Corporate Presentation

January, 2021

Targeting the powerhouse of cells to improve the lives of primary mitochondrial disease patients

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Near pivotal rare disease asset with positive Phase I safety and strong preclinical data

- KL1333, chronic oral treatment of primary mitochondrial diseases
  - Powerful small molecule NAD+ regulator
  - Strong preclinical data in patient cells and animal models
- Pivotal KL1333 phase II/III study planned to start 2H21
  - Recommended by the US FDA based upon existing documentation
  - Orphan drug designation in EU and US
  - No safety issues in ongoing Phase I a/b studies
  - Ongoing Phase 1b PMD patient study with initial results expected in 1Q21
  - Leveraging prior clinical supportive studies
- NV354, an energy replacement therapy for Leigh Syndrome
  - In IND-enabling studies
- Highly attractive USD 30m valuation
  - M&A and venture deals support Mito disease as a significant rare disease opportunity
- Cash position USD 8.6m (Sep 30, 2020)
  - New life science specialist venture investor since 2020

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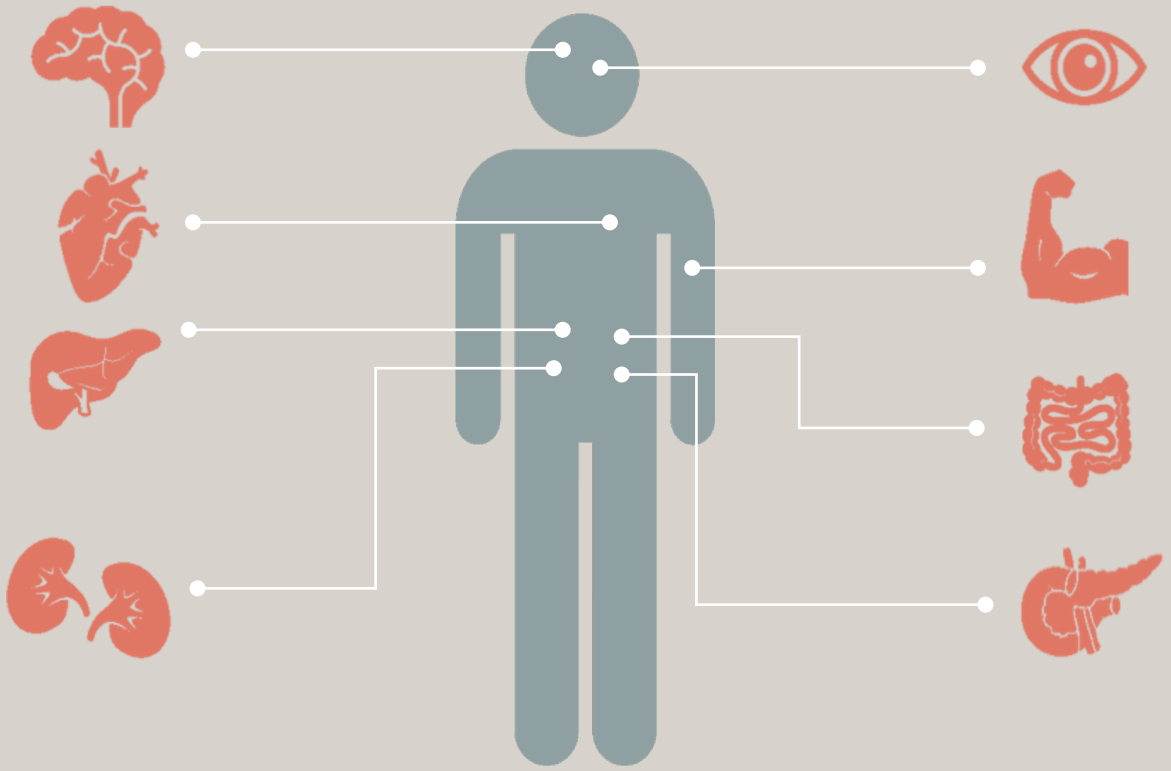
KL1333 for primary mitochondrial diseases



Genetic mutations leading to dysfunctional mitochondria

Devastating diseases with severe symptoms and continuous deterioration

- Focus on MELAS-MIDD and KSS-CPEO patients with chronic fatigue, muscle weakness, and mitochondrial diabetes
- No approved medicines for systemic PMD
- Orphan indications

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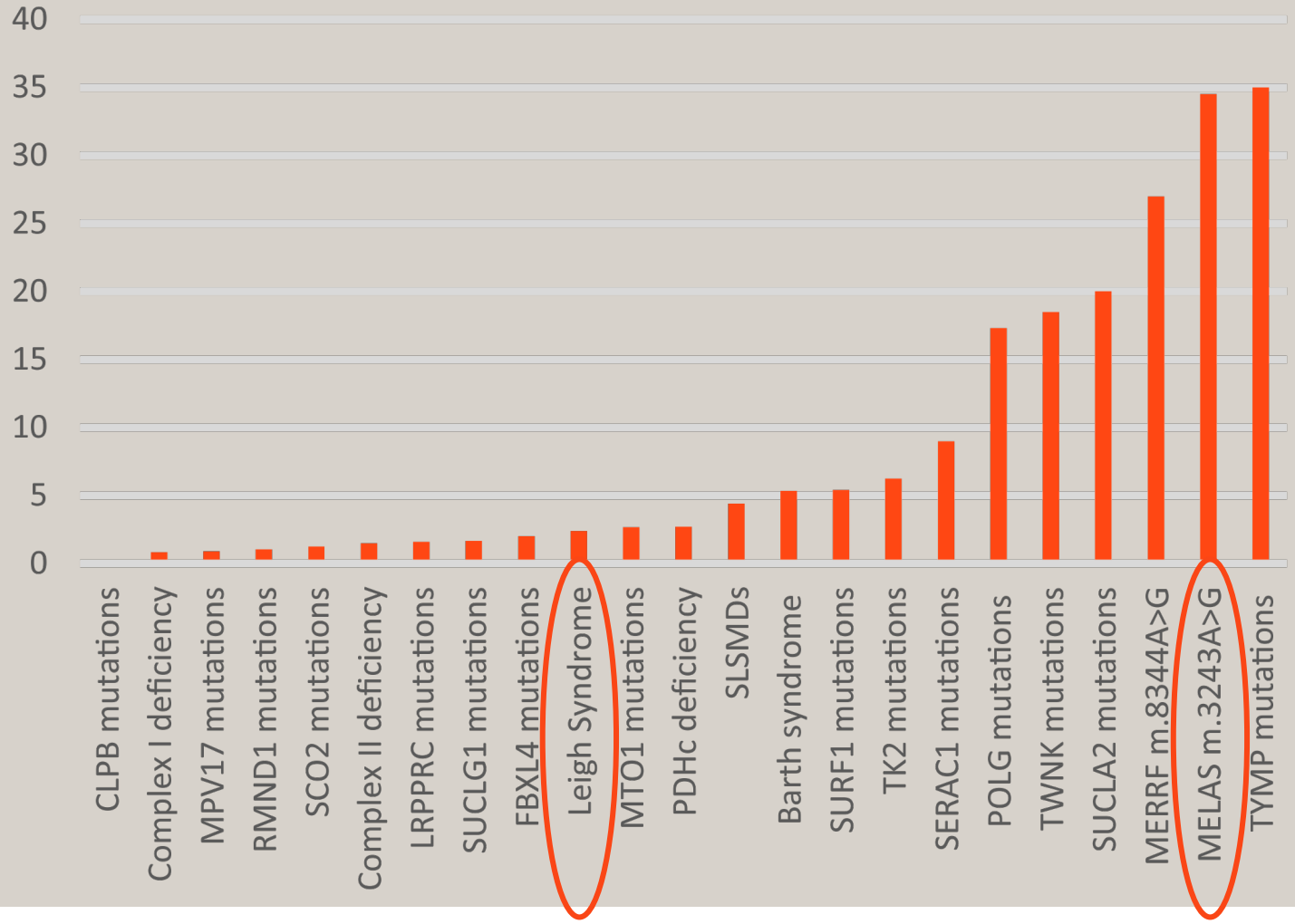
Mito disease is severe & leads to early death



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KL1333

First-in-class NAD+ disease modifying treatment
to improve the lives of patients with mito disease

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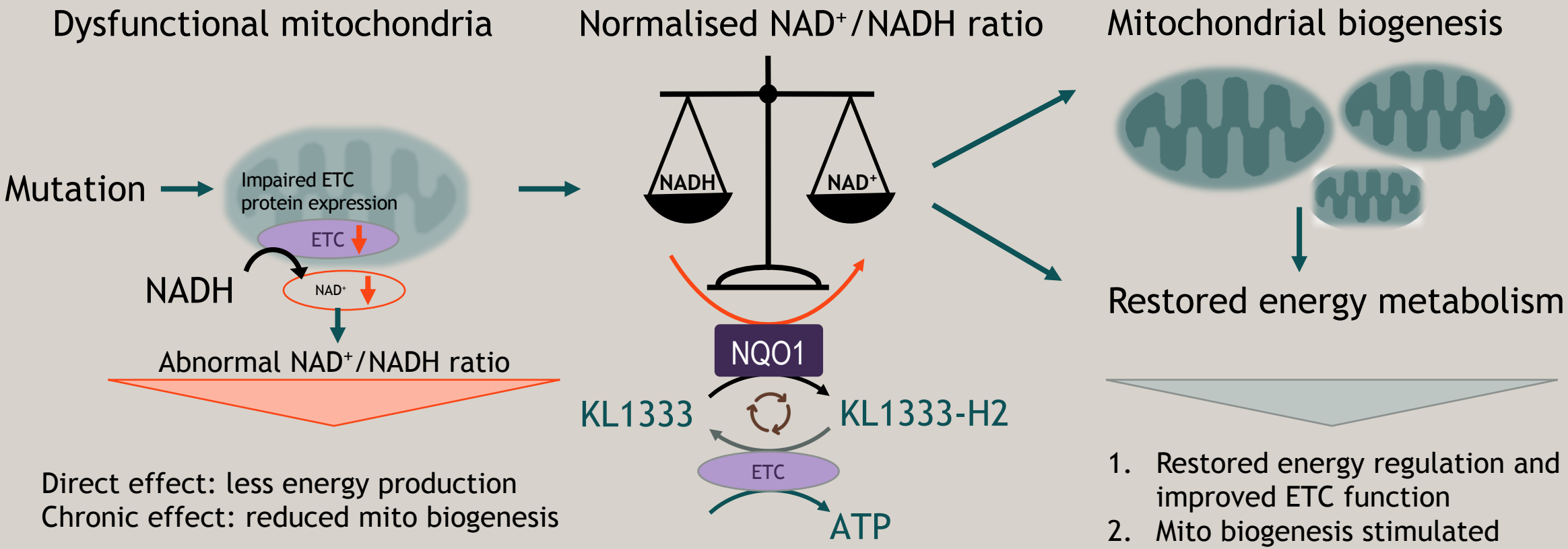
KL1333 corrects underlying pathophysiology of mito disease



Direct effect: less energy production
Chronic effect: reduced mito biogenesis

Overall results: Organ dysfunction, disease deterioration

1. Restored energy regulation and improved ETC function
2. Mito biogenesis stimulated

Overall results: Symptom reduction, disease modification

NAD= **Nicotinamide adenine dinucleotide,** NQO1= NAD(P)H quinone dehydrogenase oxidoreductases 1, ETC=Electron Transport Chain

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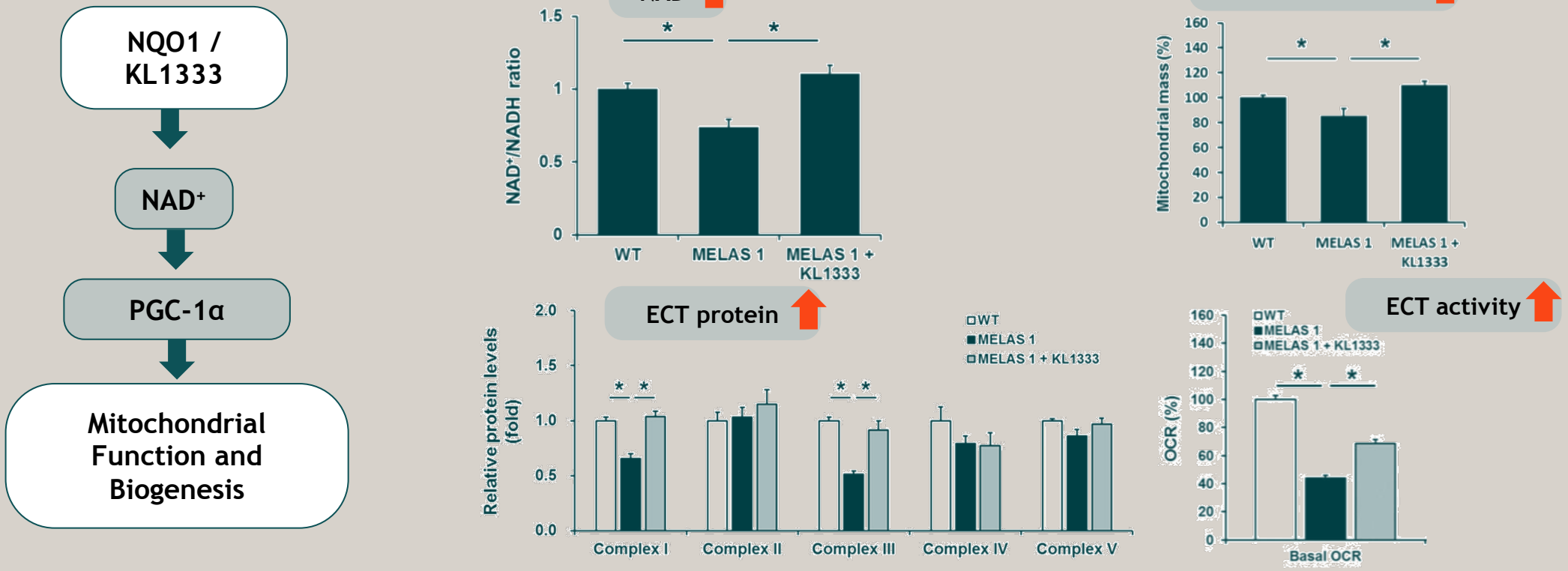
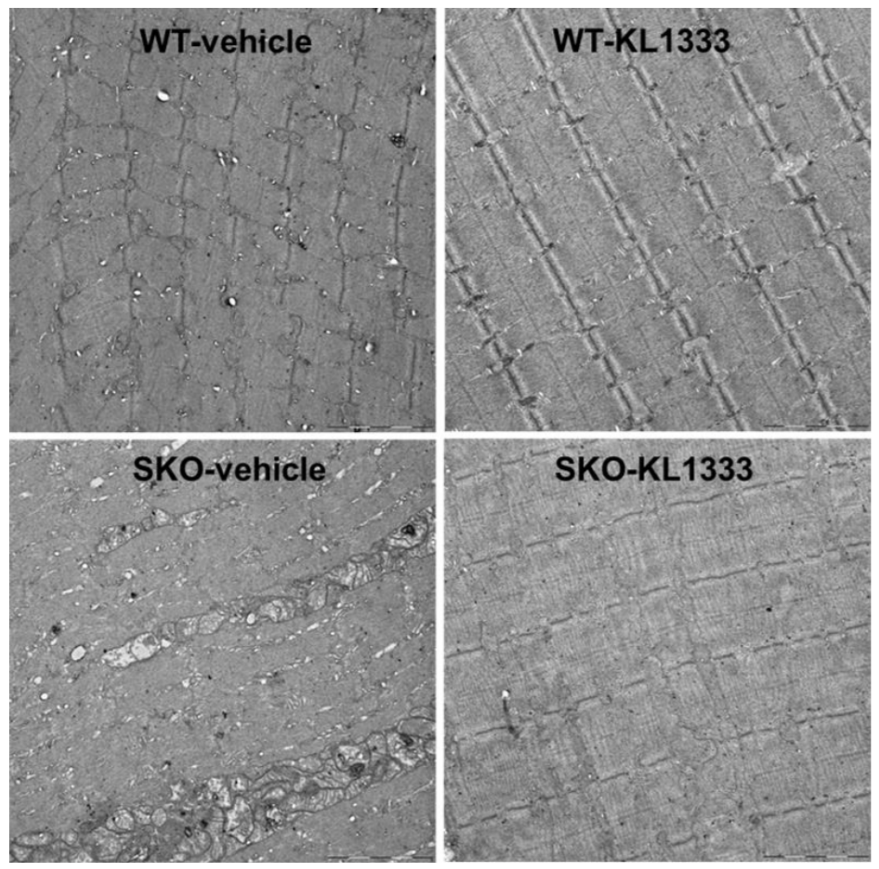
KL1333 increases NAD⁺ and mitochondrial biogenesis in MELAS patient fibroblasts

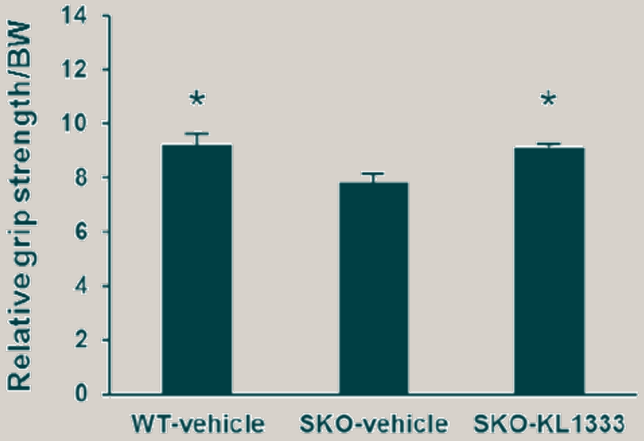


Seo et al. 2018. KL1333, a Novel NAD⁺ Modulator, Improves Energy Metabolism and Mitochondrial Dysfunction in MELAS Fibroblasts (https://www.frontiersin.org/articles/10.3389/fneur.2018.00552/)

KL1333 improves muscle function and histology in an *in vivo* mouse model



- Mito disease model *Crif1* skeletal knock-out (SKO) mice have impaired translation of mtDNA-encoded respiratory chain subunits
  - Similar to m.3243A>G mutation and large mtDNA deletions



KL1333 qd oral dosing for 4 w (n=4-5/group

- **KL1333 significantly increases grip strength and normalizes muscle histology**

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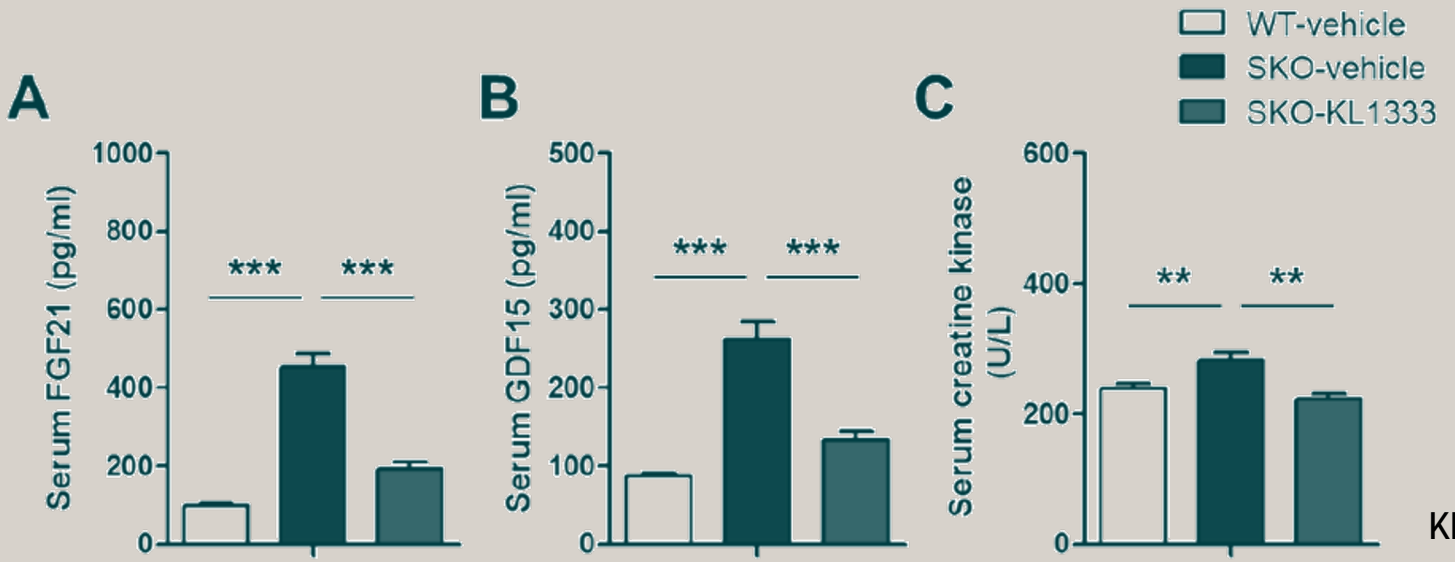
KL1333 improves PMD relevant biomarkers in an *in vivo* mouse model

Mito disease model *Crif1* skeletal knock-out mice



KL1333 qd oral dosing for 4 w (n=4-5/group)

- KL1333 significantly reduces mitochondrial disease biomarkers FGF21 & GDF15, and normalizes muscle injury marker creatine kinase

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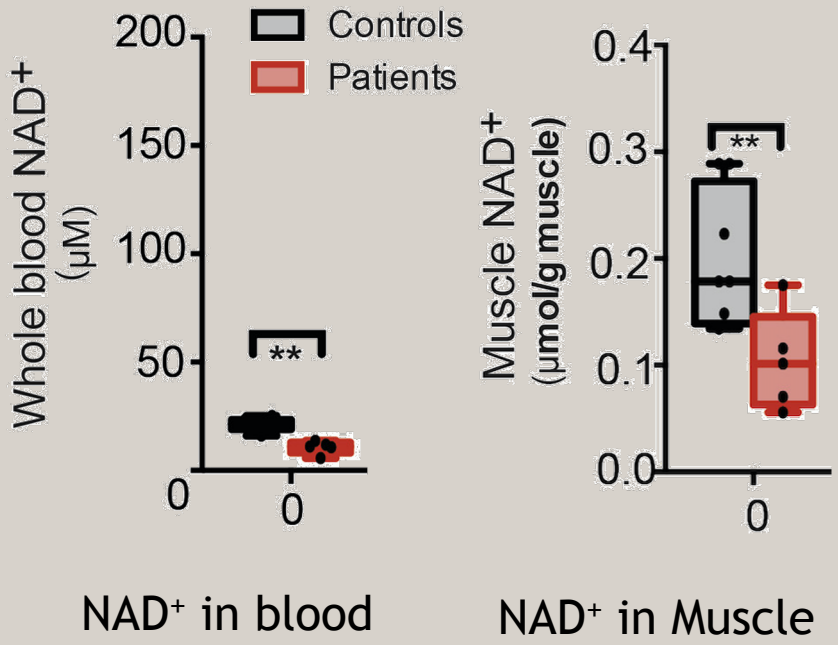
NAD+ levels are reduced in human PMD patients



NAD+ in blood

NAD+ in Muscle

- NAD+ levels were lower in blood and muscle in study of PMD patients
  - Beneficial effects seen by NAD supplement (Niacin)
  - Supports KL1333 concept that NAD+ is central to mito disease

Pirinen et al. Cell Metab. 2020;31(6):1078-1090

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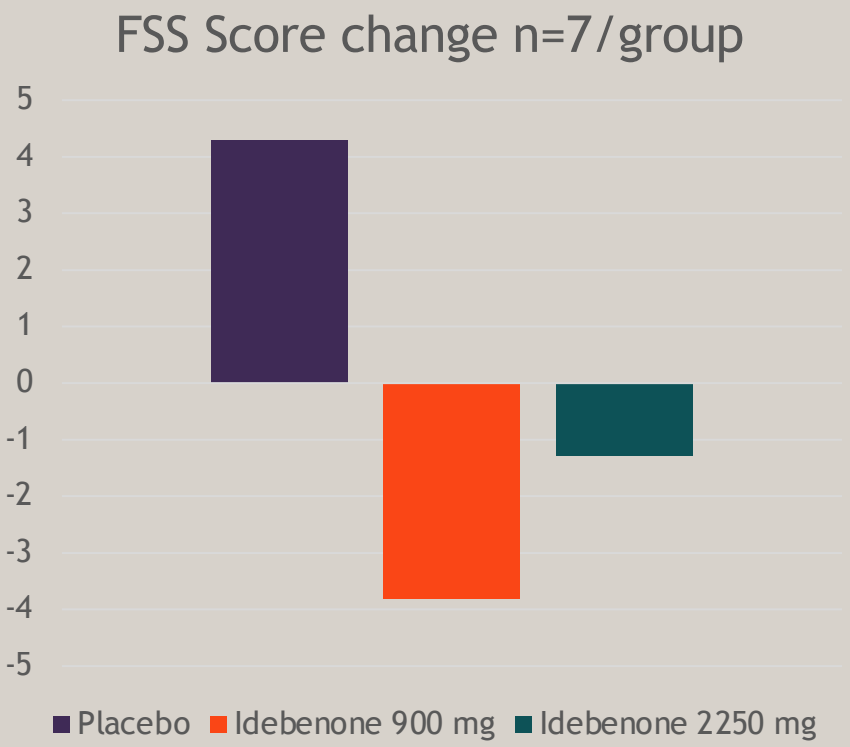
Idebenone has demonstrated a positive effect on fatigue in MELAS patients



- MELAS patient study with idebenone (Columbia Univ. investigator led)
- Mean Change in Score on the Fatigue Severity Scale (FSS) indicated clinically meaningful effect
  - 1-month treatment, n=7 in each group
- Small study of short duration with sub-optimal molecule
- But interesting support for NQO1 mechanism

Hirano et al. ClinicalTrials.gov NCT00887562

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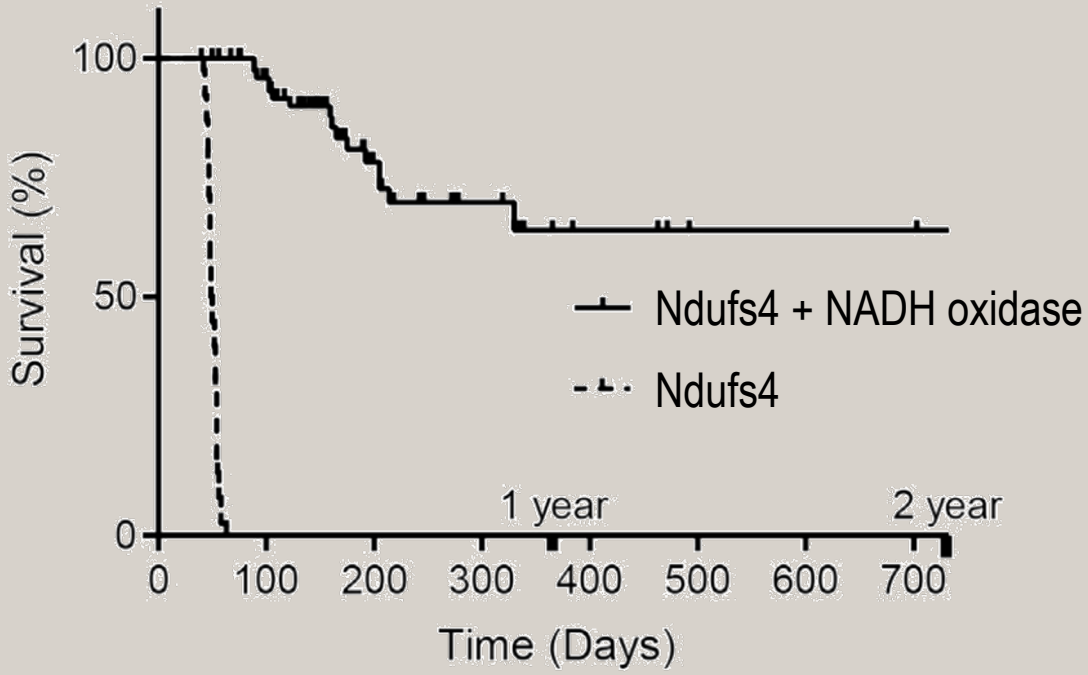
Animal model validation of NAD+/NADH correction in mito disease



- Artificial NADH oxidases raises NAD+ levels and increases lifespan in mitochondrial disease model (Ndufs4)[1]
  - Other studies show cell metabolism rescue[2] and normalization of mito disease biomarkers[3]

[1] McElroy et al. Cell Metab. 2020;32(2):301-308
[2] Titov et al. Science 2016;352(6282):231-5
[3] Goodman et al. Nature 2020;583:122–126

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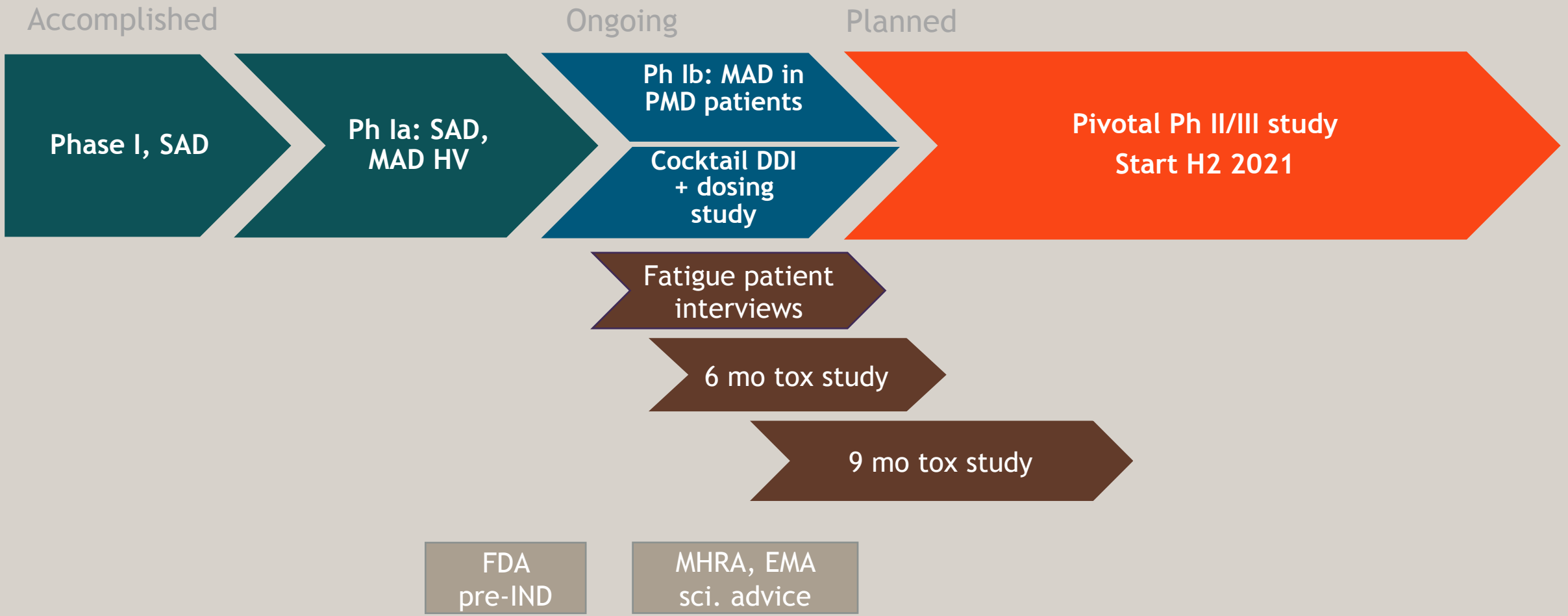
KL1333 development plan overview



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NV354 & pipeline

Targeting energy replacement therapy for Leigh syndrome and developing follow-on compounds in MELAS-MIDD

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NV354 targets high unmet need in Leigh syndrome

Leigh syndrome is a devastating PMD with severe multiorgan deterioration and a life expectancy <five years

- 25:1,000,000 live births
- Aim to develop an energy replacement that modifies disease progression
- The indication could be expanded to include LHON and MELAS
- Clinical development start targeted for end 2021

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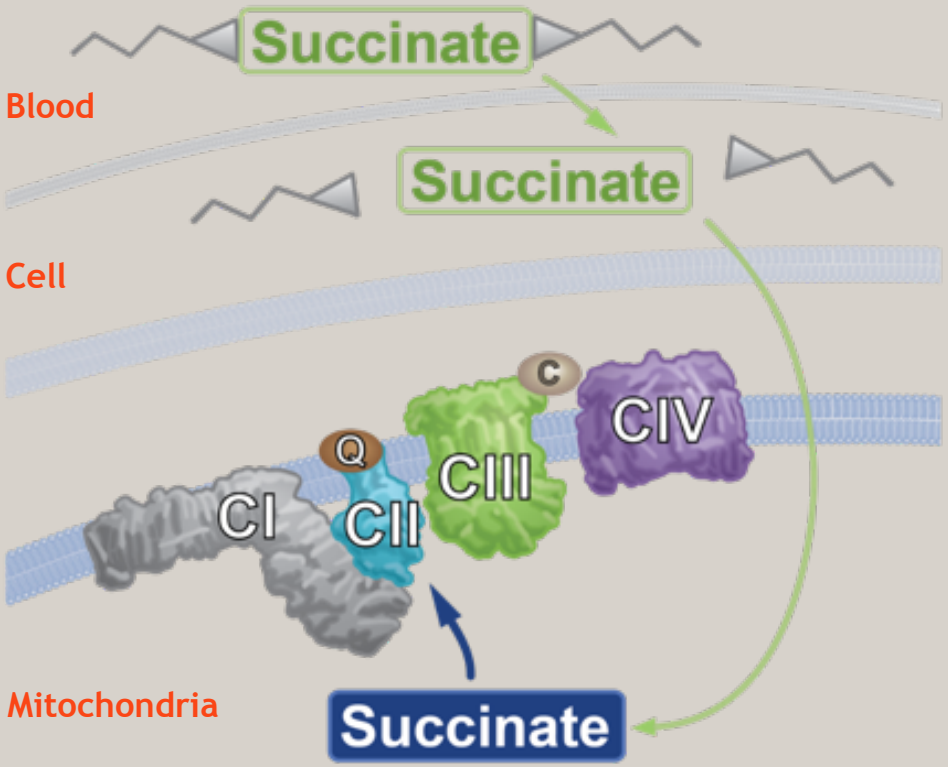
Succinate pro-drug bypasses defective Complex I



- Complex I dysfunction is one of the most common causes of mitochondrial disease
  - Key factor in Leigh Syndrome, MELAS, and LHON
- Disease modifying
  - Protects mitochondria and loss of organ function
  - Prevents complications caused by acute energy crisis
- NV354 in pre-IND phase

Ehinger et al. Nature Communications, 7:12317, August 2016

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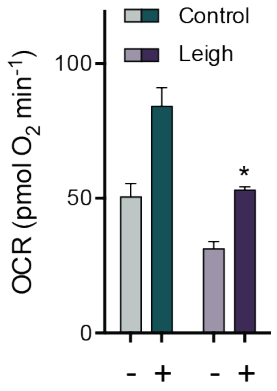
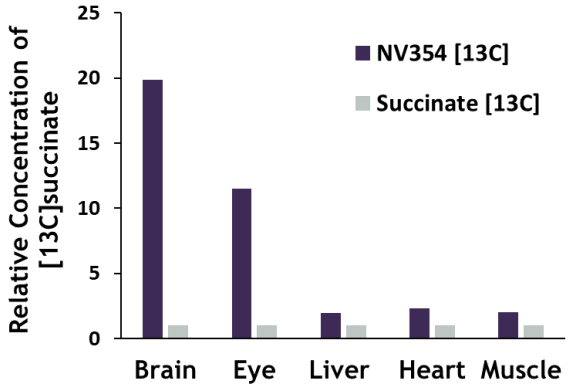
NV354 corrects underlying biochemical dysfunction in Leigh patient cells



Improved mitochondrial function in Leigh patient cells



^{13}C-labelled succinate delivery to organs

- NV354 has optimized drug properties
  - High oral bioavailability
  - High brain distribution
  - Good tolerability in initial toxicology studies
- Bypasses most common mutations in Leigh syndrome and LHON (complex I)
- Normalizes function of mitochondria in cells from Leigh patients

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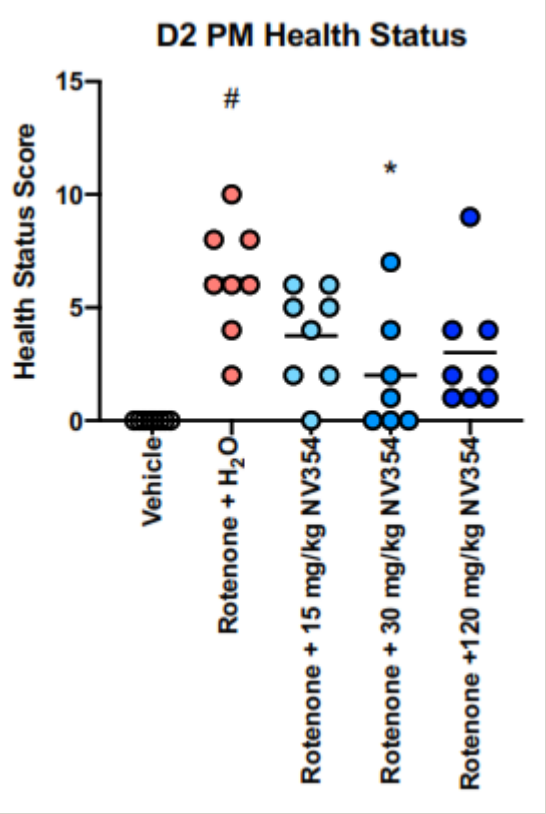
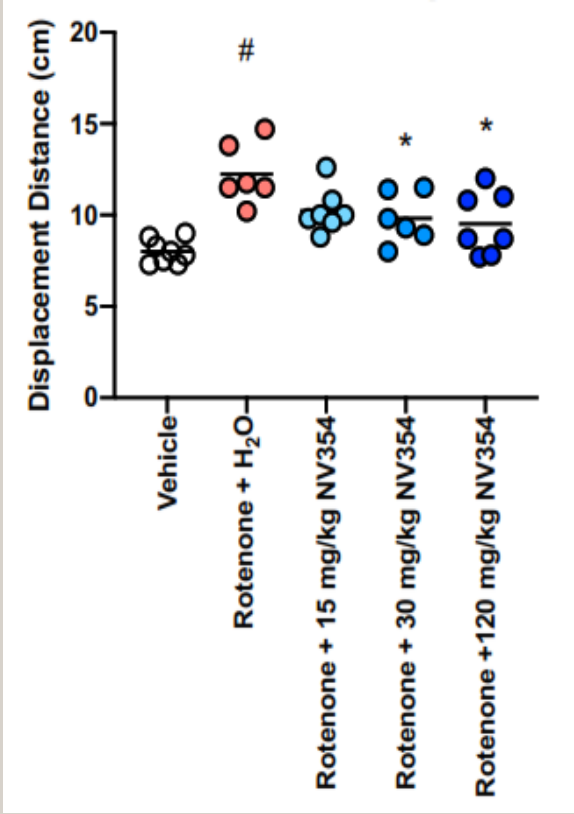
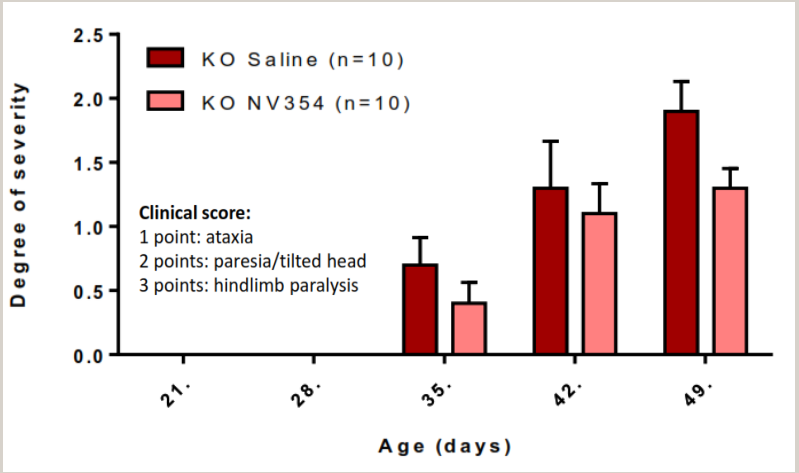
…and demonstrates efficacy in preclinical models

Toxin-induced complex I dysfunction in Rat





Genetic mouse model (Ndufs4) complex I dysfunction



- Strong early signs of preclinical efficacy in rat toxin and mouse genetic models of Leigh's Syndrome

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