. J. 83, 587–604.

Bricker, D.K., Taylor, E.B., Schell, J.C., Orsak, T., Boutron, A., Chen, Y.C., et al., 2012. A mitochondrial pyruvate carrier required for pyruvate uptake in yeast, *Drosophila*, and humans. Science 337, 96–100.

Brierley, G.P., Baysal, K., Jung, D.W., 1994. Cation transport systems in mitochondria: Na^+ and K^+ uniports and exchangers. J. Bioenerg. Biomembr. 26, 519–526.

Brini, M., Carafoli, E., 2009. Calcium pumps in health and disease. Physiol. Rev. 89, 1341–1378.

Brown, G.C., Borutaite, V., 2007. Nitric oxide and mitochondrial respiration in the heart. Cardiovasc. Res. 75, 283–290.

Buckel, W., Thauer, R.K., 2013. Energy conservation via electron bifurcating ferredoxin reduction and proton/Na(+) translocating ferredoxin oxidation. Biochim. Biophys. Acta 1827, 94–113.

Busch, A., Hippler, M., 2011. The structure and function of eukaryotic photosystem I. Biochim. Biophys. Acta 1807, 864–877.

Butler, J.A., Ventura, N., Johnson, T.E., Rea, S.L., 2010. Long-lived mitochondrial (Mit) mutants of *Caenorhabditis elegans* utilize a novel metabolism. FASEB J. 24, 4977–4988.

Cai, Q., Sheng, Z.H., 2009. Mitochondrial transport and docking in axons. Exp. Neurol. 218, 257–267.

Cai, Q., Davis, M.L., Sheng, Z.H., 2011. Regulation of axonal mitochondrial transport and its impact on synaptic transmission. Neurosci. Res. 70, 9–15.

Cairns, R.A., Harris, I.S., Mak, T.W., 2011. Regulation of cancer cell metabolism. Nat. Rev. Cancer 11, 85–95.

Caldeira da Silva, C.C., Cerqueira, F.M., Barbosa, L.F., Medeiros, M.H., Kowaltowski, A.J., 2008. Mild mitochondrial uncoupling in mice affects energy metabolism, redox balance and longevity. Aging Cell 7, 552–560.

Callaghan, R., Geroge, A.M., Kerr, I.D., 2012. Molecular aspects of the translocation process by ABC Proteins. Compr. Biophys. 8, 146–172.

Campanella, M., Parker, N., Tan, C.H., Hall, A.M., Duchen, M.R., 2009. IF(1): setting the pace of the F(1)F(o)-ATP synthase. Trends. Biochem. Sci. 34, 343–350.

Cannon, M.B., Remington, S.J., 2008. Redox-sensitive green fluorescent protein: probes for dynamic intracellular redox responses. A review. Methods Mol. Biol. 476, 51–65.

Canto, C., Gerhart-Hines, Z., Feige, J.N., Lagouge, M., Noriega, L., Milne, J.C., et al., 2009. AMPK regulates energy expenditure by modulating NAD^+ metabolism and SIRT1 activity. Nature 458, 1056–1060.

Cardaci, S., Desideri, E., Ciriolo, M.R., 2012. Targeting aerobic glycolysis: 3-Bromopyruvate as a promising anticancer drug. J. Bioenerg. Biomembr. 44, 17–29.

Carlson, A.P., Carter, R.E., Shuttleworth, C.W., 2012. Vascular, electrophysiological, and metabolic consequences of cortical spreading depression in a mouse model of simulated neurosurgical conditions. Neurol. Res. 34, 223–231.

Chalmers, S., Nicholls, D.G., 2003. The relationship between free and total calcium concentrations in the matrix of liver and brain mitochondria. J. Biol. Chem. 279, 19062–19070.

Chance, B., Williams, G.R., 1955. Respiratory enzymes in oxidative phosphorylation: III. The steady state. J. Biol. Chem. 217, 409–427.

Chaturvedi, R.K., Adhihetty, P., Shukla, S., Hennessy, T., Calingasan, N., Yang, L., et al., 2009. Impaired PGC-1alpha function in muscle in Huntington's disease. Hum. Mol. Genet. 18, 3048–3065.

Chen, C.T., Hsu, S.H., Wei, Y.H., 2009. Upregulation of mitochondrial function and antioxidant defense in the differentiation of stem cells. Biochim. Biophys. Acta 1800, 257–263.

Cherepanov, D.A., Mulkidjanian, A.Y., Junge, W., 1999. Transient accumulation of elastic energy in proton translocating ATP synthase. FEBS. Lett. 449, 1–6.

Chipuk, J.E., Green, D.R., 2008. How do BCL-2 proteins induce mitochondrial outer membrane permeabilization? Trends Cell Biol. 18, 157–164.

Choi, S., Gerencser, A.A., Nicholls, D.G., 2009. Bioenergetic analysis of isolated cerebrocortical nerve terminals on a microgram scale: spare respiratory capacity and stochastic mitochondrial failure. J. Neurochem. 109, 1179–1191.

Choi, S.W., Gerencser, A.A., Lee, D., Rajagopalan, S., Nicholls, D.G., Andersen, J.K., et al., 2011. Intrinsic bioenergetic properties and stress-sensitivity of dopaminergic synaptosomes. J. Neurosci. 31, 4524–4534.

Choi, S.W., Gerencser, A.A., Ng, R., Flynn, J.M., Melov, S., Danielson, S.R., et al., 2012. No consistent mitochondrial bioenergetic defects in presynaptic nerve terminals isolated from mouse models of Alzheimer's disease. J. Neurosci. 32, 16775–16784.

Chu, C.T., 2010. Tickled PINK1: mitochondrial homeostasis and autophagy in recessive parkinsonism. Biochim. Biophys. Acta 1802, 20–29.

Cogdell, R.J., Gall, A., Kohler, J., 2006. The architecture and function of the light-harvesting apparatus of purple bacteria: from single molecules to *in vivo* membranes. Q. Rev. Biophys. 39, 227–324.

Copeland, W.C., 2012. Defects in mitochondrial DNA replication and human disease. Crit. Rev. Biochem. Mol. Biol. 47, 64–74.

Correia, S.C., Santos, R.X., Perry, G., Zhu, X., Moreira, P.I., Smith, M.A., 2012. Mitochondrial importance in Alzheimer's, Huntington's and Parkinson's diseases. Adv. Exp. Med. Biol. 724, 205–221.

Covian, R., Balaban, R.S., 2012. Cardiac mitochondrial matrix and respiratory complex protein phosphorylation. Am. J. Physiol. Heart Circ. Physiol. 303, H940–H966.

Craigen, W.J., 2012. Mitochondrial DNA mutations: an overview of clinical and molecular aspects. Methods Mol. Biol. 837, 3–15.

Cramer, W.A., Hasan, S.S., Yamashita, E., 2011. The Q cycle of cytochrome *bc* complexes: a structure perspective. Biochim. Biophys. Acta 1807, 788–802.

Crofts, A.R., 1993. Peter Mitchell [Obituary]. Photosynth. Res. 35, 1–4.

Crofts, A.R., 2004. The cytochrome bc_1 complex: function in the context of structure. Annu. Rev. Physiol. 66, 689–733.

Crofts, A.R., Hong, S., Zhang, Z., Berry, E.A., 1999. Physicochemical aspects of the movement of the Rieske iron sulfur protein during quinol oxidation by the bc_1 complex from mitochondria and photosynthetic bacteria. Biochemistry 38, 15827–15839.

Crofts, A.R., Holland, J.T., Victoria, D., Kolling, D.R., Dikanov, S.A., Gilbreth, R., et al., 2008. The Q-cycle reviewed: how well does a monomeric mechanism of the *bc*(1) complex account for the function of a dimeric complex? Biochim. Biophys. Acta 1777, 1001–1019.

Crompton, M., Heid, I., 1978. The cycling of calcium, sodium, and protons across the inner membrane of cardiac mitochondria. Eur. J. Biochem. 91, 599–608.

Cross, R.L., 1981. The mechanism and regulation of ATP synthesis by F1-ATPases. Annu. Rev. Biochem. 50, 681–714.

Cruz-Gallardo, I., Diaz-Moreno, I., Diaz-Quintana, A., De la Rosa, M.A., 2012. The cytochrome *f*-plastocyanin complex as a model to study transient interactions between redox proteins. FEBS Lett. 586, 646–652.

Csordas, G., Renken, C., Varnai, P., Walter, L., Weaver, D., Buttle, K.F., et al., 2006. Structural and functional features and significance of the physical linkage between ER and mitochondria. J. Cell Biol. 174, 915–921.

Cunningham, J.T., Rodgers, J.T., Arlow, D.H., Vazquez, F., Mootha, V.K., Puigserver, P., 2007. mTOR controls mitochondrial oxidative function through a YY1-PGC-1alpha transcriptional complex. Nature 450, 736–740.

Cypess, A.M., Lehman, S., Williams, G., Tal, I., Rodman, D., Goldfine, A.B., et al., 2009. Identification and importance of brown adipose tissue in adult humans. N. Engl. J. Med. 360, 1509–1517.

Damiano, M., Galvan, L., Deglon, N., Brouillet, E., 2010. Mitochondria in Huntington's disease. Biochim. Biophys. Acta 1802, 52–61.

Dang, S., Sun, L., Huang, Y., Lu, F., Liu, Y., Gong, H., et al., 2010. Structure of a fucose transporter in an outward-open conformation. Nature 467, 734–738.

Darrouzet, E., Cooley, J.W., Daldal, F., 2004. The cytochrome *bc*(1) complex and its homologue the *b*(6)*f* complex: similarities and differences. Photosynth. Res. 79, 25–44.

Daum, B., Nicastro, D., Austin, J., McIntosh, J.R., Kuhlbrandt, W., 2010. Arrangement of photosystem II and ATP synthase in chloroplast membranes of spinach and pea. Plant. Cell 22, 1299–1312.

Davey, G.P., Peuchen, S., Clark, J.B., 1998. Energy thresholds in brain mitochondria: potential involvement in neurodegeneration. J. Biol. Chem. 273, 12753–12757.

De, S.D., Raffaello, A., Teardo, E., Szabo, I., Rizzuto, R., 2011. A forty-kilodalton protein of the inner membrane is the mitochondrial calcium uniporter. Nature 476, 336–340.

De Brito, O.M., Scorrano, L., 2008. Mitofusin 2 tethers endoplasmic reticulum to mitochondria. Nature 456, 605–610.

Deas, E., Wood, N.W., Plun-Favreau, H., 2011. Mitophagy and Parkinson's disease: the PINK1-parkin link. Biochim. Biophys. Acta 1813, 623–633.

Deisenhofer, J., Michel, H., 1989. Nobel lecture. The photosynthetic reaction centre from the purple bacterium *Rhodopseudomonas viridis*. EMBO J. 8, 2149–2170.

Deisenhofer, J., Epp, O., Sinning, I., Michel, H., 1995. Crystallographic refinement at 2.3 Å resolution and refined model of the photosynthetic reaction centre from *Rhodopseudomonas viridis*. J Mol. Biol. 246, 429–457.

Denton, R.M., 2009. Regulation of mitochondrial dehydrogenases by calcium ions. Biochim. Biophys. Acta 1787, 1309–1316.

Diana, F.F., Silva Esteves, A.R., Oliveira, C.R., Cardoso, S.M., 2011. Mitochondria: the common upstream driver of amyloid-β and tau pathology in Alzheimer's disease. Curr. Alzheimer Res. 8, 563–572.

Diaz, F., Moraes, C.T., 2008. Mitochondrial biogenesis and turnover. Cell Calcium 44, 24–35.

Diaz-Ruiz, R., Rigoulet, M., Devin, A., 2011. The warburg and crabtree effects: on the origin of cancer cell energy metabolism and of yeast glucose repression. Biochim. Biophys. Acta 1807, 568–576.

Dickinson, B.C., Srikun, D., Chang, C.J., 2010. Mitochondrial-targeted fluorescent probes for reactive oxygen species. Curr. Opin. Chem. Biol. 14, 50–56.

Dimroth, P., Jockel, P., Schmid, M., 2001. Coupling mechanism of the oxaloacetate decarboxylase Na^+ pump. Biochim. Biophys. Acta 1505, 1–14.

Drago, I., Pizzo, P., Pozzan, T., 2011. After half a century mitochondrial calcium in- and efflux machineries reveal themselves. EMBO J. 30, 4119–4125.

Droge, W., 2002. Free radicals in the physiological control of cell function. Physiol. Rev. 82, 47–95.

D'Souza, G.G., Wagle, M.A., Saxena, V., Shah, A., 2011. Approaches for targeting mitochondria in cancer therapy. Biochim. Biophys. Acta 1807, 689–696.

Du, H., Guo, L., Yan, S., Sosunov, A.A., McKhann, G.M., Yan, S.S., 2010. Early deficits in synaptic mitochondria in an Alzheimer's disease mouse model. Proc. Natl. Acad. Sci. USA 107, 18670–18675.

Duchen, M.R., Szabadkai, G., 2010. Roles of mitochondria in human disease. Essays Biochem. 47, 115–137.

Duchen, M.R., Surin, A., Jacobson, J., 2003. Imaging mitochondrial function in intact cells. Methods Enzymol. 361, 353–389.

Dudkina, N.V., Oostergetel, G.T., Lewejohann, D., Braun, H.P., Boekema, E.J., 2010. Row-like organization of ATP synthase in intact mitochondria determined by cryo-electron tomography. Biochim. Biophys. Acta 1797, 272–277.

Duffy, L.M., Chapman, A.L., Shaw, P.J., Grierson, A.J., 2011. The role of mitochondria in the pathogenesis of amyotrophic lateral sclerosis. Neuropathol. Appl. Neurobiol. 37, 336–352.

Dukanovic, J., Rapaport, D., 2011. Multiple pathways in the integration of proteins into the mitochondrial outer membrane. Biochim. Biophys. Acta 1808, 971–980.

Eckert, A., Schmitt, K., Gotz, J., 2011. Mitochondrial dysfunction: the beginning of the end in Alzheimer's disease? Separate and synergistic modes of tau and amyloid-beta toxicity. Alzheimer's Res. Ther. 3, 15–25.

Edlich, F., Banerjee, S., Suzuki, M., Cleland, M.M., Arnoult, D., Wang, C., et al., 2011. Bcl-x(L) retrotranslocates Bax from the mitochondria into the cytosol. Cell 145, 104–116.

Efremov, R.G., Sazanov, L.A., 2011. Structure of the membrane domain of respiratory complex I. Nature 476, 414–420.

Endo, T., Yamano, K., Kawano, S., 2011. Structural insight into the mitochondrial protein import system. Biochim. Biophys. Acta 1808, 955–970.

Ericson, N.G., Kulawiec, M., Vermulst, M., Sheahan, K., O'Sullivan, J., Salk, J.J., et al., 2012. Decreased mitochondrial DNA mutagenesis in human colorectal cancer. PLoS Genet. 8, e1002689.

Ermler, U., Fritzsch, G., Buchanan, S.K., Michel, H., 1994. Structure of the photosynthetic reaction centre from *Rhodobacter sphaeroides* at 2.65 Å resolution: cofactors and protein–cofactor interactions. Structure 2, 925–936.

Ernst, S., Duser, M.G., Zarrabi, N., Dunn, S.D., Borsch, M., 2012. Elastic deformations of the rotary double motor of single F(o)F(1)-ATP synthases detected in real time by Forster resonance energy transfer. Biochim. Biophys. Acta 1817, 1722–1731.

Erusalimsky, J.D., Moncada, S., 2007. Nitric oxide and mitochondrial signaling. From physiology to pathophysiology. Arterioscler. Thromb. Vasc. Biol. 27, 2524–2531.

Evans, D.S., Kapahi, P., Hsueh, W.C., Kockel, L., 2011. TOR signaling never gets old: aging, longevity and TORC1 activity. Ageing Res. Rev. 10, 225–237.

Exner, N., Lutz, A.K., Haass, C., Winklhofer, K.F., 2012. Mitochondrial dysfunction in Parkinson's disease: molecular mechanisms and pathophysiological consequences. EMBO J. 31, 3038–3062.

Faccenda, D., Campanella, M., 2012. Molecular regulation of the mitochondrial F(1)F(o)-ATP synthase: physiological and pathological significance of the inhibitory factor 1 (IF(1)). Int. J. Cell Biol 2012, 367934.

Falkenberg, M., Larsson, N.G., Gustafsson, C.M., 2007. DNA replication and transcription in mammalian mitochondria. Annu. Rev. Biochem. 76, 679–699.

Fan, M.M., Raymond, L.A., 2007. *N*-methyl-D-aspartate (NMDA) receptor function and excitotoxicity in Huntington's disease. Prog. Neurobiol. 81, 272–293.

Feniouk, B.A., Kozlova, M.A., Knorre, D.A., Cherepanov, D.A., Mulkidjanian, A.Y., Junge, W., 2004. The proton-driven rotor of ATP synthase: ohmic conductance (10 fS), and absence of voltage gating. Biophys. J. 86, 4094–4109.

Fercher, A., O'Riordan, T.C., Zhdanov, A.V., Dmitriev, R.I., Papkovsky, D.B., 2010. Imaging of cellular oxygen and analysis of metabolic responses of mammalian cells. Methods Mol. Biol. 591, 257–273.

Ferguson, S.J., 2000. ATP synthase: what dictates the size of the ring? Curr. Biol. 10, R804–R808.

Ferguson, S.J., Ingledew, W.J., 2008. Energetic problems faced by micro-organisms growing or surviving on parsimonious energy sources and at acidic pH: I. *Acidithiobacillus ferrooxidans* as a paradigm. Biochim. Biophys. Acta 1777, 1471–1479.

Fernandez-Marcos, P.J., Auwerx, J., 2011. Regulation of PGC-1α, a nodal regulator of mitochondrial biogenesis. Am. J. Clin. Nutr. 93, 884S–890S.

Finley, L.W., Haigis, M.C., 2009. The coordination of nuclear and mitochondrial communication during aging and calorie restriction. Ageing Res. Rev. 8, 173–188.

Folmes, C.D., Nelson, T.J., Dzeja, P.P., Terzic, A., 2012. Energy metabolism plasticity enables stemness programs. Ann. N. Y. Acad. Sci. 1254, 82–89.

Fonteriz, R.I., de la Fuente, S., Moreno, A., Lobatin, C.D., Montero, M., Alvarez, J., 2010. Monitoring mitochondrial [Ca^{2+}] dynamics with rhod-2, ratiometric pericam and aequorin. Cell Calcium 48, 61–69.

Foretz, M., Hebrard, S., Leclerc, J., Zarrinpashneh, E., Soty, M., Mithieux, G., et al., 2010. Metformin inhibits hepatic gluconeogenesis in mice independently of the LKB1/AMPK pathway via a decrease in hepatic energy state. J. Clin. Invest. 120, 2355–2369.

Frank, H.A., Cogdell, R.J., 2012. Light capture in photosynthesis. Comp. Biophys. 8, 94–114.

Frey, T.G., Mannella, C.A., 2000. The internal structure of mitochondria. Trends Biochem. Sci. 25, 319–324.

Friedman, J.R., Lackner, L.L., West, M., Dibenedetto, J.R., Nunnari, J., Voeltz, G.K., 2011. ER tubules mark sites of mitochondrial division. Science 334, 358–362.

Galter, D., Pernold, K., Yoshitake, T., Lindqvist, E., Hoffer, B., Kehr, J., et al., 2009. MitoPark mice mirror the slow progression of key symptoms and L-DOPA response in Parkinson's disease. Genes Brain Behav. 9, 173–181.

Garlid, K.D., Halestrap, A.P., 2012. The mitochondrial K(ATP) channel—Fact or fiction? J. Mol. Cell. Cardiol. 52, 578–583.

Gautier, C.A., Kitada, T., Shen, J., 2008. Loss of PINK1 causes mitochondrial functional defects and increased sensitivity to oxidative stress. Proc. Natl. Acad. Sci. USA 105, 11364–11369.

Gerencser, A.A., Nicholls, D.G., 2008. Measurement of instantaneous velocity vectors of organelle transport: mitochondrial transport and bioenergetics in hippocampal neurons. Biophys. J. 95, 3079–3099.

Gibasiewicz, K., Pajzderska, M., Karolczak, J., Dobek, A., 2009. Excitation and electron transfer in reaction centers from *Rhodobacter sphaeroides* probed and analyzed globally in

the 1-nanosecond temporal window from 330 to 700 nm. Phys. Chem. Chem. Phys. 11, 10484–10493.

Gil, J.M., Rego, A.C., 2008. Mechanisms of neurodegeneration in Huntington's disease. Eur. J. Neurosci. 27, 2803–2820.

Giorgi, C., De, S.D., Bononi, A., Rizzuto, R., Pinton, P., 2009. Structural and functional link between the mitochondrial network and the endoplasmic reticulum. Int. J. Biochem. Cell Biol. 41, 1817–1827.

Giralt, A., Villarroya, F., 2012. SIRT3, a pivotal actor in mitochondrial functions: metabolism, cell death and aging. Biochem. J. 444, 1–10.

Giuditta, A., Chun, J.T., Eyman, M., Cefaliello, C., Bruno, A.P., Crispino, M., 2008. Local gene expression in axons and nerve endings: the glia-neuron unit. Physiol. Rev. 88, 515–555.

Goehring, I., Gerencser, A.A., Schmidt, S., Brand, M.D., Mulder, H., Nicholls, D.G., 2012. Plasma membrane potential oscillations in insulin secreting ins-1 832/13 cells do not require glycolysis and are not initiated by fluctuations in mitochondrial bioenergetics. J. Biol. Chem. 287, 15706–15717.

Gomez-Duran, A., Pacheu-Grau, D., Martinez-Romero, I., Lopez-Gallardo, E., Lopez-Perez, M.J., Montoya, J., et al., 2012. Oxidative phosphorylation differences between mitochondrial DNA haplogroups modify the risk of Leber's hereditary optic neuropathy. Biochim. Biophys. Acta 1822, 1216–1222.

Griffiths, E.J., 2009. Mitochondrial calcium transport in the heart: physiological and pathological roles. J. Mol. Cell. Cardiol. 46, 789–803.

Griffiths, E.J., 2012. Mitochondria and heart disease. Adv. Exp. Med. Biol. 942, 249–267.

Guan, L., Kaback, H.R., 2006. Lessons from lactose permease. Annu. Rev. Biophys. Biomol. Struct. 35, 67–91.

Gutscher, M., Pauleau, A.L., Marty, L., Brach, T., Wabnitz, G.H., Samstag, Y., et al., 2008. Real-time imaging of the intracellular glutathione redox potential. Nat. Methods 5, 553–559.

Hajnoczky, G., Csordas, G., Das, S., Garcia-Perez, C., Saotome, M., Sinha, R.S., et al., 2006. Mitochondrial calcium signalling and cell death: approaches for assessing the role of mitochondrial Ca^{2+} uptake in apoptosis. Cell Calcium 40, 553–560.

Haldrup, A., Jensen, P.E., Lunde, C., Scheller, H.V., 2001. Balance of power: a view of the mechanism of photosynthetic state transitions. Trends Plant. Sci. 6, 301–305.

Halestrap, A.P., 2009. What is the mitochondrial permeability transition pore? J. Mol. Cell. Cardiol. 46, 821–831.

Halestrap, A.P., Pasdois, P., 2009. The role of the mitochondrial permeability transition pore in heart disease. Biochim. Biophys. Acta Bioenerg. I1787, 1402–1415.

Hancock, C.R., Han, D.H., Higashida, K., Kim, S.H., Holloszy, J.O., 2011. Does calorie restriction induce mitochondrial biogenesis? A reevaluation. FASEB J. 25, 785–791.

Handy, D.E., Loscalzo, J., 2011. Redox regulation of mitochondrial function. Antioxid. Redox Signal. 16, 1323–1367.

Hardie, D.G., 2011. AMP-activated protein kinase: a cellular energy sensor with a key role in metabolic disorders and in cancer. Biochem. Soc. Trans. 39, 1–13.

Harman, D., 1956. Aging: a theory based on free radical and radiation chemistry. J. Gerontol. 11, 298–300.

Harner, M., Korner, C., Walther, D., Mokranjac, D., Kaesmacher, J., Welsch, U., et al., 2011. The mitochondrial contact site complex, a determinant of mitochondrial architecture. EMBO J. 30, 4356–4370.

Hauser, D.N., Hastings, T.G., 2012. Mitochondrial dysfunction and oxidative stress in Parkinson's disease and monogenic parkinsonism. Neurobiol. Dis. 51, 35–42.

He, W., Newman, J.C., Wang, M.Z., Ho, L., Verdin, E., 2012. Mitochondrial sirtuins: regulators of protein acylation and metabolism. Trends Endocrinol. Metab. 23, 467–476.

Heathcote, P., Jones, M.R., 2012. The structure -function relationships of photosynthetic reaction centres. Comp. Biophys. 8, 116–144.

Hekimi, S., Lapointe, J., Wen, Y., 2011. Taking a "good" look at free radicals in the aging process. Trends Cell Biol. 21, 569–576.

Henderson, P.J., 2012. Membrane proteins for secondary active transport and their molecular mechanism. Compr. Biophys. 8, 265–288.

Herrmann, J.M., 2011. MINOS is plus: a mitofilin complex for mitochondrial membrane contacts. Dev. Cell 21, 599–600.

Herrmann, J.M., Riemer, J., 2010. The intermembrane space of mitochondria. Antioxid. Redox Signal. 13, 1341–1358.

Herzig, S., Raemy, E., Montessuit, S., Veuthey, J.L., Zamboni, N., Westermann, B., et al., 2012. Identification and functional expression of the mitochondrial pyruvate carrier. Science 337, 93–96.

Hiller, S., Garces, R.G., Malia, T.J., Orekhov, V.Y., Colombini, M., Wagner, G., 2008. Solution structure of the integral human membrane protein VDAC-1 in detergent micelles. Science 321, 1206–1210.

Hirai, T., Subramaniam, S., Lanyi, J.K., 2009. Structural snapshots of conformational changes in a seven-helix membrane protein: lessons from bacteriorhodopsin. Curr. Opin. Struct. Biol. 19, 433–439.

Hirst, J., 2010. Towards the molecular mechanism of respiratory complex I. Biochem. J. 425, 327–339.

Hirst, J., Carroll, J., Fearnley, I.M., Shannon, R.J., Walker, J.E., 2003. The nuclear encoded subunits of complex I from bovine heart mitochondria. Biochim. Biophys. Acta Bioenergetics 1604, 135–150.

Hoek, J.B., Nicholls, D.G., Williamson, J.R., 1980. Determination of the mitochondrial protonmotive force in isolated hepatocytes. J. Biol. Chem. 255, 1458–1464.

Hoffman, D.L., Brookes, P.S., 2009. Oxygen sensitivity of mitochondrial reactive oxygen species generation depends on metabolic conditions. J. Biol. Chem. 284, 16236–16245.

Hohmann-Marriott, M.F., Blankenship, R.E., 2011. Evolution of photosynthesis. Annu. Rev. Plant. Biol. 62, 515–548.

Hunte, C., Zickermann, V., Brandt, U., 2010. Functional modules and structural basis of conformational coupling in mitochondrial complex I. Science 329, 448–451.

Isaev, P.I., Liberman, E.A., Samuilov, V.D., Skulachev, V.P., Tsofina, L.M., 1970. Conversion of biomembrane-produced energy into electric form: 3. Chromatophores of *Rhodospirillum rubrum*. Biochim. Biophys. Acta 216, 22–29.

Ishmukhametov, R., Hornung, T., Spetzler, D., Frasch, W.D., 2010. Direct observation of stepped proteolipid ring rotation in *E. coli* F(0)F(1)-ATP synthase. EMBO J. 29, 3911–3923.

Iverson, T.M., 2012. Catalytic mechanisms of complex II enzymes: a structural perspective. Biochim. Biophys. Acta [Epub ahead of print]

Iwabu, M., Yamauchi, T., Okada-Iwabu, M., Sato, K., Nakagawa, T., Funata, M., et al., 2010. Adiponectin and AdipoR1 regulate PGC-1α and mitochondria by Ca(2+) and AMPK/SIRT1. Nature 464, 1313–1319.

Iwai, M., Takizawa, K., Tokutsu, R., Okamuro, A., Takahashi, Y., Minagawa, J., 2010. Isolation of the elusive supercomplex that drives cyclic electron flow in photosynthesis. Nature 464, 1210–1213.

Iwata, M., Lee, Y., Yamashita, T., Yagi, T., Iwata, S., Cameron, A.D., et al., 2012. The structure of the yeast NADH dehydrogenase (Ndi1) reveals overlapping binding sites for water- and lipid-soluble substrates. Proc. Natl. Acad. Sci. USA 109, 15247–15252.

Jackson, J.B., 2012. A review of the binding-change mechanism for proton-translocating transhydrogenase. Biochim. Biophys. Acta 1817, 1839–1846.

Jacobson, J., Duchen, M.R., 2002. Mitochondrial oxidative stress and cell death in astrocytes: requirement for stored Ca^{2+} and sustained opening of the permeability transition pore. J. Cell Sci. 115, 1175–1188.

Jagendorf, A.T., 2002. Photophosphorylation and the chemiosmotic perspective. Photosynth. Res. 73, 233–241.

Jang, Y.C., Remmen, V.H., 2009. The mitochondrial theory of aging: insight from transgenic and knockout mouse models. Exp. Gerontol. 44, 256–260.

Janssen, R.J., Nijtmans, L.G., Heuvel, L.P., Smeitink, J.A., 2006. Mitochondrial complex I: structure, function and pathology. J. Inherit. Metab. Dis. 29, 499–515.

Jekabsons, M.B., Nicholls, D.G., 2004. *In situ* respiration and bioenergetic status of mitochondria in primary cerebellar granule neuronal cultures exposed continuously to glutamate. J. Biol. Chem. 279, 32989–33000.

Jiang, D., Zhao, L., Clapham, D.E., 2009. Genome-wide RNAi screen identifies Letm1 as a mitochondrial Ca^{2+}/H^{+} antiporter. Science 326, 144–147.

Jitrapakdee, S., Wutthisathapornchai, A., Wallace, J.C., MacDonald, M.J., 2010. Regulation of insulin secretion: role of mitochondrial signalling. Diabetologia 53, 1019–1032.

Johnson-Cadwell, L.I., Jekabsons, M.B., Wang, A., Polster, B.M., Nicholls, D.G., 2007. "Mild uncoupling" does not decrease mitochondrial superoxide levels in cultured cerebellar granule neurons but decreases spare respiratory capacity and increases toxicity to glutamate and oxidative stress. J. Neurochem. 101, 1619–1631.

Jokinen, R., Battersby, B.J., 2012. Insight into mammalian mitochondrial DNA segregation. Ann. Med. 45, 149–155.

Jones, M.R., 2009. The petite purple photosynthetic powerpack. Biochem. Soc. Trans. 37, 400–407.

Jose, C., Bellance, N., Rossignol, R., 2010. Choosing between glycolysis and oxidative phosphorylation: a tumor's dilemma? Biochim. Biophys. Acta 1807, 552–561.

Junge, W., Sielaff, H., Engelbrecht, S., 2009. Torque generation and elastic power transmission in the rotary F_oF_1-ATPase. Nature 459, 364–370.

Kaila, V.R., Verkhovsky, M.I., Wikstrom, M., 2010. Proton-coupled electron transfer in cytochrome oxidase. Chem. Rev. 110, 7062–7081.

Kajimura, S., Seale, P., Spiegelman, B.M., 2010. Transcriptional control of brown fat development. Cell Metab. 11, 257–262.

Kapahi, P., et al., 2010. With TOR, Less is more: a key role for the conserved nutrient-sensing TOR pathway in aging. Cell Metab. 11, 453–465.

Karbowski, M., Neutzner, A., 2012. Neurodegeneration as a consequence of failed mitochondrial maintenance. Acta Neuropathol. 123, 157–171.

Kawakami, K., Umena, Y., Kamiya, N., Shen, J.R., 2011. Structure of the catalytic, inorganic core of oxygen-evolving photosystem II at 1.9Å resolution. J. Photochem. Photobiol. B 104, 9–18.

Kellosalo, J., Kajander, T., Kogan, K., Pokharel, K., Goldman, A., 2012. The structure and catalytic cycle of a sodium-pumping pyrophosphatase. Science 337, 473–476.

Kemp, G.J., Brindle, K.M., 2012. What do magnetic resonance-based measurements of P_i→ATP flux tell us about skeletal muscle metabolism? Diabetes 61, 1927–1934.

Kim, J.S., Jin, Y., Lemasters, J.J., 2006. Reactive oxygen species, but not Ca^{2+}, trigger pH- and mitochondrial permeability transition-dependent death of adult rat myocytes after ischemia-reperfusion. Am. J. Physiol. Heart Circ. Physiol. 290, H2024–H2034.

Klingenberg, M., 2008. The ADP and ATP transport in mitochondria and its carrier. Biochim. Biophys. Acta 1778, 1978–2021.

Komary, Z., Tretter, L., Adam-Vizi, V., 2010. Membrane potential-related effect of calcium on reactive oxygen species generation in isolated brain mitochondria. Biochim. Biophys. Acta 1797, 922–928.

Korkhov, V.M., Mireku, S.A., Locher, K.P., 2012. Structure of AMP–PNP-bound vitamin B12 transporter BtuCD-F. Nature 490, 367–372.

Korshunov, S.S., Skulachev, V.P., Starkov, A.A., 1997. High protonic potential actuates a mechanism of production of reactive oxygen species in mitochondria. FEBS Lett. 416, 15–18.

Krishnan, K.J., Turnbull, D.M., 2010. Mitochondrial DNA and genetic disease. Essays Biochem. 47, 139–151.

Kunji, E.R., 2012. Structural and mechanistic aspects of mitochondrial transport proteins. Compr. Biophys. 8, 174–205.

Lange, C., Hunte, C., 2002. Crystal structure of the yeast cytochrome bc_1 complex with its bound substrate cytochrome *c*. Proc. Natl. Acad. Sci. USA 99, 2800–2805.

Lanyi, J.K., 2004. Bacteriorhodopsin. Annu. Rev. Physiol. 66, 665–688.

Lanyi, J.K., 2012. Light capture and energy transduction in bacterial rhodopsins and related proteins. Comp. Biophys. 8, 206–227.

Lax, N.Z., Turnbull, D.M., Reeve, A.K., 2011. Mitochondrial mutations: newly discovered players in neuronal degeneration. Neuroscientist 17, 645–658.

Lee, H.C., Wei, Y.H., 2012. Mitochondria and aging. Adv. Exp. Med. Biol. 942, 311–327.

Lee, J., Giordano, S., Zhang, J., 2012. Autophagy, mitochondria and oxidative stress: cross-talk and redox signalling. Biochem. J. 441, 523–540.

Lee, W.K., Thevenod, F., 2006. A role for mitochondrial aquaporins in cellular life-and-death decisions? Am. J. Physiol. Cell Physiol. 291, C195–C202.

Lenaers, G., Reynier, P., Elachouri, G., Soukkarieh, C., Olichon, A., Belenguer, P., et al., 2009. OPA1 functions in mitochondria and dysfunctions in optic nerve. Int. J. Biochem. Cell Biol. 41, 1866–1874.

Lewin, R., 1987. The unmasking of mitochondrial eve. Science 238, 24–26.

Liao, J., Li, H., Zeng, W., Sauer, D.B., Belmares, R., Jiang, Y., 2012. Structural insight into the ion-exchange mechanism of the sodium/calcium exchanger. Science 335, 686–690.

Lin, S., Jaschke, P.R., Wang, H., Paddock, M., Tufts, A., Allen, J.P., et al., 2009. Electron transfer in the *Rhodobacter sphaeroides* reaction center assembled with zinc bacteriochlorophyll. Proc. Natl. Acad. Sci. USA 106, 8537–8542.

Lin, S.M., Tsai, J.Y., Hsiao, C.D., Huang, Y.T., Chiu, C.L., Liu, M.H., et al., 2012. Crystal structure of a membrane-embedded H^+-translocating pyrophosphatase. Nature 484, 399–403.

Liu, T., O'Rourke, B., 2009. Regulation of mitochondrial Ca^{2+} and its effects on energetics and redox balance in normal and failing heart. J. Bioenerg. Biomembr. 41, 127–132.

Liu, Z., Yan, H., Wang, K., Kuang, T., Zhang, J., Gui, L., et al., 2004. Crystal structure of spinach major light-harvesting complex at 2.72 Å resolution. Nature 428, 287–292.

Locher, K.P., Lee, A.T., Rees, D.C., 2002. The *E. coli* BtuCD structure: a framework for ABC transporter architecture and mechanism. Science 296, 1091–1098.

Locke, R.M., Rial, E., Nicholls, D.G., 1982. Fatty acids as acute regulators of the proton conductance of hamster brown fat mitochondria. Eur. J. Biochem. 129, 373–380.

Loiseau, D., Chevrollier, A., Verny, C., Guillet, V., Gueguen, N., Pou De Crescenzo, M.A., et al., 2007. Mitochondrial coupling defect in Charcot–Marie–Tooth type 2A disease. Ann. Neurol. 61, 315–323.

Lu, M., Fu, D., 2012. Structure–function relationships in P-type ATPases. Science 317, 1746–1748.

Lü, W., Du, J., Schwarzer, N.J., Andrade, S.L.A., Einsle, O., 2013. The Formate/Nitrite Transporter family of anion channels. Biol. Chem. 394, 715–727.

Luecke, H., Schobert, B., Richter, H.T., Cartailler, J.P., Lanyi, J.K., 1999. Structure of bacteriorhodopsin at 1.55 Å resolution. J. Mol. Biol. 291, 899–911.

Lyons, J.A., Aragao, D., Slattery, O., Pisliakov, A.V., Soulimane, T., Caffrey, M., 2012. Structural insights into electron transfer in caa_3-type cytochrome oxidase. Nature 487, 514–518.

Maechler, P., Wollheim, C.B., Bentzen, C.L., Niesor, E., 1992. Role of the intestinal acyl-CoA:cholesterol acyltransferase activity in the hyperresponse of diabetic rats to dietary cholesterol. J. Lipid Res. 33, 1475–1484.

Maechler, P., Li, N., Casimir, M., Vetterli, L., Frigerio, F., Brun, T., 2010. Role of mitochondria in β-cell function and dysfunction. Adv. Exp. Med. Biol. 654, 193–216.

Maklashina, E., Cecchini, G., 2010. The quinone-binding and catalytic site of complex II. Biochim. Biophys. Acta 1797, 1877–1882.

Malnoe, A., Wollman, F.-A., de Vitry, C., Rappaport, F., 2011. Photosynthetic growth despite a broken Q-cycle. Nat. Commun. 2 (301), 1–6.

Martin, L.J., 2012. Biology of mitochondria in neurodegenerative diseases. Prog. Mol. Biol. Transl. Sci. 107, 355–415.

Martinez, T.N., Greenamyre, J.T., 2012. Toxin models of mitochondrial dysfunction in Parkinson's disease. Antioxid. Redox Signal. 16, 920–934.

Matthies, D., Haberstock, S., Joos, F., Dotsch, V., Vonck, J., Bernhard, F., et al., 2011. Cell-free expression and assembly of ATP synthase. J. Mol. Biol. 413, 593–603.

McCarty, M.F., 2005. Up-regulation of PPARgamma coactivator-1α as a strategy for preventing and reversing insulin resistance and obesity. Med. Hypotheses 64, 399–407.

McKenzie, M., Liolitsa, D., Akinshina, N., Campanella, M., Sisodiya, S., Hargreaves, I., et al., 2007. Mitochondrial ND5 gene variation associated with encephalomyopathy and mitochondrial ATP consumption. J. Biol. Chem. 282, 36845–36852.

McManus, M.J., Murphy, M.P., Franklin, J.L., 2011. The mitochondria-targeted antioxidant MitoQ prevents loss of spatial memory retention and early neuropathology in a transgenic mouse model of Alzheimer's disease. J. Neurosci. 31, 15703–15715.

Menalled, L.B., Chesselet, M.F., 2002. Mouse models of Huntington's disease. Trends Pharmacol. Sci. 23, 32–39.

Menz, R.I., et al., 2001. Structure of bovine mitochondrial F1-ATPase with nucleotide bound to all three catalytic sites: implications for the mechanism of rotary catalysis. Cell 106, 331–341.

Merritt, E.A., Stout, G.H., Turley, S., Sieker, L.C., Jehsen, L.H., Orme-Johnson, W.H., 1993. Structure at pH 6.5 of ferredoxin I from *Azotobacter vinelandii* at 2.3Å resolution. Acta Crystallogr. D Biol. Crystallogr. 49, 272–281.

Mimaki, M., Wang, X., McKenzie, M., Thorburn, D.R., Ryan, M.T., 2011. Understanding mitochondrial complex I assembly in health and disease. Biochim. Biophys. Acta 1817, 851–862.

Mitchell, P., 1961. Coupling of phosphorylation to electron and hydrogen transfer by a chemiosmotic type of mechanism. Nature 191, 144–148.

Mitchell, P., 1966. Chemiosmotic Coupling in Oxidative and Photosynthetic Phosphorylation. Glynn Research, Bodmin, UK.

Mitchell, P., 2011. Chemiosmotic coupling in oxidative and photosynthetic phosphorylation. Biochim. Biophys. Acta 1807, 1507–1538.

Mitchell, P., Moyle, J., 1967. Respiration-driven proton translocation in rat liver mitochondria. Biochem. J. 105, 1147–1162.

Mitchell, P., Moyle, J., 1969. Estimation of membrane potential and pH difference across the cristae membrane of rat liver mitochondria. Eur. J. Biochem. 7, 471–484.

Mootha, V.K., Lindgren, C.M., Eriksson, K.F., Subramanian, A., Sihag, S., Lehar, J., et al., 2003. PGC-1α-responsive genes involved in oxidative phosphorylation are coordinately downregulated in human diabetes. Nat. Genet. 34, 267–273.

Morgan, B., Sobotta, M.C., Dick, T.P., 2011. Measuring E(GSH) and H_2O_2 with roGFP$_2$-based redox probes. Free Radic. Biol. Med. 51, 1943–1951.

Morgan, J.E., Gennis, R.B., Maeda, A., 2008. A role for internal water molecules in proton affinity changes in the Schiff base and Asp85 for one-way proton transfer in bacteriorhodopsin. Photochem. Photobiol. 84, 1038–1045.

Morino, K., Petersen, K.F., Shulman, G.I., 2006. Molecular mechanisms of insulin resistance in humans and their potential links with mitochondrial dysfunction. Diabetes 55 (**Suppl 2**), S9–S15.

Morino, M., Natsui, S., Ono, T., Swartz, T.H., Krulwich, T.A., Ito, M., 2010. Single site mutations in the hetero-oligomeric Mrp antiporter from alkaliphilic *Bacillus pseudofirmus* OF4 that affect Na^+/H^+ antiport activity, sodium exclusion, individual Mrp protein levels, or Mrp complex formation. J. Biol. Chem. 285, 30942–30950.

Moroni, F., 2008. Poly(ADP-ribose)polymerase 1 (PARP-1) and postischemic brain damage. Curr. Opin. Pharmacol. 8, 96–103.

Moser, C.C., Farid, T.A., Chobot, S.E., Dutton, P.L., 2006. Electron tunneling chains of mitochondria. Biochim. Biophys. Acta 1757, 1096.

Muench, S.P., Trinick, J., Harrsion, M.A., 2011. Structural divergence of rotary ATPases. Quart. Rev. Biophys. 44, 311–356.

Mulder, H., Ling, C., 2009. Mitochondrial dysfunction in pancreatic β-cells in type 2 diabetes. Mol. Cell. Endocrinol. 297, 34–40.

Muller, M., Mentel, M., Van Hellemond, J.J., Henze, K., Woehle, C., Gould, S.B., et al., 2012. Biochemistry and evolution of anaerobic energy metabolism in eukaryotes. Microbiol. Mol. Biol. Rev. 76, 444–495.

Murakami, S., Nakashima, R., Yamashita, E., Matsumoto, T., Yamaguchi, A., 2006. Crystal structures of a multidrug transporter reveal a functionally rotating mechanism. Nature 443, 173–179.

Murphy, M.P., 2009. How mitochondria produce reactive oxygen species. Biochem. J. 417, 1–13.

Murphy, M.P., 2011. Mitochondrial thiols in antioxidant protection and redox signalling: distinct roles for glutathionylation and other thiol modifications. Antioxid. Redox Signal. 16, 476–484.

Murphy, M.P., Holmgren, A., Larsson, N.G., Halliwell, B., Chang, C.J., Kalyanaraman, B., et al., 2011. Unraveling the biological roles of reactive oxygen species. Cell Metab. 13, 361–366.

Narendra, D.P., Youle, R.J., 2011. Targeting mitochondrial dysfunction: role for PINK1 and Parkin in mitochondrial quality control. Antioxid. Redox Signal. 14, 1929–1938.

Nedergaard, J., Cannon, B., 2003. The 'novel' 'uncoupling' proteins UCP2 and UCP3: what do they really do? Pros and cons for suggested functions. Exp. Physiol. 88, 65–84.

Neupert, W., Herrmann, J.M., 2007. Translocation of proteins into mitochondria. Annu. Rev. Biochem. 76, 723–749.

Newsholme, P., Gaudel, C., Krause, M., 2012. Mitochondria and diabetes: an intriguing pathogenetic role. Adv. Exp. Med. Biol. 942, 235–247.

Newstead, S., Drew, D., Cameron, A.D., Postis, V.L., Xia, X., Fowler, P.W., et al., 2011. Crystal structure of a prokaryotic homologue of the mammalian oligopeptide-proton symporters, PepT1 and PepT2. EMBO J. 30, 417–426.

Nicholls, D.G., 1974. The influence of respiration and ATP hydrolysis on the proton electrochemical potential gradient across the inner membrane of rat liver mitochondria as determined by ion distribution. Eur. J. Biochem. 50, 305–315.

Nicholls, D.G., 2005a. Commentary on: 'Old and new data, new issues: the mitochondrial $\Delta\psi$' by H. Tedeschi. Biochim. Biophys. Acta 1710, 63–65.

Nicholls, D.G., 2005b. Mitochondria and calcium signalling. Cell Calcium 38, 311–317.

Nicholls, D.G., 2006a. Simultaneous monitoring of ionophore- and inhibitor-mediated plasma and mitochondrial membrane potential changes in cultured neurons. J. Biol. Chem. 281, 14864–14874.

Nicholls, D.G., 2006b. The physiological regulation of uncoupling proteins. Biochim. Biophys. Acta 1757, 459–466.

Nicholls, D.G., 2008a. Oxidative stress and energy crises in neuronal dysfunction. Ann. N. Y. Acad. Sci. 1147, 53–60.

Nicholls, D.G., 2008b. The Peter Pitchell medal lecture: forty years of Pitchell's proton circuit: from little grey books to little grey cells. Biochim. Biophys. Acta 1777, 550–556.

Nicholls, D.G., 2012. Fluorescence measurement of mitochondrial membrane potential changes in cultured cells. Methods Mol. Biol. 810, 119–133.

Nicholls, D.G., Chalmers, S., 2004. The integration of mitochondrial calcium transport and storage. J. Bioenerg. Biomembr. 36, 277–281.

Nield, J., Barber, J., 2006. Refinement of the structural model for the photosystem II supercomplex of higher plants. Biochim. Biophys. Acta 1757, 353–361.

Nogueiras, R., Habegger, K.M., Chaudhary, N., Finan, B., Banks, A.S., Dietrich, M.O., et al., 2012. Sirtuin 1 and sirtuin 3: physiological modulators of metabolism. Physiol. Rev. 92, 1479–1514.

Noinaj, N., Guillier, M., Barnard, T.J., Buchanan, S.K., 2010. TonB-dependent transporters: regulation, structure, and function. Ann. Rev. Microbiol. 64, 43–60.

Nowikovsky, K., Schweyen, R.J., Bernardi, P., 2009. Pathophysiology of mitochondrial volume homeostasis: potassium transport and permeability transition. Biochim. Biophys. Acta 1787, 345–350.

Okita, K., Yamanaka, S., 2011. Induced pluripotent stem cells: opportunities and challenges. Philos. Trans. R. Soc. London B Biol. Sci. 366, 2198–2207.

Oliveira, J., Ellerby, L.M., Rego, A.C., Nicholls, D.G., 2007. Mitochondrial dysfunction in Huntington's disease: the bioenergetics of isolated and *in-situ* mitochondria from transgenic mice. J. Neurochem. 101, 241–249.

O'Rourke, B., Blatter, L.A., 2009. Mitochondrial Ca^{2+} uptake: tortoise or hare? J. Mol. Cell. Cardiol. 46, 767–774.

Pagani, L., Eckert, A., 2011. Amyloid-β interaction with mitochondria. Int. J. Alzheimer's Dis. 2011, 925050.

Palmer, T., Berks, B.C., 2012. The twin-arginine translocation (Tat) protein export pathway. Nat. Rev. Microbiol. 10, 483–496.

Palmieri, F., Pierri, C.L., 2010. Mitochondrial metabolite transport. Essays Biochem. 47, 37–52.

Palty, R., Sekler, I., 2012. The mitochondrial Na^{+}/Ca^{2+} exchanger. Cell Calcium 52, 9–15.

Palty, R., Silverman, W.F., Hershfinkel, M., Caporale, T., Sensi, S.L., Parnis, J., et al., 2010. NCLX is an essential component of mitochondrial Na^{+}/Ca^{2+} exchange. Proc. Natl. Acad. Sci. USA 107, 436–441.

Papa, S., Rasmo, D.D., Technikova-Dobrova, Z., Panelli, D., Signorile, A., Scacco, S., et al., 2012. Respiratory chain complex I, a main regulatory target of the cAMP/PKA pathway is defective in different human diseases. FEBS Lett. 586, 568–577.

Park, E., Rapoport, T.A., 2012. Mechanisms of Sec61/SecY-mediated protein translocation across membranes. Annu. Rev. Biophys. 41, 21–40.

Parsons, M.J., Green, D.R., 2010. Mitochondria in cell death. Essays Biochem. 47, 99–114.

Pebay-Peyroula, E., Dahout-Gonzalez, C., Kahn, R., Trézéguet, V., Lauquin, G.J.M., Brandolin, R., 2003. Structure of mitochondrial ADP/ATP carrier in complex with carboxyatractyloside. Nature 426, 39–44.

Pedrini, S., Sau, D., Guareschi, S., Bogush, M., Brown Jr., R.H., Naniche, N., et al., 2010. ALS-linked mutant SOD1 damages mitochondria by promoting conformational changes in Bcl-2. Hum. Mol. Genet. 19, 2974–2986.

Pellegrini, L., Scorrano, L., 2007. A cut short to death: parl and opa1 in the regulation of mitochondrial morphology and apoptosis. Cell Death Differ. 14, 1275–1284.

Perez-Pinzon, M.A., Stetler, R.A., Fiskum, G., 2012. Novel mitochondrial targets for neuroprotection. J. Cereb. Blood Flow Metab. 32, 1362–1376.

Perkins, G.A., Tjong, J., Brown, J.M., Poquiz, P.H., Scott, R.T., Kolson, D.R., et al., 2010. The micro-architecture of mitochondria at active zones: electron tomography reveals novel anchoring scaffolds and cristae structured for high-rate metabolism. J. Neurosci. 30, 1015–1026.

Perry, G.M., Tallaksen-Greene, S., Kumar, A., Heng, M.Y., Kneynsberg, A., van, G.T., et al., 2010. Mitochondrial calcium uptake capacity as a therapeutic target in the R6/2 mouse model of Huntington's disease. Hum. Mol. Genet. 19, 3354–3371.

Phillips, D., Aponte, A.M., Covian, R.G., Balaban, R.S., 2011. Intrinsic protein kinase activity of mitochondrial oxidative phosphorylation complexes. Biochemistry 50, 2515–2529.

Pi, J., Bai, Y., Daniel, K.W., Liu, D., Lyght, O., Edelstein, D., et al., 2009. Persistent oxidative stress due to absence of uncoupling protein 2 associated with impaired pancreatic beta-cell function. Endocrinology 150, 3040–3048.

Pilsl, A., Winklhofer, K.F., 2011. Parkin, PINK1 and mitochondrial integrity: emerging concepts of mitochondrial dysfunction in Parkinson's disease. Acta Neuropathol. 123, 173–188.

Pogoryelov, D., Yildiz, O., Faraldo-Gomez, J.D., Meier, T., 2009. High-resolution structure of the rotor ring of a proton-dependent ATP synthase. Nat. Struct. Mol. Biol. 16, 1068–1073.

Pogoryelov, D., Klyszejko, A.L., Krasnoselska, G.O., Heller, E.M., Leone, V., Langer, J.D., et al., 2012. Engineering rotor ring stoichiometries in the ATP synthase. Proc. Natl. Acad. Sci. USA 109, E1599–E1608.

Popov, V., Medvedev, N.I., Davies, H.A., Stewart, M.G., 2005. Mitochondria form a filamentous reticular network in hippocampal dendrites but are present as discrete bodies in axons: a three-dimensional ultrastructural study. J. Comp. Neurol. 492, 50–65.

Prebble, J., 2002. Peter Mitchell and the ox phos wars. Trends Biochem. Sci. 27, 209–212.

Prentki, M., Nolan, C.J., 2006. Islet beta cell failure in type 2 diabetes. J. Clin. Invest. 116, 1802–1812.

Price, N.C., Dwek, R.A., Ratcliffe, R.G., Wormald, M., 2001. Principles and Problems in Physical Chemistry for Biochemists. Oxford University Press, New York.

Pryde, K.R., Hirst, J., 2011. Superoxide is produced by the reduced flavin in mitochondrial complex I: a single, unified mechanism that applies during both forward and reverse electron transfer. J. Biol. Chem. 286, 18056–18065.

Quinlan, C.L., Gerencser, A.A., Treberg, J.R., Brand, M.D., 2011. The mechanism of superoxide production by the antimycin-inhibited mitochondrial Q-cycle. J. Biol. Chem. 286, 31361–31372.

Quinlan, C.L., Treberg, J.R., Perevoshchikova, I.V., Orr, A.L., Brand, M.D., 2012. Native rates of superoxide production from multiple sites in isolated mitochondria measured using endogenous reporters. Free Radic. Biol. Med. 53, 1807–1817.

Rabl, R., Soubannier, V., Scholz, R., Vogel, F., Mendl, N., Vasiljev-Neumeyer, A., et al., 2009. Formation of cristae and crista junctions in mitochondria depends on antagonism between Fcj1 and Su e/g. J. Cell Biol. 185, 1047–1063.

Radi, R., Cassina, A., Hodara, R., Quijano, C., Castro, L., 2002. Peroxynitrite reactions and formation in mitochondria. Free Radic. Biol. Med. 33, 1451–1464.

Ralph, S.J., Rodriguez-Enriquez, S., Neuzil, J., Saavedra, E., Moreno-Sanchez, R., 2010. The causes of cancer revisited: "Mitochondrial malignancy" and ROS-induced oncogenic transformation—Why mitochondria are targets for cancer therapy. Mol. Aspects Med. 31, 145–170.

Rapoport, S.I., 2003. Coupled reductions in brain oxidative phosphorylation and synaptic function can be quantified and staged in the course of Alzheimer's disease. Neurotox. Res. 5, 385–398.

Ravnskjaer, K., Boergesen, M., Rubi, B., Larsen, J.K., Nielsen, T., Fridriksson, J., et al., 2005. Peroxisome proliferator-activated receptor α (PPARα) potentiates, whereas PPARγ attenuates, glucose-stimulated insulin secretion in pancreatic beta-cells. Endocrinology 146, 3266–3276.

Ravussin, E., Kozak, L.P., 2009. Have we entered the brown adipose tissue renaissance? Obes. Rev. 10, 265–268.

Ray, P.D., Huang, B.W., Tsuji, Y., 2012. Reactive oxygen species (ROS) homeostasis and redox regulation in cellular signaling. Cell Signal. 24, 981–990.

Rees, D.M., Montgomery, M.G., Leslie, A.G., Walker, J.E., 2012. Structural evidence of a new catalytic intermediate in the pathway of ATP hydrolysis by F_1-ATPase from bovine heart mitochondria. Proc. Natl. Acad. Sci. USA 109, 11139–11143.

Rial, E., Poustie, E.A., Nicholls, D.G., 1983. Brown adipose tissue mitochondria: the regulation of the 32,000 Mr uncoupling protein by fatty acids and purine nucleotides. Eur. J. Biochem. 137, 197–203.

Rich, P.R., Marechal, A., 2012. Electron transfer chains: structures, mechanisms and energy coupling. Comp. Biophys. 8, 73–93.

Richardson, D.J., Butt, J.N., Fredrickson, J.K., Zachara, J.M., Shi, L., Edwards, M.J., et al., 2012. The "porin-cytochrome" model for microbe-to-mineral electron transfer. Mol. Microbiol. 85, 201–212.

Ristow, M., Zarse, K., 2010. How increased oxidative stress promotes longevity and metabolic health: the concept of mitochondrial hormesis (mitohormesis). Exp. Gerontol. 45, 410–418.

Robinson, A.J., Overy, C., Kunji, E.R., 2008. The mechanism of transport by mitochondrial carriers based on analysis of symmetry. Proc. Natl. Acad. Sci. USA 105, 17766–17771.

Rochet, J.C., Hay, B.A., Guo, M., 2012. Molecular insights into Parkinson's disease. Prog. Mol. Biol. Transl. Sci. 107, 125–188.

Rollauer, S.E., et al., 2012. Structure of the TatC core of the twin-arginine protein transport system. Nature 492, 210–214.

Rutter, G.A., 2004. Visualising insulin secretion: the minkowski lecture 2004. Diabetologia 47, 1861–1872.

Sack, G.H., 2011. Introduction to the minireviews series on mitochondrial matters in amyotrophic lateral sclerosis, Lou Gehrig's disease. J. Bioenerg. Biomembr. 43, 565–567.

Safiulina, D., Kaasik, A., Seppet, E., Peet, N., Zharkovsky, A., Seppet, E., 2004. Method for *in situ* detection of the mitochondrial function in neurons. J. Neurosci. Methods 137, 87–95.

Saft, C., Zange, J., Andrich, J., Muller, K., Lindenberg, K., Landwehrmeyer, B., et al., 2005. Mitochondrial impairment in patients and asymptomatic mutation carriers of Huntington's disease. Mov. Disord. 20, 674–679.

Sahin, E., DePinho, R.A., 2012. Axis of ageing: telomeres, p53 and mitochondria. Nat. Rev. Mol. Cell Biol. 13, 397–404.

Santos, R., Lefevre, S., Sliwa, D., Seguin, A., Camadro, J.M., Lesuisse, E., 2010. Friedreich's ataxia: molecular mechanisms, redox considerations and therapeutic opportunities. Antioxid. Redox Signal. 13, 651–690.

Sanz, A., Fernandez-Ayala, D.J., Stefanatos, R.K., Jacobs, H.T., 2010. Mitochondrial ROS production correlates with, but does not directly regulate lifespan in *Drosophila*. Aging 2, 200–223.

Saraste, M., 1999. Oxidative phosphorylation at the *fin de siècle*. Science 283, 1488–1493.

Saroussi, S., Schushan, M., Ben-Tal, N., Junge, W., Nelson, N., 2012. Structure and flexibility of the C-ring in the electromotor of rotary F_oF_1-ATPase of pea chloroplasts. PLoS ONE 7, e43045.

Satrustegui, J., Pardo, B., Del, A.A., 2007. Mitochondrial transporters as novel targets for intracellular calcium signaling. Physiol. Rev. 87, 29–67.

Sazanov, L.A., Hinchliffe, P., 2006. Structure of the hydrophilic domain of respiratory complex I from *Thermus thermophilus*. Science 311, 1430–1436.

Scaduto, R.C., Grotyohann, L.W., 1999. Measurement of mitochondrial membrane potential using fluorescent rhodamine derivatives. Biophys. J. 76, 469–477.

Scarpulla, R.C., 2008. Nuclear control of respiratory chain expression by nuclear respiratory factors and PGC-1-related coactivator. Ann. N. Y. Acad. Sci. 1147, 321–334.

Scarpulla, R.C., 2011. Metabolic control of mitochondrial biogenesis through the PGC-1 family regulatory network. Biochim. Biophys. Acta 1813, 1269–1278.

Schafer, F.Q., Buettner, G.R., 2001. Redox environment of the cell as viewed through the redox state of the glutathione disulfide/glutathione couple. Free Radic. Biol. Med. 30, 1191–1212.

Schapira, A.H., 2012. Mitochondrial diseases. Lancet 379, 1825–1834.

Schieke, S.M., Phillips, D., McCoy Jr., J.P., Aponte, A.M., Shen, R.F., Balaban, R.S., et al., 2006. The mTOR pathway regulates mitochondrial oxygen consumption and oxidative capacity. J. Biol. Chem. 281, 27643–27652.

Scott, I., Webster, B.R., Li, J.H., Sack, M.N., 2012. Identification of a molecular component of the mitochondrial acetyl transferase program; a novel role for GCN5L1. Biochem. J. 443, 655–661.

Sears, I.B., MacGinnitie, M.A., Kovacs, L.G., Graves, R.A., 1996. Differentiation-dependent expression of the brown adipocyte uncoupling protein gene: regulation by peroxisome proliferator-activated receptor gamma. Mol. Cell Biol. 16, 3410–3419.

Semenza, G.L., 2007. Oxygen-dependent regulation of mitochondrial respiration by hypoxia-inducible factor 1. Biochem. J. 405, 1–9.

Serviddio, G., Sastre, J., 2010. Measurement of mitochondrial membrane potential and proton leak. Methods Mol. Biol. 594, 107–121.

Shiba, T., Kido, Y., Sakamoto, K., et al., 2013. Structure of the trypanosome cyanide-insensitive alternative oxidase. Proc. Natl. Acad. Sci. U.S.A. 110, 4580–4585.

Shibata, N., Inoue, T., Nagano, C., Nishio, N., Kohzuma, T., Onodera, K., et al., 1999. Novel insight into the copper-ligand geometry in the crystal structure of *Ulva pertusa* plastocyanin at 1.6-A resolution: structural basis for regulation of the copper site by residue 88. J. Biol. Chem. 274, 4225–4230.

Shigeto, M., Katsura, M., Matsuda, M., Ohkuma, S., Kaku, K., 2006. First phase of glucose-stimulated insulin secretion from MIN 6 cells does not always require extracellular calcium influx. J. Pharmacol. Sci. 101, 293–302.

Shimamura, T., Weyand, S., Beckstein, O., Rutherford, N.G., Hadden, J.M., Sharples, D., et al., 2010. Molecular basis of alternating access membrane transport by the sodium-hydantoin transporter Mhp1. Science 328, 470–473.

Shimomura, K., Galvanovskis, J., Goldsworthy, M., Hugill, A., Kaizak, S., Lee, A., et al., 2009. Insulin secretion from beta-cells is affected by deletion of nicotinamide nucleotide transhydrogenase. Methods Enzymol. 457, 451–480.

Shin, J.H., Ko, H.S., Kang, H., Lee, Y., Lee, Y.I., Pletinkova, O., et al., 2011. PARIS (ZNF746) repression of PGC-1α contributes to neurodegeneration in Parkinson's disease. Cell 144, 689–702.

Shoshan-Barmatz, V., De, P.V., Zweckstetter, M., Raviv, Z., Keinan, N., Arbel, N., 2010. VDAC, a multi-functional mitochondrial protein regulating cell life and death. Mol. Aspects Med. 31, 227–285.

Shoubridge, E.A., Wai, T., 2007. Mitochondrial DNA and the mammalian oocyte. Curr. Top. Dev. Biol. 77, 87–111.

Shoubridge, E.A., Wai, T., 2008. Medicine. Sidestepping mutational meltdown. Science 319, 914–915.

Shuttleworth, C.W., 2010. Use of NAD(P)H and flavoprotein autofluorescence transients to probe neuron and astrocyte responses to synaptic activation. Neurochem. Int. 56, 379–386.

Smith, J.C., 1990. Potential-sensitive molecular probes in membranes of bioenergetic relevance. Biochim. Biophys. Acta 1016, 1–28.

Smith, R.A., Murphy, M.P., 2011. Mitochondria-targeted antioxidants as therapies. Discov. Med. 11, 106–114.

Soares, P., Ermini, L., Thomson, N., Mormina, M., Rito, T., Rohl, A., et al., 2009. Correcting for purifying selection: an improved human mitochondrial molecular clock. Am. J. Hum. Genet. 84, 740–759.

Solaini, G., Harris, D.A., 2005. Biochemical dysfunction in heart mitochondria exposed to ischaemia and reperfusion. Biochem. J. 390, 377–394.

Solcan, N., Kwok, J., Fowler, P.W., Cameron, A.D., Drew, D., Iwata, S., et al., 2012. Alternating access mechanism in the POT family of oligopeptide transporters. EMBO J. 31, 3411–3421.

Soubannier, V., McBride, H.M., 2009. Positioning mitochondrial plasticity within cellular signaling cascades. Biochim. Biophys. Acta 1793, 154–170.

Sowa, Y., Berry, R.M., 2012. The rotary bacterial flagella motor. Comp. Biophys. 8, 50–71.

Spetzler, D., Ishmukhametov, R., Hornung, T., Day, L.J., Martin, J., Frasch, W.D., 2009. Single molecule measurements of F1-ATPase reveal an interdependence between the power stroke and the dwell duration. Biochemistry 48, 7979–7985.

Stanika, R.I., Winters, C.A., Pivovarova, N.B., Andrews, S.B., 2010. Differential NMDA receptor-dependent calcium loading and mitochondrial dysfunction in CA1 vs. CA3 hippocampal neurons. Neurobiol. Dis. 37, 403–411.

Stavrovskaya, I.G., Kristal, B.S., 2005. The powerhouse takes control of the cell: is the mitochondrial permeability transition a viable therapeutic target against neuronal dysfunction and death? Free Radic. Biol. Med. 38, 687–697.

Stewart, J.B., Freyer, C., Elson, J.L., Wredenberg, A., Cansu, Z., Trifunovic, A., et al., 2008. Strong purifying selection in transmission of mammalian mitochondrial DNA. PLoS Biol. 6, e10.

Supale, S., Li, N., Brun, T., Maechler, P., 2012. Mitochondrial dysfunction in pancreatic beta cells. Trends. Endocrinol. Metab. 23, 477–487.

Swerdlow, R.H., 2011. Does mitochondrial DNA play a role in Parkinson's disease? A review of cybrid and other supportive evidence. Antioxid. Redox Signal. 16, 950–964.

Swierczek, M., Cieluch, E., Sarewicz, M., Borek, A., Moser, C.C., Dutton, P.L., et al., 2010. An electronic bus bar lies in the core of cytochrome bc_1. Science 329, 451–454.

Symersky, J., Osowski, D., Walters, D.E., Mueller, D.M., 2012a. Oligomycin frames a common drug-binding site in the ATP synthase. Proc. Natl. Acad. Sci. USA 109, 13961–13965.

Symersky, J., Pagadala, V., Osowski, D., Krah, A., Meier, T., Faraldo-Gomez, J.D., et al., 2012b. Structure of the c_{10} ring of the yeast mitochondrial ATP synthase in the open conformation. Nat. Struct. Mol. Biol. 19, 485–491. S1

Szendroedi, J., Phielix, E., Roden, M., 2012. The role of mitochondria in insulin resistance and type 2 diabetes mellitus. Nat. Rev. Endocrinol. 8, 92–103.

Takeda, K., Matsui, Y., Kamiya, N., Adachi, S., Okumura, H., Kouyama, T., 2004. Crystal structure of the M intermediate of bacteriorhodopsin: allosteric structural changes mediated by sliding movement of a transmembrane helix. J. Mol. Biol. 341, 1023–1037.

Taylor, C.T., 2008. Mitochondria and cellular oxygen sensing in the HIF pathway. Biochem. J. 409, 19–26.

Tedeschi, H., 2005. Old and new data, new issues: the mitochondrial DeltaPsi 17. Biochim. Biophys. Acta 1709, 195–202.

Terzioglu, M., Galter, D., 2008. Parkinson's disease: genetic versus toxin-induced rodent models. FEBS J. 275, 1384–1391.

Thauer, R.K., Kaster, A.K., Seedorf, H., Buckel, W., Hedderich, R., 2008. Methanogenic archaea: ecologically relevant differences in energy conservation. Nat. Rev. Microbiol. 6, 579–591.

Toyabe, S., Watanabe-Nakayama, T., Okamoto, T., Kudo, S., Muneyuki, E., 2011. Thermodynamic efficiency and mechanochemical coupling of F_1-ATPase. Proc. Natl. Acad. Sci. USA 108, 17951–17956.

Treberg, J.R., Quinlan, C.L., Brand, M.D., 2011. Evidence for two sites of superoxide production by mitochondrial NADH-ubiquinone oxidoreductase (complex I). J. Biol. Chem. 286, 27103–27110.

Trifunovic, A., Wredenberg, A., Falkenberg, M., Spelbrink, J.N., Rovio, A.T., Bruder, C.E., et al., 2004. Premature ageing in mice expressing defective mitochondrial DNA polymerase. Nature 429, 417–423.

Trouillard, M., Meunier, B., Rappaport, F., 2011. Questioning the functional relevance of mitochondrial supercomplexes by time-resolved analysis of the respiratory chain. Proc. Natl. Acad. Sci. U.S.A. 108, E1027–E1034.

Tsika, E., Moore, D.J., 2012. Mechanisms of LRRK2-mediated neurodegeneration. Curr. Neurol. Neurosci. Rep. 12, 251–260.

Twig, G., Graf, S.A., Wikstrom, J.D., Mohamed, H., Haigh, S.E., Elorza, A.G., et al., 2006. Tagging and tracking individual networks within a complex mitochondrial web using photoactivatable GFP. Am. J. Physiol. Cell Physiol. 291, C176–C184.

Twig, G., Elorza, A., Molina, A.J., Mohamed, H., Wikstrom, J.D., Walzer, G., et al., 2008a. Fission and selective fusion govern mitochondrial segregation and elimination by autophagy. EMBO J. 27, 433–446.

Twig, G., Hyde, B., Shirihai, O.S., 2008b. Mitochondrial fusion, fission and autophagy as a quality control axis: the bioenergetic view. Biochim. Biophys. Acta 1777, 1092–1097.

Umena, Y., Kawakami, K., Shen, J.R., Kamiya, N., 2011. Crystal structure of oxygen-evolving photosystem II at a resolution of 1.9Å. Nature 473, 55–60.

Usukura, E., Suzuki, T., Furuike, S., Soga, N., Saita, E., Hisabori, T., et al., 2012. Torque generation and utilization in motor enzyme F_0F_1-ATP synthase: half-torque F_1 with short-sized pushrod helix and reduced ATP synthesis by half-torque F_0F_1. J. Biol. Chem. 287, 1884–1891.

van Oven, M., Kayser, M., 2009. Updated comprehensive phylogenetic tree of global human mitochondrial DNA variation. Hum. Mutat. 30, E386–E394.

van Spanning, R.J., Richardson, D.J., Ferguson, S.J., 2012. Introduction to the biochemistry and molecular biology of denitrification. In: Bothe, H., Ferguson, S.J., Newton, W.E. (Eds.), Biology of the Nitrogen Cycle. Elsevier, New York, pp. 3–20.

Vander Heiden, M.G., Cantley, L.C., Thompson, C.B., 2009. Understanding the warburg effect: the metabolic requirements of cell proliferation. Science 324, 1029–1033.

Vaseva, A.V., Moll, U.M., 2009. The mitochondrial p53 pathway. Biochim. Biophys. Acta 1787, 414–420.

Vaubel, R.A., Isaya, G., 2012. Iron–sulfur cluster synthesis, iron homeostasis and oxidative stress in Friedreich ataxia. Mol. Cell Neurosci. [Epub ahead of print]

Vendelbo, M.H., Nair, K.S., 2011. Mitochondrial longevity pathways. Biochim. Biophys. Acta 1813, 634–644.

Venter, J.C., Remington, K., Heidelberg, J.F., Halpern, A.L., Rusch, D., Eisen, J.A., et al., 2004. Environmental genome shotgun sequencing of the Sargasso Sea. Science 304, 66–74.

Verdin, E., Hirschey, M.D., Finley, L.W., Haigis, M.C., 2010. Sirtuin regulation of mitochondria: energy production, apoptosis, and signaling. Trends Biochem. Sci. 35, 669–675.

Viollet, B., Guigas, B., Sanz, G.N., Leclerc, J., Foretz, M., Andreelli, F., 2012. Cellular and molecular mechanisms of metformin: an overview. Clin. Sci.(London) 122, 253–270.

Vithayathil, S.A., Ma, Y., Kaipparettu, B.A., 2012. Transmitochondrial cybrids: tools for functional studies of mutant mitochondria. Methods Mol. Biol. 837, 219–230.

Vives-Bauza, C., Zhou, C., Huang, Y., Cui, M., de Vries, R.L., Kim, J., et al., 2010. PINK1-dependent recruitment of Parkin to mitochondria in mitophagy. Proc. Natl. Acad. Sci. USA 107, 378–383.

von Ballmoos, C., Brunner, J., Dimroth, P., 2004. The ion channel of F-ATP synthase is the target of toxic organotin compounds. Proc. Natl. Acad. Sci. USA 101, 11239–11244.

Walker, J.E., 2013. The ATP synthase: the understood, the uncertain and the unknown. Biochem. Soc. Trans. 41, 1–16.

Wang, C., Youle, R.J., 2009. The role of mitochondria in apoptosis. Annu. Rev. Genet. 43, 95–118.

Wang, X., Petrie, T.G., Liu, Y., Liu, J., Fujioka, H., Zhu, X., 2012. Parkinson's disease-associated DJ-1 mutations impair mitochondrial dynamics and cause mitochondrial dysfunction. J. Neurochem. 121, 830–839.

Warburg, O., Wind, F., Negelein, E., 1927. The metabolism of tumors in the body. J. Gen. Physiol. 8, 519–530.

Ward, A., Reyes, C.L., Yu, J., Roth, C.B., Chang, G., 2007. Flexibility in the ABC transporter MsbA: alternating access with a twist. Proc. Natl. Acad. Sci. USA 104, 19005–19010.

Ward, M.W., Rego, A.C., Frenguelli, B.G., Nicholls, D.G., 2000. Mitochondrial membrane potential and glutamate excitotoxicity in cultured cerebellar granule cells. J. Neurosci. 20, 7208–7219.

Wasilewski, M., Scorrano, L., 2009. The changing shape of mitochondrial apoptosis. Trends Endocrinol. Metab. 20, 287–294.

Watmough, N.J., Frerman, F.E., 2010. The electron transfer flavoprotein: ubiquinone oxidoreductases. Biochim. Biophys. Acta 1797, 1910–1916.

Watt, I.N., Montgomery, M.G., Runswick, M.J., Leslie, A.G., Walker, J.E., 2010. Bioenergetic cost of making an adenosine triphosphate molecule in animal mitochondria. Proc. Natl. Acad. Sci. USA 107, 16823–16827.

Wei, A.C., Liu, T., Winslow, R.L., O'Rourke, B., 2012. Dynamics of matrix-free Ca^{2+} in cardiac mitochondria: two components of Ca^{2+} uptake and role of phosphate buffering. J. Gen. Physiol. 139, 465–478.

Weisova, P., Anilkumar, U., Ryan, C., Concannon, C.G., Prehn, J.H., Ward, M.W., 2012. "Mild mitochondrial uncoupling" induced protection against neuronal excitotoxicity requires AMPK activity. Biochim. Biophys. Acta 1817, 744–753.

Weyand, S., Shimamura, T., Yajima, S., Suzuki, S., Mirza, O., Krusong, K., et al., 2008. Structure and molecular mechanism of a nucleobase-cation-symport-1 family transporter. Science 322, 709–713.

Wiederkehr, A., Park, K.S., Dupont, O., Demaurex, N., Pozzan, T., Cline, G.W., et al., 2009. Matrix alkalinization: a novel mitochondrial signal for sustained pancreatic beta-cell activation. EMBO J. 28, 417–428.

Wilson, C.Y., Rubinstein, J.L., 2012. Subnanometre-resolution structure of the intact *Thermus thermophilus* H^+-driven ATP synthase. Nature 481, 214–218.

Winklhofer, K.F., Haass, C., 2010. Mitochondrial dysfunction in Parkinson's disease. Biochim. Biophys. Acta 1802, 29–44.

Wraight, C.A., 2004. Proton and electron transfer in the acceptor quinone complex of photosynthetic reaction centers from *Rhodobacter sphaeroides*. Front Biosci. 9, 309–337.

Yao, J., Irwin, R.W., Zhao, L., Nilsen, J., Hamilton, R.T., Brinton, R.D., 2009. Mitochondrial bioenergetic deficit precedes Alzheimer's pathology in female mouse model of Alzheimer's disease. Proc. Natl. Acad. Sci. USA 106, 14670–14675.

Yoshikawa, S., 1999. X-ray structure and reaction mechanism of bovine heart cytochrome *c* oxidase. Biochem. Soc. Trans. 27, 351–362.

Zeuthen, T., 2001. How water molecules pass through aquaporins. Trends Biochem. Sci. 26, 77–79.

Zhang, C.Y., Baffy, G., Perret, P., Krauss, S., Peroni, O., Grujic, D., et al., 2001. Uncoupling protein-2 negatively regulates insulin secretion and is a major link between obesity, beta cell dysfunction, and type 2 diabetes. Cell 105, 745–755.

Zhou, N., Gordon, G.R., Feighan, D., MacVicar, B.A., 2010. Transient swelling, acidification, and mitochondrial depolarization occurs in neurons but not astrocytes during spreading depression. Cereb. Cortex 20, 2614–2624.

Zoccarato, F., Nicholls, D.G., 1982. The role of phosphate in the regulation of the Ca efflux pathway of liver mitochondria. Eur. J. Biochem. 127, 333–338.

Zouni, A., Witt, H.T., Kern, J., Fromme, P., Krauss, N., Saenger, W., et al., 2001. Crystal structure of photosystem II from *Synechococcus elongatus* at 3.8 Å resolution. Nature 409, 739–743.