NEUROMICS

Human iPSC - Motor Neurons (ALS Patient, Sporadic)

Catalog #: IPS014

Cell #: >1.0x10^6 cells

Storage: Liquid Nitrogen until ready for culture.
While Culturing keep in 37°C CO_2 incubator

Product Format: Frozen Vial

GENERAL INFORMATION

These human spinal motor neurons (MNs) are derived from induced Pluripotent Stem Cells (iPSCs) reprogrammed from a donor with a confirmed diagnosis of sporadic Amyotrophic Lateral Sclerosis (sALS). The differentiation process utilizes a directed, floor-plate based protocol to generate a highly pure population of cells expressing key motor neuron markers.

This product provides a physiologically relevant in vitro model of sALS, which accounts for ~90% of all ALS cases. These neurons recapitulate key disease phenotypes, including increased susceptibility to cellular stress, altered mitochondrial function, and potential TDP-43 proteinopathy. They are an essential tool for investigating the mechanisms of motor neuron degeneration, screening neuroprotective compounds, and developing personalized medicine approaches for ALS.

*Differentiation efficiency may vary due to the inherent biological variability of the donor phenotype.

These cells exhibit biological characteristics associated with the donor phenotype, which may impact their growth, differentiation, and metabolic behavior compared to cells from health donors. It is the user's responsibility to determine the suitability of this product for their specific application.

Product is for Research use only.

Frozen Vials are shipped in a Dry Ice Package.

KEY FEATURES & BENEFITS

- **Authentic Disease Model:** Captures the complex genetic and pathological background of sporadic ALS in a human-relevant system.
- **High Purity:** Differentiated population is highly enriched for spinal motor neurons, expressing characteristic markers.
- **Key Disease Phenotypes:** Exhibits hallmarks of ALS pathology, providing a robust platform for mechanistic and therapeutic studies.
- **Ready-to-Use:** Cryopreserved at a post-mitotic stage, ready for functional assays upon recovery and maturation.
- **Comprehensive Characterization:** Each lot is validated for motor neuron identity, purity, and viability.

APPLICATIONS

- **ALS Disease Modeling:** Study mechanisms of motor neuron vulnerability, axonal degeneration, and glial cell interactions in co-culture.
- **Neurotoxicity & Drug Screening:** Identify compounds that protect against stress-induced death or modulate excitability.

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- **Electrophysiology:** Characterize electrical properties and synaptic activity using patch-clamp or multi-electrode array (MEA) systems.
- **Biomarker Discovery:** Investigate protein aggregation (TDP-43), RNA metabolism, and secretory profiles.
- **Gene Expression Analysis:** Profile transcriptomic changes in a patient-specific background.

QUALITY CONTROL SPECIFICATIONS

Each lot is tested to meet the following release criteria:

Parameter	Specification	Test Method
Viability (Post-Thaw)	≥ 70%	Trypan Blue Exclusion
Cell Yield per Vial	≥ 1 Million Cells	Cell Count
Purity (MN Marker Expression)	≥ 70% HB9+/ChAT+ or ISL1+/Tuj1+	Immunocytochemistry (ICC)
Neuronal Marker Expression	Positive: β-III-Tubulin (Tuj1), MAP2 Motor Neuron Markers: ISL1, HB9 (MNX1), ChAT (functional)	ICC/Flow Cytometry
Pluripotency Marker Absence	Negative: Oct3/4, Nanog	ICC
Microbiological Sterility	No Growth	USP <71>
Mycoplasma	Negative	PCR Method

TYPICAL DONOR DEMOGRAPHICS & CLINICAL HISTORY

Donors are carefully selected and characterized. A typical profile includes:

- **Diagnosis:** Sporadic Amyotrophic Lateral Sclerosis (sALS).
- **Genetics:** No known mutations in SOD1, C9orf72, TARDBP, or FUS (specific details per lot).
- **Age at Collection:** 18 - 65 years
- **Sex:** Male and Female donors available
- **Source Cell Type:** Dermal Fibroblasts or Peripheral Blood Mononuclear Cells (PBMCs).

HANDLING OF ARRIVING CELLS

1. Check all containers for leakage or breakage.

2. Remove the frozen cells from the dry ice packaging and immediately place the cells into a -80°C freezer for short-term storage or liquid nitrogen tank for long-term storage.

3. To ensure the highest level of viability, thaw the vial and initiate the culture as soon as possible upon receipt. If upon arrival, continued storage of the frozen culture is necessary, it should be stored in liquid nitrogen vapor phase and not at -70°C. Storage at -70°C will result in loss of viability.

4. When handling cell cultures, the application of stringent aseptic techniques is imperative. Maintaining a contamination-free environment is crucial for preserving the integrity and reliability of experimental results. This involves not only cleanliness but also the use of sterile tools, media, and working environments to prevent contamination from airborne particles, microbial agents, and other sources.

MEDIUM

We recommend Motor Neuron Maintenance Medium Kit (cat. HNM013) for these cells. The kit contains ingredients to support cell growth.

PROTOCOL FOR THAWING THE CELLS

Note: If you have any questions or need clarification regarding the protocol for culturing these cells, please reach out to Dr. Jensen Auguste at (978) 608-1766 with your questions before beginning.

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1. Pre-coat culture vessels overnight at 4°C or 1-2 hours at 37°C with AlphaBioCoat Solution (cat. AC001), followed by rinses with PBS before plating.

2. Quick Thaw: Remove the vial from liquid nitrogen and immediately place in a 37°C water bath. Gently agitate until only a small ice crystal remains (≈1-2 minutes).

3. Decontaminate: Spray the vial with 70% ethanol before transferring to a biosafety cabinet.

4. Transfer and Dilute: Gently transfer the cell suspension to a 15 mL conical tube. Slowly add 5-9 mL of pre-warmed IPSC Neuron Plating Medium (cat. HNM003) drop-wise to dilute the cryoprotectant (DMSO).

5. Centrifuge: Spin at 200 x g for 5 minutes.

6. Resuspend and Plate: Aspirate the supernatant and gently resuspend the cell pellet in fresh, pre-warmed IPSC Neuron Plating Medium.

    a. Recommended Seeding Density: 50,000 - 100,000 cells/cm².

7. Incubate: Place the culture vessel in a humidified 37°C incubator with 5% CO_2.

8. Medium Change: After 24-48 hours, carefully replace 50% of the medium with fresh Motor Neuron Maintenance Medium. Thereafter, perform a 50% medium change every 2-3 days.

MAINTENANCE & MATURATION

- **Maturation:** Cells are cryopreserved at an early post-mitotic stage. Allow 7-14 days in culture for full maturation, including robust neurite outgrowth and expression of synaptic markers.
- **Handling:** These are delicate, post-mitotic cells. Avoid harsh pipetting and frequent, full medium changes to prevent shear stress.
- **Morphology:** Expect to see phase-bright cell bodies with extensive, branching neurite networks.

NOTICE ON CO-CULTURES

For co-culture with human iPSC-derived astrocytes or microglia (available separately), please contact technical support for optimized protocols to ensure proper cell ratio and medium formulation.

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