

BIOGRAPHICAL SKETCH

Provide the following information for the key personnel and other significant contributors in the order listed on Form Page 2.
Follow this format for each person. **DO NOT EXCEED FOUR PAGES.**

NAME Don Hong, Ph.D.		POSITION TITLE Assistant Professor, Veterinary Pathobiology, College of Veterinary Medicine, Texas A&M University	
eRA COMMONS USER NAME			
EDUCATION/TRAINING <i>(Begin with baccalaureate or other initial professional education, such as nursing, and include postdoctoral training.)</i>			
INSTITUTION AND LOCATION	DEGREE <i>(if applicable)</i>	YEAR(s)	FIELD OF STUDY
Korea University, Seoul, Korea	B.S.	1984	Agricultural Chemistry
Oregon State University	Ph.D.	1993	Biochemistry/Biophysics

NOTE: The Biographical Sketch may not exceed four pages. Items A and B may not exceed two of the four-page limit.

A. Positions and Honors. List in chronological order previous positions, concluding with your present position. List any honors. Include present membership on any Federal Government public advisory committee.

PROFESSIONAL EXPERIENCE:

1998-1999 Research Fellow, Harvard Medical School, Boston, MA
1999-2005 Instructor in Ophthalmology, Harvard Medical School, Boston, MA
2005 Assistant Professor, Texas A&M University, College Station, TX

RESEARCH EXPERIENCE:

1993-1997 Hospital for Sick Children, Biochemistry Research, University of Toronto, Toronto, Canada, M5G 1X8
1990-1993 Oregon State University, Corvallis, Oregon, Graduate Research Assistant

PROFESSIONAL MEMBERSHIPS:

Association for Research in Vision and Ophthalmology (ARVO)
American Society for Cell Biology (ASCB)
American Association for the Advancement of Science (AAAS)

HONORS AND AWARDS:

1994-1997 Canadian Cystic Fibrosis Foundation, Post-doctoral Fellowship.

B. Selected peer-reviewed publications (in reverse chronological order). Do not include publications submitted or in preparation.

- Hong, D.H., Forsberg, N.E. Effects of serum and insulin-like growth factor I on protein degradation and protease gene expression in rat L8 myotubes. *J. Anim.Sci.* 72:2279-2288, 1994.
- Hong, D.H., Forsberg, N.E. Effects of dexamethasone on protein degradation and protease gene expression in rat L8 myotube cultures. *Molecular and Cellular Endocrinology.* 108:199-209, 1995.
- Hong, D.H., Huan, J., Ou, B.R., Yeh, J., Saido, T.C., Cheek, P.R., Forsberg, N.E. Protein kinase C isoforms in muscle cells and their regulation by phorbol ester and calpain. *Biochem. Biophys. Acta.*1267:45-54, 1995.
- Hong, D.H., Forstner, J.F., Forstner, G.G. PKC epsilon is the likely mediator of mucin exocytosis in human colonic cell lines. *Am. J. Physiology* 272:G31-G37, 1997.
- Hong, D.H., Lehel, C., Petrovic, G, W. Anderson, Forstner, J.F., Forstner, G.G. Induction of mucin gene expression in human colonic cell lines by PMA is dependent on PKC-epsilon. *Am. J. Physiology*, 277:G1041-G1047, 1999
- Hong, D, B. Pawlyk, E. Berson, T. Li. A retinitis pigmentosa GTPase regulator (RPGR)-deficient mouse model for X-linked retinitis pigmentosa (RP3). *P.N.A.S.* 97(7):3649-3654, 2000.
- Hong, D., G.Yue, M. Adamian, T. Li. Retinitis pigmentosa GTPase regulator (RPGR)-interacting protein is stably associated with the photoreceptor ciliary axoneme and anchors RPGR to the connecting cilium. *J Biol Chem.* 276(15): 12091-9, 2001.
- Dryja, TP, SM Adams, JL Grimsby, TL McGee, DH Hong, T Li, S. Andreasson, EL Berson. Null RPGRIP1 alleles in patients with Leber congenital amaurosis. *Am J Hum Genet.* 68(5): 1295-8, 2001.
- Hong DH, Li T. Complex expression pattern of RPGR reveals a role for purine-rich exonic splicing enhancers. *Invest Ophthalmol Vis Sci.* 43(11): 3373-82, 2002.
- Zhao Y, Hong DH, Pawlyk B, Yue G, Adamian M, Grynberg M, Godzik A, Li T. The retinitis pigmentosa GTPase regulator (RPGR)-interacting protein: subserving RPGR function and participating in disk morphogenesis. *Proc Natl Acad Sci U S A.* 100(7): 3965-70, 2003.
- Hong DH, Pawlyk B, Sokolov M, Strissel KJ, Yang J, Tulloch B, Wright AF, Arshavsky VY, Li T. RPGR isoforms in photoreceptor connecting cilia and the transitional zone of motile cilia. *Invest Ophthalmol Vis Sci.* 44(6): 2413-21, 2003.
- Bulgakov OV, JT. Eggenschwiler, DH Hong, KV. Anderson and T Li. FKBP8 is a negative regulator of mouse Sonic hedgehog signaling in neural tissues. *Development.*131(9):2149-59, 2004.
- Hong DH, Pawlyk BS, Adamian M, Li T. Dominant, gain-of-function mutant produced by truncation of RPGR. *Invest Ophthalmol Vis Sci.* 45(1): 36-41, 2004.

Principal Investigator/Program Director (Last, First, Middle): Hong, Don

14. Shaoul R, D Hong, Y Okada, E Cutz, MA Marcon. Lineage development in a patient without goblet, Paneth and enteroendocrine cells, a clue for intestinal epithelial differentiation. *Pediatric Research*. 2005 *Accepted*.
15. Hong DH, Basil S. Pawlyk, Michael Adamian, Michael A. Sandberg, and Tiansen Li. A Single, Abbreviated RPGR-ORF15 Variant Reconstitutes RPGR Function In Vivo. *Invest Ophthalmol Vis Sci*. 46(2): 435-441, 2005.
16. Adamian M, B. S. Pawlyk, DH Hong and E.L. Berson. Rod and cone mislocalization in an autopsy eye from a carrier of X-linked RP with a Gly436Asp mutation in the RPGR gene. *American Journal of Ophthalmology*. 142 (3): 515-8, 2006.

RECENT ABSTRACTS:

17. D. Hong, B. Pawlyk, M. Adamian and T. Li. A single abbreviated ORF15 variant appears sufficient to reconstitute RPGR function *in vivo* ARVO 2004.
18. B.S. Pawlyk, A.J. Smith, P.Buch, M.Adamian, D.-H.Hong, M.A. Sandberg, R.R. Ali, T.Li. Replacement Gene Therapy Rescues Photoreceptor Degeneration in a Murine Model of Leber Congenital Amaurosis (LCA) Lacking RPGRIP. ARVO 2005.
19. M.Adamian, B.Pawlyk, D.-H.Hong, E.L. Berson. Rod and Cone Opsin Mislocalization in an Autopsy Eye From a Female Carrier of X-linked RP With a Gly436Asp Mutation in the RPGR Gene. ARVO 2005.

C. Research Support. List selected ongoing or completed (during the last three years) research projects (federal and non-federal support). Begin with the projects that are most relevant to the research proposed in this application. Briefly indicate the overall goals of the projects and responsibilities of principal investigator identified above.

National Institute of Health. 2003-2006. RO3 EY014188. "Significance of RPGR Heterogeneity in Photoreceptors" P.I.: D-H. Hong.

Fight for Sight (Prevent Blindness America) Grant-in-Aid, 2001-2002. "Functional Studies of the Retinitis Pigmentosa GTPase Regulator (RPGR) and Its Interacting Protein". P.I.: D-H. Hong.

Knight Templar Eye Foundation Research Grant, 2001-2002. "Functional Studies of the Retinitis Pigmentosa GTPase Regulator (RPGR) and Its Interacting Protein". P.I.: D-H. Hong.