Publications Stemming From

The Progeria Research Foundation Cell and Tissue Bank

The Progeria Research Foundation Cell and Tissue Bank has contributed to the following medical publications, categorized by cell line for researcher convenience:

HGADFN001

Anti-hsa-miR-59 alleviates premature senescence associated with Hutchinson-Gilford progeria syndrome in mice

Hu Q, Zhang N, Sui T, et al. [published online ahead of print, 2022 Nov 16]. *EMBO J*. 2022;e110937. doi:10.15252/embj.2022110937

Age-dependent loss of MMP-3 in Hutchinson-Gilford progeria syndrome.
Harten IA, Zahr RS, Lemire JM, Machan JT, Moses MA, Doiron RJ, Curatolo AS, Rothman FG, Wight TN, Toole BP, Gordon LB. *J Gerontol A Biol Sci Med Sci*. 2011 Nov;66(11):1201-7.

The mutant form of lamin A that causes Hutchinson-Gilford progeria is a biomarker of cellular aging in human skin.
McClintock D, Ratner D, Lokuge M, Owens DM, Gordon LB, Collins FS, Djabali K. *PLoS One*. 2007 Dec 5;2(12):e1269.

Hutchinson-Gilford progeria mutant lamin A primarily targets human vascular cells as detected by an anti-Lamin A G608G antibody.
McClintock D, Gordon LB, Djabali K. *Proc Natl Acad Sci U S A.* 2006 Feb 14;103(7):2154-9.

Aggrecan expression is substantially and abnormally upregulated in Hutchinson-Gilford Progeria Syndrome dermal fibroblasts.
Lemire JM, Patis C, Gordon LB, Sandy JD, Toole BP, Weiss AS. *Mech Ageing Dev.* 2006 Aug;127(8):660-9.

Rescue of heterochromatin organization in Hutchinson-Gilford progeria by drug treatment.
Columbaro M, Capanni C, Mattioli E, Novelli G, Parnaik VK, Squarzoni S, Maraldi NM, Lattanzi G. *Cell Mol Life Sci.* 2005 Nov;62(22):2669-78.

Recurrent de novo point mutations in lamin A cause Hutchinson-Gilford progeria syndrome.
Eriksson M, Brown WT, Gordon LB, Glynn MW, Singer J, Scott L, Erdos MR, Robbins CM, Moses TY, Berglund P, Dutra A, Pak E, Durkin S, Csoka AB, Boehnke M, Glover TW, Collins FS. *Nature.* 2003 May 15;423(6937):293-8.

HGADFN003

Ghrelin delays premature aging in Hutchinson-Gilford progeria syndrome
Ferreira-Marques M, Carvalho A, Franco AC, et al. Ghrelin delays premature aging in Hutchinson-Gilford progeria syndrome [published online ahead of print, 2023 Oct 19]. *Aging Cell*. 2023;e13983. doi:10.1111/acel.13983

Impact of Combined Baricitinib and FTI Treatment on Adipogenesis in Hutchinson-Gilford Progeria Syndrome and Other Lipodystrophic Laminopathies
Hartinger R, Lederer EM, Schena E, Lattanzi G, Djabali K. Cells. 2023;12(10):1350. Published 2023 May 9. doi:10.3390/cells12101350

Unique progerin C-terminal peptide ameliorates Hutchinson-Gilford progeria syndrome phenotype by rescuing BUBR1.
Zhang N, Hu Q, Sui T, Fu L, Zhang X, Wang Y, Zhu X, Huang B, Lu J, Li Z, Zhang Y. Nat Aging. 2023 Feb;3(2):185-201. doi: 10.1038/s43587-023-00361-w. Epub 2023 Feb 2. Erratum in: Nat Aging. 2023 May 2;: PMID: 37118121; PMCID: PMC10154249.

Establishment and Characterization of hTERT Immortalized Hutchinson-Gilford Progeria Fibroblast Cell Lines
Lin H, Mensch J, Haschke M, et al. Published 2022 Sep 6. doi:10.3390/cells11182784

Impact of MnTBAP and Baricitinib Treatment on Hutchinson-Gilford Progeria Fibroblasts
Vehns E, at R, Djabali K. Pharmaceuticals (Basel). 2022;15(8):945. Published 2022 Jul 29. doi:10.3390/ph15080945

SerpinE1 drives a cell-autonomous pathogenic signaling in Hutchinson-Gilford progeria syndrome
Catarinella G, Nicoletti C, Bracaglia A, et al. Cell Death Dis. 2022;13(8):737. Published 2022 Aug 26. doi:10.1038/s41419-022-05168-y

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Nucleoplasmic lamins define growth-regulating functions of lamina-associated polypeptide 2α in progeria cells.
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Lemire JM, Patis C, Gordon LB, Sandy JD, Toole BP, Weiss AS. *Mech Ageing Dev.* 2006 Aug;127(8):660-9.

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Columbaro M, Capanni C, Mattioli E, Novelli G, Parnaik VK, Squarzoni S, Maraldi NM, Lattanzi G. *Cell Mol Life Sci.* 2005 Nov;62(22):2669-78.

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Accumulation of mutant lamin A causes progressive changes in nuclear architecture in Hutchinson-Gilford progeria syndrome.
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Recurrent de novo point mutations in lamin A cause Hutchinson-Gilford progeria syndrome.
Eriksson M, Brown WT, Gordon LB, Glynn MW, Singer J, Scott L, Erdos MR, Robbins CM, Moses TY, Berglund P, Dutra A, Pak E, Durkin S, Csoka AB, Boehnke M, Glover TW, Collins FS. *Nature.* 2003 May 15;423(6937):293-8.

HGADFN004

Incomplete processing of mutant lamin A in Hutchinson-Gilford progeria leads to nuclear abnormalities, which are reversed by farnesyltransferase inhibition.
Glynn MW, Glover TW. *Hum Mol Genet.* 2005 Oct 15;14(20):2959-69.

HGADFN005

Recurrent de novo point mutations in lamin A cause Hutchinson-Gilford progeria syndrome.
Eriksson M, Brown WT, Gordon LB, Glynn MW, Singer J, Scott L, Erdos MR, Robbins CM,

Moses TY, Berglund P, Dutra A, Pak E, Durkin S, Csoka AB, Boehnke M, Glover TW, Collins FS. *Nature.* 2003 May 15;423(6937):293-8.

HGADFN008

Recurrent de novo point mutations in lamin A cause Hutchinson-Gilford progeria syndrome.
Eriksson M, Brown WT, Gordon LB, Glynn MW, Singer J, Scott L, Erdos MR, Robbins CM, Moses TY, Berglund P, Dutra A, Pak E, Durkin S, Csoka AB, Boehnke M, Glover TW, Collins FS. *Nature.* 2003 May 15;423(6937):293-8.

HGADFN014

Recurrent de novo point mutations in lamin A cause Hutchinson-Gilford progeria syndrome.
Eriksson M, Brown WT, Gordon LB, Glynn MW, Singer J, Scott L, Erdos MR, Robbins CM, Moses TY, Berglund P, Dutra A, Pak E, Durkin S, Csoka AB, Boehnke M, Glover TW, Collins FS. *Nature.* 2003 May 15;423(6937):293-8.

HGMDFN090

Activation of endoplasmic reticulum stress in premature aging via the inner nuclear membrane protein SUN2
Vidak S, Serebryannyy LA, Pegoraro G, Misteli T. *Cell Rep*. 2023;42(5):112534. doi:10.1016/j.celrep.2023.112534

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Epigenetic Deregulation of Lamina-Associated Domains in Hutchinson-Gilford Progeria

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Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies
Horvath S, Oshima J, Martin GM, et al. *Aging* (Albany NY). 2018;10(7):1758-1775. doi:10.18632/aging.101508

Nucleoplasmic lamins define growth-regulating functions of lamina-associated polypeptide 2α in progeria cells.
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Glynn MW, Glover TW. *Hum Mol Genet.* 2005 Oct 15;14(20):2959-69.

HGADFN122

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PML2-mediated Thread-Like Nuclear Bodies Mark Late Senescence in Hutchinson-Gilford Progeria Syndrome
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Correction acknowledging PRF for cell lines is pending

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Lamin A Is an Endogenous SIRT6 Activator and Promotes SIRT6-Mediated DNA Repair.
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HGADFN127

Aberrant migration features in primary skin fibroblasts of Huntington's disease patients hold potential for unraveling disease progression using an image based machine learning tool
Gharaba S, Shalem A, Paz O, Muchtar N, Wolf L, Weil M. *Comput Biol Med*. Published online August 2, 2024. doi:10.1016/j.compbiomed.2024.108970

Activation of endoplasmic reticulum stress in premature aging via the inner nuclear membrane protein SUN2
Vidak S, Serebryannyy LA, Pegoraro G, Misteli T. *Cell Rep*. 2023;42(5):112534. doi:10.1016/j.celrep.2023.112534

Ghrelin delays premature aging in Hutchinson-Gilford progeria syndrome
Ferreira-Marques M, Carvalho A, Franco AC, et al. Ghrelin delays premature aging in Hutchinson-Gilford progeria syndrome [published online ahead of print, 2023 Oct 19]. *Aging Cell*. 2023;e13983. doi:10.1111/acel.13983

Perturbed actin cap as a new personalized biomarker in primary fibroblasts of Huntington's disease patients
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HGADFN367

The NLRP3 inhibitor Dapansutrile improves the therapeutic action of lonafarnib on progeroid mice

Muela-Zarzuela I, Suarez-Rivero JM, Boy-Ruiz D, et al. *Aging Cell*. Published online August 27, 2024. doi:10.1111/acel.14272

Aberrant migration features in primary skin fibroblasts of Huntington's disease patients hold potential for unraveling disease progression using an image based machine learning tool
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Hutchinson-Gilford progeria patient-derived cardiomyocyte model of carrying LMNA gene variant c.1824 C > T
Perales S, Sigamani V, Rajasingh S, Czirok A, Rajasingh J. [published online ahead of print, 2023 Aug 12]. *Cell Tissue Res*. 2023;10.1007/s00441-023-03813-2. doi:10.1007/s00441-023-03813-2

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Inhibition of the NLRP3 inflammasome improves lifespan in animal murine model of Hutchinson-Gilford Progeria
González-Dominguez A, Montañez R, Castejón-Vega B, et al. [published online ahead of print, 2021 Aug 27]. *EMBO Mol Med*. 2021;e14012. doi:10.15252/emmm.202114012

A targeted antisense therapeutic approach for Hutchinson-Gilford progeria syndrome
Erdos MR, Cabral WA, Tavarez UL, Cao K, Gvozdenovic-Jeremic J, Narisu N, Zerfas PM, Crumley S, Boku Y, Hanson G, Mourich DV, Kole R, Eckhaus MA, Gordon LB, Collins FS. Nat Med. 2021 Mar;27(3):536-545. doi: 10.1038/s41591-021-01274-0. Epub 2021 Mar 11. PMID: 33707773.

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HGMDFN368

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HGFDFN369

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HGADFN370

Imbalanced Nucleocytoskeletal Connections Create Common Polarity Defects in Progeria and Physiological Aging
Chang W, Wang Y, Luxton GWG, Östlund C, Worman HJ, Gundersen GG. *Proc Natl Acad Sci U S A*. 2019;116(9):3578-3583. doi:10.1073/pnas.1809683116

HGMDFN371

Imbalanced Nucleocytoskeletal Connections Create Common Polarity Defects in Progeria and Physiological Aging
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A mutation abolishing the ZMPSTE24 cleavage site in prelamin A causes a progeroid disorder
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HGADFN496

A targeted antisense therapeutic approach for Hutchinson-Gilford progeria syndrome
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HGMDFN717

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HGMDFN718

A targeted antisense therapeutic approach for Hutchinson-Gilford progeria syndrome
Erdos MR, Cabral WA, Tavarez UL, Cao K, Gvozdenovic-Jeremic J, Narisu N, Zerfas PM, Crumley S, Boku Y, Hanson G, Mourich DV, Kole R, Eckhaus MA, Gordon LB, Collins FS. Nat Med. 2021 Mar;27(3):536-545. doi: 10.1038/s41591-021-01274-0. Epub 2021 Mar 11. PMID: 33707773.

PSADFN086 (formally HGADFN086)

Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies
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Increased progerin expression associated with unusual LMNA mutations causes severe progeroid syndromes.
Moulson CL, Fong LG, Gardner JM, Farber EA, Go G, Passariello A, Grange DK, Young SG, Miner JH. *Hum Mutat.* 2007 Sep;28(9):882-9.

PSADFN257

A Cell-Intrinsic Interferon-like Response Links Replication Stress to Cellular Aging Caused by Progerin.
Kreienkamp R, Graziano S, Coll-Bonfill N, Bedia-Diaz G, Cybulla E, Vindigni A, Dorsett D, Kubben N, Batista LFZ, Gonzalo S. *Cell Rep*. 2018 Feb 20;22(8):2006-2015.

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PSADFN317

Impact of Combined Baricitinib and FTI Treatment on Adipogenesis in Hutchinson-Gilford Progeria Syndrome and Other Lipodystrophic Laminopathies
Hartinger R, Lederer EM, Schena E, Lattanzi G, Djabali K. Cells. 2023;12(10):1350. Published 2023 May 9. doi:10.3390/cells12101350

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PSADFN318

Impact of Combined Baricitinib and FTI Treatment on Adipogenesis in Hutchinson-Gilford Progeria Syndrome and Other Lipodystrophic Laminopathies
Hartinger R, Lederer EM, Schena E, Lattanzi G, Djabali K. Cells. 2023;12(10):1350. Published 2023 May 9. doi:10.3390/cells12101350

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PSFDFN319

Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies
Horvath S, Oshima J, Martin GM, et al. *Aging* (Albany NY). 2018;10(7):1758-1775. doi:10.18632/aging.101508

Rapamycin reverses cellular phenotypes and enhances mutant protein clearance in Hutchinson-Gilford progeria syndrome cells.
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Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies
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Cao K, Graziotto JJ, Blair CD, Mazzulli JR, Erdos MR, Krainc D, Collins FS. *Sci Transl Med.* 2011 Jun 29;3(89):89ra58.

PSMDFN326

Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies
Horvath S, Oshima J, Martin GM, et al. *Aging* (Albany NY). 2018;10(7):1758-1775. doi:10.18632/aging.101508

PSFDFN327

A Cell-Intrinsic Interferon-like Response Links Replication Stress to Cellular Aging Caused by Progerin.
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Kreienkamp R, Croke M, Neumann MA, et al. *Oncotarget* 2016;7(21):30018-30031. doi:10.18632/oncotarget.9065

PSMDFN346

A Cell-Intrinsic Interferon-like Response Links Replication Stress to Cellular Aging Caused by Progerin.

Kreienkamp R, Graziano S, Coll-Bonfill N, Bedia-Diaz G, Cybulla E, Vindigni A, Dorsett D, Kubben N, Batista LFZ, Gonzalo S. *Cell Rep*. 2018 Feb 20;22(8):2006-2015.

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Vitamin D Receptor Signaling Improves Hutchinson-Gilford Progeria Syndrome Cellular Phenotypes

Kreienkamp R, Croke M, Neumann MA, et al. *Oncotarget* 2016;7(21):30018-30031. doi:10.18632/oncotarget.9065

PSADFN363

The farnesyl transferase inhibitor (FTI) lonafarnib improves nuclear morphology in ZMPSTE24-deficient fibroblasts from patients with the progeroid disorder MAD-B

Odinammadu KO, Shilagardi K, Tuminelli K, Judge DP, Gordon LB, Michaelis S. *Nucleus*. 2023;14(1):2288476. doi:10.1080/19491034.2023.2288476

PSADFN373

The farnesyl transferase inhibitor (FTI) lonafarnib improves nuclear morphology in ZMPSTE24-deficient fibroblasts from patients with the progeroid disorder MAD-B

Odinammadu KO, Shilagardi K, Tuminelli K, Judge DP, Gordon LB, Michaelis S. *Nucleus*. 2023;14(1):2288476. doi:10.1080/19491034.2023.2288476

Targeting RAS-converting enzyme 1 overcomes senescence and improves progeria-like phenotypes of ZMPSTE24 deficiency

Yao H, Chen X, Kashif M, Wang T, Ibrahim MX, Tüksammel E, Revêchon G, Eriksson M, Wiel C, Bergo MO. Aging Cell. 2020 Aug;19(8):e13200. doi: 10.1111/acel.13200. Epub 2020 Jul 24. PMID: 32910507; PMCID: PMC7431821.

PSADFN386

MG132 Induces Progerin Clearance and Improves Disease Phenotypes in HGPS-like Patients' Cells

Harhouri K, Cau P, Casey F, et al. *Cells*. 2022;11(4):610. Published 2022 Feb 10. doi:10.3390/cells11040610

A novel somatic mutation achieves partial rescue in a child with Hutchinson-Gilford progeria syndrome

Bar DZ, Arlt MF, Brazier JF, et al. *J Med Genet*. 2017;54(3):212-216. doi:10.1136/jmedgenet-2016-104295

PSMDFN387

A novel somatic mutation achieves partial rescue in a child with Hutchinson-Gilford progeria syndrome

Bar DZ, Arlt MF, Brazier JF, et al. *J Med Genet*. 2017;54(3):212-216. doi:10.1136/jmedgenet-2016-104295

PSFDFN388

A novel somatic mutation achieves partial rescue in a child with Hutchinson-Gilford progeria syndrome

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PSADFN392

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PSADFN425

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PSADFN485

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PSADFN542

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HGADFN003 iPS1B

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HGMDFN090 iPS1B

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HGADFN167 iPS1J

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HGADFN167 iPS1Q

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HGFDFN168 iPS1D2

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HGFDFN168 iPS1P

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HGALBV009

Inhibition of the NLRP3 inflammasome improves lifespan in animal murine model of Hutchinson-Gilford Progeria
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HGMLBV010

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HGMLBV013

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HGFLBV021

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HGMLBV023

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HGFLBV031

Stem cell depletion in Hutchinson-Gilford progeria syndrome.
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HGFLBV050

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Eriksson M, Brown WT, Gordon LB, Glynn MW, Singer J, Scott L, Erdos MR, Robbins CM, Moses TY, Berglund P, Dutra A, Pak E, Durkin S, Csoka AB, Boehnke M, Glover TW, Collins FS. *Nature*. 2003 May 15;423(6937):293-8. Epub 2003 Apr 25.

HGALBV057

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HGMLBV058

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Eriksson M, Brown WT, Gordon LB, Glynn MW, Singer J, Scott L, Erdos MR, Robbins CM,

Moses TY, Berglund P, Dutra A, Pak E, Durkin S, Csoka AB, Boehnke M, Glover TW, Collins FS. *Nature*. 2003 May 15;423(6937):293-8. Epub 2003 Apr 25.

HGSLBV059

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HGMLBV066

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HGFLBV067

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HGALBV071

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Eriksson M, Brown WT, Gordon LB, Glynn MW, Singer J, Scott L, Erdos MR, Robbins CM, Moses TY, Berglund P, Dutra A, Pak E, Durkin S, Csoka AB, Boehnke M, Glover TW, Collins FS. *Nature*. 2003 May 15;423(6937):293-8. Epub 2003 Apr 25.

HGMLBV081

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HGFLBV082

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