

1. iPSC Background information for the non-scientist

Stem cells are “immature” cells that have not yet committed to becoming any one cell type. They are pliable because they have the potential to develop into many different types of mature cells in the body, such as cells that make up the heart or blood vessels, and other tissues and organs. In 2007, researchers discovered a strategy for creating stem cells in the laboratory by reprogramming mature adult cells that we commonly grow for research purposes.^{1,2} These artificially created stem cells are called Induced Pluripotent Stem Cells (“iPSCs”). For the field of Progeria, this is a huge breakthrough. For the first time, scientists can now make Progeria stem cells and ask questions about how stem cells function and develop in Progeria. Previously there was no source of human Progeria stem cells, and there was therefore a void of information about how Progeria stem cells function compared with stem cells from people without Progeria. In addition, scientists can re-program the Progeria stem cells to create, for the first time, mature Progeria blood vessels, heart cells, and other cell types. Until now, there was no source of human Progeria heart or blood vessel cells. **We can now ask key** questions about the heart disease that leads to early death in Progeria from heart attacks and strokes. We can compare these discoveries with heart disease and aging in the general population and discover more about what influences aging in all of us. Already there have been several excellent studies published using Progeria stem cells.³⁻⁴ Our goal at The Progeria Research Foundation is to facilitate many more discoveries using this invaluable tool. For a primer on stem cells, please see this US government website: [Stem Cell Basics | STEM Cell Information](#)

1. [Induction of pluripotent stem cells from adult human fibroblasts by defined factors](#)
Takahashi K, Tanabe K, Ohnuki M, et al. *Cell*. 2007;131(5):861-872.
doi:10.1016/j.cell.2007.11.019
2. [Induced pluripotent stem cell lines derived from human somatic cells](#)
Yu J, Vodyanik MA, Smuga-Otto K, et al. *Science*. 2007;318(5858):1917-1920.
doi:10.1126/science.1151526
3. [Recapitulation of premature ageing with iPSCs from Hutchinson-Gilford progeria syndrome](#)
Liu GH, Barkho BZ, Ruiz S, et al. *Nature*. 2011;472(7342):221-225.
doi:10.1038/nature09879
4. [HGPS-derived iPSCs for the ages](#)
Misteli T. *Cell Stem Cell*. 2011;8(1):4-6. doi:10.1016/j.stem.2010.12.014

2. Purpose of induced pluripotent stem cell (iPSC) generation and distribution by The Progeria Research Foundation

The mission of The Progeria Research Foundation is to discover treatments and the cure for Hutchinson-Gilford Progeria Syndrome and its aging-related disorders. In 2009, PRF entered into a collaboration with an expert team of scientists at the University of Toronto, Canada, under the direction of William Stanford, PhD, to generate high-quality Progeria iPSCs. Dr. Stanford is the Canada Research Chair in Integrative Stem Cell Biology. As of 2026, PRF continues to collaborate with Dr. Stanford at the University of Ottawa, Canada where he is Professor of Cellular and Molecular Medicine, Faculty of Medicine, and Senior Scientist at Ottawa Hospital Research Institute's Sprott Centre for Stem Cell Research.

Our goal is to provide this invaluable tool to researchers throughout the world. This new research tool will be used to generate new and innovative research in Progeria, as well as its relationship to heart disease and aging.

3. Generation of Hutchinson-Gilford Progeria Syndrome Induced-Pluripotent Stem Cells (iPSCs)

Induced-Pluripotent Stem Cells (iPSCs) were derived using VSVG-pseudotyped retroviral transduction of four human factors, Oct4, Sox2, Klf4, and c-Myc into fibroblasts. iPSC colonies were derived on mouse-embryonic fibroblasts (MEFs). The procedure used was essentially as previously described but without the use of the EOS reporter ([Nature Protocols 4: 1828-1844, 2009](#)).

4. Quality Control: Validation and Characterization

The lines that are currently available have undergone several validation steps:

1. Mycoplasma Testing for each line: Dr. Stanford's lab has performed mycoplasma analysis by PCR for each cell line. In addition, after expansion and prior to shipping cells, the lines will be retested for mycoplasma.
2. Immunostaining for pluripotency markers Tra-1-60, Tra-1-81, and SSEA4.
3. Alkaline Phosphatase Staining as an indicator of pluripotency
4. Embryoid body formation and subsequent immunostaining for markers of the three germ-layers. Markers tested were β III-Tubulin (Ectoderm), Smooth-Muscle Actin (Mesoderm), and Gata4 or AFP (Endoderm)
5. Karyotype analysis.
6. Re-expression of lamin A in differentiated cells
7. Teratoma assays

Additional validation in process:

Some lines have completed teratoma assays as shown in supporting data. For all other lines, teratoma assays are in process and status will be updated as these assays are completed.

5. Original starting material from which these iPS cells were derived

iPSCs were derived from PRF Cell & Tissue Bank non-transformed fibroblast cell lines. The transduction method used for all iPS lines was Retrovirus MKOS.

iPSC Line ID	Mutation	Gender	Age at Donation	Originating Cell Line and Type	Supporting Data
HGADFN003 iPS 1B	LMNA Exon 11, heterozygous c.1824C>T (p.Gly608Gly)	Male	2 yr 0 mo	HGADFN003 Dermal Fibroblasts	003 iPS 1B
HGADFN003 iPS 1C	LMNA Exon 11, heterozygous c.1824C>T (p.Gly608Gly)	Male	2 yr 0 mo	HGADFN003 Dermal Fibroblasts	003 iPS 1C
HGADFN003 iPS 1D	LMNA Exon 11, heterozygous c.1824C>T (p.Gly608Gly)	Male	2 yr 0 mo	HGADFN003 Dermal Fibroblasts	003 iPS 1D
HGADFN167 iPS 1J	LMNA Exon 11, heterozygous c.1824C>T (p.Gly608Gly)	Male	8 yr 5 mo	HGADFN167 Dermal Fibroblasts	167 iPS 1J
HGADFN167 iPS 1Q	LMNA Exon 11, heterozygous c.1824C>T (p.Gly608Gly)	Male	8 yr 5 mo	HGADFN167 Dermal Fibroblasts	167 iPS 1Q
HGMDFN090 iPS 1B	LMNA Exon 11, Negative Mother of HGADFN167	Female	37 yr 10 mo	HGMDFN090 Dermal Fibroblasts	090 iPS 1B
HGMDFN090 iPS 1C	LMNA Exon 11, Negative Mother of HGADFN167	Female	37 yr 10 mo	HGMDFN090 Dermal Fibroblasts	090 iPS 1C
HGFDFN168 iPS 1D2	LMNA Exon 11, Negative Father of HGADFN167	Male	40 yr 5 mo	HGFDFN168 Dermal Fibroblasts	168 iPS 1D2
HGFDFN168 iPS 1P	LMNA Exon 11, Negative Father of HGADFN167	Male	40 yr 5 mo	HGFDFN168 Dermal Fibroblasts	168 iPS 1P

6. Questions?

Please contact Leslie Gordon, MD, PhD, Medical Director, with any questions or needs, at lgordon@progeriaresearch.org or 978-535-2594

7. Progeria iPSC Cell Line Distribution Process

Since 2014, the Progeria Research Foundation (PRF) has maintained a policy of no modifications to its Material Transfer Agreement (MTA). This policy reflects more than 12 years of experience with contractual arrangements involving approximately 70 research teams at institutions in 14 countries. During this time, PRF and its legal counsel have reviewed issues that have arisen and revised the agreement as needed. The current MTA reflects terms that PRF considers fair and reasonable for the distribution of these research materials.

For U.S. federal government institutions, or if you have questions about the process, please contact:

Wendy Norris

wnorris@brownhealth.org

401-274-1122, ext. 48063

1. Review the Guidelines

Review the [guidelines](#) developed by the Ottawa Hospital Research Institute (OHRI) and PRF.

2. Complete the Application and MTA

Complete the required application and Material Transfer Agreement.

The application and MTA are available on the PRF website:

[Cell & Tissue Bank – The Progeria Research Foundation](#)

3. Submit the Completed Documents

Return the completed application and MTA to Wendy Norris at

wnorris@brownhealth.org.

Once your application has been approved, you will receive an email confirming your order and anticipated shipping date.

4. Shipment and Training

Dr. Stanford's laboratory currently distributes iPSC lines as frozen cryovials. The laboratory will notify you by email when the culture has been shipped and will provide the shipping and tracking information.

Researchers who are inexperienced with human embryonic stem cell or iPSC culture are strongly encouraged to obtain appropriate training before working with these materials.

The Human Pluripotent Stem Cell Facility, directed by Dr. Stanford at the Ottawa Hospital Research Institute, offers virtual, one-on-one training in basic iPSC culture techniques specific to the Progeria iPSC lines. Training format and content can be tailored to the researcher's level of experience.

For more information about training, contact hpscf@ohri.ca.

5. Payment and Shipping Costs

The University of Ottawa will invoice the recipient directly for each iPSC line, as well as any applicable courier or shipping costs.

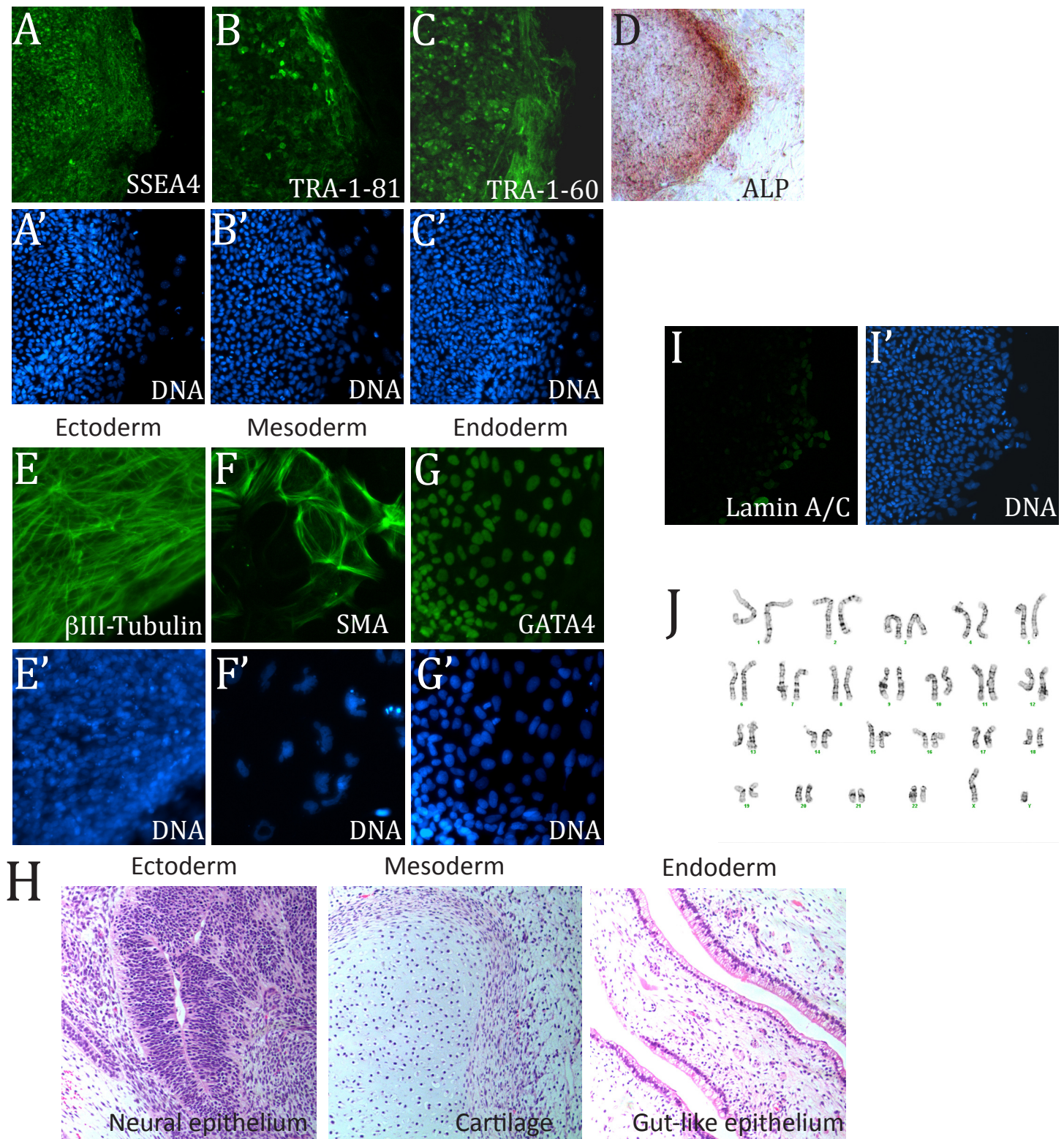


Figure Legend. Hutchinson-Gilford Progeria induced-pluripotent stem cell line HGADFN003 iPS1B exhibits characteristics of pluripotency. (A-D) HGADFN003 iPS1B cells express markers for pluripotency, SSEA4, Tra-1-81, Tra-1-60, and alkaline phosphatase. (E-G') In vitro differentiation of HGADFN003 iPS1B cells shows that it has the capacity to differentiate to cells of all 3-germ layers, β III-tubulin (ectoderm), smooth-muscle actin (mesoderm), Gata4 (endoderm). (H) In vivo differentiation of HGADFN003 iPS1B cells shows that it has the capacity to differentiate to tissues of all 3-germ layers, neural epithelium (ectoderm), cartilage (mesoderm), Gut-like epithelium (endoderm) (I-I') HGADFN003 iPS1B cells exhibit repressed Lamin A/C expression in undifferentiated cells but detectable expression in differentiated cells at the border of the colony. (J) The HGADFN003 iPS1B cell line exhibits normal karyotype.

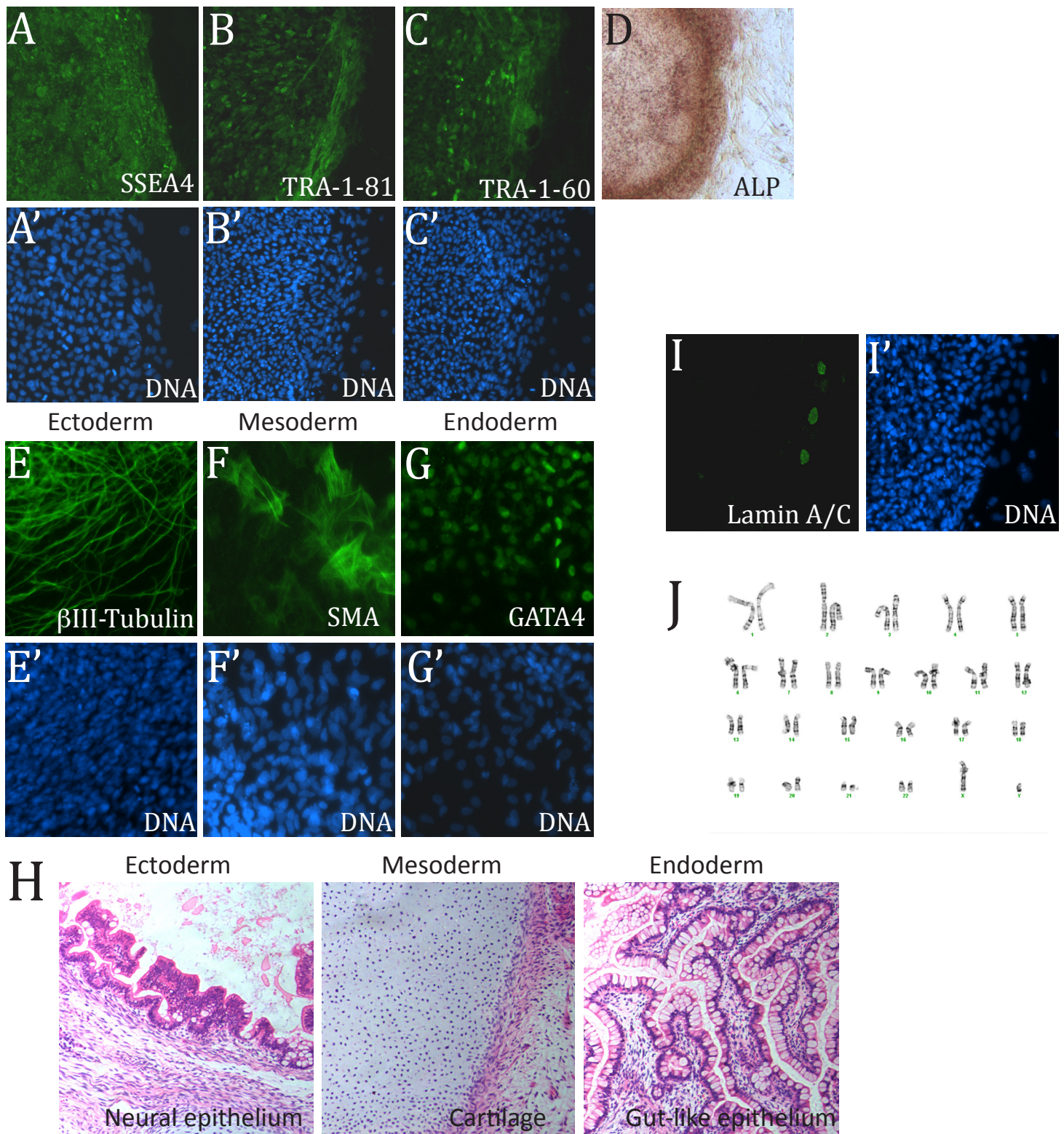


Figure Legend. Hutchinson-Gilford Progeria induced-pluripotent stem cell line HGADFN003 iPS1C exhibits characteristics of pluripotency. (A-D) HGADFN003 iPS1C cells express markers for pluripotency, SSEA4, Tra-1-81, Tra-1-60, and alkaline phosphatase. (E-G') In vitro differentiation of HGADFN003 iPS1C cells shows that it has the capacity to differentiate to cells of all 3-germ layers, β III-tubulin (ectoderm), smooth-muscle actin (mesoderm), Gata4 (endoderm). (H) In vivo differentiation of HGADFN003 iPS1C cells shows that it has the capacity to differentiate to tissues of all 3-germ layers, neural epithelium (ectoderm), cartilage (mesoderm), Gut-like epithelium (endoderm) (I-I') HGADFN003 iPS1C cells exhibit repressed Lamin A/C expression in undifferentiated cells but detectable expression in differentiated cells at the border of the colony. (J) The HGADFN003 iPS1C cell line exhibits normal karyotype.

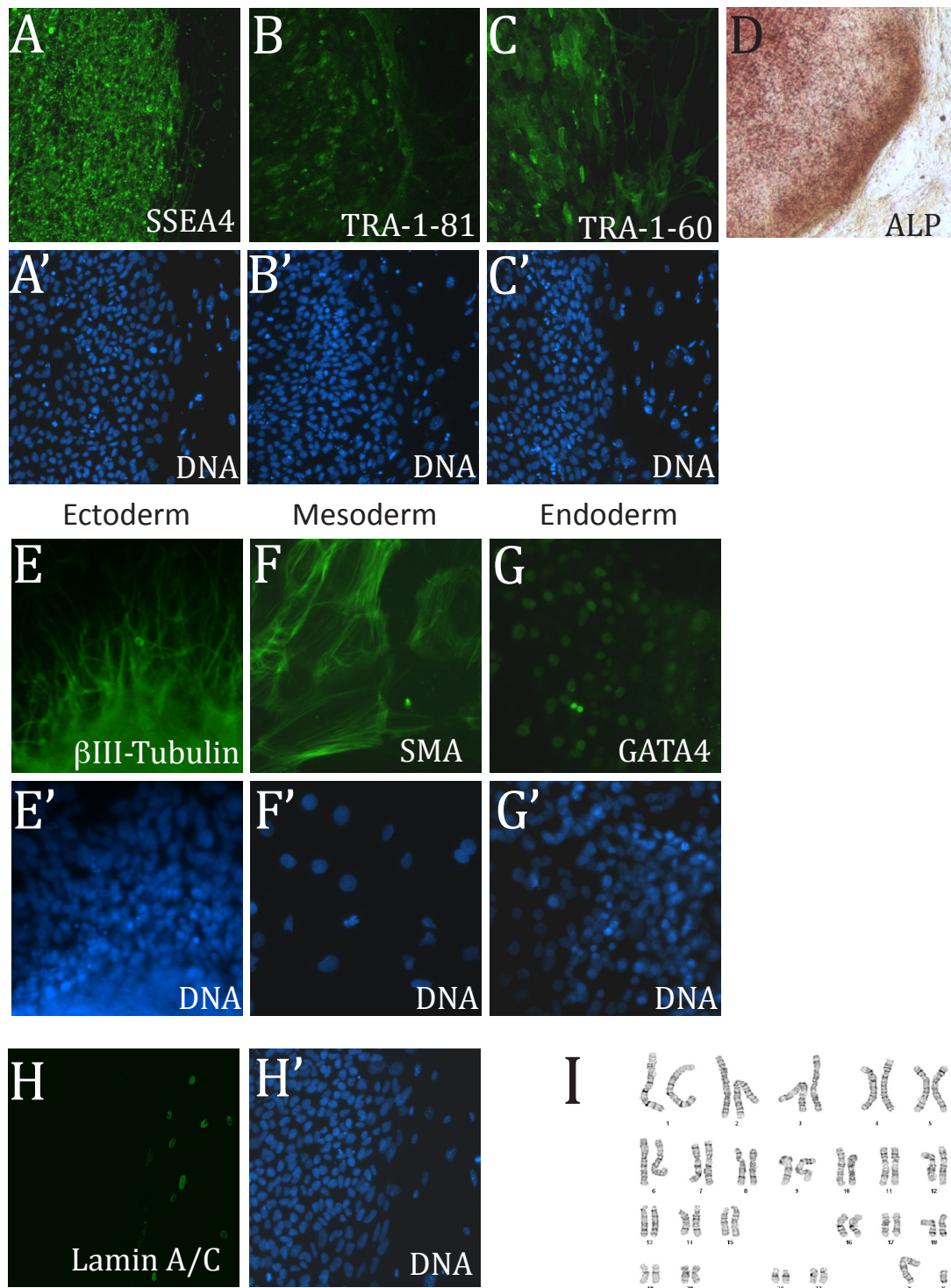


Figure Legend. Hutchinson-Gilford Progeria induced-pluripotent stem cell line HGDFN003 iPS1D exhibits characteristics of pluripotency. (A-D) HGDFN003 iPS1D cells express markers for pluripotency, SSEA4, Tra-1-81, Tra-1-60, and alkaline phosphatase. (E-G') In vitro differentiation of HGDFN003 iPS1D cells shows that it has the capacity to differentiate to cells of all 3-germ layers, β III-tubulin (ectoderm), smooth-muscle actin (mesoderm), Gata4 (endoderm). (H-H') HGDFN003 iPS1D cells exhibit repressed Lamin A/C expression in undifferentiated cells but detectable expression in differentiated cells at the border of the colony. (I) The HGDFN003 iPS1D cell line exhibits normal karyotype.

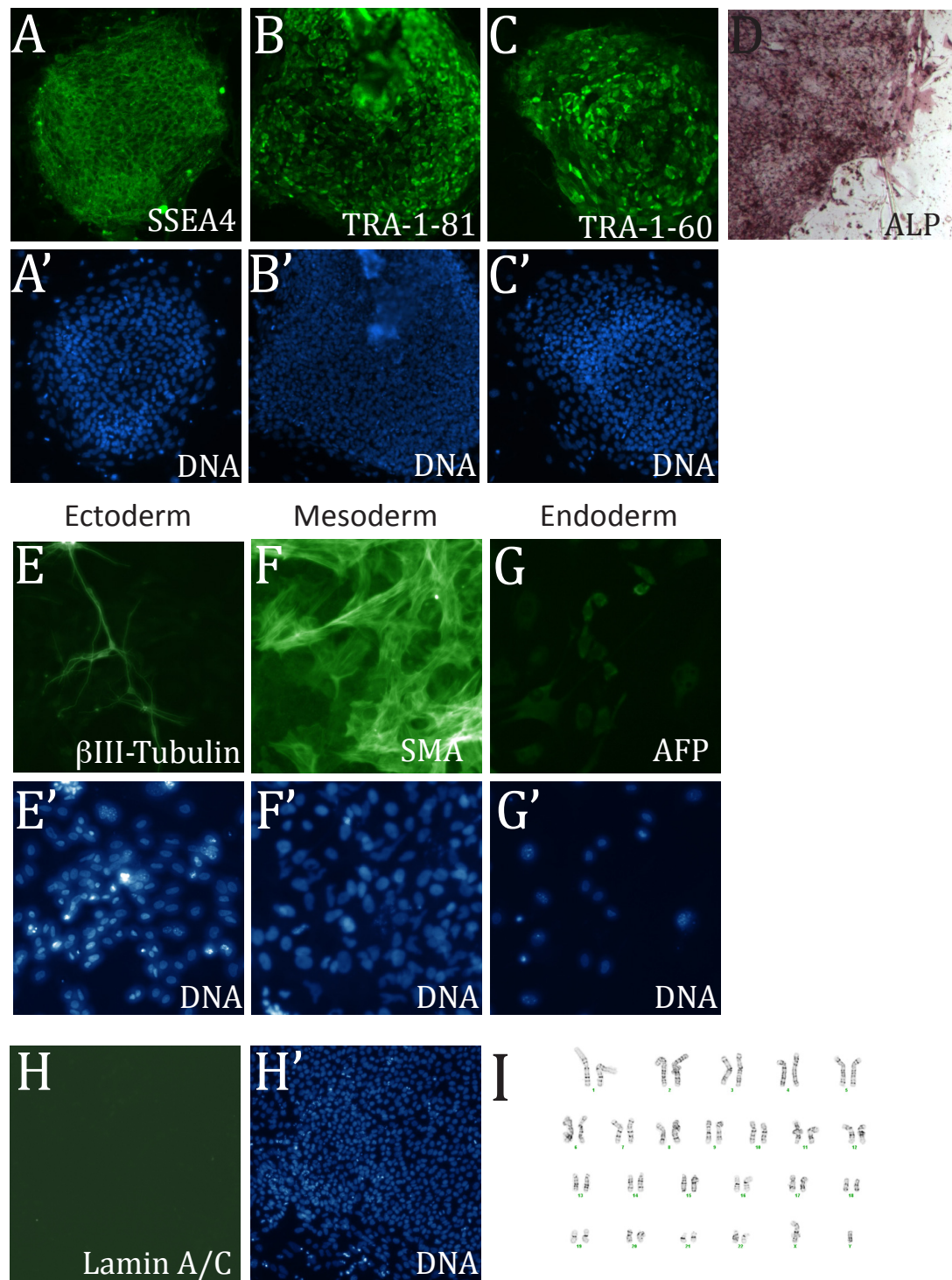


Figure Legend. Hutchinson-Gilford Progeria induced-pluripotent stem cell line HGADFN167 iPS1J exhibits characteristics of pluripotency. (A-D) HGADFN167 iPS1J cells express markers for pluripotency, SSEA4, Tra-1-81, Tra-1-60, and alkaline phosphatase. (E-G') In vitro differentiation of HGADFN167 iPS1J cells shows that it has the capacity to differentiate to cells of all 3-germ layers, β III-tubulin (ectoderm), smooth-muscle actin (mesoderm), AFP (endoderm). (H-H') HGADFN167 iPS1J cells exhibit repressed Lamin A/C expression in undifferentiated cells but detectable expression in differentiated cells at the border of the colony. (I) Karyotypic analysis of HGADFN167 iPS1J revealed normal karyotype (95%), with a single cell exhibiting trisomy 20.

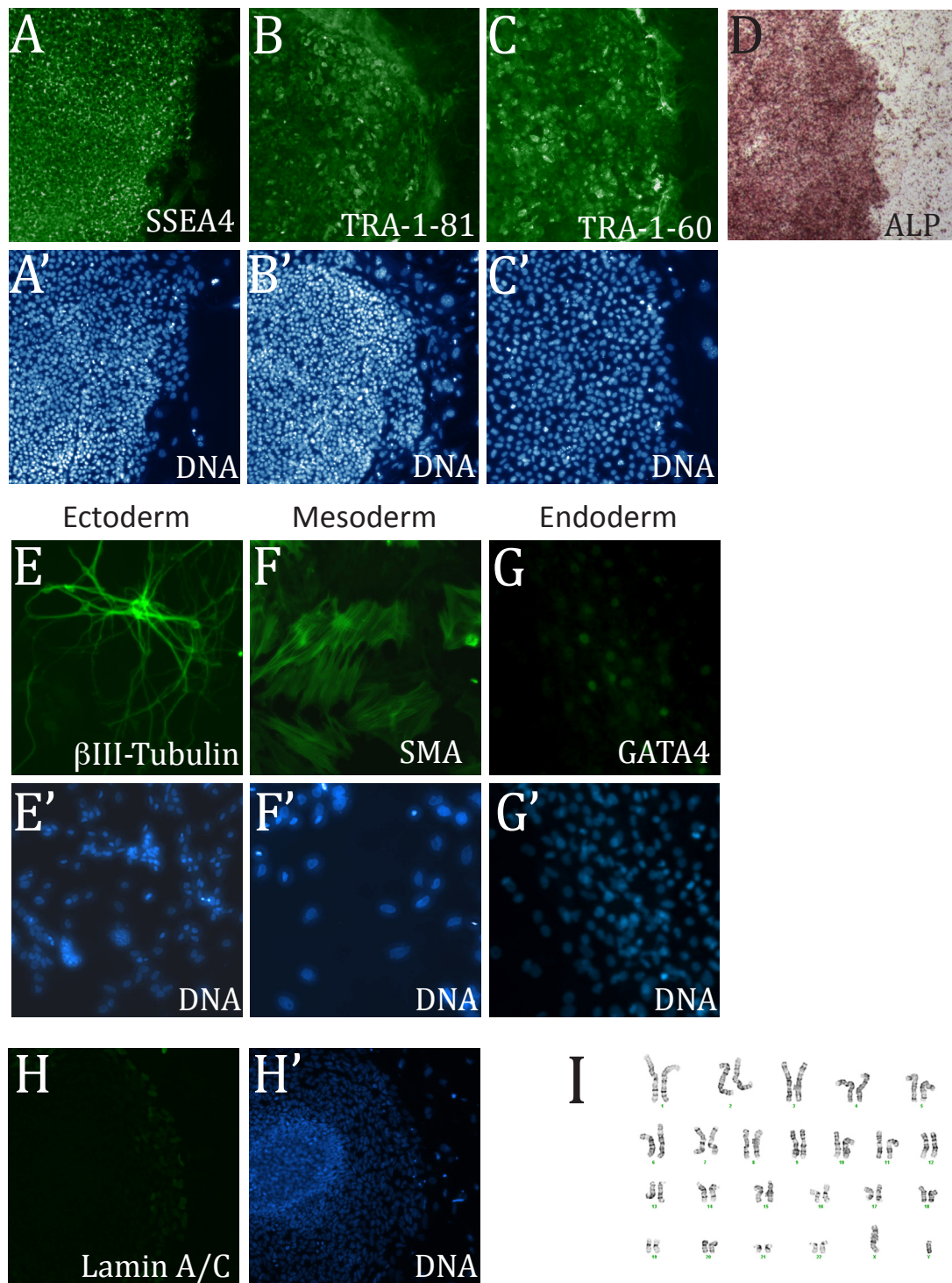


Figure Legend. Hutchinson-Gilford Progeria induced-pluripotent stem cell line HGADFN167 iPS1Q exhibits characteristics of pluripotency. (A-D) HGADFN167 iPS1Q cells express markers for pluripotency, SSEA4, Tra-1-81, Tra-1-60, and alkaline phosphatase. (E-G') In vitro differentiation of HGADFN167 iPS1Q cells shows that it has the capacity to differentiate to cells of all 3-germ layers, β III-tubulin (ectoderm), smooth-muscle actin (mesoderm), Gata4 (endoderm). (H-H') HGADFN167 iPS1Q cells exhibit repressed Lamin A/C expression in undifferentiated cells but detectable expression in differentiated cells at the border of the colony. (I) The HGADFN167 iPS1Q cell line exhibits normal karyotype.

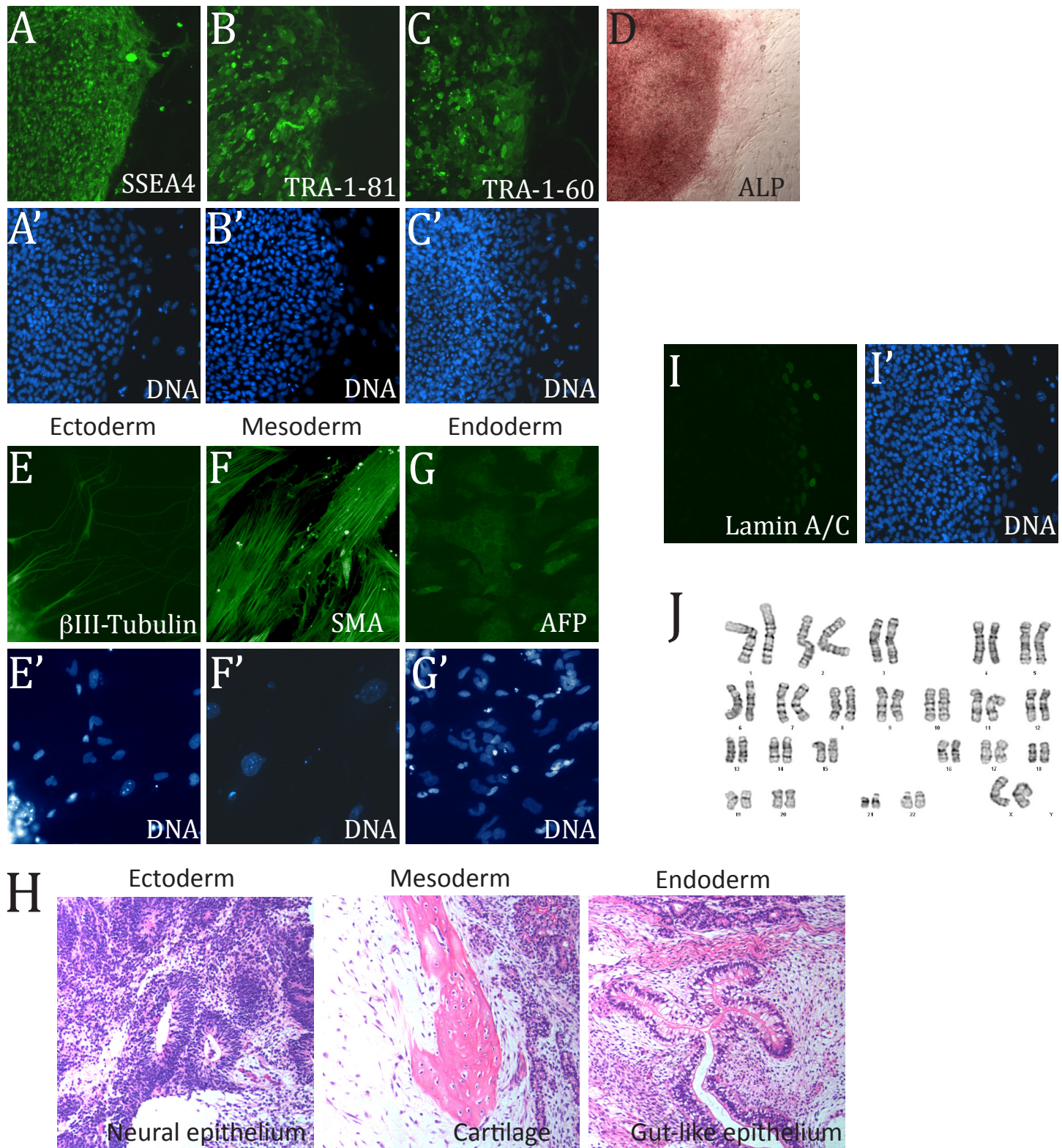


Figure Legend. Hutchinson-Gilford Progeria induced-pluripotent stem cell line HGMDFN090 iPS1B exhibits characteristics of pluripotency. (A-D) HGMDFN090 iPS1B cells express markers for pluripotency, SSEA4, Tra-1-81, Tra-1-60, and alkaline phosphatase. (E-G') In vitro differentiation of HGADFN003 iPS1B cells shows that it has the capacity to differentiate to cells of all 3-germ layers, β III-tubulin (ectoderm), smooth-muscle actin (mesoderm), AFP (endoderm). (H) In vivo differentiation of HGMDFN090 iPS1B cells shows that it has the capacity to differentiate to tissues of all 3-germ layers, neural epithelium (ectoderm), cartilage (mesoderm), Gut-like epithelium (endoderm) (I-I') HGMDFN090 iPS1B cells exhibit repressed Lamin A/C expression in undifferentiated cells but detectable expression in differentiated cells at the border of the colony. (J) The HGMDFN090 iPS1B cell line exhibits normal karyotype.

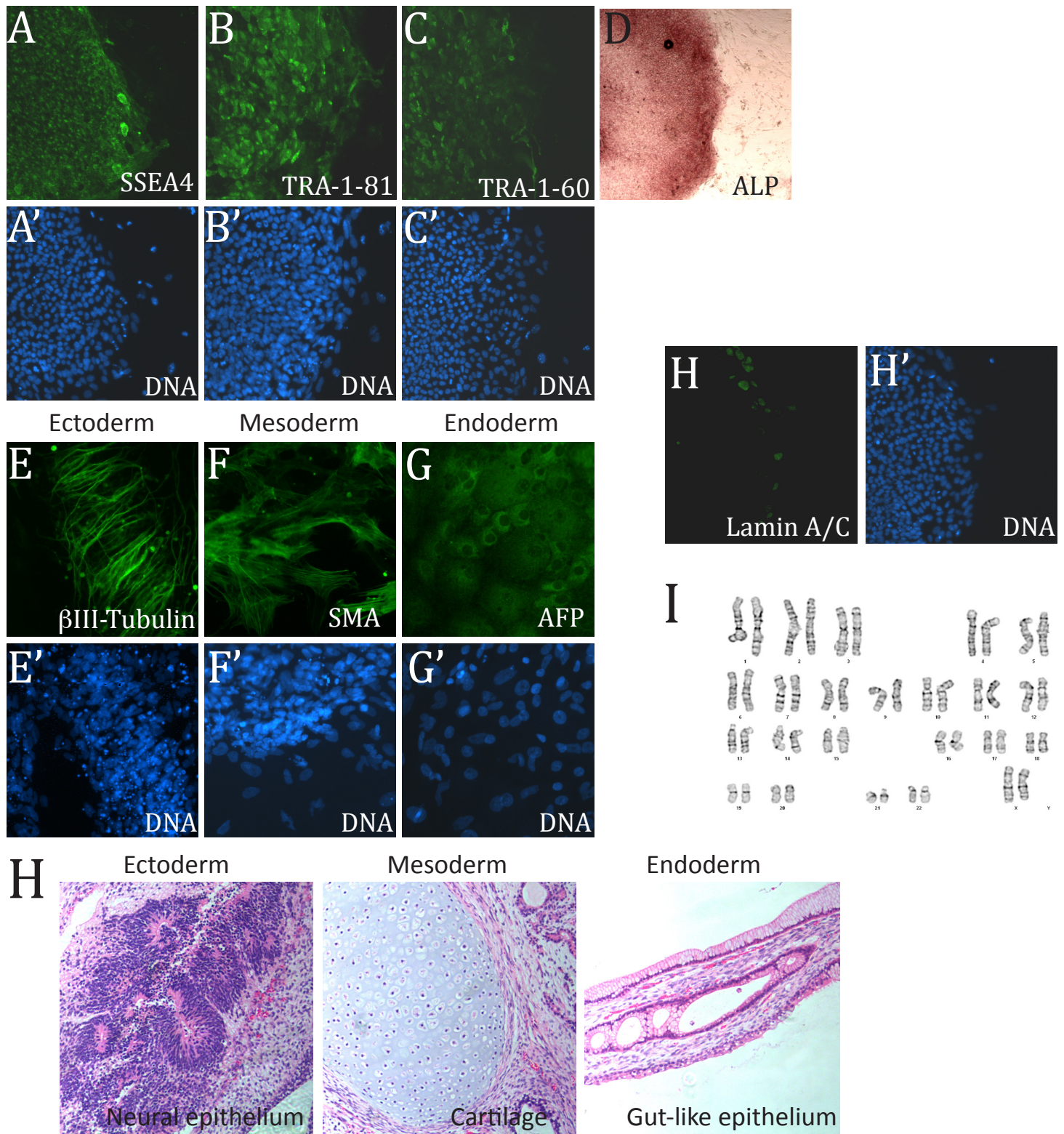


Figure Legend. Hutchinson-Gilford Progeria induced-pluripotent stem cell line HGMDFN090 iPS1C exhibits characteristics of pluripotency. (A-D) HGMDFN090 iPS1C cells express markers for pluripotency, SSEA4, Tra-1-81, Tra-1-60, and alkaline phosphatase. (E-G') In vitro differentiation of HGADFN003 iPS1C cells shows that it has the capacity to differentiate to cells of all 3-germ layers, βIII-tubulin (ectoderm), smooth-muscle actin (mesoderm), AFP (endoderm). (H) In vivo differentiation of HGMDFN090 iPS1C cells shows that it has the capacity to differentiate to tissues of all 3-germ layers, neural epithelium (ectoderm), cartilage (mesoderm), Gut-like epithelium (endoderm) (I-I') HGMDFN090 iPS1C cells exhibit repressed Lamin A/C expression in undifferentiated cells but detectable expression in differentiated cells at the border of the colony. (J) The HGMDFN090 iPS1C cell line exhibits normal karyotype.

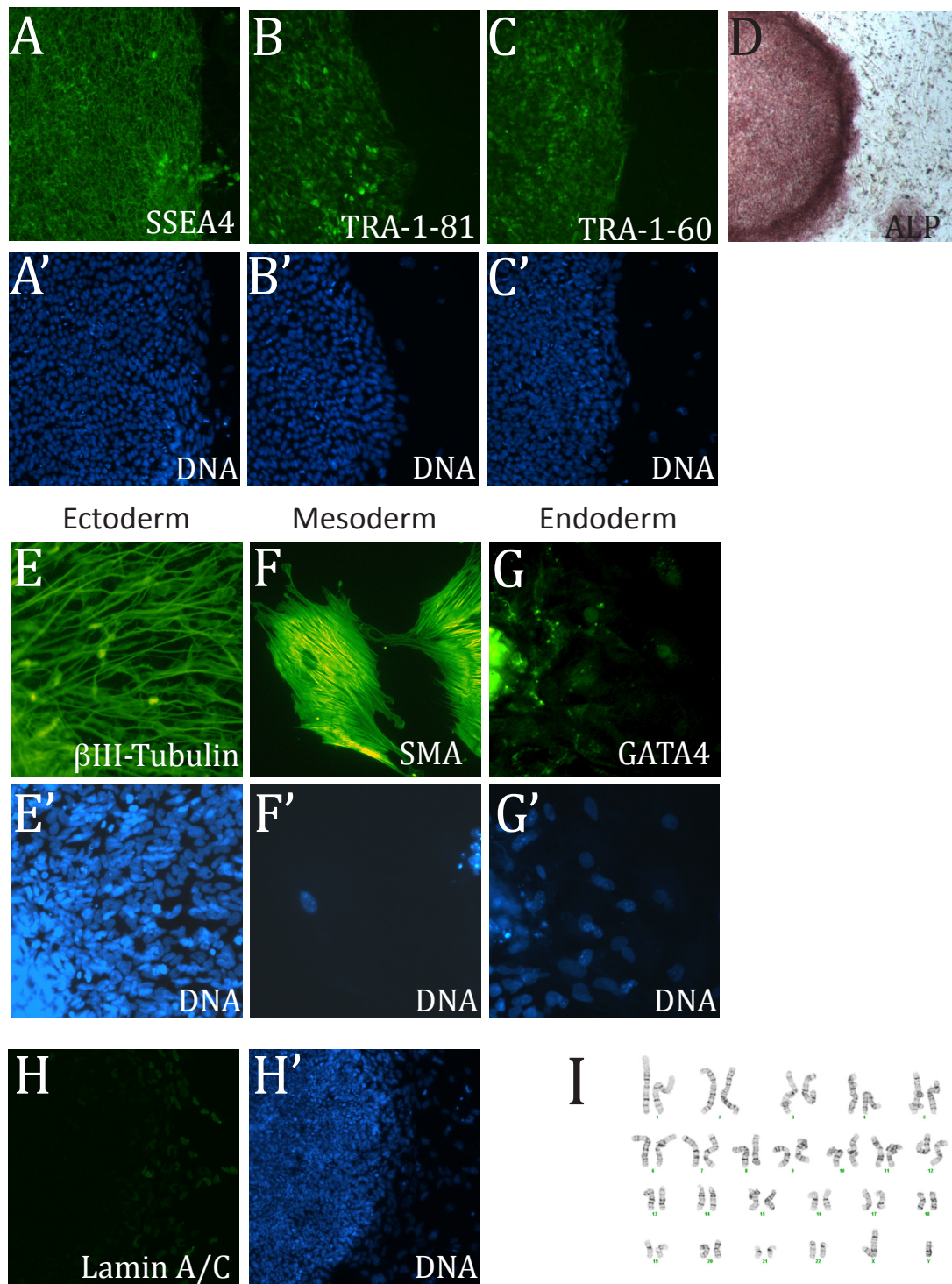


Figure Legend. Hutchinson-Gilford Progeria induced-pluripotent stem cell line HGFDNF168 iPS1D2 exhibits characteristics of pluripotency. (A-D) HGFDNF168 iPS1D2 cells express markers for pluripotency, SSEA4, Tra-1-81, Tra-1-60, and alkaline phosphatase. (E-G') In vitro differentiation of HGFDNF168 iPS1D2 cells shows that it has the capacity to differentiate to cells of all 3-germ layers, βIII-tubulin (ectoderm), smooth-muscle actin (mesoderm), Gata4 (endoderm). (H-H') HGFDNF168 iPS1D2 cells exhibit repressed Lamin A/C expression in undifferentiated cells but detectable expression in differentiated cells at the border of the colony. (I) The HGFDNF168 iPS1D2 cell line exhibits normal karyotype.

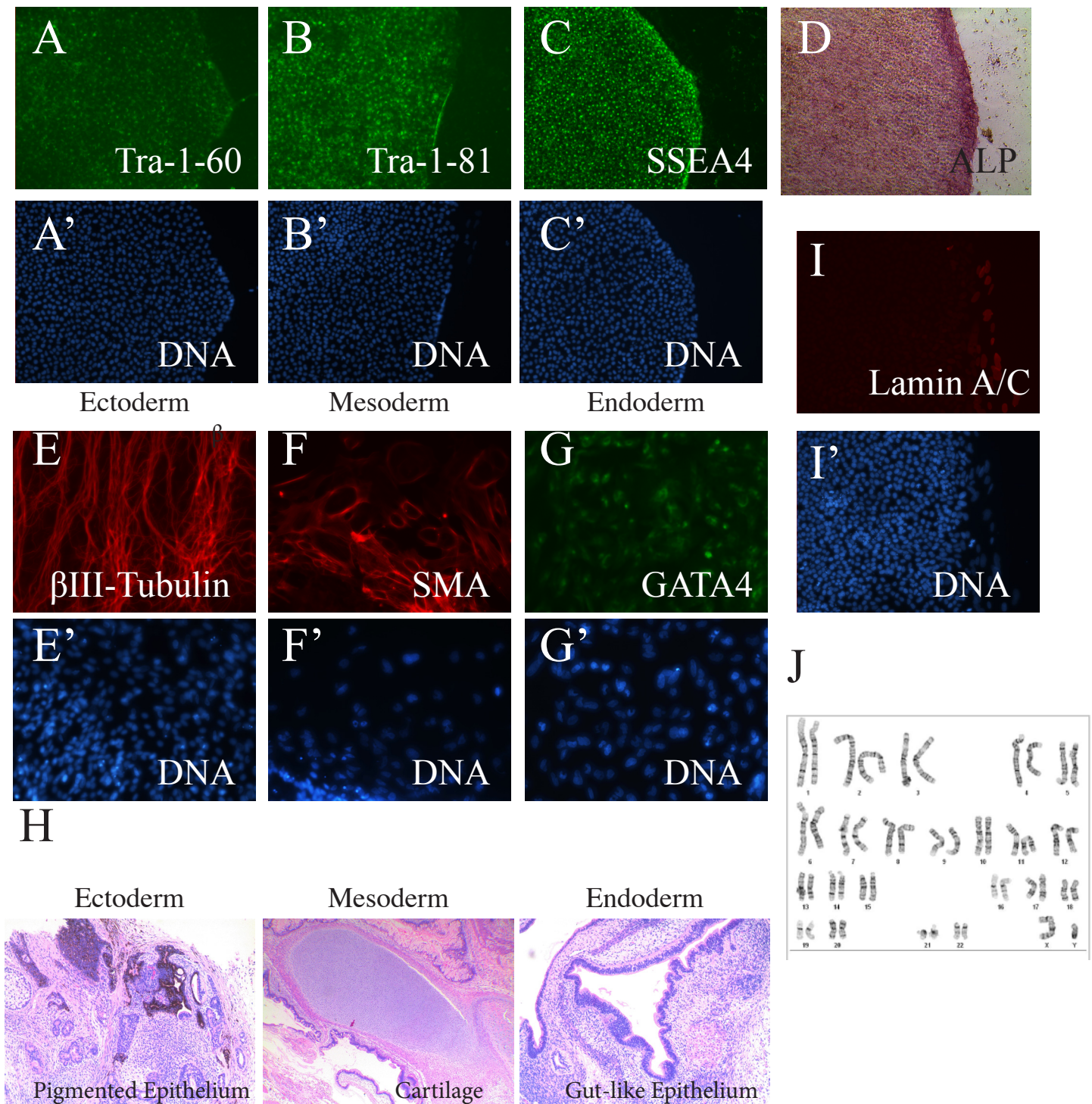


Figure Legend. Hutchinson-Gilford Progeria induced-pluripotent stem cell line HGADFN168 iPS1P exhibits characteristics of pluripotency. (A-D) HGADFN168 iPS1P cells express markers for pluripotency, Tra-1-6-, Tra-1-81, SSEA4, and alkaline phosphatase. (E-G') In vitro differentiation of HGADFN168 iPS1P cells shows that it has the capacity to differentiate to cells of all 3-germ layers, bIII-tubulin (ectoderm), smooth-muscle actin (mesoderm), Gata4 (endoderm). (H) In vivo differentiation of HGADFN168 iPS1P cells shows that it has the capacity to differentiate to tissues of all 3-germ layers, pigmented epithelium (ectoderm), cartilage (mesoderm), Gut-like epithelium (endoderm) (I-I') HGADFN168 iPS1P cells exhibit repressed Lamin A/C expression in undifferentiated cells but detectable expression in differentiated cells at the border of the colony. (J) The HGADFN168 iPS1P cell line exhibits normal karyotype.

The Progeria Research Foundation
and
Ottawa Hospital Research Institute
Induced Pluripotent Stem Cell Program

The Progeria Research Foundation (PRF) is pleased to provide Progeria researchers with induced pluripotent stem cell (iPSC) lines from patients with HGPS and control donors. The iPSC lines were created and are maintained, stored and shipped by our collaborator, The Human Pluripotent Stem Cell Facility at Ottawa Hospital Research Institute (OHRI), directed by Dr William Stanford.

iPSC lines are valuable resources for Progeria research, and PRF is committed to helping researchers who wish to utilize these cell lines for their research.

We have found that researchers who have not worked with iPSC lines previously have found it challenging to establish these cell lines in their labs. Therefore, we ask that you carefully read the following.

PRF and OHRI offer the following guidelines for researchers before ordering iPSC lines.

1. iPSC lines are among the most difficult cell lines to work with. Each cell line behaves differently, and it is important that the individual actually working with the cells has experience working with several different iPSC lines.
2. If you have not worked with iPSC lines before, please find an experienced individual at your institution who can advise you on proper techniques.
3. It is essential that you follow OHRI's protocol exactly when thawing and expanding the cells. The protocol can be found here: <https://www.progeriaresearch.org/cell-tissue-bank>.
4. The Human Pluripotent Stem Cell Facility at OHRI offers virtual one-on-one training on basics of iPSC culture techniques specific to the Progeria iPSC lines. Training options and format are flexible depending on the scientists' level of experience. For more information, please email hpscf@ohri.ca.
5. We recommend that you order 2 vials of each line and thaw one vial at a time. If you are unable to grow the first vial, you will have a second vial on hand without having to incur additional shipping costs for a replacement vial.
6. Some of the lines are easier to maintain undifferentiated than others. We recommend that you begin with the HGPS line HGADFN167 iPSC 1J and the control line HGFDFN168 iPSC 1D2. HGADFN003 iPSCs are difficult to grow and should only be used after you have gained experience with the other lines.

7. We recommend that you expand and freeze vials of the undifferentiated cells before beginning differentiations. Pluripotent stem cells are self-renewing. After differentiation, self-renewal capacity is lost and with it viability through freeze thaws.
8. We have found that the longer cells spend in transit, the more difficult it is to recover the cells even when kept on dry ice.
9. It is the responsibility of the recipient to contact brokers, customs officials and/or the shipping carrier to ensure the package is not delayed in customs and to make necessary arrangements in the event of any delays. This includes paying any additional fees for brokers or the shipping carrier to add dry ice to a package experiencing delays. For shipments outside of Canada or the US, OHRI recommends World Courier as they assist with the delivery including customs issues and having dry ice added as needed. Other couriers can also be used but be aware you are responsible for following the shipment, resolving customs issues and arranging to have dry ice added if necessary.
10. Should you experience issues with the cell lines, please contact Wendy Norris (wnorris@brownhealth.org) at PRF's Cell and Tissue Bank, and she will put you in contact with OHRI. Please be certain to include Wendy in any correspondence with OHRI.