

7. BIBLIOGRAPHY

- Aijaz, S., L. Erskine, G. Jeffery, S.S. Bhattacharya, and M. Votruba. 2004. Developmental expression profile of the optic atrophy gene product: OPA1 is not localized exclusively in the mammalian retinal ganglion cell layer. *Invest Ophthalmol Vis Sci.* 45:1667-73.
- Alexander, C., M. Votruba, U.E. Pesch, D.L. Thiselton, S. Mayer, A. Moore, M. Rodriguez, U. Kellner, B. Leo-Kottler, G. Auburger, S.S. Bhattacharya, and B. Wissinger. 2000. OPA1, encoding a dynamin-related GTPase, is mutated in autosomal dominant optic atrophy linked to chromosome 3q28. *Nat Genet.* 26:211-5.
- Amati-Bonneau, P., A. Guichet, A. Olichon, A. Chevrollier, F. Viala, S. Miot, C. Ayuso, S. Odent, C. Arrouet, C. Verny, M.N. Calmels, G. Simard, P. Belenguer, J. Wang, J.L. Puel, C. Hamel, Y. Malthiery, D. Bonneau, G. Lenaers, and P. Reynier. 2005. OPA1 R445H mutation in optic atrophy associated with sensorineural deafness. *Ann Neurol.* 58:958-63.
- Amati-Bonneau, P., S. Odent, C. Derrien, L. Pasquier, Y. Malthiery, P. Reynier, and D. Bonneau. 2003. The association of autosomal dominant optic atrophy and moderate deafness may be due to the R445H mutation in the OPA1 gene. *Am J Ophthalmol.* 136:1170-1.
- Amchenkova, A.A., L.E. Bakeeva, Y.S. Chentsov, V.P. Skulachev, and D.B. Zorov. 1988. Coupling membranes as energy-transmitting cables. I. Filamentous mitochondria in fibroblasts and mitochondrial clusters in cardiomyocytes. *J Cell Biol.* 107:481-95.
- Antonsson, B., F. Conti, A. Ciavatta, S. Montessuit, S. Lewis, I. Martinou, L. Bernasconi, A. Bernard, J.J. Mermod, G. Mazzei, K. Maundrell, F. Gambale, R. Sadoul, and J.C. Martinou. 1997. Inhibition of Bax channel-forming activity by Bcl-2. *Science.* 277:370-2.
- Arnoult, D., A. Grodet, Y.J. Lee, J. Estaquier, and C. Blackstone. 2005. Release of OPA1 during apoptosis participates in the rapid and complete release of cytochrome c and subsequent mitochondrial fragmentation. *J Biol Chem.* 280:35742-50.
- Ashkenazi, A., and V.M. Dixit. 1998. Death receptors: signaling and modulation. *Science.* 281:1305-8.
- Attardi, G., and G. Schatz. 1988. Biogenesis of mitochondria. *Annu Rev Cell Biol.* 4:289-333.
- Ausubel, F.M., R. Brent, R.E. Kingston, D.D. Moore, J.G. Seidman, J.A. Smith, and K. Struhl. 2000. Current Protocols in Molecular Biology. John Wiley & Sons, Inc., New York, USA.

- Bay, D.C., and D.A. Court. 2002. Origami in the outer membrane: the transmembrane arrangement of mitochondrial porins. *Biochem Cell Biol.* 80:551-62.
- Beaudoing, E., S. Freier, J.R. Wyatt, J.M. Claverie, and D. Gautheret. 2000. Patterns of variant polyadenylation signal usage in human genes. *Genome Res.* 10:1001-10.
- Beaudoing, E., and D. Gautheret. 2001. Identification of alternate polyadenylation sites and analysis of their tissue distribution using EST data. *Genome Res.* 11:1520-6.
- Belmokhtar, C.A., J. Hillion, and E. Segal-Bendirdjian. 2001. Staurosporine induces apoptosis through both caspase-dependent and caspase-independent mechanisms. *Oncogene.* 20:3354-62.
- Benz, R. 1994. Permeation of hydrophilic solutes through mitochondrial outer membranes: review on mitochondrial porins. *Biochim Biophys Acta.* 1197:167-96.
- Bereiter-Hahn, J., and M. Voth. 1994. Dynamics of mitochondria in living cells: shape changes, dislocations, fusion, and fission of mitochondria. *Microsc Res Tech.* 27:198-219.
- Bernardi, P. 1992. Modulation of the mitochondrial cyclosporin A-sensitive permeability transition pore by the proton electrochemical gradient. Evidence that the pore can be opened by membrane depolarization. *J Biol Chem.* 267:8834-9.
- Bernardi, P., R. Colonna, P. Costantini, O. Eriksson, E. Fontaine, F. Ichas, S. Massari, A. Nicolli, V. Petronilli, and L. Scorrano. 1998. The mitochondrial permeability transition. *Biofactors.* 8:273-81.
- Bleazard, W., J.M. McCaffery, E.J. King, S. Bale, A. Mozdy, Q. Tieu, J. Nunnari, and J.M. Shaw. 1999. The dynamin-related GTPase Dnm1 regulates mitochondrial fission in yeast. *Nat Cell Biol.* 1:298-304.
- Boatright, K.M., and G.S. Salvesen. 2003. Mechanisms of caspase activation. *Curr Opin Cell Biol.* 15:725-31.
- Bossy-Wetzel, E., D.D. Newmeyer, and D.R. Green. 1998. Mitochondrial cytochrome c release in apoptosis occurs upstream of DEVD-specific caspase activation and independently of mitochondrial transmembrane depolarization. *Embo J.* 17:37-49.
- Bouchier-Hayes, L., L. Lartigue, and D.D. Newmeyer. 2005. Mitochondria: pharmacological manipulation of cell death. *J Clin Invest.* 115:2640-7.
- Boyer, P.D. 1997. The ATP synthase--a splendid molecular machine. *Annu Rev Biochem.* 66:717-49.
- Brookes, P.S. 2005. Mitochondrial H(+) leak and ROS generation: an odd couple. *Free Radic Biol Med.* 38:12-23.

- Brown, M.D. 1999. The enigmatic relationship between mitochondrial dysfunction and Leber's hereditary optic neuropathy. *J Neurol Sci.* 165:1-5.
- Buchet, K., and C. Godinot. 1998. Functional F1-ATPase essential in maintaining growth and membrane potential of human mitochondrial DNA-depleted rho degrees cells. *J Biol Chem.* 273:22983-9.
- Cai, J., J. Yang, and D.P. Jones. 1998. Mitochondrial control of apoptosis: the role of cytochrome c. *Biochim Biophys Acta.* 1366:139-49.
- Carelli, V., F.N. Ross-Cisneros, and A.A. Sadun. 2002a. Optic nerve degeneration and mitochondrial dysfunction: genetic and acquired optic neuropathies. *Neurochem Int.* 40:573-84.
- Carelli, V., F.N. Ross-Cisneros, and A.A. Sadun. 2004a. Mitochondrial dysfunction as a cause of optic neuropathies. *Prog Retin Eye Res.* 23:53-89.
- Carelli, V., M. Rugolo, G. Sgarbi, A. Ghelli, C. Zanna, A. Baracca, G. Lenaz, E. Napoli, A. Martinuzzi, and G. Solaini. 2004b. Bioenergetics shapes cellular death pathways in Leber's hereditary optic neuropathy: a model of mitochondrial neurodegeneration. *Biochim Biophys Acta.* 1658:172-9.
- Carelli, V., L. Vergani, B. Bernazzi, C. Zampieron, L. Bucchi, M. Valentino, C. Rengo, A. Torroni, and A. Martinuzzi. 2002b. Respiratory function in cybrid cell lines carrying European mtDNA haplogroups: implications for Leber's hereditary optic neuropathy. *Biochim Biophys Acta.* 1588:7-14.
- Cerritelli, S.M., E.G. Frolova, C. Feng, A. Grinberg, P.E. Love, and R.J. Crouch. 2003. Failure to produce mitochondrial DNA results in embryonic lethality in Rnaseh1 null mice. *Mol Cell.* 11:807-15.
- Chan, D.C. 2006. Mitochondrial fusion and fission in mammals. *Annu Rev Cell Dev Biol.* 22:79-99.
- Chen, C.Y., and A.B. Shyu. 1995. AU-rich elements: characterization and importance in mRNA degradation. *Trends Biochem Sci.* 20:465-70.
- Chen, H., A. Chomyn, and D.C. Chan. 2005. Disruption of fusion results in mitochondrial heterogeneity and dysfunction. *J Biol Chem.* 280:26185-92.
- Chen, H., S.A. Detmer, A.J. Ewald, E.E. Griffin, S.E. Fraser, and D.C. Chan. 2003. Mitofusins Mfn1 and Mfn2 coordinately regulate mitochondrial fusion and are essential for embryonic development. *J Cell Biol.* 160:189-200.

- Chester, N., F. Kuo, C. Kozak, C.D. O'Hara, and P. Leder. 1998. Stage-specific apoptosis, developmental delay, and embryonic lethality in mice homozygous for a targeted disruption in the murine Bloom's syndrome gene. *Genes Dev.* 12:3382-93.
- Cipolat, S., O. Martins de Brito, B. Dal Zilio, and L. Scorrano. 2004. OPA1 requires mitofusin 1 to promote mitochondrial fusion. *Proc Natl Acad Sci U S A.* 101:15927-32.
- Cipolat, S., T. Rudka, D. Hartmann, V. Costa, L. Serneels, K. Craessaerts, K. Metzger, C. Frezza, W. Annaert, L. D'Adamio, C. Derkx, T. Dejaegere, L. Pellegrini, R. D'Hooge, L. Scorrano, and B. De Strooper. 2006. Mitochondrial rhomboid PARL regulates cytochrome c release during apoptosis via OPA1-dependent cristae remodeling. *Cell.* 126:163-75.
- Clayton, D.A. 2000. Vertebrate mitochondrial DNA-a circle of surprises. *Exp Cell Res.* 255:4-9.
- Colgan, D.F., and J.L. Manley. 1997. Mechanism and regulation of mRNA polyadenylation. *Genes Dev.* 11:2755-66.
- Collins, T.J., M.J. Berridge, P. Lipp, and M.D. Bootman. 2002. Mitochondria are morphologically and functionally heterogeneous within cells. *Embo J.* 21:1616-27.
- Colombini, M. 1979. A candidate for the permeability pathway of the outer mitochondrial membrane. *Nature.* 279:643-5.
- Colombini, M. 1983. Purification of VDAC (voltage-dependent anion-selective channel) from rat liver mitochondria. *J Membr Biol.* 74:115-21.
- Cory, S., and J.M. Adams. 2002. The Bcl2 family: regulators of the cellular life-or-death switch. *Nat Rev Cancer.* 2:647-56.
- D'Herde, K., B. De Prest, S. Mussche, P. Schotte, R. Beyaert, R.V. Coster, and F. Roels. 2000. Ultrastructural localization of cytochrome c in apoptosis demonstrates mitochondrial heterogeneity. *Cell Death Differ.* 7:331-7.
- Daems, W.T., and E. Wisse. 1966. Shape and attachment of the cristae mitochondriales in mouse hepatic cell mitochondria. *J Ultrastruct Res.* 16:123-40.
- Danial, N.N., and S.J. Korsmeyer. 2004. Cell death: critical control points. *Cell.* 116:205-19.
- Danino, D., and J.E. Hinshaw. 2001. Dynamin family of mechanoenzymes. *Curr Opin Cell Biol.* 13:454-60.
- De Giorgi, F., L. Lartigue, and F. Ichas. 2000. Electrical coupling and plasticity of the mitochondrial network. *Cell Calcium.* 28:365-70.

- de Oliveira Miguel, N.C., V.B. Meyer-Rochow, and S. Allodi. 2003. A structural study of the retinal photoreceptor, plexiform and ganglion cell layers following exposure to UV-B and UV-C radiation in the albino rat. *Micron*. 34:395-404.
- De Vos, K., V. Goossens, E. Boone, D. Vercammen, K. Vancompernolle, P. Vandenabeele, G. Haegeman, W. Fiers, and J. Grootenhuis. 1998. The 55-kDa tumor necrosis factor receptor induces clustering of mitochondria through its membrane-proximal region. *J Biol Chem*. 273:9673-80.
- Delettre, C., J.M. Griffoin, J. Kaplan, H. Dollfus, B. Lorenz, L. Faivre, G. Lenaers, P. Belenguer, and C.P. Hamel. 2001. Mutation spectrum and splicing variants in the OPA1 gene. *Hum Genet*. 109:584-91.
- Delettre, C., G. Lenaers, P. Belenguer, and C.P. Hamel. 2003. Gene structure and chromosomal localization of mouse Opa1 : its exclusion from the Bst locus. *BMC Genet*. 4:8.
- Delettre, C., G. Lenaers, J.M. Griffoin, N. Gigarel, C. Lorenzo, P. Belenguer, L. Pelloquin, J. Grosgeorge, C. Turc-Carel, E. Perret, C. Astarie-Dequeker, L. Lasquellec, B. Arnaud, B. Ducommun, J. Kaplan, and C.P. Hamel. 2000. Nuclear gene OPA1, encoding a mitochondrial dynamin-related protein, is mutated in dominant optic atrophy. *Nat Genet*. 26:207-10.
- Desagher, S., A. Osen-Sand, A. Nichols, R. Eskes, S. Montessuit, S. Lauper, K. Maundrell, B. Antonsson, and J.C. Martinou. 1999. Bid-induced conformational change of Bax is responsible for mitochondrial cytochrome c release during apoptosis. *J Cell Biol*. 144:891-901.
- Duchen, M.R. 2000. Mitochondria and calcium: from cell signalling to cell death. *J Physiol*. 529 Pt 1:57-68.
- Eiberger, J., M. Kibschull, N. Strenzke, A. Schober, H. Bussow, C. Wessig, S. Djahed, H. Reucher, D.A. Koch, J. Lautermann, T. Moser, E. Winterhager, and K. Willecke. 2006. Expression pattern and functional characterization of connexin29 in transgenic mice. *Glia*. 53:601-11.
- Ekstrand, M.I., M. Falkenberg, A. Rantanen, C.B. Park, M. Gaspari, K. Hultenby, P. Rustin, C.M. Gustafsson, and N.G. Larsson. 2004. Mitochondrial transcription factor A regulates mtDNA copy number in mammals. *Hum Mol Genet*. 13:935-44.
- Erickson, R.P. 1972. Leber's optic atrophy, a possible example of maternal inheritance. *Am J Hum Genet*. 24:348-9.

- Ferre, M., P. Amati-Bonneau, Y. Tourmen, Y. Malthiery, and P. Reynier. 2005. eOPA1: an online database for OPA1 mutations. *Hum Mutat.* 25:423-8.
- Finsterer, J. 2004. Mitochondriopathies. *Eur J Neurol.* 11:163-86.
- Foran, D.R., and A.C. Peterson. 1992. Myelin acquisition in the central nervous system of the mouse revealed by an MBP-Lac Z transgene. *J Neurosci.* 12:4890-7.
- Frank, S., B. Gaume, E.S. Bergmann-Leitner, W.W. Leitner, E.G. Robert, F. Catez, C.L. Smith, and R.J. Youle. 2001. The role of dynamin-related protein 1, a mediator of mitochondrial fission, in apoptosis. *Dev Cell.* 1:515-25.
- Frayne, E.G., E.J. Leys, G.F. Crouse, A.G. Hook, and R.E. Kellems. 1984. Transcription of the mouse dihydrofolate reductase gene proceeds unabated through seven polyadenylation sites and terminates near a region of repeated DNA. *Mol Cell Biol.* 4:2921-4.
- Frey, T.G., and C.A. Mannella. 2000. The internal structure of mitochondria. *Trends Biochem Sci.* 25:319-24.
- Frezza, C., S. Cipolat, O. Martins de Brito, M. Micaroni, G.V. Beznousenko, T. Rudka, D. Bartoli, R.S. Polishuck, N.N. Danial, B. De Strooper, and L. Scorrano. 2006. OPA1 controls apoptotic cristae remodeling independently from mitochondrial fusion. *Cell.* 126:177-89.
- Garesse, R., and C.G. Vallejo. 2001. Animal mitochondrial biogenesis and function: a regulatory cross-talk between two genomes. *Gene.* 263:1-16.
- Germain, M., J.P. Mathai, H.M. McBride, and G.C. Shore. 2005. Endoplasmic reticulum BIK initiates DRP1-regulated remodelling of mitochondrial cristae during apoptosis. *Embo J.* 24:1546-56.
- Ghelli, A., C. Zanna, A.M. Porcelli, A.H. Schapira, A. Martinuzzi, V. Carelli, and M. Rugolo. 2003. Leber's hereditary optic neuropathy (LHON) pathogenic mutations induce mitochondrial-dependent apoptotic death in transmtochondrial cells incubated with galactose medium. *J Biol Chem.* 278:4145-50.
- Gotz, J., L.M. Ittner, and S. Kins. 2006. Do axonal defects in tau and amyloid precursor protein transgenic animals model axonopathy in Alzheimer's disease? *J Neurochem.* 98:993-1006.
- Graber, J.H., C.R. Cantor, S.C. Mohr, and T.F. Smith. 1999. In silico detection of control signals: mRNA 3'-end-processing sequences in diverse species. *Proc Natl Acad Sci U S A.* 96:14055-60.

- Grafstein, B. 1969. Axonal transport: communication between soma and synapse. *Adv Biochem Psychopharmacol.* 1:11-25.
- Green, D.R., and G. Kroemer. 2004. The pathophysiology of mitochondrial cell death. *Science.* 305:626-9.
- Green, D.R., and J.C. Reed. 1998. Mitochondria and apoptosis. *Science.* 281:1309-12.
- Griparic, L., and A.M. van der Bliek. 2001. The many shapes of mitochondrial membranes. *Traffic.* 2:235-44.
- Griparic, L., N.N. van der Wel, I.J. Orozco, P.J. Peters, and A.M. van der Bliek. 2004. Loss of the intermembrane space protein Mgm1/OPA1 induces swelling and localized constrictions along the lengths of mitochondria. *J Biol Chem.* 279:18792-8.
- Guillou, E., C. Bousquet, M. Daloyau, L.J. Emorine, and P. Belenguer. 2005. Msp1p is an intermembrane space dynamin-related protein that mediates mitochondrial fusion in a Dnm1p-dependent manner in *S. pombe*. *FEBS Lett.* 579:1109-16.
- Hales, K.G., and M.T. Fuller. 1997. Developmentally regulated mitochondrial fusion mediated by a conserved, novel, predicted GTPase. *Cell.* 90:121-9.
- Hance, N., M.I. Ekstrand, and A. Trifunovic. 2005. Mitochondrial DNA polymerase gamma is essential for mammalian embryogenesis. *Hum Mol Genet.* 14:1775-83.
- Herlan, M., F. Vogel, C. Bornhovd, W. Neupert, and A.S. Reichert. 2003. Processing of Mgm1 by the rhomboid-type protease Pcp1 is required for maintenance of mitochondrial morphology and of mitochondrial DNA. *J Biol Chem.* 278:27781-8.
- Hermann, G.J., J.W. Thatcher, J.P. Mills, K.G. Hales, M.T. Fuller, J. Nunnari, and J.M. Shaw. 1998. Mitochondrial fusion in yeast requires the transmembrane GTPase Fzo1p. *J Cell Biol.* 143:359-73.
- Herrmann, J.M., and W. Neupert. 2000. Protein transport into mitochondria. *Curr Opin Microbiol.* 3:210-4.
- Hinshaw, J.E. 2000. Dynamin and its role in membrane fission. *Annu Rev Cell Dev Biol.* 16:483-519.
- Hollenbeck, P.J. 1996. The pattern and mechanism of mitochondrial transport in axons. *Front Biosci.* 1:d91-102.
- Hooper, M., K. Hardy, A. Handyside, S. Hunter, and M. Monk. 1987. HPRT-deficient (Lesch-Nyhan) mouse embryos derived from germline colonization by cultured cells. *Nature.* 326:292-5.
- Howell, N., J.L. Elson, P.F. Chinnery, and D.M. Turnbull. 2005. mtDNA mutations and common neurodegenerative disorders. *Trends Genet.* 21:583-6.

- Hoyt, C.S. 1980. Autosomal dominant optic atrophy. A spectrum of disability. *Ophthalmology*. 87:245-51.
- Huang, D.C., and A. Strasser. 2000. BH3-Only proteins-essential initiators of apoptotic cell death. *Cell*. 103:839-42.
- Hubener, M. 2003. Mouse visual cortex. *Curr Opin Neurobiol*. 13:413-20.
- Innis, M.A., Gelfand, D.H. and Sninsky, J.J. . 1989. PCR Protocols: A Guide To Methods And Applications. Academic Press, San Diego, CA 92101-4495, USA.
- Ishihara, N., Y. Eura, and K. Mihara. 2004. Mitofusin 1 and 2 play distinct roles in mitochondrial fusion reactions via GTPase activity. *J Cell Sci*. 117:6535-46.
- Ishihara, N., A. Jofuku, Y. Eura, and K. Mihara. 2003. Regulation of mitochondrial morphology by membrane potential, and DRP1-dependent division and FZO1-dependent fusion reaction in mammalian cells. *Biochem Biophys Res Commun*. 301:891-8.
- Jacobson, D.M., and E.M. Stone. 1991. Difficulty differentiating Leber's from dominant optic neuropathy in a patient with remote visual loss. *J Clin Neuroophthalmol*. 11:152-7.
- James, D.I., P.A. Parone, Y. Mattenberger, and J.C. Martinou. 2003. hFis1, a novel component of the mammalian mitochondrial fission machinery. *J Biol Chem*. 278:36373-9.
- Johnston, P.B., R.N. Gaster, V.C. Smith, and R.C. Tripathi. 1979. A clinicopathologic study of autosomal dominant optic atrophy. *Am J Ophthalmol*. 88:868-75.
- Johnston, R.L., M.J. Seller, J.T. Behnam, M.A. Burdon, and D.J. Spalton. 1999. Dominant optic atrophy. Refining the clinical diagnostic criteria in light of genetic linkage studies. *Ophthalmology*. 106:123-8.
- Joyner, A.L. 1999. Gene Targeting: A Practical Approach. Oxford University Press, Oxford, United Kingdom.
- Ju, W.K., T. Misaka, Y. Kushnareva, S. Nakagomi, N. Agarwal, Y. Kubo, S.A. Lipton, and E. Bossy-Wetzel. 2005. OPA1 expression in the normal rat retina and optic nerve. *J Comp Neurol*. 488:1-10.
- Kamei, S., M. Chen-Kuo-Chang, C. Cazevieille, G. Lenaers, A. Olichon, P. Belenguer, G. Roussignol, N. Renard, M. Eybalin, A. Michelin, C. Delettre, P. Brabet, and C.P. Hamel. 2005. Expression of the Opa1 mitochondrial protein in retinal ganglion cells: its downregulation causes aggregation of the mitochondrial network. *Invest Ophthalmol Vis Sci*. 46:4288-94.

- Karbowski, M., D. Arnoult, H. Chen, D.C. Chan, C.L. Smith, and R.J. Youle. 2004. Quantitation of mitochondrial dynamics by photolabeling of individual organelles shows that mitochondrial fusion is blocked during the Bax activation phase of apoptosis. *J Cell Biol.* 164:493-9.
- Karbowski, M., Y.J. Lee, B. Gaume, S.Y. Jeong, S. Frank, A. Nechushtan, A. Santel, M. Fuller, C.L. Smith, and R.J. Youle. 2002. Spatial and temporal association of Bax with mitochondrial fission sites, Drp1, and Mfn2 during apoptosis. *J Cell Biol.* 159:931-8.
- Karbowski, M., and R.J. Youle. 2003. Dynamics of mitochondrial morphology in healthy cells and during apoptosis. *Cell Death Differ.* 10:870-80.
- Kijima, K., C. Numakura, H. Izumino, K. Umetsu, A. Nezu, T. Shiiki, M. Ogawa, Y. Ishizaki, T. Kitamura, Y. Shozawa, and K. Hayasaka. 2005. Mitochondrial GTPase mitofusin 2 mutation in Charcot-Marie-Tooth neuropathy type 2A. *Hum Genet.* 116:23-7.
- Kim, J.Y., J.M. Hwang, H.S. Ko, M.W. Seong, B.J. Park, and S.S. Park. 2005. Mitochondrial DNA content is decreased in autosomal dominant optic atrophy. *Neurology.* 64:966-72.
- Kim, M., P. Qiu, R. Abuodeh, J. Chen, and D. Yuan. 1999. Differential regulation of transcription termination occurring at two different sites on the micro-delta gene complex. *Int Immunol.* 11:813-24.
- Kivlin, J.D., E.W. Lovrien, D.T. Bishop, and I.H. Maumenee. 1983. Linkage analysis in dominant optic atrophy. *Am J Hum Genet.* 35:1190-5.
- Kjer, B., H. Eiberg, P. Kjer, and T. Rosenberg. 1996. Dominant optic atrophy mapped to chromosome 3q region. II. Clinical and epidemiological aspects. *Acta Ophthalmol Scand.* 74:3-7.
- Kjer, P. 1959. Infantile optic atrophy with dominant mode of inheritance: a clinical and genetic study of 19 Danish families. *Acta Ophthalmol Suppl.* 164:1-147.
- Kjer, P., O.A. Jensen, and L. Klinken. 1983. Histopathology of eye, optic nerve and brain in a case of dominant optic atrophy. *Acta Ophthalmol (Copenh).* 61:300-12.
- Kolarov, J., and M. Klingenberg. 1974. The adenine nucleotide translocator in genetically and physiologically modified yeast mitochondria. *FEBS Lett.* 45:320-3.
- Kosaka, T., and K. Ikeda. 1983. Possible temperature-dependent blockage of synaptic vesicle recycling induced by a single gene mutation in Drosophila. *J Neurobiol.* 14:207-25.
- Koshiba, T., S.A. Detmer, J.T. Kaiser, H. Chen, J.M. McCaffery, and D.C. Chan. 2004. Structural basis of mitochondrial tethering by mitofusin complexes. *Science.* 305:858-62.

- Krajewski, S., S. Tanaka, S. Takayama, M.J. Schibler, W. Fenton, and J.C. Reed. 1993. Investigation of the subcellular distribution of the bcl-2 oncprotein: residence in the nuclear envelope, endoplasmic reticulum, and outer mitochondrial membranes. *Cancer Res.* 53:4701-14.
- Kroemer, G., and J.C. Reed. 2000. Mitochondrial control of cell death. *Nat Med.* 6:513-9.
- Kuhn, R., K. Rajewsky, and W. Muller. 1991. Generation and analysis of interleukin-4 deficient mice. *Science.* 254:707-10.
- Labrousse, A.M., D.L. Shurland, and A.M. van der Bliek. 1998. Contribution of the GTPase domain to the subcellular localization of dynamin in the nematode *Caenorhabditis elegans*. *Mol Biol Cell.* 9:3227-39.
- Larsson, N.G., J. Wang, H. Wilhelmsson, A. Oldfors, P. Rustin, M. Lewandoski, G.S. Barsh, and D.A. Clayton. 1998. Mitochondrial transcription factor A is necessary for mtDNA maintenance and embryogenesis in mice. *Nat Genet.* 18:231-6.
- Lawson, V.H., B.V. Graham, and K.M. Flanigan. 2005. Clinical and electrophysiologic features of CMT2A with mutations in the mitofusin 2 gene. *Neurology.* 65:197-204.
- Lee, Y.J., S.Y. Jeong, M. Karbowski, C.L. Smith, and R.J. Youle. 2004. Roles of the mammalian mitochondrial fission and fusion mediators Fis1, Drp1, and Opa1 in apoptosis. *Mol Biol Cell.* 15:5001-11.
- Legros, F., A. Lombes, P. Frachon, and M. Rojo. 2002. Mitochondrial fusion in human cells is efficient, requires the inner membrane potential, and is mediated by mitofusins. *Mol Biol Cell.* 13:4343-54.
- Leonard, J.A., R.K. Wayne, J. Wheeler, R. Valadez, S. Guillen, and C. Vila. 2002. Ancient DNA evidence for Old World origin of New World dogs. *Science.* 298:1613-6.
- Li, C., G. Kosmorsky, K. Zhang, B.J. Katz, J. Ge, and E.I. Traboulsi. 2005. Optic atrophy and sensorineural hearing loss in a family caused by an R445H OPA1 mutation. *Am J Med Genet A.* 138:208-11.
- Li, K., Y. Li, J.M. Shelton, J.A. Richardson, E. Spencer, Z.J. Chen, X. Wang, and R.S. Williams. 2000. Cytochrome c deficiency causes embryonic lethality and attenuates stress-induced apoptosis. *Cell.* 101:389-99.
- Li, Z., K. Okamoto, Y. Hayashi, and M. Sheng. 2004. The importance of dendritic mitochondria in the morphogenesis and plasticity of spines and synapses. *Cell.* 119:873-87.
- Lim, M.L., T. Minamikawa, and P. Nagley. 2001. The protonophore CCCP induces mitochondrial permeability transition without cytochrome c release in human osteosarcoma cells. *FEBS Lett.* 503:69-74.

- Liu, X., C.N. Kim, J. Yang, R. Jemmerson, and X. Wang. 1996. Induction of apoptotic program in cell-free extracts: requirement for dATP and cytochrome c. *Cell.* 86:147-57.
- Lodish, H., Berk, A., Matsudaira, P., Kaiser, C.A., Krieger, M., Scott, M.P., Zipursky, S.L., Darnell, J. . 2004. Molecular Cell Biology. W.H. Freeman and Company, New York.
- Lonsbury-Martin, B.L., and G.K. Martin. 1990. The clinical utility of distortion-product otoacoustic emissions. *Ear Hear.* 11:144-54.
- Luo, X., I. Budihardjo, H. Zou, C. Slaughter, and X. Wang. 1998. Bid, a Bcl2 interacting protein, mediates cytochrome c release from mitochondria in response to activation of cell surface death receptors. *Cell.* 94:481-90.
- Lyle, W.M. 1990. Genetic Risks. University of Waterloo Press, Waterloo, Ontario.
- Malka, F., O. Guillery, C. Cifuentes-Diaz, E. Guillou, P. Belenguer, A. Lombes, and M. Rojo. 2005. Separate fusion of outer and inner mitochondrial membranes. *EMBO Rep.* 6:853-9.
- Mannella, C.A. 2006. The relevance of mitochondrial membrane topology to mitochondrial function. *Biochim Biophys Acta.* 1762:140-7.
- Mannella, C.A., M. Marko, P. Penczek, D. Barnard, and J. Frank. 1994. The internal compartmentation of rat-liver mitochondria: tomographic study using the high-voltage transmission electron microscope. *Microsc Res Tech.* 27:278-83.
- Marchbank, N.J., J.E. Craig, J.P. Leek, M. Toohey, A.J. Churchill, A.F. Markham, D.A. Mackey, C. Toomes, and C.F. Inglehearn. 2002. Deletion of the OPA1 gene in a dominant optic atrophy family: evidence that haploinsufficiency is the cause of disease. *J Med Genet.* 39:e47.
- Marmor, M.F., G.E. Holder, M.W. Seeliger, and S. Yamamoto. 2004. Standard for clinical electroretinography (2004 update). *Doc Ophthalmol.* 108:107-14.
- McNiven, M.A., H. Cao, K.R. Pitts, and Y. Yoon. 2000. The dynamin family of mechanoenzymes: pinching in new places. *Trends Biochem Sci.* 25:115-20.
- McQuibban, G.A., S. Saurya, and M. Freeman. 2003. Mitochondrial membrane remodelling regulated by a conserved rhomboid protease. *Nature.* 423:537-41.
- Meeusen, S., J.M. McCaffery, and J. Nunnari. 2004. Mitochondrial fusion intermediates revealed in vitro. *Science.* 305:1747-52.
- Misaka, T., T. Miyashita, and Y. Kubo. 2002. Primary structure of a dynamin-related mouse mitochondrial GTPase and its distribution in brain, subcellular localization, and effect on mitochondrial morphology. *J Biol Chem.* 277:15834-42.

- Mozdy, A.D., J.M. McCaffery, and J.M. Shaw. 2000. Dnm1p GTPase-mediated mitochondrial fission is a multi-step process requiring the novel integral membrane component Fis1p. *J Cell Biol.* 151:367-80.
- Neupert, W., and M. Brunner. 2002. The protein import motor of mitochondria. *Nat Rev Mol Cell Biol.* 3:555-65.
- Neuspiel, M., R. Zunino, S. Gangaraju, P. Rippstein, and H. McBride. 2005. Activated mitofusin 2 signals mitochondrial fusion, interferes with Bax activation, and reduces susceptibility to radical induced depolarization. *J Biol Chem.* 280:25060-70.
- Newman, S.E. 1978. The EEG manifestations of chronic ethanol abuse: relation to cerebral cortical atrophy. *Ann Neurol.* 3:299-304.
- Olichon, A., L. Baricault, N. Gas, E. Guillou, A. Valette, P. Belenguer, and G. Lenaers. 2003. Loss of OPA1 perturbs the mitochondrial inner membrane structure and integrity, leading to cytochrome c release and apoptosis. *J Biol Chem.* 278:7743-6.
- Olichon, A., L.J. Emorine, E. Descoids, L. Pelloquin, L. Brichese, N. Gas, E. Guillou, C. Delettre, A. Valette, C.P. Hamel, B. Ducommun, G. Lenaers, and P. Belenguer. 2002. The human dynamin-related protein OPA1 is anchored to the mitochondrial inner membrane facing the inter-membrane space. *FEBS Lett.* 523:171-6.
- Olichon, A., E. Guillou, C. Delettre, T. Landes, L. Arnaune-Pelloquin, L.J. Emorine, V. Mils, M. Daloyau, C. Hamel, P. Amati-Bonneau, D. Bonneau, P. Reynier, G. Lenaers, and P. Belenguer. 2006. Mitochondrial dynamics and disease, OPA1. *Biochim Biophys Acta.* 1763:500-9.
- Ono, T., K. Isobe, K. Nakada, and J.I. Hayashi. 2001. Human cells are protected from mitochondrial dysfunction by complementation of DNA products in fused mitochondria. *Nat Genet.* 28:272-5.
- Orozco, I.J., S.J. Kim, and H.G. Martinson. 2002. The poly(A) signal, without the assistance of any downstream element, directs RNA polymerase II to pause in vivo and then to release stochastically from the template. *J Biol Chem.* 277:42899-911.
- Otsuga, D., B.R. Keegan, E. Brisch, J.W. Thatcher, G.J. Hermann, W. Bleazard, and J.M. Shaw. 1998. The dynamin-related GTPase, Dnm1p, controls mitochondrial morphology in yeast. *J Cell Biol.* 143:333-49.
- Pacher, P., and G. Hajnoczky. 2001. Propagation of the apoptotic signal by mitochondrial waves. *Embo J.* 20:4107-21.
- Palade, G.E. 1952. The fine structure of mitochondria. *Anat Rec.* 114:427-51.

- Palay, S.L. 1956. Synapses in the central nervous system. *J Biophys Biochem Cytol.* 2:193-202.
- Park, M.K., M.C. Ashby, G. Erdemli, O.H. Petersen, and A.V. Tepikin. 2001. Perinuclear, perigranular and sub-plasmalemmal mitochondria have distinct functions in the regulation of cellular calcium transport. *Embo J.* 20:1863-74.
- Parone, P.A., D. James, and J.C. Martinou. 2002. Mitochondria: regulating the inevitable. *Biochimie.* 84:105-11.
- Payne, M., Z. Yang, B.J. Katz, J.E. Warner, C.J. Weight, Y. Zhao, E.D. Pearson, R.L. Treft, T. Hillman, R.J. Kennedy, F.M. Meire, and K. Zhang. 2004. Dominant optic atrophy, sensorineural hearing loss, ptosis, and ophthalmoplegia: a syndrome caused by a missense mutation in OPA1. *Am J Ophthalmol.* 138:749-55.
- Pelloquin, L., P. Belenguer, Y. Menon, N. Gas, and B. Ducommun. 1999. Fission yeast Msp1 is a mitochondrial dynamin-related protein. *J Cell Sci.* 112 (Pt 22):4151-61.
- Perfettini, J.L., T. Roumier, and G. Kroemer. 2005. Mitochondrial fusion and fission in the control of apoptosis. *Trends Cell Biol.* 15:179-83.
- Perkins, G.A., C.W. Renken, J.Y. Song, T.G. Frey, S.J. Young, S. Lamont, M.E. Martone, S. Lindsey, and M.H. Ellisman. 1997. Electron tomography of large, multicomponent biological structures. *J Struct Biol.* 120:219-27.
- Perotti, M.E., W.A. Anderson, and H. Swift. 1983. Quantitative cytochemistry of the diaminobenzidine cytochrome oxidase reaction product in mitochondria of cardiac muscle and pancreas. *J Histochem Cytochem.* 31:351-65.
- Pesch, U.E., J.E. Fries, S. Bette, H. Kalbacher, B. Wissinger, C. Alexander, and K. Kohler. 2004. OPA1, the disease gene for autosomal dominant optic atrophy, is specifically expressed in ganglion cells and intrinsic neurons of the retina. *Invest Ophthalmol Vis Sci.* 45:4217-25.
- Pesch, U.E., B. Leo-Kottler, S. Mayer, B. Jurklies, U. Kellner, E. Apfelstedt-Sylla, E. Zrenner, C. Alexander, and B. Wissinger. 2001. OPA1 mutations in patients with autosomal dominant optic atrophy and evidence for semi-dominant inheritance. *Hum Mol Genet.* 10:1359-68.
- Petit, P.X., S.A. Susin, N. Zamzami, B. Mignotte, and G. Kroemer. 1996. Mitochondria and programmed cell death: back to the future. *FEBS Lett.* 396:7-13.
- Pinton, P., D. Ferrari, E. Rapizzi, F. Di Virgilio, T. Pozzan, and R. Rizzuto. 2001. The Ca²⁺ concentration of the endoplasmic reticulum is a key determinant of ceramide-induced

- apoptosis: significance for the molecular mechanism of Bcl-2 action. *Embo J.* 20:2690-701.
- Pitts, K.R., Y. Yoon, E.W. Krueger, and M.A. McNiven. 1999. The dynamin-like protein DLP1 is essential for normal distribution and morphology of the endoplasmic reticulum and mitochondria in mammalian cells. *Mol Biol Cell.* 10:4403-17.
- Praefcke, G.J., and H.T. McMahon. 2004. The dynamin superfamily: universal membrane tubulation and fission molecules? *Nat Rev Mol Cell Biol.* 5:133-47.
- Proudfoot, N., and J. O'Sullivan. 2002. Polyadenylation: a tail of two complexes. *Curr Biol.* 12:R855-7.
- Rapaport, D., M. Brunner, W. Neupert, and B. Westermann. 1998. Fzo1p is a mitochondrial outer membrane protein essential for the biogenesis of functional mitochondria in *Saccharomyces cerevisiae*. *J Biol Chem.* 273:20150-5.
- Reed, J.C. 1997. Cytochrome c: can't live with it--can't live without it. *Cell.* 91:559-62.
- Reynolds, E.S. 1963. The use of lead citrate at high pH as an electron-opaque stain in electron microscopy. *J Cell Biol.* 17:208-12.
- Rizzuto, R., P. Bernardi, and T. Pozzan. 2000. Mitochondria as all-round players of the calcium game. *J Physiol.* 529 Pt 1:37-47.
- Rizzuto, R., P. Pinton, W. Carrington, F.S. Fay, K.E. Fogarty, L.M. Lifshitz, R.A. Tuft, and T. Pozzan. 1998. Close contacts with the endoplasmic reticulum as determinants of mitochondrial Ca²⁺ responses. *Science.* 280:1763-6.
- Rojo, M., F. Legros, D. Chateau, and A. Lombes. 2002. Membrane topology and mitochondrial targeting of mitofusins, ubiquitous mammalian homologs of the transmembrane GTPase Fzo. *J Cell Sci.* 115:1663-74.
- Rowland, K.C., N.K. Irby, and G.A. Spiro. 2000. Specialized synapse-associated structures within the calyx of Held. *J Neurosci.* 20:9135-44.
- Rube, D.A., and A.M. van der Bliek. 2004. Mitochondrial morphology is dynamic and varied. *Mol Cell Biochem.* 256-257:331-9.
- Sadun, A.A., and V. Carelli. 2003. Mitochondrial function and dysfunction within the optic nerve. *Arch Ophthalmol.* 121:1342-3.
- Sadun, A.A., J.F. Martone, R. Muci-Mendoza, L. Reyes, L. DuBois, J.C. Silva, G. Roman, and B. Caballero. 1994. Epidemic optic neuropathy in Cuba. Eye findings. *Arch Ophthalmol.* 112:691-9.
- Sambrook, J.a.R., D.W. . 2001. Molecular Cloning: A Laboratory Manual. Cold Spring Harbor Laboratory Press, Cold Spring Harbor, New York 11803-2500, USA.

- Santel, A., and M.T. Fuller. 2001. Control of mitochondrial morphology by a human mitofusin. *J Cell Sci.* 114:867-74.
- Satoh, M., T. Hamamoto, N. Seo, Y. Kagawa, and H. Endo. 2003. Differential sublocalization of the dynamin-related protein OPA1 isoforms in mitochondria. *Biochem Biophys Res Commun.* 300:482-93.
- Scheffler, I.E. 1999. Mitochondria. New York. Wiley-Liss. Inc.
- Schlesinger, P.H., A. Gross, X.M. Yin, K. Yamamoto, M. Saito, G. Waksman, and S.J. Korsmeyer. 1997. Comparison of the ion channel characteristics of proapoptotic BAX and antiapoptotic BCL-2. *Proc Natl Acad Sci U S A.* 94:11357-62.
- Seeliger, M.W., S.C. Beck, N. Pereyra-Munoz, S. Dangel, J.Y. Tsai, U.F. Luhmann, S.A. van de Pavert, J. Wijnholds, M. Samardzija, A. Wenzel, E. Zrenner, K. Narfstrom, E. Fahl, N. Tanimoto, N. Acar, and F. Tonagel. 2005. In vivo confocal imaging of the retina in animal models using scanning laser ophthalmoscopy. *Vision Res.* 45:3512-9.
- Seeliger, M.W., C. Grimm, F. Stahlberg, C. Friedburg, G. Jaissle, E. Zrenner, H. Guo, C.E. Reme, P. Humphries, F. Hofmann, M. Biel, R.N. Fariss, T.M. Redmond, and A. Wenzel. 2001. New views on RPE65 deficiency: the rod system is the source of vision in a mouse model of Leber congenital amaurosis. *Nat Genet.* 29:70-4.
- Sesaki, H., and R.E. Jensen. 1999. Division versus fusion: Dnm1p and Fzo1p antagonistically regulate mitochondrial shape. *J Cell Biol.* 147:699-706.
- Sesaki, H., S.M. Southard, M.P. Yaffe, and R.E. Jensen. 2003. Mgm1p, a dynamin-related GTPase, is essential for fusion of the mitochondrial outer membrane. *Mol Biol Cell.* 14:2342-56.
- Shaw, J.M., and J. Nunnari. 2002. Mitochondrial dynamics and division in budding yeast. *Trends Cell Biol.* 12:178-84.
- Shepard, K.A., and M.P. Yaffe. 1999. The yeast dynamin-like protein, Mgm1p, functions on the mitochondrial outer membrane to mediate mitochondrial inheritance. *J Cell Biol.* 144:711-20.
- Shepherd, G.M., and K.M. Harris. 1998. Three-dimensional structure and composition of CA3->CA1 axons in rat hippocampal slices: implications for presynaptic connectivity and compartmentalization. *J Neurosci.* 18:8300-10.
- Shimohama, S., H. Sawada, Y. Kitamura, and T. Taniguchi. 2003. Disease model: Parkinson's disease. *Trends Mol Med.* 9:360-5.

- Shoshan-Barmatz, V., A. Israelson, D. Brdiczka, and S.S. Sheu. 2006. The voltage-dependent anion channel (VDAC): function in intracellular signalling, cell life and cell death. *Curr Pharm Des.* 12:2249-70.
- Shy, M.E. 2004. Charcot-Marie-Tooth disease: an update. *Curr Opin Neurol.* 17:579-85.
- Silver, L.M. 1995. Mouse Genetics: Concepts and Applications. Oxford University Press.
- Sjostrand, F.S. 1953. Electron microscopy of mitochondria and cytoplasmic double membranes. *Nature.* 171:30-2.
- Skulachev, V.P. 2001. Mitochondrial filaments and clusters as intracellular power-transmitting cables. *Trends Biochem Sci.* 26:23-9.
- Smaili, S.S., Y.T. Hsu, K.M. Sanders, J.T. Russell, and R.J. Youle. 2001. Bax translocation to mitochondria subsequent to a rapid loss of mitochondrial membrane potential. *Cell Death Differ.* 8:909-20.
- Smeitink, J., L. van den Heuvel, and S. DiMauro. 2001. The genetics and pathology of oxidative phosphorylation. *Nat Rev Genet.* 2:342-52.
- Smirnova, E., L. Griparic, D.L. Shurland, and A.M. van der Bliek. 2001. Dynamin-related protein Drp1 is required for mitochondrial division in mammalian cells. *Mol Biol Cell.* 12:2245-56.
- Smirnova, E., D.L. Shurland, S.N. Ryazantsev, and A.M. van der Bliek. 1998. A human dynamin-related protein controls the distribution of mitochondria. *J Cell Biol.* 143:351-8.
- Smith, A.G., J.K. Heath, D.D. Donaldson, G.G. Wong, J. Moreau, M. Stahl, and D. Rogers. 1988. Inhibition of pluripotential embryonic stem cell differentiation by purified polypeptides. *Nature.* 336:688-90.
- Smith, R.S. 2002. Systematic Evaluation of the Mouse Eye: Anatomy, Pathology, and Biomethods. CRC Press.
- Stojanovski, D., O.S. Koutsopoulos, K. Okamoto, and M.T. Ryan. 2004. Levels of human Fis1 at the mitochondrial outer membrane regulate mitochondrial morphology. *J Cell Sci.* 117:1201-10.
- Sugioka, R., S. Shimizu, and Y. Tsujimoto. 2004. Fzo1, a protein involved in mitochondrial fusion, inhibits apoptosis. *J Biol Chem.* 279:52726-34.
- Szabadkai, G., A.M. Simoni, M. Chami, M.R. Wieckowski, R.J. Youle, and R. Rizzuto. 2004. Drp-1-dependent division of the mitochondrial network blocks intraorganellar Ca²⁺ waves and protects against Ca²⁺-mediated apoptosis. *Mol Cell.* 16:59-68.

- Takagaki, Y., and J.L. Manley. 1998. Levels of polyadenylation factor CstF-64 control IgM heavy chain mRNA accumulation and other events associated with B cell differentiation. *Mol Cell.* 2:761-71.
- Tanimoto, K., S. Handa, N. Ueno, K. Murakami, and A. Fukamizu. 1991. Structure and sequence analysis of the human activin beta A subunit gene. *DNA Seq.* 2:103-10.
- van der Bliek, A.M. 1999. Functional diversity in the dynamin family. *Trends Cell Biol.* 9:96-102.
- Vermes, I., C. Haanen, H. Steffens-Nakken, and C. Reutelingsperger. 1995. A novel assay for apoptosis. Flow cytometric detection of phosphatidylserine expression on early apoptotic cells using fluorescein labelled Annexin V. *J Immunol Methods.* 184:39-51.
- Vo, L.T., M. Minet, J.M. Schmitter, F. Lacroute, and F. Wyers. 2001. Mpe1, a zinc knuckle protein, is an essential component of yeast cleavage and polyadenylation factor required for the cleavage and polyadenylation of mRNA. *Mol Cell Biol.* 21:8346-56.
- Vogel, H. 2001. Mitochondrial myopathies and the role of the pathologist in the molecular era. *J Neuropathol Exp Neurol.* 60:217-27.
- Votruba, M., F.W. Fitzke, G.E. Holder, A. Carter, S.S. Bhattacharya, and A.T. Moore. 1998. Clinical features in affected individuals from 21 pedigrees with dominant optic atrophy. *Arch Ophthalmol.* 116:351-8.
- Wada, T., T. Takagi, Y. Yamaguchi, A. Ferdous, T. Imai, S. Hirose, S. Sugimoto, K. Yano, G.A. Hartzog, F. Winston, S. Buratowski, and H. Handa. 1998. DSIF, a novel transcription elongation factor that regulates RNA polymerase II processivity, is composed of human Spt4 and Spt5 homologs. *Genes Dev.* 12:343-56.
- Wang, X. 2001. The expanding role of mitochondria in apoptosis. *Genes Dev.* 15:2922-33.
- Wei, M.C., W.X. Zong, E.H. Cheng, T. Lindsten, V. Panoutsakopoulou, A.J. Ross, K.A. Roth, G.R. MacGregor, C.B. Thompson, and S.J. Korsmeyer. 2001. Proapoptotic BAX and BAK: a requisite gateway to mitochondrial dysfunction and death. *Science.* 292:727-30.
- Westermann, B. 2003. Mitochondrial membrane fusion. *Biochim Biophys Acta.* 1641:195-202.
- Wickens, M., P. Anderson, and R.J. Jackson. 1997. Life and death in the cytoplasm: messages from the 3' end. *Curr Opin Genet Dev.* 7:220-32.
- Wittwer, C.T., M.G. Herrmann, A.A. Moss, and R.P. Rasmussen. 1997a. Continuous fluorescence monitoring of rapid cycle DNA amplification. *Biotechniques.* 22:130-1, 134-8.

- Wittwer, C.T., K.M. Ririe, R.V. Andrew, D.A. David, R.A. Gundry, and U.J. Balis. 1997b. The LightCycler: a microvolume multisample fluorimeter with rapid temperature control. *Biotechniques*. 22:176-81.
- Wong, E.D., J.A. Wagner, S.W. Gorsich, J.M. McCaffery, J.M. Shaw, and J. Nunnari. 2000. The dynamin-related GTPase, Mgm1p, is an intermembrane space protein required for maintenance of fusion competent mitochondria. *J Cell Biol*. 151:341-52.
- Wong, E.D., J.A. Wagner, S.V. Scott, V. Okreglak, T.J. Holewinski, A. Cassidy-Stone, and J. Nunnari. 2003. The intramitochondrial dynamin-related GTPase, Mgm1p, is a component of a protein complex that mediates mitochondrial fusion. *J Cell Biol*. 160:303-11.
- Wynshaw-Boris, A. 1996. Model mice and human disease. *Nat Genet*. 13:259-60.
- Yeung, G., L.M. Choi, L.C. Chao, N.J. Park, D. Liu, A. Jamil, and H.G. Martinson. 1998. Poly(A)-driven and poly(A)-assisted termination: two different modes of poly(A)-dependent transcription termination. *Mol Cell Biol*. 18:276-89.
- Yoon, Y., E.W. Krueger, B.J. Oswald, and M.A. McNiven. 2003. The mitochondrial protein hFis1 regulates mitochondrial fission in mammalian cells through an interaction with the dynamin-like protein DLP1. *Mol Cell Biol*. 23:5409-20.
- Young, P., and U. Suter. 2003. The causes of Charcot-Marie-Tooth disease. *Cell Mol Life Sci*. 60:2547-60.
- Yu, D.Y., and S.J. Cringle. 2001. Oxygen distribution and consumption within the retina in vascularised and avascular retinas and in animal models of retinal disease. *Prog Retin Eye Res*. 20:175-208.
- Yu, T., R.J. Fox, L.S. Burwell, and Y. Yoon. 2005. Regulation of mitochondrial fission and apoptosis by the mitochondrial outer membrane protein hFis1. *J Cell Sci*. 118:4141-51.
- Zanna, C., A. Ghelli, A.M. Porcelli, A. Martinuzzi, V. Carelli, and M. Rugolo. 2005. Caspase-independent death of Leber's hereditary optic neuropathy cybrids is driven by energetic failure and mediated by AIF and Endonuclease G. *Apoptosis*. 10:997-1007.
- Zhang, H., J. Hu, M. Recce, and B. Tian. 2005a. PolyA_DB: a database for mammalian mRNA polyadenylation. *Nucleic Acids Res*. 33:D116-20.
- Zhang, H., J.Y. Lee, and B. Tian. 2005b. Biased alternative polyadenylation in human tissues. *Genome Biol*. 6:R100.
- Zhang, J., M.C. Reedy, Y.A. Hannun, and L.M. Obeid. 1999. Inhibition of caspases inhibits the release of apoptotic bodies: Bcl-2 inhibits the initiation of formation of apoptotic bodies in chemotherapeutic agent-induced apoptosis. *J Cell Biol*. 145:99-108.

Bibliography

- Zhang, X.D., S.K. Gillespie, and P. Hersey. 2004. Staurosporine induces apoptosis of melanoma by both caspase-dependent and -independent apoptotic pathways. *Mol Cancer Ther.* 3:187-97.
- Zhao, J., L. Hyman, and C. Moore. 1999. Formation of mRNA 3' ends in eukaryotes: mechanism, regulation, and interrelationships with other steps in mRNA synthesis. *Microbiol Mol Biol Rev.* 63:405-45.
- Zhuang, J., D. Dinsdale, and G.M. Cohen. 1998. Apoptosis, in human monocytic THP.1 cells, results in the release of cytochrome c from mitochondria prior to their ultracondensation, formation of outer membrane discontinuities and reduction in inner membrane potential. *Cell Death Differ.* 5:953-62.
- Zuchner, S., I.V. Mersyanova, M. Muglia, N. Bissar-Tadmouri, J. Rochelle, E.L. Dadali, M. Zappia, E. Nelis, A. Patitucci, J. Senderek, Y. Parman, O. Evgrafov, P.D. Jonghe, Y. Takahashi, S. Tsuji, M.A. Pericak-Vance, A. Quattrone, E. Battaloglu, A.V. Polyakov, V. Timmerman, J.M. Schroder, and J.M. Vance. 2004. Mutations in the mitochondrial GTPase mitofusin 2 cause Charcot-Marie-Tooth neuropathy type 2A. *Nat Genet.* 36:449-51.