March 2026

Yosef Shiloh, Ph.D.

Publications

No. of Citations		h-index	
Web of Science:	32,710	79	
Scopus:	33,591	79	
Google Scholar:	51,237	93	(March 15, 2026)

ORCID No. https://orcid.org/0000-0002-1538-6060
PubMed: https://pubmed.ncbi.nlm.nih.gov/?term=shiloh+y&sort=date&size=200

Books

1. **Shiloh, Y.** (1991) The New Genetics. Publications of the Ministry of Defense, Government of Israel (Hebrew).
2. Khanna, K.K. and **Shiloh, Y.** (Eds.) (2010) The DNA Damage Response: Implications on Cancer Formation and Treatment. Springer.

Guest Editor

2004: "Bridge over Broken Ends", Special issue of *DNA Repair* on the Cellular Response to DNA Breaks.

Featured Interview in a Scientific Journal

The A-T Gene Hunt. An interview with Yossi Shiloh on decision making, the discovery of the *ATM* gene and lessons from genetics. By Esther Schnapp and Holger Breithaupt. EMBO Rep. (2019) 20:e48947. https://doi.org/10.15252/embr.201948947

Featured Profile in a Scientific Journal

Profile: Yosef Shiloh. By Jennifer Viegas. Proc. Natl. Acad. Sci. USA (2025) 122 (3) e2426242122. https://doi.org/10.1073/pnas.2426242122

Key Publications (1982 – 2026)

Shiloh, Y., Tabor, E. and Becker, Y. (1982) Colony forming ability of ataxia-telangiectasia skin fibroblasts is an indicator of their early senescence and increased demand for growth factors. Exp. Cell Res., 140:191-199.

Shiloh, Y., Tabor, E. and Becker, Y. (1983) Abnormal response of ataxia-telangiectasia cells to agents that break the deoxyribose moiety of DNA via a targeted free radical mechanism. Carcinogenesis, 4:1317-1322.

Shiloh, Y., Shipley, J., Brodeur, G.M., Bruns, G., Korf, B., Donlon, T., Seeger, R., Sakai, K. and Latt, S. (1985) Differential amplification, assembly and relocation of multiple DNA sequences in human neuroblastomas and neuroblastoma cell lines. Proc. Natl. Acad. Sci. USA, 82:3761-3765.

Shiloh, Y., Sanford, K.K., Parshad, R. and Jones, G.M. (1986) Carrier detection in ataxia-telangiectasia. The Lancet, I:689.

Avigad, S., Cohen, B.E., Bauer, S., Schwartz, G., Frydman, M., Woo, S.L.C., Niny, Y. and **Shiloh, Y.** (1990) A single origin of phenylketonuria in Yemenite Jews. Nature, 344:168-170.

Savitsky, K., Bar-Shira, A., Gilad, S., Rotman, G., Ziv, Y., Vanagaite, L., Tagle, D.A., Smith, S., Uziel, T., Sfez, S., Ashkenazi, M., Pecker, I., Frydman, M., Harnik, R., Patanjali, S.R., Simmons, A., Clines, G.A., Sartiel, A., Gatti, R.A., Chessa, L., Sanal, O., Lavin, M.F., Jaspers, N.G.J., Taylor, A.M.R., Arlett, C.F., Miki, T., Weissman, S., Lovett, M., Collins, F.S. and **Shiloh, Y.** (1995) A single ataxia telangiectasia gene with a product similar to PI-3 kinase. Science, 268:1749-1753.

Savitsky, K., Sfez, S., Tagle, D., Ziv, Y., Sartiel, A., Collins, F.S. **Shiloh, Y.**, and Rotman, G*. (1995) The complete sequence of the coding region of the ATM gene reveals similarity to cell cycle regulators in different species. Hum. Mol. Genet., 4:2025-2032. (*Member of the Shiloh lab)

Gilad, S., Bar-Shira, A., Harnik, R., Shkedy, D., Ziv, Y., Khosravi, R., Brown, K., Vanagaite, L., Xu, G., Frydman, M., Lavin, M.F., Hill, D., Tagle, D., and **Shiloh, Y.** (1996) Ataxia-telangiectasia: founder effect among North African Jews. Hum. Mol. Genet., 5:2033-2038.

Ziv, Y., Bar-Shira, A., Pecker, I., Russell, P., Jorgensen, T.J., Tsarfaty, I., and **Shiloh, Y.** (1997) Recombinant ATM protein complements the cellular A-T phenotype. Oncogene, 15:159-167.

Rotman, G., and **Shiloh, Y.** (1997) Ataxia-telangiectasia and ATM: Possible involvement in cellular responses to oxidative damage and stress. BioEssays, 19:911-917

Gilad, S., Chessa, L., Khosravi R., Russell, P., Galanty, Y., Piane, M., Gatti, R.A., Jorgensen, T.J. **Shiloh, Y.**, and Bar-Shira, A.* (1998) Genotype-phenotype relationships in ataxia-telangiectasia (A-T) and A-T variants. Am. J. Hum. Genet., 62:551-561. (*Member of the Shiloh lab).

Banin, S., Moyal. L., Shieh, S.-Y., Taya, Y., Anderson, C.W., Chessa, L., Smorodinsky, N.I., Prives, C., Reiss, Y., **Shiloh, Y.,** and Ziv, Y.* (1998) Enhanced phosphorylation of p53 by ATM in response to DNA damage. Science, 281:1674-1677. (*Member of the Shiloh lab).

Uziel, T., Lerenthal, Y., Moyal, L., Andegeko, Y., Mittelman, L., and **Shiloh, Y.** (2003) Requirement of the MRN complex for ATM activation. EMBO J. 22:5612-5621.

Pereg, Y., Shkedy, D., de Graaf, P., Meulmeester, E., Edelson-Averbukh, M., Salek, M., Biton, S., Teunisse, A.F.A.S., Lehmann, W.D., Jochemsen, A.G., and **Shiloh, Y.** (2005) Multiple phosphorylations of Hdmx mediate its Hdm2- and ATM-dependent degradation in response to DNA damage. Proc. Natl. Acad. Sci. USA 102:5056-5061.

Ziv, Y., Bielopolski, D., Galanty, Y., Lukas, C., Taya, Y., Schultz, D.C., Lukas, J., Bekker-Jensen, S., Bartek, J., and **Shiloh, Y**. (2006) Chromatin relaxation in response to DNA double strand breaks: a novel ATM- and KAP-1-dependent pathway. Nature Cell Biol., 8:870-876.

Alterman, N., Fattal-Valevski, A., Moyal, L., Crawford, T., Lederman, H.M., Ziv, Y., and **Shiloh, Y.** (2007) Ataxia-telangiectasia: mild neurological presentation despite null *ATM* mutation and severe cellular phenotype. Am. J. Med. Genet. 143:1827-1834.

Bensimon, A., Schmidt, A., Ziv, Y., Elkon, R., Wang, S.-Y., Chen, D., Aebersold, R., and **Shiloh, Y**. (2010) ATM-dependent and independent dynamics of the nuclear phosphoproteome following DNA damage. Sci. Signal. 3(151) rs3.

Moyal, L., Gana-Weisz, M., Lerenthal, Y., Mass, G., So, S., Wang, S.-Y., Eppink, B., Chung, Y.-M., Shalev, G., Shema, E., Shkedy, D., Smorodinsky, N.I., van-Vliet, N., Kuster, B., Mann, M., Ciechanover, A., Dahm-Daphi, J., Kanaar, R., Hu, M.C-T., Chen, D.J., Oren, M., and **Shiloh, Y.** (2011) Requirement of ATM-dependent monoubiquitylation of histone H2B for timely repair of DNA double strand break. Mol. Cell, 41:529-542. **Featured article.**

Segal-Raz, H., Mass, G., Ziv-Lehrman, S., Wang, S.-Y., Strom, C., Helleday, T., Chen, D.J., and **Shiloh, Y.** (2011) ATM-mediated phosphorylation of polynucleotide kinase is required for effective DNA double-strand break repair. EMBO Reports, 12:713-719.

Rashi-Elkeles, S. Warnatz, H.-J., Elkon, R., Kupershtein, A., Chobod, Y., Paz, A., Amstislavskiy, V., Sultan, M., Safer, H., Nietfeld, W., Lehrach, H., Shamir, R., Yaspo, M.-L., and **Shiloh, Y.** (2014) Parallel profiling of cellular Responses to ionizing radiation at the levels of the transcriptome, cistrome and epigenome. Sci. Signal., 7:rs3.

Meir, M., Galanty, Y., Kashani, L., Blank, M., Khosravi, R., Fernández-Ávila, M.J., Cruz-Garcia, A., Star, A., Shochat, L., Thomas, Y., Garrett, L.J., Chamovitz, D.A., Bodine, D.M., Kurz, T., Huertas, P., Ziv, Y., and **Shiloh, Y.** (2015) The COP9 signalosome is vital for timely repair of DNA double-strand breaks. Nucleic Acids Res., 43: 4517-4530.

Gavish-Izakson, M., Bhavana, VB, Elkon, R., Prados-Carvajal, R., Barnabas, G.D., Pineiro Ugalde, A., Agami, A., Geiger, T., Huertas, P., Ziv, Y., and **Shiloh, Y.** (2018) Nuclear poly(A)-binding protein 1 is an ATM target and essential for DNA double-strand break repair. Nucleic Acids Res., 46:730-747.

Baranes-Bachar, K., Levy-Barda, A., Oehler, J., Reid, D.A., Soria-Bretones, I., Vos, T.C., Chung, D., Park, Y., Liu, C., Yoon, J.-B., Li, W., Dellaire, G., Misteli, T., Huertas, P., Rothenberg, E., Ramadan, K., Ziv, Y., and **Shiloh, Y.** (2018) The ubiquitin E3/E4 ligase, UBE4A, fine-tunes protein ubiquitylation and accumulation at sites of DNA damage facilitating double-strand break repair. Mol. Cell, 69:866-878.

Jachimowicz, R.D., Belleggia, F., Isensee, J., Velpula, V.B., Georgens, J., Bustos, M.A., Doll, M.A., Shenoy, A., Checa-Rodriguez, C., Wiederstain, J.L., Baranes-Bachar, K., Bartenhagen, C., Hertwig, F., Teper, N., Nishi, T., Schmitt, A., Distelmaier, F., Lüdecke, H.-J., Albreach, B., Krüger, M., Schumacher, B., Geiger, T., Hoon D.S.B., Huertas, P., Fischer, M., Hucho, T., Peifer, M., Ziv, Y., Reinhardt, H.C., Wieczorek, D., and **Shiloh, Y.** (2019) UBQLN4 represses homologous recombination and is overexpressed in aggressive tumors. Cell, 176:505-519 (Highlighted in Cancer Discovery).

Shiloh, Y. (2020) The cerebellar degeneration in ataxia-telangiectasia: a case for genome instability. DNA Repair, doi: 10.1016/j.dnarep.2020.102950.

Schlam-Babayov, S., Bensimon, A., Harel, M., Geiger, T., Aebersold, R., Ziv, Y., and **Shiloh, Y**. (2021) Phosphoproteomics reveals novel modes of function and inter-relationships among PIKKs in response to genotoxic stress. The EMBO Journal, 40(2):e104400. doi: 10.15252/embj.2020104400.

Haj, M., Levon, A., Frey, Y., Hourvitz, N., Campisi, J., Tzfati, Y., Elkon, R., Ziv, Y., and **Shiloh, Y.** (2023) Accelerated replicative senescence of ataxia-telangiectasia skin fibroblasts is retained at physiologic oxygen levels, with unique and common transcriptional patterns. Aging Cell, e13869. doi: 10.1111/acel.13869.

Frey, Y., Haj, M., Ziv, Y., Elkon, R., and **Shiloh, Y**. (2025) Broad repression of DNA repair genes in senescent cells identified by integration of transcriptomic data. Nucleic Acids Research doi: 10.1093/nar/gkae1257/7935007.

Haj, M., Frey, Y., Levon, A., Maliah, A., Ben-Yishai, T., Slutsky, R., Smoom, R., Tzfati, Y., Ben-David, U., Levy, C., Elkon, R., Ziv, Y., and **Shiloh, Y**. (2025) The cGAS-STING, p38 MAPK and p53 pathways link genome instability to accelerated cellular senescence in ATM-deficient murine lung fibroblasts. Proc. Natl. Acad. Sci. USA doi: 10.1073/pnas.2419196122 (**PNAS Inaugural Article**)

Frey, Y., Goehring, L., Haj, M., Rona, G., Fijen, C., Pagano, M., Huang, T.T., Rothenberg, E., Ziv, Y., and **Shiloh, Y**. (2025) ZC3H4 safeguards genome integrity by preventing transcription-replication conflicts at non-coding RNA loci. Science Advances 11(25):eadt8346. doi: 10.1126/sciadv.adt8346

Chetrit, A., Forer, R., Ziv, Y., Sadetzki, S., and **Shiloh, Y.** (2026) Increased chronic morbidity in ataxia-telangiectasia carriers identified in a population-based study. (In revision for PNAS).

Complete List of Peer-Reviewed Research Publications

1. **Shiloh, Y.** and Cohen, M.M. (1978) An improved technique for preparing bone marrow specimens for cytogenetic analysis. In Vitro, 14:510-515.
2. Cohen, M.M. and **Shiloh, Y.** (1978) Genetic toxicology of LSD. Mutation Res., 47:183-209.
3. **Shiloh, Y.**, Naparstek, B. and Cohen, M.M. (1979) Chromosomal aberrations in bone marrow specimens of malignant and pre-leukemic states. Isr. J. Med. Sci., 15:500-506.
4. **Shiloh, Y.** and Becker, Y. (1981) Kinetics of O^6-methylguanine repair in human normal and ataxia-telangiectasia cell lines and correlation of repair capacity with cellular sensitivity to methylating agents. Cancer Res., 41:5114-5120.
5. Ben-Hur, E., Kol, R., Heimer, Y. M., **Shiloh, Y.**, Tabor, E. and Becker, Y. (1981) An apparent correlation between the inhibition of induced ornithine decarboxylase activity by radiation and the capacity for DNA repair synthesis in normal and ataxia- telangiectasia fibroblasts. Radiat. Environ. Biophysics, 20:21-28.
6. **Shiloh, Y.**, Tabor, E. and Becker, Y. (1982) Cellular hypersensitivity to neocarzinostatin in ataxia-telangiectasia skin fibroblasts. Cancer Res., 42:2247-2249.
7. **Shiloh, Y.**, Tabor, E. and Becker, Y. (1982) Colony forming ability of ataxia-telangiectasia skin fibroblasts is an indicator of their early senescence and increased demand for growth factors. Exp. Cell Res., 140:191-199.
8. **Shiloh, Y.**, Tabor, E. and Becker, Y. (1982) The response of ataxia-telangiectasia homozygous and heterozygous skin fibroblasts to neocarzinostatin. Carcinogenesis, 3:815-820.
9. **Shiloh, Y.**, and Becker, Y. (1982) Reduced inhibition of replicon initiation and chain elongation by neocarzinostatin in skin fibroblasts from patients with ataxia-telangiectasia. Biochem. Biophys. Acta, 721:485-488.
10. **Shiloh, Y.**, Tabor, E. and Becker, Y. (1983) Similar repair of o^6-methylguanine in normal and ataxia-telangiectasia fibroblast strains: deficient repair capacity of lymphoblastoid cell lines does not reflect a genetic polymorphism. Mutation Res., 112:47-58.
11. **Shiloh, Y.**, Tabor, E. and Becker, Y. (1983) Repair of potentially lethal and sublethal damage induced by neocarzinostatin in normal and ataxia-telangiectasia skin fibroblasts. Biochem. Biophys. Res. Commun., 110:483-490.
12. Heimer, Y., Kol, R., **Shiloh, Y.** and Riklis, E. (1983) Psoralen plus near ultraviolet light: a possible new method for measuring DNA repair synthesis. Radiation Res., 95:541-549.
13. **Shiloh, Y.**, van der Schans, G.P., Lohman, P.H.M. and Becker, Y. (1983) Induction and repair of DNA damage in normal and ataxia-telangiectasia fibroblasts treated with neocarzinostatin. Carcinogenesis, 4:917-921.
14. **Shiloh, Y.**, Tabor, E. and Becker, Y. (1983) Abnormal response of ataxia-telangiectasia cells to agents that break the deoxyribose moiety of DNA via a targeted free radical mechanism. Carcinogenesis, 4:1317-1322.
15. **Shiloh, Y.**, Tabor, E. and Becker, Y. (1984) Cells from patients with ataxia-telangiectasia are abnormally sensitive to the cytotoxic effect of a tumor promoter, phorbol-12-myristate-13-acetate. Mutation Res., 149:283-286.
16. Sakai, K., Kanda, N., **Shiloh, Y.**, Donlon, T., Shipley, J., Dryja, T. and Latt, S. (1985) Molecular and cytological analysis of DNA amplification in retinoblastoma. Cancer Genet. Cytogenet., 17:95-112.
17. **Shiloh, Y.**, Shipley, J., Brodeur, G.M., Bruns, G., Korf, B., Donlon, T., Seeger, R., Sakai, K. and Latt, S. (1985) Differential amplification, assembly and relocation of multiple DNA sequences in human neuroblastomas and neuroblastoma cell lines. Proc. Natl. Acad. Sci. USA, 82:3761-3765.

18. Bruns, G., Kavathas, P., **Shiloh, Y.**, Sakai, K., Latt, S.A. and Herzenberg, L.A. (1985) The human T cell antigen Leu-2 (T8) is encoded on chromosome 2 near IgK. Hum. Genet., 71:311-314.
19. **Shiloh, Y.**, Donlon, T., Bruns, G., Breitman, M.L. and Tsui, L.-C. (1985) Assignment of the human gamma crystallin gene cluster to the long arm of chromosome #2, region q33-36. Hum. Genet., 73:17-19.
20. **Shiloh, Y.**, Kanda, N., Bruns, G., Sakai, K. and Latt, S. (1985) Two RFLPs identified by a human chromosome #2 clone at 2p15-2p16. Nucleic Acids Res. 13:5403.
21. **Shiloh, Y.**, Korf, B., Sakai, K., Brodeur, G., Seeger, R., Harris, P., Kanda, N., Alt, F. and Latt, S.A. (1986) Amplification and rearrangement of DNA sequences from the chromosomal region 2p24 in human neuroblastomas. Cancer Res., 46:5297-5301.
22. **Shiloh, Y.**, Sanford, K.K., Parshad, R. and Jones, G.M. (1986) Carrier detection in ataxia-telangiectasia. The Lancet, I:689.
23. Nagasawa, H., Kraemer, K., **Shiloh, Y.** and Little, J.B. (1987) Detection of ataxia-telangiectasia heterozygous cell lines by postirradiation cumulative labeling index: measurements with coded samples. Cancer Res., 47:398-402.
24. **Shiloh, Y.**, Kunkel, L.M. Rose, E., Korf, B. and Latt, S.A. (1987) Rapid cloning of multiple amplified DNA sequences from human neuroblastoma cell lines by competitive DNA reassociation. Gene, 51:53-59.
25. Tal, M., Wetzler, M., Josefsberg, Z., Deutsch, A., Assaf, D., Gutman, M., Givol, D., **Shiloh, Y.** and Schlessinger, Y. (1988) Sporadic amplification of the HER2/neu proto-oncogene in adenocarcinomas of various tissues. Cancer Res., 48:1517-1520.
26. Ziv, Y., Amiel, A., Jaspers, N.G.J., Berkel, A.I. and **Shiloh, Y.** (1989) Ataxia telangiectasia: a variant with altered in vitro phenotype of fibroblast cells. Mutation Res., 210:211-219.
27. Ziv, Y., Etkin, S., Danieli, T., Amiel, A., Ravia, Y., Jaspers, N.G.J., and **Shiloh, Y.**, (1989) Cellular and biochemical characteristics of an immortalized ataxia-telangiectasia (group AB) cell line. Cancer Res., 49:2495-2501.
28. Goodman, R.M., Bonne-Tamir, B., Adam, A., Voss, R., Bach, G., **Shiloh, Y.**, Bat-Miriam Katznelson, Barkai, G., Goldman, B., Padeh, B., Chemke, J. and Legum, C. (1989) Human genetics in Israel. J. Med. Genet., 26: 179-189.
29. Orgad, S., Yaar, L., Barkai, G., Avigad, S., **Shiloh, Y.**, Cohen, B., Yahav, I., Katznelson, D., Lieberman, A., Goldman, B. and Gazit, E. (1989) Carrier detection and prenatal diagnosis in phenylketonuria, cystic fibrosis and adrenal hyperplasia using molecular biology techniques. Harefuah, 116: 297-300 (Hebrew).
30. Gutman, M., Ravia, Y., Assaf, D., Yamamoto, T., Rozin, R. and **Shiloh, Y.** (1989) Structural alterations of c-myc and c-erbB-2 proto-oncogenes in human solid tumors: frequency and clinical significance. Int. J. Cancer, 44:802-805.
31. **Shiloh, Y.**, Parshad, R., Frydman, M., Sanford, K.K., Portnoi, S., Ziv, Y. and Jones, G.M. (1989) G_2 chromosomal radio- sensitivity in families with ataxia-telangiectasia. Hum. Genet., 84:15-18.
32. Avigad, S., Cohen, B.E., Bauer, S., Schwartz, G., Frydman, M., Woo, S.L.C., Niny, Y. and **Shiloh, Y.** (1990) A single origin of phenylketonuria in Yemenite Jews. Nature, 344:168-170.
33. Yeger, H., Mor, O., Pawlin, G., Kaplinsky, C. and **Shiloh, Y.** (1990) Importance of phenotypic and molecular characterization for identification of a neuroepithelioma cell line, NUB-20. Cancer Res., 50:2794-2802.
34. **Shiloh, Y.**, Litvak, G., Ziv, Y., Sandkuyl, L., Lehner, T., Hildesheimer, M., Buchris, V., Cremers, F., Szabo, P., White, B.N., Holden, .J.A. and Ott, J. (1990) Genetic mapping of X-linked albinism-deafness syndrome (ADFN) to Xq26.3-q27.1. Am. J. Hum. Genet., 47:20-27.
35. Ziv. Y., Rotman, G., Frydman, M., Foroud, T., Gatti, R.A. and **Shiloh, Y.** (1991) The ATC (ataxia-telangiectasia complementation group C) locus localizes to 11q22-q23. Genomics, 9:373-375.

36. Mor, O., Messinger, Y., Rotman, G., Bar-Am, I., Ravia, Y., Eddy, R.L., Shows, T.B., Park, J.-G., Gazdar, A.F. and **Shiloh, Y.** (1991) Novel DNA sequences at 10q26 are amplified in human gastric carcinoma cell lines: molecular cloning by competitive DNA reassociation. Nucleic Acids Res., 19:117-123.

37. Avigad, S., Kleiman, S., Weinstein, M., Cohen, B.E., Schwartz, G., Woo, S.L.C. and **Shiloh, Y.** (1991) Compound heterozygosity in non-PKU hyperphenyl-alaninemia: the contribution of mutations for classical PKU. Am. J. Hum. Genet., 49:393-399.

38. Foroud, T., Sobel, E., Ziv, Y., Goradia, T., Wei, S., Charmley, P., McConville, C., Chao, A., Chessa, L., Tolun A., Sanal, O., Julier, C., Concannon, P., Fiorilli, M., Taylor, M., **Shiloh, Y.**, Lange, K. and Gatti, R.A. (1991) Localization of the AT locus to an 8 cM interval defined by STMY and S132. Am. J. Hum. Genet., 49:1263-1279.

39. Shomrat, R., Driks, N., Legum, C. and **Shiloh, Y.** (1992) The use of dystrophin genomic and cDNA probes for solving difficulties in carrier detection and prenatal diagnosis of Duchenne muscular dystrophy. Am. J. Med. Genet., 42:281-287.

40. Ziv, Y., Frydman, M., Lange, E., Zelnik, N., Rotman, G., Julier, C., Jaspers, N.G.J., Dagan, Y., Abeliovicz, D., Dar, H., Borochowitz, Z., Lathrop, M., Gatti, R.A. and **Shiloh, Y.** (1992) Ataxia-telangiectasia: linkage analysis in highly inbred Arab and Druze families and differentiation from an ataxia-microcephaly-cataract syndrome. Hum. Genet., 88:619-626.

41. **Shiloh, Y.**, Mor, O., Manor, A., Bar-Am, I., Rotman, G., Eubanks, J., Gutman, M., Ranzani, G.N., Houldsworth, J., Evans, G. and Avivi, L. (1992) DNA sequences amplified in cancer cells: an interface between tumor biology and human genome analysis. Mutat. Res., 276:329-337.

42. Bar-Am, I., Mor, O., Yeger, H., **Shiloh, Y.** and Avivi, L. (1992) Detection of amplified DNA sequences in human tumor cell lines by fluorescent in situ hybridization. Genes, Chromosomes and Cancer, 4:314- 320.

43. Sobel, E., Lange, E., Jaspers, N.G.J., Chessa, L., Sanal, O., **Shiloh, Y.**, Taylor, A.M.R., Weemaes, C.M.A., Lange, K. and Gatti, R.A. (1992) Ataxia-telangiectasia: evidence for genetic heterogeneity. Am. J. Hum. Genet., 50:1343-1348.

44. Kleiman, S., Schwartz, G., Akawi, Y., Woo, S.L.C. and **Shiloh, Y.** (1992) A 22-bp deletion at the phenylalanine hydroxylase gene causing phenylketonuria in an Arab family. Human Mutation, 1:344-346.

45. Kleiman, S., Bernstein, J., Schwartz, G., Woo, S.L.C. and **Shiloh, Y.** (1992) A defective splice site at the phenylalanine hydroxylase gene in phenylketonuria and benign hyperphenyl-alaninemia among Palestinian Arabs. Human Mutation, 1:340-343.

46. Weinstein, M., Eisensmith, R.C., Abadie, V., Avigad, S., Lyonnet, S., Schwartz, G., Munnich, A., Woo, S.L.C. and **Shiloh, Y.** (1993) A missense mutation, S349P, completely inactivates phenylalanine hydroxylase in North African Jews with phenylketonuria. Hum. Genet., 90:645-649.

47. Mor, O., Ranzani, G.N., Ravia, Y., Rotman, G., Gutman, M., Manor, A., Amadori, D., Houldsworth, J., Hollstein, M., van der Bosch, K., Schwab, M. and **Shiloh, Y.** (1993) DNA amplification in human gastric carcinomas. Cancer Genet. Cytogenet., 65:111-114.

48. Kleiman, S., Bernstein, J., Schwartz, G., Brand, N., Elitzur, A., Woo, S.L.C. and **Shiloh, Y.** (1993) Phenylketonuria: variable phenotypic outcomes of the R261Q mutation, and maternal PKU in the offspring of a healthy homozygote. J. Med. Genet., 30:284- 288.

49. Kleiman, S., Li, J., Schwartz, G., Eisensmith, R.C., Woo, S.L.C. and **Shiloh, Y.** (1993) Inactivation of phenylalanine hydroxylase by a missense mutation, R270S, in a Palestinian kinship with phenylketonuria. Hum. Mol. Genet., 2:605-606.

50. McConville, C.M., Byrd, P.J., Ambrose, H.J., Stankovic, T., Ziv, Y., Bar-Shira, A., Vanagaite, L., Rotman, G., **Shiloh, Y.**, Gillett, G.T., Riley, J.H. and Taylor, A.M.R. (1993) Paired STSs amplified from radiation hybrids, and from associated YACs, identify highly polymorphic loci

flanking the ataxia-telangiectasia locus on chromosome 11q22-23. Hum. Mol. Genet., 2:969-974.

51. Shomrat, R., Gluck, E., Legum, C. and **Shiloh, Y.** (1994) A relatively low proportion of dystrophin gene deletions in Israeli DMD and BMD patients. Am. J. Med. Genet., 49:369-373.
52. Legum, C., Shomrat, R., Glassner, M.C. and **Shiloh, Y.** (1994) A molecular survey of Israeli Duchenne and Becker muscular dystrophy patients. Biomed. Pharmacol. 48:359-364.
53. Kleiman, S., Avigad, S., Vanagaite, L., Shmuelevitz, A., David, M., Eisensmith, R., Brand, N., Schwartz, G., Rey, F., Munnich, A., Woo, S.L.C. and **Shiloh, Y.** (1994) Origins of hyperphenyl-alaninemia in Israel. Europ. J. Hum. Genet., 2:24-34.
54. Vanagaite, L., Savitsky, K., Rotman, G., Ziv, Y., Gerken, S.C., White, R., Weissenbach, J., Gillett, G., Benham, F.J., Richard, C.W., James, M.R., Collins, F.S. and **Shiloh, Y.** (1994) Physical localization of microsatellite markers at the ataxia-telangiectasia locus at 11q22-23. Genomics, 22:231-233.
55. Ambrose, H.J., Byrd, P.J., McConville, C.M., Cooper, P.R., Stankovitz, T., Riley, J.H., **Shiloh, Y.**, McNamara, J.O., Fuko, T. and Taylor, A.M.R. (1994) A physical map across chromosome 11q22-23 containing the major locus for ataxia telangiectasia. Genomics, 21:612-619.
56. Rotman, G., Vanagaite, L., Collins, F.S. and **Shiloh, Y.** (1994) Three dinucleotide repeat polymorphisms at the ataxia-telangiectasia locus. Hum. Mol. Genet., 3:2079.
57. Rotman, G., Savitsky, K., Ziv, Y., Cole, C.G., Higgins, M.J., Bar-Am, I., Dunham, I., Bar-Shira, A., Vanagaite, L., Shinzen, Q., Zhang, J., Nowak, N.J., Chandrasekharappa, S.C., Lehrach, H., Avivi, L., Shows, T.B., Collins, F.S., Bentley, D.R. and **Shiloh, Y.** (1994) A YAC contig spanning the ataxia-telangiectasia locus (groups A and C) at 11q22-23. Genomics, 24:234-242.
58. Taylor, A.M.R., Rotman, G., **Shiloh, Y.**, Byrd, P.J., and McConville, C.M. (1994) A haplotype common to intermediate radiosensitivity variants of ataxia-telangiectasia. Int. J. Radiat. Biol., 66:S35-S41.
59. Rotman, G., Savitsky, K., Vanagaite, L., Bar-Shira, A., Ziv, Y., Gilad, S., Uchenik, V., Smith, S. and **Shiloh, Y.** (1994) Physical and genetic mapping at the ATA/ATC locus on chromosome 11q22-23. Int. J. Radiat. Biol., 66:S63-S66.
60. Gatti, R.A., Lange, E., Rotman, G., Chen, X., Uhrhammer, N., Liang, T., Chiplunkar, S., Yang, L., Udar, N., Dandekar, S., Sheikhavandi, S., Wang, Z., Yang, H.-M., Polikow, J., Elashoff, M., Teletar, M., Sanal, O., Chessa, L., McConville, C., Taylor, M., **Shiloh, Y.**, Porras, O., Borreson, A.-L., Wegner, R.-D., Curry, C., Gerken, S., Lange, K. and Concannon, P. (1994) Genetic haplotyping of ataxia-telangiectasia families localizes the major gene to an 850 kb region on chromosome 11q23.1. Int. J. Radiat. Biol., 66:S57-S62.
61. Rotman, G., Vanagaite, L., Collins, F.S. and **Shiloh, Y.** (1995) Rapid identification of polymorphic CA-repeats in YAC clones spanning the ataxia-telangiectasia locus on chromosome 11q22-23. Mol. Biotechnol., 3:85-92.
62. Vanagaite, L., James, M.R., Rotman, G., Savitsky, K., Bar-Shira, A., Gilad, S., Ziv, Y., Uchenik, V., Sartiel, A., Collins, F.S., Sheffield, V.C., Weissenbach, J. and **Shiloh, Y.** (1995) A high- density microsatellite map of the ataxia-telangiectasia locus. Hum. Genet., 95:451-454.
63. Ben Arush, M.W., Rosenthal, J., Dale, J., Horovitch, Y., Herzl, G., Ben Arie, J., Ziv, Y. and **Shiloh, Y.** (1995) Ataxia telangiectasia and lymphoma: An indication for individualized chemotherapy dosing - report of treatment in a highly inbred Arab family. Pediatr. Hematol. Oncol., 12:163-169.
64. Lange, E., Borreson, A.-L., Chen, X., Chessa, L., Chiplunkar, S., Concannon, P., Dandekar, S., Gerken, S., Lange, K., Liang, T., McConville, C., Polakow, J., Porras, O., Rotman, G., Sanal, O., Telatar, M., Sheikhavandi, S., **Shiloh, Y.**, Sobel, E., Taylor, M., Udar, N., Uhrhammer, N., Vanagaite, L., Wang, Z., Yang, H.-M., Yang, L., Ziv, Y. and Gatti, R.A. (1995) Localization of an ataxia-telangiectasia gene to a 850 kb interval on chromosome 11q23.1 by linkage analysis of 176 families in an international consortium. Am. J. Hum. Genet., 57:112-119.

65. Savitsky, K., Bar-Shira, A., Gilad, S., Rotman, G., Ziv, Y., Vanagaite, L., Tagle, D.A., Smith, S., Uziel, T., Sfez, S., Ashkenazi, M., Pecker, I., Frydman, M., Harnik, R., Patanjali, S.R., Simmons, A., Clines, G.A., Sartiel, A., Gatti, R.A., Chessa, L., Sanal, O., Lavin, M.F., Jaspers, N.G.J., Taylor, A.M.R., Arlett, C.F., Miki, T., Weissman, S., Lovett, M., Collins, F.S. and **Shiloh, Y.** (1995) A single ataxia telangiectasia gene with a product similar to PI-3 kinase. Science, 268:1749-1753.

66. Frydman, M., Staussberg, Shomrat, R., Legum, C. and **Shiloh, Y.** (1995) Duchenne muscular dystrophy and idiopathic hyperckemia segregating in a family. Am. J. Med. Genet., 58:209-213.

67. Ziv, Y., Bar-Shira, A., Jorgensen, T.J., Russell, P.S., Sartiel, A., Shows, T.B., Eddy, R.L., Buchwald, M., Legerski, R., Schimke, R.T. and **Shiloh, Y.** (1995) Human cDNA clones that modify radiomimetic sensitivity of ataxia-telangiectasia (group A) cells. Somatic Cell Mol. Genet., 21:99-111.

68. Negrini, M., Rasio, D., Hampton, G.M., Sabbioni, S., Rattan, S., Carter, S.L., Rosenberg, A.L., Schwartz, G.F., **Shiloh, Y.**, Cavenee, W.K. and Croce, C.M. (1995) Definition and refinement of chromosome 11 regions of LOH in breast cancer: identification of a new region at 11q23.3. Cancer Res., 55:3003-3007.

69. Morrow, D.M., Tagle, D.A., **Shiloh, Y.**, Collins, F.S. and Hieter, P. (1995) TEL1, a Saccharomyces cerevisiae homologue of the human gene mutated in ataxia telangiectasia, is functionally related to the yeast checkpoint gene MEC1/ESR1. Cell, 82:831- 840.

70. Savitsky, K., Sfez, S., Tagle, D., Ziv, Y., Sartiel, A., Collins, F.S. **Shiloh, Y.**, and Rotman, G. (1995) The complete sequence of the coding region of the ATM gene reveals similarity to cell cycle regulators in different species. Hum. Mol. Genet., 4:2025-2032.

71. Savitsky, K., Ziv, Y., Bar-Shira, A., Gilad, S., Tagle, D.A., Smith, S., Uziel, T., Sfez, S., Nahmias, J., Sartiel, A., Eddy, R.L., Shows, T.B., Povey, S., Collins, F.S., **Shiloh, Y.** and Rotman, G.* (1996) A human gene (DDX10) encoding a putative DEAD-box RNA helicase at 11q22-23. Genomics, 33:199-206. (*Member of the Shiloh lab).

72. Gilad, S., Khosravi, R., Uziel, T., Ziv, Y., Rotman, G., Savitsky, K., Smith, S., Chessa, L., Harnik, R., Shkedi, D., Frydman, M., Sanal, O., Portnoi, S., Goldwicz, Z., Jaspers, N.G.J., Gatti, R.A., Lenoir, G., Lavin, M.F., Tatsumi, K., Wegner, R.D., **Shiloh, Y.** and Bar-Shira, A.* (1996) Predominance of null mutations in ataxia-telangiectasia. Hum. Mol. Genet., 5:433-439. (*Member of the Shiloh lab).

73. Uziel, T., Savitsky, K., Platzer, M., Ziv, Y., Helbitz, T., Nehls, M., Boehm, T., Rosenthal, A., **Shiloh, Y.** and Rotman, G.* (1996) Genomic organization of the ATM gene. Genomics, 33:317- 320. (*Member of the Shiloh lab).

74. Pecker, I., Avraham, K., Gilbert, D.J., Savitsky, K., Rotman, G., Harnik, R., Fukao, T., Schrock, E., Hirotsume, S., Tagle, D.A., Collins, F.S., Wynshaw-Boris, A., Ried, T., Copeland, N.G., Jenkins, N.A., **Shiloh, Y.** and Ziv, Y.* (1996) Identification and chromosomal localization of Atm, the mouse homolog of the ataxia-telangiectasia gene. Genomics, 35:39-45. (*Member of the Shiloh lab).

75. Telatar, M., Wang, Z., Udar, N., Liang, T., Bernatowska-Matsuszkiewicz, E., Lavin, M., **Shiloh, Y.**, Concannon, P., Good, R.A., and Gatti, R.A. (1996) Ataxia-telangiectasia: Mutations in ATM cDNA detected by protein truncation screening. Am. J. Hum. Genet., 59:40-44.

76. Barlow, C., Hirotsune, S., Paylor, R., Liyanage, M., Eckhaus, M., Collins, F.S., **Shiloh, Y.**, Crawley, J.N., Ried, T., Tagle, D., and Wynshaw-Boris, A. (1996) Atm-deficient mice: A paradigm of ataxia-telangiectasia. Cell, 86:159-171.

77. Beamish, H., Williams, R., Chen, P., Khanna, K.K., Hobson, D., Watters, D., **Shiloh, Y.**, and Lavin, M.F. (1996) Rapamycin resistance in ataxia-telangiectasia. Oncogene, 13:963-970.

78. Gilad, S., Bar-Shira, A., Harnik, R., Shkedy, D., Ziv, Y., Khosravi, R., Brown, K., Vanagaite, L., Xu, G., Frydman, M., Lavin, M.F., Hill, D., Tagle, D., and **Shiloh, Y.** (1996) Ataxia-telangiectasia: founder effect among North African Jews. Hum. Mol. Genet., 5:2033-2038.

79. Jongmans, W., Verhaegh, G.W.C.T., Jaspers, N.G.J., Demant, P., Natarajan, A., **Shiloh, Y.**, Oshimura, M., Stanbridge, E.J., Athwal, R.S., Newbold, R.F., Lohman, P.H.M., and Zdzienicka, M.Z. (1996) The defect in the AT-like hamster cell mutants is complemented by mouse chromosome 9 but not by any of the human chromosomes. Mutat. Res., 364:91-102.

80. Laake, K., Odegard, A., Andersen, T.I., Bukholm, I., Karesen, R., Nsaland, J.M., Ottestad, L., **Shiloh, Y.**, and Borreson, A.-L. (1997) Loss of heterozygosity at 11q23.1 in breast carcinomas: Indication for involvement of a gene distal and close to ATM. Genes, Chromosomes and Cancer, 18:175-180.

81. Brown, K., Ziv, Y., Sadanandan, S.N., Chessa, L., Collins, F.S., **Shiloh, Y.**, and Tagle, D.A. (1997) The ataxia-telangiectasia gene product, a constitutively expressed nuclear protein that is not upregulated following genome damage. Proc. Natl. Acad. Sci. USA, 94:1840-1845.

82. Morgan, S.E., Lovly, C., Pandita, T., **Shiloh, Y.**, and Kastan, M.B. (1997) Fragments of ATM which have dominant-negative or complementing activity. Mol. Cell. Biol., 17:2020-2029.

83. Khanna, K.K., Yan, J., Watters, D., Hobson, K., Beamish, H., Spring, K., **Shiloh, Y.**, Gatti, R.A., and Lavin, M.F. (1997) Defective signaling through the B cell antigen receptor in Epstein-Barr virus-transformed ataxia-telangiectasia cells. J. Biol. Chem., 272:9489-9495.

84. Savitsky, K., Platzer, M., Uziel, T., Gilad, S., Sartiel, A., Rosenthal, A., Elroy-Stein, O., **Shiloh, Y.**, and Rotman, G.* (1997) Ataxia-telangiectasia: Structural diversity of untranslated sequences suggests complex posttranscriptional regulation of ATM gene expression. Nucleic Acids Res., 25:1678- 1684. (*Member of the Shiloh lab).

85. Shafman, T., Khanna, K.K., Kedar, P., Spring, K., Kozlov, S., Yen, T., Hobson, K., Gatei, M., Zhang, N., Watters, D., Egerton, M., **Shiloh, Y.**, Kharbanda, S., Kufe, D. and Lavin, M. (1997) Role of ATM protein in stress response to DNA damage: Evidence for interaction with c-Abl. Nature, 387:520-523.

86. Ziv, Y., Bar-Shira, A., Pecker, I., Russell, P., Jorgensen, T.J., Tsarfaty, I., and **Shiloh, Y.** (1997) Recombinant ATM protein complements the cellular A-T phenotype. Oncogene, 15:159-167.

87. Platzer, M., Rotman, G., Bauer, D., Uziel, T., Savitsky, K., Bar-Shira, A., Gilad, S., **Shiloh, Y.** and Rosenthal, A. (1997) Ataxia-telangiectasia locus: Analysis of 184 kb of genomic DNA containing the entire ATM gene. Genome Res.,7:592-605.

88. Cali, F., Piazza, A., Dianzani I., Desviat L.R., Perez, B., Ugarte, M., Ozguc, M., **Shiloh, Y.**, Giammattasio, S., Carducci, C., and Romano, V. (1997) The STR 252-IVS10nt546-VNTR 7 phenylalanine hydroxylase minihaplotype in five Mediterranean samples. Hum. Genet., 100:350-355.

89. Gilad, S., Khosravi, R., Harnik, R., Ziv, Y., Shkedy, D., Galanti, Y., Frydman, Y., Carmi, R., Sanal, O., Chessa, L., Smeets, D., **Shiloh, Y.** and Bar-Shira, A.* (1998) Identification of ATM mutations using extended RT-PCR and restriction endonuclease fingerprinting, and elucidation of the repertoire of A-T mutations in Israel. Hum. Mutation., 11:69-75. (*Member of the Shiloh lab).

90. Fukao, T., Tashita, H., Teramoto, T., Inoue, R., Kaneko, H., Komiyama, K., Bar-Shira, A., Gilad, S., **Shiloh, Y.**, Nishimura, M. and Kondo, N. (1998) A novel exonic mutation (5319 G to A) resulting in two aberrantly spliced transcripts of the ATM gene in a Japanese patient with ataxia-telangiectasia. Hum. Mutation , Suppl. 1:S223-S225.

91. Gilad, S., Chessa, L., Khosravi R., Russell, P., Galanty, Y., Piane, M., Gatti, R.A., Jorgensen, T.J., **Shiloh, Y.**, and Bar-Shira, A.* (1998) Genotype-phenotype relationships in ataxia-telangiectasia (A-T) and A-T variants. Am. J. Hum. Genet., 62:551-561. (*Member of the Shiloh lab).

92. Lim, D.-S., Kirsch, D.G., Canman, C.E., Ahn, A.-H., Ziv, Y., Newman, L.S., Darnell, R.B., Shiloh, Y., and Kastan, M.B. (1998) ATM binds to β-adaptin in cytoplamic vesicles. Proc. Natl. Acad. Sci. USA , 95:10146- 10151.

93. Banin, S., Moyal. L., Shieh, S.-Y., Taya, Y., Anderson, C.W., Chessa, L., Smorodinsky, N.I., Prives, C., Reiss, Y., **Shiloh, Y**., and Ziv, Y.* (1998) Enhanced phosphorylation of p53 by ATM in response to DNA damage. Science, 281:1674-1677. (*Member of the Shiloh lab).

94. Fukao, T., Song, X.-Q., Yoshida, T., Tashita, H., Kaneko, H., Teramoto, T., Inoue, R., Hiratani, M., Taniguchi, A., Arai J., Wakiguchi, H., Bar-Shira, A., **Shiloh, Y**., and Kondo, N. (1998) Ataxia-telangiectasia (A-T) in the Japanese population: Identification of R1917X, W2491R, R2909G, IVS33(+2) gt to ga, and 7883del5, the latter two being relatively common mutations. Hum. Mutation, 12:338-343.

95. Eilam, R., Peter, Y., Elson, A., Rotman, G., **Shiloh, Y.,** Leder, P., Groner, Y., and Segal, M. (1998) Selective loss of dopaminergic nigro-striatal neurons in brains of Atm-deficient mice. Proc. Natl. Acad. Sci. USA, 95:12653-12656.

96. Rhodes, N., D'Souza, T., Foster, C.D., Ziv, Y., Kirsch, D.G., **Shiloh, Y.**, Kastan, M.B., Reinhart, P.H., and Gilmer, T.M. (1998) Defective potassium currents in ataxia-telangiectasia fibroblasts. Genes. Dev., 12:3686-3692.

97. Sandoval, N., Platzer, M., Rosenthal, A., Doerk, T., Bendix, R., Skwaran, B., Stuhrmann, M., Wegner, R-.D., Sperling, K., Banin, S., **Shiloh, Y**., Baumer, A., Bernthaler, U., Sennefelder, H., Brohm, M., Weber, B.H.F., and Schindler, D. (1999) Characterization of ATM gene mutations in 67 ataxia-telangiectasia families. Hum. Mol. Genet., 8:69-79.

98. Hassin-Baer, S., Bar-Shira, A., Gilad, S., Galanty, Y., Khosravi, R., Lossos, A., Giladi, N., Weitz, R., Gldhammer, Y., and **Shiloh, Y**. (1999) No mutations in *ATM*, the gene responsible for ataxia-telangiectasia, in patients with cerebellar ataxias. J. Neurol., 246:716-719.

99. Araki, R., Fukumura, R., Fujimori, A., Taya, Y., **Shiloh, Y**., Kurimasa, A., Burma, S., Li, G.C., Chen, D.J., Sato, K., Hoki, Y., Tatsumi, K., and Abe, M. (1999) Enhanced phosphorylation of Ser18p53 following DNA damage to dna-pkcs-deficient cells. Cancer Res., 59:3543-3546.

100. Johnson, R.T., Gotoh, E., Mullinger, A.M., Ryan, A.J., **Shiloh, Y**., Ziv, Y., and Squires, S. (1999) Targeting double-strand breaks to replicative DNA identifies a subpathway of DSB repair that is defective in ataxia- telangiectasia. Biochem. Biophys. Res. Commun., 261:317-325.

101. Khosravi, R., Maya, R., Gottlieb, T., Oren, M., **Shiloh, Y**. and Shkedy, D.* (1999) Rapid ATM-dependent phosphorylation of MDM2 precedes p53 accumulation in response to DNA damage. Proc. Natl. Acad. Sci. USA, 96:14973-14977. (*Member of the Shiloh lab).

102. Zhao, S., Weng, Y.-C., Yuan, S.-S.F., Lin, Y.-T., Hsu, H.-C., Lin, S.-C.J., Gerbino, E., Song, M.-H., Zdzienicka, M.Z., Gatti, R.A., Ziv, Y., **Shiloh, Y.**, and Lee, Y.-H.P. (2000) A functional link between ataxia-telangiectasia and Nijmegen breakage syndrome gene products. Nature, 405:473-477.

103. O'Neil, T., Dwyer, A., Ziv, Y., Chan, D.W., Lees-Miller, S.P., Abraham, R.T., Lai, J.H., Hill, D., **Shiloh, Y.**, Cantley, L.C., and Rathbun, G. (2000) Utilization of oriented peptide libraries to identify substrate motifs selected by ATM. J. Biol. Chem., 275:22719-22727.

104. Li, S., Ting, N.S., Zheng, L., Chen, P.L., Ziv, Y., **Shiloh, Y.**, Lee, E.Y., and Lee, W.H. (2000) Functional link of BRCA1 and ataxia telangiectasia gene product in DNA damage response. Nature, 406:210-215.

105. Matsuoka, S., Rotman, G., Ogawa, A., **Shiloh, Y.**, Tamai, K., and Elledge, S. (2000) ATM phosphorylates Chk2 *in vivo* and *in vitro*. Proc. Natl. Acad. Sci. USA, 97:10389-10394.

106. Gatei, M., Shkedy, D., Khanna, K.K., Uziel, T., **Shiloh, Y**., Pandita, T.K., Lavin, M., and Rotman, G. (2001) Ataxia-telangiectasia: chronic activation of damage-responsive functions is reduced by α–lipoic acid. Oncogene, 20:289-294.

107. Li, N., Banin, S., Ouyang, H., Li, G.C., Courtois, G., **Shiloh, Y**., Karin, M., and Rotman, G. (2001) ATM is required for IKK activation in response to DNA double strand breaks. J. Biol. Chem., 276:8898-8903.

108. Peter, Y., Rotman, G., Lotem, K., Elson, A., **Shiloh, Y.**, and Groner, Y. (2001) Elevated Cu/Zn-SOD exacerbates radiation sensitivity and hematopoietic abnormalities of Atm-deficient mice. EMBO J., 20:1538- 1546.

109. Kamsler, a., Daily, D., Hochman, A., Stern, N., **Shiloh, Y**., Rotman, G., and Barzilai, A. (2001) Increased oxidative stress in ataxia telangiectasia evidenced by alterations in redox state of brains from Atm-deficient mice. Cancer Res., 61:1849-1854.

110. Buschmann, T., Potapova, O., Bar-Shira, A., Ivanov, V.N., Fuchs, S.Y., Henderson, S., Fried, V., Minamoto, T., Alarcon-Vargas, D., Pincus, M., Gaarde, W., Holbrook, N.J., **Shiloh, Y.**, and Ronai, Ze'ev (2001) Jun NH_2-terminal kinase phosphorylation of p53 on Thr-81 is important for p53 stabilization and transcriptional activities in response to stress. Mol. Cell Biol. 21:2743-2754.

111. Maya, R., Balass, M., Kim, S.-T., Shkedy, D., Leal, J., Shifman, O., Moas, M., Buschmann, T., Ronai, Z., **Shiloh, Y.,** Kastan, M.B., Katzir, E., and Oren, M. (2001) ATM-dependent phosphorylation of Mdm2 on serine 395: role in p53 activation by DNA damage. Genes Dev.15:1067-1077.

112. Spring, K., Cross, S., Li, C., Ben-Senior, L., Watters, D., Waring, P., Ahangari, F., Lu, S., Chen, P., Misko, I., Paterson, C., Kay, G., Smorodinsky, N.I., **Shiloh, Y.**, and Lavin, M.F. (2001) *Atm*-knock-in mice harboring an in-frame deletion corresponding to the human ATM7636del9 common mutation exhibit a variant phenotype. Cancer Res. 61:4561-4568.

113. Macaulay, V., Salisbury, A.J., Bohula, E.A., Playford, M.P., Smorodinsky, N.I., and **Shiloh, Y.** (2001) Downregulation of the type 1 insulin-like growth factor receptor in mouse melanoma cells is associated with enhanced radiosensitivity and impaired activation of Atm kinase. Oncogene 20:4029-4030.

114. Kishi, S., Zhou, X.Z., Ziv, Y., Khoo, C., Hill, D.E., **Shiloh, Y.**, and Lu, K.P. (2001) Telomeric protein Oin2/TRF1 is an important ATM target in response to double strand DNA breaks. J. Biol. Chem. 276:29282-29291.

115. Andegeko, Y., Moyal, L., Mittelman, L., Tsarfaty, I., **Shiloh, Y.**, and Rotman, G.* (2001) Nuclear retention of ATM at sites of DNA double strand breaks. J. Biol. Chem., 276:38224-38230. (*Member of the Shiloh lab).

116. Bar-Shira, A., Rashi-Elkeles, S., Zlochover, L., Moyal, L., Smorodonsky, N.I., Seger, R., and **Shiloh, Y.** (2002) ATM-dependent activation of the gene encoding MAP kinase phosphatase 5 by radiomimetic DNA damage. Oncogene, 21:849-855.

117. Stern, N., Hochman, A., Zemach, N., Weizman, N., Hammel, I., **Shiloh, Y.**, Rotman, G., and Bazrilai, A. (2002) Accumulation of DNA damage and reduced levels of nicotine adenine dinucleotide in the brains of Atm-deficient mice. J. Biol. Chem., 277:602-608.

118. Taylor, A., Shang, F., Nowell, T., Galanty, Y., and **Shiloh, Y.** (2002) Ubiquitination capabilities in response to neocarzinostatin and H_2O_2 stress in cell lines from patients with ataxia-telangiectasia. Oncogene 21:4363-4373.

119. Sapkota, G.P., Deak, M., Kieloch, A., Morrice, N., Goodarzi, A., Smyth, C., **Shiloh, Y**., Lees-Miller, S.P., and Alessi, D.R. (2002) Ionising radiation induces ATM-mediated phosphorylation of LKB1/STK11 at Thr366. Biochem. J. 368:507-516.

120. Weizman, N., **Shiloh, Y**., and Barzilai, A. (2003) Contribution of the Atm protein to maintaining cellular homeostasis evidenced by continuous activation of the AP-1 pathway in Atm-deficient brains. J. Biol. Chem., 278:6741-6747.

121. Elkon, R., Linhart, C., Sharan, R., Shamir, R., and **Shiloh, Y**. (2003) Genome-wide *in-silico* identification of transcriptional regulators controlling cell cycle in human cells. Genome Res. 13:773-780.

122. Uziel, T., Lerenthal, Y., Moyal, L., Andegeko, Y., Mittelman, L., and **Shiloh, Y.** (2003) Requirement of the MRN complex for ATM activation. EMBO J. 22:5612-5621.

123. Orlev, N., Shamir, R., and **Shiloh, Y**. (2004) PIVOT: **P**rotein **I**nteraction **V**isualizati**O**n **T**ool (Bioinformatic Application Note) Bioinformatics, 20:424-425.

124. Elkon, R., Zeller, K., Linhart, C., Dang, C.V., Shamir, R., and **Shiloh, Y.** (2004) *In silico* identification of transcriptional regulators associated with c-Myc. Nucleic Acids Res., 32:4955-4961.

125. **Shiloh, Y**., Andegeko, Y., and Tsarfaty, I. (2004) In search of drug treatment for genetic defects in the DNA damage responses: the example of ataxia-telangiectasia. Seminars in Cancer Biology, 14:295-305.

126. Pereg, Y., Shkedy, D., de Graaf, P., Meulmeester, E., Edelson-Averbukh, M., Salek, M., Biton, S., Teunisse, A.F.A.S., Lehmann, W.D., Jochemsen, A.G., and **Shiloh, Y.** (2005) Multiple phosphorylations of Hdmx mediate its Hdm2- and ATM-dependent degradation in response to DNA damage. Proc. Natl. Acad. Sci. USA 102:5056-5061.

127. Elkon, R., Rashi-Elkeles, S., Lerenthal, Y., Linhart, C., Tenne, T., Amariglio, N., Rechavi, G., Shamir, R., and **Shiloh, Y.** (2005) Dissection of a DNA damage-induced transcriptional network using a combination of microarrays, RNAi, and computational promoter analysis. Genome Biology Vol. 6, paper R43.

128. Bhoumik, A., Takahashi, S., Breitwieser, W., **Shiloh, Y**., Jones, N., and Ronai, Z. (2005) ATM-dependent phosphorylation of ATF2 is required in the DNA damage response. Mol. Cell 18:577-587.

129. Okamoto, K., Kashima, K., Pereg, Y., Ishida, M., Yamazaki, S., Nota, A., Teuniss, A., Migliorini, D., Kitabayashi, I., Marine, J.-C., Prives, C., **Shiloh, Y**., Jochemsen, A.G., and Taya, Y. (2005) DNA damage-induced phosphorylation of MdmX at serine-367 activates p53 by targeting MdmX for Mdm2-dependent degradation. Mol. Cell. Biol., 25:9608-9620.

130. Ziv, S., Brenner, O., Amariglio, N., Smorodinsky, N.I., Carrion, D.V., Sharma, G.G., Pandita, T.R., Sharma, M., Elkon, R., Katzin, N., Bar-Am, I., Pandita, T.K., Kucherlapati, R., Rechavi, G., **Shiloh, Y**., and Barzilai, A. (2005) Impaired genomic stability but not oxidative stress enhances cancer predisposition in Atm-deficient mice. Hum. Mol. Genet., 14:2922-2943.

131. Shamir, R., Maron-Katz, A., Tanay, A., Linhart, C., Steinfeld, U., Sgaran, R., **Shiloh, Y**., and Elkon, R. (2005) EXPANDER - an integrative suite for microarray data analysis. BMC Bioinformatics, vol. 6, paper #232.

132. Linhart, C., Elkon, R., **Shiloh, Y.**, and Shamir, R. (2005) Deciphering transcriptional regulatory elements that encode specific cell-cycle phasing by comparative genomics analysis. Cell Cycle, 4:1788-1797.

133. Rashi-Elkeles, S., Elkon, R., Weizman, N., Linhart, C., Amariglio, N., Sternberg, G., Rechavi, G., Barzilai, A., Shamir, R., and **Shiloh, Y.** (2006) Parallel induction of ATM-dependent pro- and anti-apoptotic signals in murine lymphoid tissue in response to ionizing radiation. Oncogene 25:1584-1592.

134. Biton, S., Dar, I., Mittelman, L., Pereg, Y., Barzilai, A., and **Shiloh, Y.** (2006) Nuclear ATM Mediates the Cellular Response to DNA Double Strand Breaks in Human Neuron-Like Cells. J. Biol. Chem. 281:17482-17491.

135. Blank, M., Lerenthal, Y., Mittelman, L., and **Shiloh Y.** (2006) Condensin I recruitment and unscheduled chromatin condensation precede mitotic cell death in response to DNA damage". J. Cell Biol., 174:185-206.

136. Ziv, Y., Bielopolski, D., Galanty, Y., Lukas, C., Taya, Y., Schultz, D.C., Lukas, J., Bekker-Jensen, S., Bartek, J., and **Shiloh, Y**. (2006) Chromatin relaxation in response to DNA double strand breaks: a novel ATM- and KAP-1-dependent pathway. Nature Cell Biol., 8:870-876.

137. Dar, I., Biton, S., **Shiloh, Y.**, and Barzilai, A. (2006) Analysis of the Atm-mediated DNA damage response in murine cerebellar neurons. J. Neurosci. 26:7767-7774.

138. Pereg, Y., Lam, S., Teunisse, A., Biton, S., Meulmeester, E., Mittelman, L., Buscemi, G., Okamoto, K., Taya, Y., **Shiloh, Y.**, and Jochemsen, A.G. (2006) Differential Roles of ATM- and Chk2-Mediated Phosphorylations of Hdmx in Response to DNA Damage. Mol. Cell. Biol., 26:6819-6831.

139. Kirshner, J., Jobling, M.F., Pajares, M.-J., Ravani, S.A., Glick, A., Lavin, M.F., Kozlov, S., **Shiloh, Y.**, and Barcellos-Hoff, M.H. (2006) Inhibition of TGFβ1 signaling attenuates ATM activity in response to genotoxic stress. Cancer Res., 66:10861-10869.

140. Biton, S., Itsykson, P., Gropp, M., Pereg, Y., Mittelman, L., Johe, K., Reubinoff, B., and **Shiloh, Y.** (2007) ATM-mediated response to DNA double strand breaks in human neurons derived from stem cells. DNA Repair, 6:128-134.

141. Chen, B.P.C., Uematsu, N., Kobayashi, J., Lerenthal, Y., Krempler, A., Yajima, H., Lobrich, M., **Shiloh, Y.,** and Chen, D.J. (2007) ATM is essential for DNA-PKcs phosphorylations at T2609 cluster upon DNA double strand breaks. J. Biol. Chem., 282:6582-6587.

142. Alterman, N., Fattal-Valevski, A., Moyal, L., Crawford, T., Lederman, H.M., Ziv, Y., and **Shiloh, Y.** (2007) Ataxia-telangiectasia: mild neurological presentation despite null *ATM* mutation and severe cellular phenotype. Am. J. Med. Genet. 143:1827-1834.

143. Matsuoka, S., Ballif, B.A., Smogorzewska, A., McDonald, E.R., Hurov, K.E., Luo, J., Bakalarski, C.E., Zhou, J., Solimini, N., Lerenthal, Y., **Shiloh, Y**., Gygi, S.P. and Elledge, S.E. (2007) ATM and ATR substrate analysis reveals extensive protein networks responsive to DNA damage. Science, 316:1160-1166.

144. Assaf, Y., Galron, R., Shapira, I., Nitzan, A., Blumenfeld-Katzir, T., Solomon, A.S., Holdegreber, V., Wang, Zhao-Qi, **Shiloh, Y**., and Barzilai, A. (2008) MRI evidence of white matter damage in a mouse model of Nijmegen breakage syndrome. Exp. Neurol., 209:181-191.

145. Elkon, R., Linhart, C., Halperin, Y., **Shiloh, Y**., and Shamir, R. (2008) Functional genomic delineation of TLR-induced transcriptional networks. BMC Genomics, 8:394.

146. Elkon, R., Vesterman, R., Amit, N., Ulitzky, I., Zohar, I., Weisz, M., Mass, G., Orlev, N., Sternberg, G., Blekhman, R., Assa, J., **Shiloh, Y**., and Shamir, R. (2008) SPIKE – a database, visualization and analysis tool of cellular signaling pathways. BMC Bioinformatics, 9:110.

147. Goodarzi, A., Noon, A.T., Deckbar, D., Ziv, Y., **Shiloh, Y**., Löbrich, M., and Jeggo, P. A. (2008) ATM signalling facilitates repair of DNA double strand breaks associated with heterochromatin. Molecular Cell, 31:167-177.

148. Cheng, W.-H., Muftic, D., Muftuoglu, M., Dawut, L., Morris, C., Helleday, T., **Shiloh, Y.**, and Bohr, V.A. (2008) WRN is required for ATM activation and the S-phase checkpoint in response to interstrand crosslink-induced DNA double strand breaks. Mol. Biol. Cell., 19:3923-3933.

149. Shema, E., Tirosh, I., Aylon, Y., Huang, J., Ye, C., Moskovitch, N., Raver-Shapira, N., Minsky, N., Pirngruber, Y., Tarcic, G., Hublatrove, P., Moyal, L., Gana-weisz, M., **Shiloh, Y**., Yarden, Y., Johnson, S.A., Vojtsek, B., Berger, S.L., and Oren, M. (2008) The histone H2B-specific ubiquitin ligase RNF20/hBRE1 acts as a putative tumor suppressor through selective regulation of gene expression. Genes Dev., 22:2664-2676.

150. Kim, Y.-C., Gerlitz, G., Furusawa, T., Nussenzwig, A., **Shiloh, Y.**, and Bustin, M. (2009) The nucleosome-binding protein HMGN1 affects ATM activation by modulating its chromatin interactions prior and after DNA damage. Nature Cell Biol., 11:92-96.

151. Kirshner, M., Finkel, M., Nizan, A., Kanaar, R., **Shiloh, Y**, and Barzilai, A. (2009) Analysis of the relationships between ATM and the Rad54 proteins, players in homologous recombination repair. DNA Repair, 8:253-261.

152. Winter, D., Seidler, J., Ziv, Y., **Shiloh, Y.**, and Lehmann, W.D. (2009) Citrate boosts the performance of phosphopeptide analysis by UPLC-ESI-Ms/MS. J. Proteome Res., 8:418-424.

153. Baranes, K., Raz-Prag, D., Nitzan, A., Galron, R., Asjery-Padan, R., Rotenstreich, Y., Assaf, Y., **Shiloh, Y**., Wang, Z.-Q., Solomon, A.S., and Barzilai, A. (2009) Conditional inactivation of the NBS1 gene in the mouse central nervous system leads to neurodegeneration and disorganization of the visual system. Exp. Neurol. 218:24-32.

154. Das, B.B., Antony, S., Gupta, S., Dexheimer, T.S., Redon, C.E., Garfield, S**., Shiloh, Y**., and Pommier, Y. (2009) Optimal function of the DNA repair enzyme TDP1 requires its phosphorylation by ATM and/or DNA-PK. EMBO J. 28:3667-2680.

155. Winter, D., Seidler, J., Ziv-Lehrman, S., **Shiloh, Y**., and Lehmann, W.D. (2009) Simultaneous identification and quantification of proteins by differential (16)O/(18)O labeling and UPLC-MS/MS applied to mouse cerebellar phosphoproteome following irradiation. Anticancer Res. 29:4949-4958.

156. Ulitzky, I., Maron-Katz, A., Shavit, S., Sagir, D., Linhart, C., Elkon, R., Tamay, A., Sharan, R., **Shiloh, Y.,** and Shamir, R. (2010) Expander: From Expression Microarrays to Networks and Functions. Nature Protocols 5:303-322.

157. Barash, H., Gross, E., Edrei, Y., Ella, E., Cohen, I., Corchia, N., Ben-Moshe, T., Pappo, O., Pikarsky, E., Goldenberg, D., **Shiloh, Y.,** Galun, E., and Abramovitch, R. (2010) Accelerated carcinogenesis following liver regeneration is associated with chronic inflammation-induced double-strand DNA breaks. Proc. Natl. Acad. Sci. USA 107:2207-2217.

158. Salton, M., Lerenthal, Y., Wang, S.-Y., Chen, D.J., and **Shiloh, Y.** (2010) Involvement of Matrin 3 and SFPQ/NONO in the DNA damage response. Cell Cycle 9:1568-1576.

159. Bensimon, A., Schmidt, A., Ziv, Y., Elkon, R., Wang, S.-Y., Chen, D., Aebersold, R., and **Shiloh, Y**. (2010) ATM-dependent and independent dynamics of the nuclear phosphoproteome following DNA damage. Sci. Signal. 3(151) rs3.

160. Paz, A., Brownstein, Z., Ber, Y., Bialik, S., David, E., Sagir, D., Ulitsky, I., Elkon, R., Kimchi, A., Avraham, K., **Shiloh, Y**., and Shamir, R. (2011) SPIKE: A database of highly curated human signaling pathways. Nucleic Acids Res. 39:D793-799.

161. Kepkay, R., Attwood, K.M., Ziv, Y., **Shiloh, Y**., and Dellaire, G. (2011) KAP1 depletion increases PML nuclear body number in concert with ultrastructural changes in chromatin. Cell Cycle 10:308-322.

162. Galron, N., Gruber, R., Lifshitz, V., Lu, H., Kirshner, M., Ziv, N., Wang, Z.-Q., **Shiloh, Y**., Barzilai, A., and Frenkel, D. (2011) Astrocyte dysfunction associated with cerebellar attrition in a Nijmegen breakage syndrome animal model. J. Mol. Neurosci., 45:202-211.

163. Dar, I., Yosha, G., Elfassy, R., Galron, R., Wang, Z.-Q., **Shiloh, Y.,** and Barzilai, A. (2011) Investigation of the functional link between ATM and NBS1 in the DNA damage response in the mouse cerebellum. J. Biol. Chem. 286:15361-15376.

164. Moyal, L., Gana-Weisz, M., Lerenthal, Y., Mass, G., So, S., Wang, S.-Y., Eppink, B., Chung, Y.-M., Shalev, G., Shema, E., Shkedy, D., Smorodinsky, N.I., van-Vliet, N., Kuster, B., Mann, M., Ciechanover, A., Dahm-Daphi, J., Kanaar, R., Hu, M.C-T., Chen, D.J., Oren, M., and **Shiloh, Y.** (2011) Requirement of ATM-dependent monoubiquitylation of histone H2B for timely repair of DNA double strand break. Mol. Cell, 41:529-542. **Featured article.**

165. Segal-Raz, H., Mass, G., Ziv-Lehrman, S., Wang, S.-Y., Strom, C., Helleday, T., Chen, D.J., and **Shiloh, Y.** (2011) ATM-mediated phosphorylation of polynucleotide kinase is required for effective DNA double-strand break repair. EMBO Reports, 12:713-719.

166. Raz-Prag, D., Galron, R., Segev-Amzaleg, N., Barzilai, A., **Shiloh, Y**., and Frenkel, D. (2011) A role for vascular deficiency in retinal pathology in a mouse model of ataxia-telangiectasia. Am. J. Pathol., 179:1533-1541.

167. Rashi-Elkeles, S., Elkon, R., Shavit, S., Lerenthal, Y., Linhart, C., Kupershtein, A., Amariglio, N., Rechavi, G., Shamir, R., and **Shiloh, Y**. (2011) Transcriptional modulation induced by ionizing radiation: p53 remains a central player. Mol. Oncol., 5:336-348.

168. Salton-Morgenstern, M., Elkon, R., Borodina, T., Davydov, A., Yaspo, M.-L., Halperin, E., and **Shiloh, Y.** (2011) Matrin 3 binds and stabilizes mRNA. PLoS ONE, 6(8):e23882.

169. Kirshner, M., Galron, R., Frenkel, D., Mandelbaum, G., **Shiloh, Y**., Wang, Z.-Q., and Barzilai, A. (2011) Malfunctioning DNA damage response (DDR) leads to the degeneration of nigro-straiatal pathway in mouse brain. J. Mol. Neurosci., 46:554-68.

170. Levy-Barda, A., Lerenthal, Y., Davis, A.J., Chung, Y.M., Essers, J., Shao, Z., van Vliet, N., Chen, D.J., Hu, M.C-T., Kanaar, R., Ziv, Y., and **Shiloh, Y**. (2011) Involvement of the nuclear proteasome activator PA28γ in the cellular response to DNA double-strand breaks. Cell Cycle, 10:4300-4310.

171. Tzur-Gilat, A., Ziv, Y., Dusart, I., Mittelman, L., Barzilai, A., and **Shiloh, Y**. (2013) Studying the cerebellar DNA damage response in the tissue culture dish. Mech. Age. Dev., 134:496-505.

172. Rashi-Elkeles, S. Warnatz, H.-J., Elkon, R., Kupershtein, A., Chobod, Y., Paz, A., Amstislavskiy, V., Sultan, M., Safer, H., Nietfeld, W., Lehrach, H., Shamir, R., Yaspo, M.-L., and **Shiloh, Y.** (2014) Parallel profiling of cellular Responses to ionizing radiation at the levels of the transcriptome, cistrome and epigenome. Sci. Signal., 7:rs3.

173. Meir, M., Galanty, Y., Kashani, L., Blank, M., Khosravi, R., Fernández-Ávila, M.J., Cruz-Garcia, A., Star, A., Shochat, L., Thomas, Y., Garrett, L.J., Chamovitz, D.A., Bodine, D.M., Kurz, T., Huertas, P., Ziv, Y., and **Shiloh, Y.** (2015) The COP9 signalosome is vital for timely repair of DNA double-strand breaks. Nucleic Acids Res., 43: 4517-4530.

174. Gavish-Izakson, M., Bhavana, VB, Elkon, R., Prados-Carvajal, R., Barnabas, G.D., Pineiro Ugalde, A., Agami, A., Geiger, T., Huertas, P., Ziv, Y., and **Shiloh, Y.** (2018) Nuclear poly(A)-binding protein 1 is an ATM target and essential for DNA double-strand break repair. Nucleic Acids Res., 46:730-747.

175. Baranes-Bachar, K., Levy-Barda, A., Oehler, J., Reid, D.A., Soria-Bretones, I., Vos, T.C., Chung, D., Park, Y., Liu, C., Yoon, J.-B., Li, W., Dellaire, G., Misteli, T., Huertas, P., Rothenberg, E., Ramadan, K., Ziv, Y., and **Shiloh, Y.** (2018) The ubiquitin E3/E4 ligase, UBE4A, fine-tunes protein ubiquitylation and accumulation at sites of DNA damage facilitating double-strand break repair. Mol. Cell, 69:866-878.

176. Tal, E., Alfo, M., Zha, S., Barzilai, A., De Zeeuw, C., Ziv, Y., **Shiloh, Y.** (2018) Inactive Atm abrogates DSB repair in mouse cerebellum more than does Atm loss, without causing a neurological phenotype. DNA Repair, 72:10-17.

177. Jachimowicz, R.D., Belleggia, F., Isensee, J., Velpula, B.B., Georgens, J., Bustos, M.A., Doll, M.A., Shenoy, A., Checa-Rodriguez, C., Wiederstain, J.L., Baranes-Bachar, K., Bartenhagen, C., Hertwig, F., Teper, N., Nishi, T., Schmitt, A., Distelmaier, F., Lüdecke, H.-J., Albreach, B., Krüger, M., Schumacher, B., Geiger, T., Hoon D.S.B., Huertas, P., Fischer, M., Hucho, T., Peifer, M., Ziv, Y., Reinhardt, H.C., Wieczorek, D., and **Shiloh, Y.** (2019) UBQLN4 represses homologous recombination and is overexpressed in aggressive tumors. Cell, 176:505-519. (**Highlighted in Cancer Discovery).**

178. Hait, T.A., Maron-Katz, A, Sagir, D., Amar, D., Ulitzky, I., Linhart, C., Sharan, R., **Shiloh, Y**., Elkon, R., Shamir, R. (2019) The EXPANDER integrated platform for transcriptome analysis. J. Mol. Biol., 431:2398-2406.

179. Antoniou, N., Lagopadi, N., Balourdas, D.I., Nikolaou, M., Papalampros, A., Vasileiou, P.V.S., Myrianthopoulos, V., Kostinas, A., **Shiloh, Y.,** Liontos, M., and Gorgoulis, V.G., (2019) The role of E3, E4 ubiquitin ligase (UBE4B) in human pathologies. Cancers, 12(1). pii: E62. doi: 10.3390/cancers12010062.

180. Schlam-Babayov, S., Bensimon, A., Harel, M., Geiger, T., Aebersold, R., Ziv, Y., and **Shiloh, Y**. (2021) Phosphoproteomics reveals novel modes of function and inter-relationships among PIKKs in response to genotoxic stress. The EMBO Journal, 40(2):e104400. doi: 10.15252/embj.2020104400.

181. Murakami, T., Shoji, Y., Nishi, T., Chang, S.-C., Jachimowicz, R., Hoshimoto, S., Ono, S., **Shiloh, Y.,** Takeuchi, H., Kitagawa, Y., Hoon, D.S.B., and Buston, M.A. (2021) MRE11A regulation by UBQLN4 leads to cisplatin-resistance in esophageal squamous cell carcinoma patients. Molecular Oncology, 15:1069-1087.

182. Shoji, Y., Yokoe, T., Kobayashi, Y., Murakami, T., Bostick, P.J., **Shiloh, Y**., Hoon, D.S.B., and Bustos, M. (2022) UBQLN4 promotes proteasomal degradation of STING during cisplatin-induced DNA damage in triple-negative breast cancer. Clinical and Translational Medicine, 12:e985. doi: 10.1002/ctm2.985.

183. Kobayashi, Y., Bustos, M.A., Shoji, Y., Jachimowicz, R.D., **Shiloh, Y**., and Hoon, D.S.B. (2022) Genomic amplification of *UBQLN4* is a prognostic and treatment resistance factor. Cells,

11(20):3311. doi: 10.3390/cells11203311.

184. Elkoshi, N., Parikh, S., Malcov-Brog, H., Parikh, R., Manich, P., Netti, F., Maliah, A., Frand, J., Perluk, T., Sarfstein, R., Brenner, R., Gonen, P., Dror, I., Levi, H., Hameiri, O., Ast, G., Nizri, E., Ziv, Y., Elkon, R., Khaled, M., Ebenstein, Y., **Shiloh, Y**., and Levy, C. (2023) ATM signaling delays skin pigmentation upon UV exposure by mediating MITF function towards DNA repair mode. J. Invest. Dermatol.,143:2494-2506. doi: 10.1016/j.jid.2023.03.1686.

185. Haj, M., Levon, A., Frey, Y., Hourvitz, N., Campisi, J., Tzfati, Y., Elkon, R., Ziv, Y., and **Shiloh, Y.** (2023) Accelerated replicative senescence of ataxia-telangiectasia skin fibroblasts is retained at physiologic oxygen levels, with unique and common transcriptional patterns. Aging Cell, e13869. doi: 10.1111/acel.13869.

186. Elitzur, S., Shiloh, R., Loeffen, JLC, Pastorczak, A., Takagi, M., Bomken, S., Baruche, A., Lehrnbecher, T., Tasian, KT., Abla, O., Arad-Cohen, N., Astigarraga, I., Ben-Harosh, M., Bodmer, N., Brozou, T., Ceppi., F., Chugaeva, L., Dalla Pozza, L., Ducassou, S., Farah, R., Gibson, A., Hasle, H., Jacoby, E., Jazbec, J., Kolenova, A., Lazic, J., Nigro, L., Mahlaoui, N., Miller, L., Papadakis, L., Pecheux, L., Pillon, M., Sarouk, Y., Stary, J., Stiakaki, E., Strullu, M., Tran, T.T., Ussowicz, M., Verdu-Amoros, J., Wakulinska, A., Zawitkowska, J., Stoppa-Lyonnet, D., Taylor, M., **Shiloh, Y**., Izraeli, S., Minard-Colin, V., Schmiegelow, K., Nirel, R., Attarbaschi, A., and Borkhardt, A. (2024) *ATM* Germline pathogenic variants affect treatment outcomes in children with ataxia-telangiectasia and hematological malignancies. Blood doi: 10.1182/blood.2024024283.

187. Naor, S., Ziv, Y., **Shiloh, Y**. (2024) Visualizing the DNA Damage Response in Purkinje Cells Using Cerebellar Organotypic Cultures. J. Vis. Exp. (214), e67167, doi:10.3791/67167.

188. Frey, Y., Haj, M., Ziv, Y., Elkon, R., and **Shiloh, Y**. (2025) Broad repression of DNA repair genes in senescent cells identified by integration of transcriptomic data. Nucleic Acids Research doi: 10.1093/nar/gkae1257/7935007.

189. Haj, M., Frey, Y., Levon, A., Maliah, A., Ben-Yishai, T., Slutsky, R., Smoom, R., Tzfati, Y., Ben-David, U., Levy, C., Elkon, R., Ziv, Y., and **Shiloh, Y**. (2025) The cGAS-STING, p38 MAPK and p53 pathways link genome instability to accelerated cellular senescence in ATM-deficient murine lung fibroblasts. Proc. Natl. Acad. Sci. USA doi: 10.1073/pnas.2419196122 **(PNAS Inaugural Article).**

190. Garcia, D.D., Giri, S., Chattaerjee, R., Nieves, A.S., Rios, M., Lindzen, M., Sekar, A., Gupta, N., Schwarzmüller, L., Tarade, N., Zhu, R., Karatekin, F., Roncato, R., Eyal-Lubling, Y., Keidar, T., Nof, Y., Nataraj, B., Alon, R., Seger, R., Wagner, B., Nair, N., Sanghiv, N., Pikarsky, E., Blandino, G., Wiemann, S., Lev, S., Rueda, O., Caldas, C., Ruppin, E., **Shiloh, Y**., Domany, E., Dahlhoff, M., and Yarden, Y. (2025) By inducing ROS and autophagy, the novel OVOL1-C1ORF116-TRX/GCLC pathway might control escape from cancer dormancy. Science Signaling,18(883):eado3473; doi: 10.1126/scisignal.ado3473.

191. Frey, Y., Goehring, L., Haj, M., Rona, G., Fijen, C., Pagano, M., Huang, T.T., Rothenberg, E., Ziv, Y., and **Shiloh, Y**. (2025) ZC3H4 safeguards genome integrity by preventing transcription-replication conflicts at non-coding RNA loci. Science Advances 11(25):eadt8346. doi: 10.1126/sciadv.adt8346.

In revision:

Chetrit, A., Forer, R., Ziv, Y., Sadetzki, S., and Shiloh, Y. (2026) Increased chronic morbidity in ataxia-telangiectasia carriers identified in a population-based study. Proc. Natl. Acad. Sci. USA.

Submitted:

Hoon, D., Takamatsu, D., Chong, K., Jimenez, J., Ramos, R.I., Krasne, D., Twardowski, P., Linehan, J., **Shiloh, Y.,** and Wilson, T. (2026) WNK1-UBQLN4-DDR Axis: an oncogenic hub driving aggressiveness and therapeutic resistance in urological malignancies.

Review Articles and Commentaries

1. **Shiloh, Y.** (1995) Ataxia-telangiectasia: Closer to unraveling the mystery. Europ. J. Hum. Genet. 3:116-138.

2. Lavin, M.F., Khanna, K.K., Beamish, H., Spring, K., Watters, D. and **Shiloh, Y.** (1995) Relationship of the ataxia-telangiectasia gene, ATM, to phosphatidylinositol 3-kinase. Trends Biochem. Sci., 20:382-383.

3. Jorgensen, T.J. and **Shiloh, Y.** (1996) The *ATM* gene and the radiobiology of ataxia-telangiectasia. Int. J. Radiat. Biol., 69:527-537.

4. **Shiloh, Y.**, and Rotman, G. (1996) Ataxia-telangiectasia and the ATM gene: Linking neurodegeneration, immunodeficiency, and cancer to cell cycle checkpoints. J. Clin. Immunol., 16:254-260.

5. Lavin, M.F., and **Shiloh, Y.** (1996) Ataxia-telangiectasia: A multifaceted genetic disorder associated with defective signal transduction. Curr. Opin. Immunol., 8:459-464.

6. Lavin, M.F., and **Shiloh, Y.** (1997) The genetic defect in ataxia- telangiectasia. Ann. Rev. Immunol., 15:177-202.

7. Shiloh, Y. (1997) Ataxia-telangiectasia and the ATM gene. Encyclopedia of Human Biology, 2nd Edition, Academic Press, NY, vol. 1, pp. 545-554.

8. Rotman, G., and **Shiloh, Y.** (1997) The ATM gene and protein: Possible roles in genome surveillance, checkpoint controls and cellular Defense against oxidative stress. Cancer Surveys, 29:285-304.

9. **Shiloh, Y.** (1997) Ataxia-telangiectasia and the Nijmegen breakage syndrome: related disorders but genes apart. Ann. Rev. Genet., 31:635-662.

10. Rotman, G., and **Shiloh, Y.** (1997) Ataxia-telangiectasia and ATM: Possible involvement in cellular responses to oxidative damage and stress. BioEssays, 19:911-917.

11. **Shiloh, Y.** (1998) Ataxia-telangiectasia, ATM and genome stability. Maintaining a delicate balance. Biochim. Biophys. Acta Reviews on Cancer, 1378:R11-R18.

12. Rotman, G., and **Shiloh, Y.** (1998) ATM: from gene to function. Hum. Mol. Genet., 7:1555-1563.

13. **Shiloh, Y.** (1999) Ataxia-telangiectasia: from disease to gene, protein and physiological function (translated to Japanese). Experimental Medicine (Japan), 17:10-15.

14. Rotman, G., and **Shiloh, Y.** (1999) ATM: a mediator of multiple responses to genotoxic stress Oncogene, 18:6135-6144.

15. Halazonetis, T.D. and **Shiloh, Y.** (1999) Many faces of ATM: Eighth International Workshop on Ataxia-Telangiectasia Biochim. Biophys. Acta (Reviews on Cancer), 1424:R45-R55.

16. **Shiloh, Y.** (2000) ATM: Sounding the double-strand break alarm. Cold Spring Harbor Symposia on Quantitative Biology, "Biological Responses to DNA Damage", Vol. 65, pp. 527-533.

17. **Shiloh, Y.** (2001) ATM and ATR: Networking cellular responses to DNA damage. Curr. Opin. Genet. Dev., 11:71-77.

18. **Shiloh, Y.** and Kastan, M.B. (2001) ATM: Genomic stability, neuronal development and cancer cross paths. Adv. Cancer Res., 83:210-253.

19. **Shiloh, Y.** (2001) ATM: Expanding roles in the DNA damage response and cellular homeostasis. Biochem. Soc. Transact. 29:661-666.

20. Barzilai, A., Rotman, G., and **Shiloh, Y.** (2002) ATM deficiency and oxidative stress; a new dimension of defective response to DNA damage. DNA Repair, 1:3-26.

21. **Shiloh, Y.** (2003) ATM and related protein kinases: safeguarding genome integrity. Nature Reviews Cancer , 3:155-168. **(Noted by *ISI* as "Hot paper of the Month in Clinical Medicine, July 2004).**

22. **Shiloh, Y**. (2003) ATM: Ready, Set, Go. Cell Cycle 2:116-117.

23. **Shiloh, Y.,** and Lehmann, A.R. (2004) Maintaining integrity. Nature Cell Biology, 6:923-928.

24. Meulmeester, E., Pereg, Y., **Shiloh, Y**., and Jochemsen, A.G. (2005) ATM-mediated phosphorylations inhibit Mdmx/Mdm2 stabilization by HAUSP in favor of p53 activation. Cell Cycle, 4:1166-1170.

25. **Shiloh, Y.** (2006) The ATM-mediated DNA damage response: taking shape. Trends Biochem. Sci., 31:402-410.

26. Blank, M. and **Shiloh, Y**. (2007) Programs for cell death: Apoptosis is only one way to go. Cell Cycle, 6:686-695.

27. Barzilai, A., Biton, S., and **Shiloh, Y.** (2008) The role of the DNA damage response in neuronal development, organization and maintenance. DNA Repair, 7:1010-1027.

28. Biton, S., Barzilai, A., and **Shiloh, Y.** (2008) The neurological phenotype of ataxia-telangiectasia: solving a persistent puzzle. DNA Repair, 7:1028-1038.

29. **Shiloh, Y.** (2009) FBXO31: a new player in the ever-expanding DNA damage orchestra. Sci. Signal., 2(96):pe73.

30. Jochemsen, A.A., and **Shiloh, Y**. (2010) USP10: Friend or foe? Cell, 140:308-310.

31. Sander, M., Begley, T.J., Desaintes, C., Gavin, A.-C., Pelroy, R., Pothof, J., **Shiloh, Y**., van Gent, D., van Houten, B., Yaffe, M., and Mullenders, L. (2010) 3rd US-EU Workshop on Systems Level Understanding of DNA Damage Responses. Mutat. Res., 692:53-60.

32. Bensimon, A., Aebersold, R., and **Shiloh, Y.** (2011) Beyond ATM: the protein kinase landscape of the DNA damage response. FEBS Letters, 585:1625-1639.

33. **Shiloh, Y**., Shema, E., Moyal, L., and Oren, M., (2011) RNF20-RNF40: a ubiquitin-driven link between gene expression and the DNA damage response. FEBS Letters, 585:2795-2802.

34. **Shiloh, Y**. and Ziv, Y. (2012) The ATM protein: the importance of being active. J. Cell Biol., 193:273-275.

35. Rasmussen, L.J., **Shiloh, Y**., Bergersen, L.H., Sander, M., Bohr, V.A., and Tønjum, T. (2013) DNA damage response, bioenergetics, and neurological disease: the challenge of maintaining brain health in an aging human population. Mech. Ageing Dev., 134:427-33.

36. **Shiloh, Y**., and Ziv, Y. (2013) The ATM protein: regulating the DNA damage response, and more. Nature Rev. Mol. Cell Biol., 14:197-210.

37. **Shiloh, Y.** (2014) ATM: expanding roles as a chief guardian of genome stability. Exp. Cell Res., 329:154-161.

38. Ribezzo, F., **Shiloh, Y**., and Schumacher, B. (2016) Systemic DNA damage responses in aging and diseases. Seminars in Cancer Biology, 37-38:26-35.

39. Tal, E. and **Shiloh, Y.** (2017) Monitoring the ATM-mediated DNA damage response in the cerebellum using organotypic cultures. Methods in Molecular Biology, 1599:419-430.

40. Barzilai, A., Schumacher, B., and **Shiloh, Y.** (2017) Genome instability: linking ageing and brain degeneration. Mechanisms of Ageing and Development ,161(Pt A):4-18.

41. **Shiloh, Y**., and Lederman, H. (2017) Ataxia-telangiectasia (A-T): an emerging dimension of premature ageing. Ageing Research Reviews, 33:76-88.

42. **Shiloh, Y.** (2017) The ATM Protein. In: Encyclopedia of Cancer, 4th Edition (M. Schwab, Ed.) Springer Verlag, Berlin Heidelberg, Vol. 1, pp. 395-397.

43. **Shiloh, Y.** (2020) The cerebellar degeneration in ataxia-telangiectasia: a case for genome instability. DNA Repair, doi: 10.1016/j.dnarep.2020.102950.

44. **Shiloh, Y.** (2020) The serendipitous dawn of DNA repair. Nature Reviews Molecular Cell Biology ('Journal Club') doi: 10.1038/s41580-020-00286-x.

45. Schlam-Babayov, S., Ziv, Y., and **Shiloh, Y.** (2021) It takes three to the DNA damage response tango. Molecular and Cellular Oncology, 8, Article1881395.

46. Aguado, J., Gomez-Inclan, C., Leeson, H.C., Lavin, M.F., **Shiloh, Y**., and Wolvetang, E.J., (2022) The hallmarks of aging in ataxia-telangiectasia. Aging Research Reviews, 79:101653. doi: 10.1016/j.arr.2022.101653.

47. Lavin, M., Taylor, A.M.R., and **Shiloh, Y**. (2026) Richard Gatti: From concert pianist to bone marrow transplants and the DNA damage response syndrome, ataxia-telangiectasia. DNA Repair (obituary; in press)

In preparation:

Frey, Y., Ziv, Y., and **Shiloh, Y**. (2026) From barrier to burden: cellular senescence as a short-term safeguard that becomes a genomic ticking bomb.

Meeting Reports

1. **Shiloh, Y** (1998) Ataxia-telangiectasia, ATM and genomic stability: maintaining a delicate balance. Two international workshops on ataxia-telangiectasia, related disorders and the ATM protein. Biochim. Biophys. Acta, 1378:R11-R18.

2. Schmauck-Medina, T., Molière, A., Lautrup, S., Zhang, J., Chlopicki, S., Borland Madsen, H., Søndenbro, S., Casper, C., Mansell, E., Vestergaad, M.B., L, Z., **Shiloh, Y**., Opresko, P., Egly, J.-M., Kirkwood, T., Verdin, E., Bohr, V.A., Cox, L.S., Stevnsner, T.,Rasmussen, L.J., and Fang, E.F. (2022) New Hallmarks of Aging: a 2022 Copenhagen Ageing meeting summary. Aging, doi: 10.18632/aging.204248.

Book Chapters

1. **Shiloh, Y.**, Cohen, M. M. and Becker, Y. (1981) Ataxia-telangiectasia: studies on DNA repair synthesis. In: Chromosome Damage and Repair (K. Kleppe and E. Seeberg, eds.) Plenum Press, New York, pp. 361-366.

2. **Shiloh, Y.** and Becker, Y. (1981) Ataxia-telangiectasia lymphoblastoid lines resemble non A-T cell lines in their ability to remove MNNG-induced methylation products. In: Ataxia-Telangiectasia: A Cellular and Molecular Link Between Cancer, Neuropathology and Immune Deficiency (B.A. Bridges and D.G. Harnden, eds.) John Wiley & Sons, London, pp. 177-187.

3. Becker, Y., Shaham, M., **Shiloh, Y.** and Voss, R. (1982) Chromosome breakage and sensitivity to DNA breaking agents in ataxia-telangiectasia and their possible association with predisposition to cancer. In: Biochemical and Biophysical Markers of Neoplastic Transformation (P. Chandra, ed.), Plenum Press, New York, pp. 237-244.

4. Becker, Y., Shaham, M., Tabor, E. and **Shiloh, Y.** (1984) Ataxia-telangiectasia - a human genetic disorder predisposing to cancer. In: The Role of Viruses in Human Cancer (G. Giraldo and E. Beth, eds.), Vol II, Elsevier/North Holland Publishers, pp. 189-209.

5. **Shiloh, Y.**, Tabor, E. and Becker, Y. (1985) In-vitro phenotype of A-T fibroblast strains: clues to the nature of the "A-T DNA lesion" and the molecular defect in A-T. In: Ataxia-Telangiectasia: Genetics, Neuropathology and Immunology of a Degenerative Disease of Childhood (R.A. Gatti and M. Swift, eds.), Alan R. Liss, Inc., New York, pp. 111-121.

6. Latt, S.A., **Shiloh, Y.**, Sakai, K., Brodeur, G. Donlon, T., Korf, B., Shipley, J., Bruns, G., Kohl, N., Alt, F. and Seeger, R. (1986) Novel DNA rearrangement phenomena associated with DNA amplification in human neuroblastomas and neuroblastoma cell lines. In: Genetic Toxicology of Environmental Chemicals (C. Ramel and B. Lambert, eds.), Alan R. Liss, Inc., New-York, pp. 601-612.

7. Latt, S.A., Lalande, M., Donlon, T., Wyman, A., Rose, E., **Shiloh, Y.**, Korf, B., Sakai, K., Harris, P., Bruns, G. and Kaplan, L. (1987) DNA-based detection of chromosome deletion and amplification: diagnostic and mechanistic significance. Cold Spring Harbor Symposia on Quantitative Biology, 51:299-307.

8. **Shiloh, Y.**, Rose, E., Korf, B., Shipley, J., Sakai, K., Brodeur, G., Seeger, R.C. and Latt, S.A. (1987) Analysis of DNA amplification in tumor cells: the neuroblastoma model. In: Accomplishments in Oncology: The Role of DNA Amplification in Carcinogenesis (H. zur Hausen and J. Schlehofer, eds.), J.B. Lippincott Co., Philadelphia, pp. 186-197.

9. Latt, S.A., **Shiloh, Y.**, Sakai, K., Rose, E., Brodeur, G., Korf, B., Heartline, M., Kang, J., Harris, P., Bruns, G. and Seeger, R. (1988) DNA rearrangement, relocation and amplification in neuroblastoma cell lines and primary tumors. In: Cellular and Molecular Biology of Tumors and Potential Clinical Applications (ICN-UCLA Symposia on Cancer Biology, J. Minna and M. Kuehl, eds.), Alan R. Liss, Inc., New York, pp. 167-178.

10. **Shiloh, Y.**, Avigad, S., Kleiman, S., Weinstein, M., Schwartz, G., Woo, S.L.C., and Cohen, B.E. (1992) Molecular analysis of hyperphenylalaninemia in Israel: a study of Jewish genetic diversity. In: Genetic Diversity Among Jews: Diseases and Markers at the DNA Level (B. Bonne-Tamir and A. Adam, eds.), Oxford University Press, pp. 237-247.

11. Ziv, Y., Danieli, T., Rotman, G., Sartiel, A., Bar-Shira, A., Swirski, R., Schimke, R.T., Eddy, R.L., Shows, T.B. and **Shiloh, Y.** (1993) Complementation of the cellular A-T phenotype by gene transfer. In: Ataxia-Telangiectasia (R.A. Gatti and R.B. Painter, eds.), NATO ASI Series, Series H: Cell Biology, Vol. 77. Springer-Verlag Publishers, New York, pp. 65-74.

12. Rotman, G., and **Shiloh, Y.** (1996) Ataxia-telangiectasia: Linking immunodeficiency, neurodegeneration and cancer to defects in signal transduction and cell cycle checkpoints. Progress in Immunodeficiency VI (A. Fasth and J. Bjorkander, eds.), Elsevier Sciences, Amsterdam, pp. 41-52.

13. **Shiloh, Y**., Bar-Shira, A., Galanty, Y., and Ziv, Y. (1998) Cloning and expression of large mammalian cDNAs: Lessons from ATM. In: Genetic Engineering, Principles and Methods, (J. Setlow, ed.), Vol. 20, Plenum Press, N.Y., pp. 239-248.

14. Lavin, M.F., and **Shiloh, Y.** (1999; 2002) Ataxia-telangiectasia. In: Primary Immunodeficiency Diseases, A Molecular and Genetic Approach (H.D. Ochs, C.I.E. Smith and J. Puck, eds.) Oxford University Press, Oxford, pp. 306-323.

15. Rathbun, G.A., Ziv, Y., Lai, J.H., Hill, D., Abraham, R.H., **Shiloh, Y.**, and Cantley, L.C. (1999) ATM and lymphoid malignancies: use of oriented peptide libraries to identify novel substrates of ATM critical in downstream signaling pathways. In: Mechanisms of B Cell Neoplasia (F. Melchers and M. Potter, eds.), Curr. Top. Microbiol. Immunol., 246: 267-273, Springer, Berlin, pp. 267-273.

16. Rotman, G. and **Shiloh, Y.** (2000) ATM: At the crossroads of DNA damage response, cell cycle control, genome stability and cancer. In: DNA Alterations in Cancer: Genetic and Epigenetic Changes, (M. Erlich, ed.), Eaton Publishing, Natick, MA, pp. 227-240.

17. Ziv, Y., Banin, S., Lim, D.-S., Kastan, M.B., and **Shiloh, Y.** (2000) Expression and assay of recombinant ATM kinase. In: Stress Responses: Methods and Protocols, (S. Keyse, ed.), Methods Mol. Biol., 99:99-108, Humana Press, N.J.

18. **Shiloh, Y.** (2002) ATM: From phenotype to functional genomics - and back. In: The Human Genome: Biology and Medicine (A. Rosenthal and L. Vakalopoulou, eds.) Ernst Schering Research Foundation Workshop 36, Springer, Berlin, pp. 51-70.

19. Crawford, T.O. and **Shiloh, Y.** (2006) Ataxia-Telangiectasia. In: Spinocerebellar Degenerations: The Ataxias and Spastic Paraplegias. (A. Brice and S-M. Pulst, eds.) Elsevier, Amsterdam, pp. 724-739.

20. Elkon, R., Rashi-Elkeles, S., Shamir, R., and **Shiloh, Y**. (2006) Transcriptional responses to DNA Damage: Systems-level delineation by functional genomics. In: Microarrays and Transcription Networks. (Shannon, M. F. and Rao, S., eds.) Landes Bioscience (Georgetown), pp. 98-113.

21. **Shiloh, Y.** (2010) The ATM-mediated DNA damage response. In: Molecular Oncology (Gelmann, I., Sawyers, G.L. and Rauscher, F., eds.) Cambridge University Press, pp. 297-309.