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.**

**Age-dependent loss of MMP-3 in Hutchinson-Gilford progeria syndrome.**

**The mutant form of lamin A that causes Hutchinson-Gilford progeria is a biomarker of cellular aging in human skin.**

**Hutchinson-Gilford progeria mutant lamin A primarily targets human vascular cells as detected by an anti-Lamin A G608G antibody.**

**Aggrecan expression is substantially and abnormally upregulated in Hutchinson-Gilford Progeria Syndrome dermal fibroblasts.**

**Rescue of heterochromatin organization in Hutchinson-Gilford progeria by drug treatment.**

**Recurrent de novo point mutations in lamin A cause Hutchinson-Gilford progeria syndrome.**

**HGADFN003**

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Ghrelin delays premature aging in Hutchinson-Gilford progeria syndrome

Impact of Combined Baricitinib and FTI Treatment on Adipogenesis in Hutchinson-Gilford Progeria Syndrome and Other Lipodystrophic Laminopathies

Unique progerin C-terminal peptide ameliorates Hutchinson-Gilford progeria syndrome phenotype by rescuing BUBR1.

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Isoprenylcysteine Carboxymethyltransferase-Based Therapy for Hutchinson-Gilford Progeria Syndrome

Baricitinib, a JAK-STAT Inhibitor, Reduces the Cellular Toxicity of the Farnesyltransferase Inhibitor Lonafarnib in Progeria Cells

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Nuclear Pore Complexes Cluster in Dysmorphic Nuclei of Normal and Progeria Cells during Replicative Senescence.

Self-assembly of multi-component mitochondrial nucleoids via phase separation.

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Autophagic Removal of Farnesylated Carboxy-Terminal Lamin Peptides

Targeting the Phospholipase A2 Receptor Ameliorates Premature Aging Phenotypes

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

Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies

Nucleoplasmic lamins define growth-regulating functions of lamina-associated polypeptide 2α in progeria cells.
Intermittent treatment with farnesyltransferase inhibitor and sulforaphane improves cellular homeostasis in Hutchinson-Gilford progeria fibroblasts.

Temsirolimus Partially Rescues the Hutchinson-Gilford Progeria Cellular Phenotype.

Progerin Impairs Chromosome Maintenance by Depleting CENP-F From Metaphase Kinetochores in Hutchinson-Gilford Progeria Fibroblasts

Permanent farnesylation of lamin A mutants linked to progeria impairs its phosphorylation at serine 22 during interphase.

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

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

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Harten IA, Zahr RS, Lemire JM, Machan JT, Moses MA, Doiron RJ, Curatolo AS, Rothman FG,

**Progerin and telomere dysfunction collaborate to trigger cellular senescence in normal human fibroblasts.**  

**Defective lamin A-Rb signaling in Hutchinson-Gilford Progeria Syndrome and reversal by farnesyltransferase inhibition.**  

**Effect of progerin on the accumulation of oxidized proteins in fibroblasts from Hutchinson Gilford progeria patients.**  

**Ageing-related chromatin defects through loss of the NURD complex.**  

**Lamin A-dependent misregulation of adult stem cells associated with accelerated ageing.**  

**Perturbation of wild-type lamin A metabolism results in a progeroid phenotype.**  

**Alterations in mitosis and cell cycle progression caused by a mutant lamin A known to accelerate human aging.**  

**The mutant form of lamin A that causes Hutchinson-Gilford progeria is a biomarker of cellular aging in human skin.**  

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Genomic instability in laminopathy-based premature aging.

Incomplete processing of mutant lamin A in Hutchinson-Gilford progeria leads to nuclear abnormalities, which are reversed by farnesyltransferase inhibition.

Accumulation of mutant lamin A causes progressive changes in nuclear architecture in Hutchinson-Gilford progeria syndrome.

Recurrent de novo point mutations in lamin A cause Hutchinson-Gilford progeria syndrome.

HGADFN004

Incomplete processing of mutant lamin A in Hutchinson-Gilford progeria leads to nuclear abnormalities, which are reversed by farnesyltransferase inhibition.

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,
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**Correlated alterations in genome organization, histone methylation, and DNA-lamin A/C interactions in Hutchinson-Gilford progeria syndrome.**

**Comparison of constitutional and replication stress-induced genome structural variation by SNP array and mate-pair sequencing.**

**Hydroxyurea induces de novo copy number variants in human cells.**

**Progerin and telomere dysfunction collaborate to trigger cellular senescence in normal human fibroblasts.**

**CTP:phosphocholine cytidylyltransferase α (CCTα) and lamins alter nuclear membrane structure without affecting phosphatidylcholine synthesis.**

**Effect of progerin on the accumulation of oxidized proteins in fibroblasts from Hutchinson Gilford progeria patients.**

**Replication stress induces genome-wide copy number changes in human cells that resemble polymorphic and pathogenic variants.**

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Park SK, Shin OS. Exp Dermatol. 2017 Feb 13. [Epub ahead of print]

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the aging cell model.

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HGADFN127
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**Replicative Senescence.**
PMID: 33466669; PMCID: PMC7828780.

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doi:10.3390/cells8101276

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doi:10.18632/aging.101508

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doi:10.1371/journal.pone.0168988

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Kinetochores in Hutchinson-Gilford Progeria Fibroblasts
doi:10.18632/oncotarget.8267

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Genomic instability in laminopathy-based premature aging.

Novel progerin-interactive partner proteins hnRNP E1, EGF, Mel 18, and UBC9 interact with lamin A/C.

**HGADFN136**

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**HGADFN143**

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HGADFN155

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HGADFN164

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**HGADFN167**

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**Lonafarnib and everolimus reduce pathology in iPSC-derived tissue engineered blood vessel model of Hutchinson-Gilford Progeria Syndrome.**

**Achieving single nucleotide sensitivity in direct hybridization genome imaging**
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Nucleolar expansion and elevated protein translation in premature aging.

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Loss of H3K9me3 Correlates with ATM Activation and Histone H2AX Phosphorylation Deficiencies in Hutchinson-Gilford Progeria Syndrome.

NANOG reverses the Myogenic Differentiation Potential of Senescent Stem Cells by Restoring ACTIN Filamentous Organization and SRF-Dependent Gene Expression.

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

**Inhibition of JAK-STAT Signaling With Baricitinib Reduces Inflammation and Improves Cellular Homeostasis in Progeria Cells**

**Analysis of Somatic Mutations Identifies Signs of Selection During in Vitro Aging of Primary Dermal Fibroblasts**

**Predicting Age From the Transcriptome of Human Dermal Fibroblasts**

**p53 isoforms regulate premature aging in human cells.**

**Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies**

**Temsirolimus Partially Rescues the Hutchinson-Gilford Progeria Cellular Phenotype.**

Version date December 2023

**Progerin Impairs Chromosome Maintenance by Depleting CENP-F From Metaphase Kinetochores in Hutchinson-Gilford Progeria Fibroblasts**

**Sulforaphane enhances progerin clearance in Hutchinson-Gilford progeria fibroblasts.**

**Depleting the methyltransferase Suv39h1 improves DNA repair and extends lifespan in a progeria mouse model.**

**Naïve adult stem cells from patients with Hutchinson-Gilford progeria syndrome express low levels of progerin in vivo.**

**Defective lamin A-Rb signaling in Hutchinson-Gilford Progeria Syndrome and reversal by farnesyltransferase inhibition.**

**HGADFN271**

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

**SAMMY-seq reveals early alteration of heterochromatin and deregulation of bivalent genes in Hutchinson-Gilford Progeria Syndrome**

**Epigenetic Deregulation of Lamina-Associated Domains in Hutchinson-Gilford Progeria Syndrome**
Transient Introduction of Human Telomerase mRNA Improves Hallmarks of Progeria Cells

Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies

HGADFN367

Hutchinson-Gilford progeria patient-derived cardiomyocyte model of carrying LMNA gene variant c.1824 C > T

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

SerpinE1 drives a cell-autonomous pathogenic signaling in Hutchinson-Gilford progeria syndrome

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

A targeted antisense therapeutic approach for Hutchinson-Gilford progeria syndrome

Direct reprogramming of human smooth muscle and vascular endothelial cells reveals defects associated with aging and Hutchinson-Gilford progeria syndrome
Transient Introduction of Human Telomerase mRNA Improves Hallmarks of Progeria Cells

Predicting Age From the Transcriptome of Human Dermal Fibroblasts

Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies

HGMDFN368
Hutchinson-Gilford progeria patient-derived cardiomyocyte model of carrying LMNA gene variant c.1824 C > T

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

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Version date December 2023
HGFDFN369

Hutchinson-Gilford progeria patient-derived cardiomyocyte model of carrying LMNA gene variant c.1824 C > T

Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies

HGADFN370

Imbalanced Nucleocytoskeletal Connections Create Common Polarity Defects in Progeria and Physiological Aging

HGMDFN371

Imbalanced Nucleocytoskeletal Connections Create Common Polarity Defects in Progeria and Physiological Aging

HGADFN496

A targeted antisense therapeutic approach for Hutchinson-Gilford progeria syndrome

HGMDFN717

Hutchinson-Gilford progeria patient-derived cardiomyocyte model of carrying LMNA gene variant c.1824 C > T

Version date December 2023
HGMDFN718

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

PSADFN086
(formally HGADFN086)

**Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies**

**Increased progerin expression associated with unusual LMNA mutations causes severe progeroid syndromes**

PSADFN257

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

**Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies**

**Vitamin D Receptor Signaling Improves Hutchinson-Gilford Progeria Syndrome Cellular Phenotypes**
PSADFN317

Impact of Combined Baricitinib and FTI Treatment on Adipogenesis in Hutchinson-Gilford Progeria Syndrome and Other Lipodystrophic Laminopathies

Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies

PSADFN318

Impact of Combined Baricitinib and FTI Treatment on Adipogenesis in Hutchinson-Gilford Progeria Syndrome and Other Lipodystrophic Laminopathies

Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies

PSFDFN319

Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies

Rapamycin reverses cellular phenotypes and enhances mutant protein clearance in Hutchinson-Gilford progeria syndrome cells.

PSMDFN320

Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies

Version date December 2023
Rapamycin reverses cellular phenotypes and enhances mutant protein clearance in Hutchinson-Gilford progeria syndrome cells.

**PSMDFN326**

Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies
doi:10.18632/aging.101508

**PSFDFN327**

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

Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies
doi:10.18632/aging.101508

Vitamin D Receptor Signaling Improves Hutchinson-Gilford Progeria Syndrome Cellular Phenotypes
doi:10.18632/oncotarget.9065

**PSMDFN346**

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

Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies
Vitamin D Receptor Signaling Improves Hutchinson-Gilford Progeria Syndrome Cellular Phenotypes

PSADFN363

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

PSADFN373

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

PSADFN386

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

PSADFN392

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

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Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies

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

PSMDFN393
Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies

PSFDFN394
Epigenetic clock for skin and blood cells applied to Hutchinson Gilford Progeria Syndrome and ex vivo studies

PSADFN414
Everolimus Rescues Multiple Cellular Defects in Laminopathy-Patient Fibroblasts

PSADFN425
Everolimus Rescues Multiple Cellular Defects in Laminopathy-Patient Fibroblasts

Version date December 2023
The farnesyl transferase inhibitor (FTI) lonafarnib improves nuclear morphology in ZMPSTE24-deficient fibroblasts from patients with the progeroid disorder MAD-B

Gaussian curvature dilutes the nuclear lamina, favoring nuclear rupture, especially at high strain rate

iPSC-Derived Endothelial Cells Affect Vascular Function in a Tissue-Engineered Blood Vessel Model of Hutchinson-Gilford Progeria Syndrome

Progerin Phosphorylation in Interphase Is Lower and Less Mechanosensitive Than lamin-A,C in iPS-derived Mesenchymal Stem Cells

Reprogramming Progeria Fibroblasts Re-Establishes a Normal Epigenetic Landscape
Telomerase therapy reverses vascular senescence and extends lifespan in progeria mice

Reprogramming Progeria Fibroblasts Re-Establishes a Normal Epigenetic Landscape

**HGADFN003 iPS1D**

Lonafarnib and everolimus reduce pathology in iPSC-derived tissue engineered blood vessel model of Hutchinson-Gilford Progeria Syndrome.

iPSC-Derived Endothelial Cells Affect Vascular Function in a Tissue-Engineered Blood Vessel Model of Hutchinson-Gilford Progeria Syndrome

Dysfunction of iPSC-derived Endothelial Cells in Human Hutchinson-Gilford Progeria Syndrome

**HGMDFN090 iPS1B**

Telomerase therapy reverses vascular senescence and extends lifespan in progeria mice

Dysfunction of iPSC-derived Endothelial Cells in Human Hutchinson-Gilford Progeria Syndrome

Reprogramming Progeria Fibroblasts Re-Establishes a Normal Epigenetic Landscape

**HGMDFN090 iPS1C**

Aging Model for Analyzing Drug-Induced Proarrhythmia Risks Using Cardiomyocytes

Version date December 2023
Differentiated from Progeria-Patient-Derived Induced Pluripotent Stem Cells

Reprogramming Progeria Fibroblasts Re-Establishes a Normal Epigenetic Landscape

HGADFN167 iPS1J

Aging Model for Analyzing Drug-Induced Proarrhythmia Risks Using Cardiomyocytes Differentiated from Progeria-Patient-Derived Induced Pluripotent Stem Cells

Modelling premature cardiac aging with induced pluripotent stem cells from a Hutchinson-Gilford Progeria Syndrome patient

Reprogramming Progeria Fibroblasts Re-Establishes a Normal Epigenetic Landscape

Mechanisms Controlling the Smooth Muscle Cell Death in Progeria via Down-Regulation of poly(ADP-ribose) Polymerase 1

HGADFN167 iPS1Q

Vascular senescence in progeria: role of endothelial dysfunction

Telomerase therapy reverses vascular senescence and extends lifespan in progeria mice

Dysfunction of iPSC-derived Endothelial Cells in Human Hutchinson-Gilford Progeria Syndrome

Reprogramming Progeria Fibroblasts Re-Establishes a Normal Epigenetic Landscape

Version date December 2023

**HGFDFN168 iPS1D2**

Reprogramming Progeria Fibroblasts Re-Establishes a Normal Epigenetic Landscape

Mechanisms Controlling the Smooth Muscle Cell Death in Progeria via Down-Regulation of poly(ADP-ribose) Polymerase 1

**HGFDFN168 iPS1P**

Vascular senescence in progeria: role of endothelial dysfunction

Telomerase therapy reverses vascular senescence and extends lifespan in progeria mice

Dysfunction of iPSC-derived Endothelial Cells in Human Hutchinson-Gilford Progeria Syndrome

Reprogramming Progeria Fibroblasts Re-Establishes a Normal Epigenetic Landscape

**HGALBV009**

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

Stem cell depletion in Hutchinson-Gilford progeria syndrome

Version date December 2023
Low and high expressing alleles of the LMNA gene: implications for laminopathy disease development.

Recurrent de novo point mutations in lamin A cause Hutchinson-Gilford progeria syndrome.

HGMLBV010

Stem cell depletion in Hutchinson-Gilford progeria syndrome.

Recurrent de novo point mutations in lamin A cause Hutchinson-Gilford progeria syndrome.

HGALBV011

Low and high expressing alleles of the LMNA gene: implications for laminopathy disease development.

Recurrent de novo point mutations in lamin A cause Hutchinson-Gilford progeria syndrome.

HGMLBV013

Recurrent de novo point mutations in lamin A cause Hutchinson-Gilford progeria syndrome.
Inhibition of the NLRP3 inflammasome improves lifespan in animal murine model of Hutchinson-Gilford Progeria

Stem cell depletion in Hutchinson-Gilford progeria syndrome.

Recurrent de novo point mutations in lamin A cause Hutchinson-Gilford progeria syndrome.
Recurrent de novo point mutations in lamin A cause Hutchinson-Gilford progeria syndrome.

Stem cell depletion in Hutchinson-Gilford progeria syndrome.

Recurrent de novo point mutations in lamin A cause Hutchinson-Gilford progeria syndrome.

Stem cell depletion in Hutchinson-Gilford progeria syndrome.

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Recurrent de novo point mutations in lamin A cause Hutchinson-Gilford progeria syndrome.

HGFLBV067

Stem cell depletion in Hutchinson-Gilford progeria syndrome.

Recurrent de novo point mutations in lamin A cause Hutchinson-Gilford progeria syndrome.

HGALBV071

Recurrent de novo point mutations in lamin A cause Hutchinson-Gilford progeria syndrome.

HGMLBV081

Recurrent de novo point mutations in lamin A cause Hutchinson-Gilford progeria syndrome.

HGFLBV082

Recurrent de novo point mutations in lamin A cause Hutchinson-Gilford progeria syndrome.

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