UNITED STATES
SECURITIES AND EXCHANGE COMMISSION
WASHINGTON, D.C. 20549

FORM 8-K

CURRENT REPORT

Pursuant to Section 13 or 15(d) of the Securities Exchange Act of 1934

Date of Report (Date of earliest event reported): August 07, 2024

Larimar Therapeutics, Inc.

(Exact name of Registrant as Specified in Its Charter)

Delaware	**001-36510**	**20-3857670**
(State or Other Jurisdiction of Incorporation)	**(Commission File Number)**	**(IRS Employer Identification No.)**
Three Bala Plaza East **Bala Cynwyd, Pennsylvania**		**19004**
(Address of Principal Executive Offices)		**(Zip Code)**

Registrant's Telephone Number, Including Area Code: (844) 511-9056

(Former Name or Former Address, if Changed Since Last Report)

Check the appropriate box below if the Form 8-K filing is intended to simultaneously satisfy the filing obligation of the registrant under any of the following provisions:

☐ Written communications pursuant to Rule 425 under the Securities Act (17 CFR 230.425)

☐ Soliciting material pursuant to Rule 14a-12 under the Exchange Act (17 CFR 240.14a-12)

☐ Pre-commencement communications pursuant to Rule 14d-2(b) under the Exchange Act (17 CFR 240.14d-2(b))

☐ Pre-commencement communications pursuant to Rule 13e-4(c) under the Exchange Act (17 CFR 240.13e-4(c))

Securities registered pursuant to Section 12(b) of the Act:

Title of each class	**Trading Symbol(s)**	**Name of each exchange on which registered**
Common Stock, par value $0.001 per share	LRMR	Nasdaq Global Market

Indicate by check mark whether the registrant is an emerging growth company as defined in Rule 405 of the Securities Act of 1933 (§230.405 of this chapter) or Rule 12b-2 of the Securities Exchange Act of 1934 (§240.12b-2 of this chapter).

Emerging growth company ☐

If an emerging growth company, indicate by check mark if the registrant has elected not to use the extended transition period for complying with any new or revised financial accounting standards provided pursuant to Section 13(a) of the Exchange Act. ☐

Item 2.02 Results of Operations and Financial Condition.

On August 7, 2024, Larimar Therapeutics, Inc. (the "***Company***") announced its financial results and operational highlights for the second quarter ended June 30, 2024. A copy of the press release is being furnished as Exhibit 99.1 to this Current Report on Form 8-K and is incorporated herein by reference.

The information furnished pursuant to this Item 2.02, including Exhibit 99.1 attached hereto, is intended to be furnished and shall not be deemed "filed" for purposes of Section 18 of the Securities Exchange Act of 1934, as amended (the "***Exchange Act***"), or otherwise subject to the liabilities of that section, nor shall it be deemed incorporated by reference in any filing under the Securities Act of 1933, as amended, or the Exchange Act, except as expressly set forth by specific reference in such filing.

Item 8.01 Other Events.

On August 7, 2024, the Company posted on its website an updated slide presentation, which is attached as Exhibit 99.2 to this Current Report on Form 8-K and is incorporated herein by reference. Representatives of the Company will use the presentation in various meetings with investors, analysts and other parties from time to time.

Item 9.01 Financial Statements and Exhibits.

(d) Exhibits

Below is a list of exhibits included with this Current Report on Form 8-K.

Exhibit No.	Document
99.1	Press Release issued by Larimar Therapeutics, Inc. on August 7, 2024*
99.2	Larimar Therapeutics, Inc. Corporate Presentation, dated August 7, 2024
104	Cover Page Interactive Data File (embedded within the Inline XBRL document)

* Furnished herewith

SIGNATURES

Pursuant to the requirements of the Securities Exchange Act of 1934, the registrant has duly caused this report to be signed on its behalf by the undersigned hereunto duly authorized.

Larimar Therapeutics. Inc.

Date: August 7, 2024

By: /s/ Carole S. Ben-Maimon, M.D.

Name: Carole S. Ben-Maimon, M.D.
Title: President and Chief Executive Officer

Exhibit 99.1

Larimar Therapeutics Reports Second Quarter 2024 Operating and Financial Results

- *Open label extension (OLE) study is progressing with all 7 sites activated; interim data planned for Q4 2024*
- *Selected by Food and Drug Administration (FDA) to participate in Support for Clinical Trials Advancing Rare Disease Therapeutics (START) pilot program for nomlabofusp*
- *Joined TRACK-FA Neuroimaging Consortium as an industry partner; TRACK-FA collects natural history data to establish disease-specific neuroimaging biomarkers for potential use in clinical trials*
- *Planning initiation of pharmacokinetic (PK) run-in study in adolescents by year-end 2024; plan to transition adolescents into ongoing OLE study upon completion of PK study*
- *Planning initiation of global confirmatory study by mid-2025 with potential sites in the U.S., Europe, U.K., Canada, and Australia*
- *Biologics License Application (BLA) filing targeted for 2H 2025 to support accelerated approval*
- *Strong balance sheet of $226.1 million cash, cash equivalents and marketable securities as of June 30, 2024, with projected cash runway into 2026*

Bala Cynwyd, PA, August 7, 2024 – Larimar Therapeutics, Inc. (Larimar) (Nasdaq: LRMR), a clinical-stage biotechnology company focused on developing treatments for complex rare diseases, today reported its second quarter 2024 operating and financial results.

“We made significant achievements in our nomlabofusp program this quarter that strongly position us for successful execution across important catalysts over the next 12 months. We were honored to be selected by the FDA to participate in the START pilot program which may be invaluable in helping us achieve our timeline for BLA submission targeted for the second half of 2025 to support accelerated approval. We are actively pursuing clinical sites in the U.S., Europe, U.K. Canada, and Australia in anticipation of initiating a global confirmatory study in mid-2025. We are excited to have recently joined the TRACK-FA Neuroimaging Consortium as an industry partner to support research to define disease-specific neuroimaging biomarkers for potential use in clinical trials.” said Carole Ben-Maimon, MD, President, and Chief Executive Officer of Larimar. “Our OLE study continues to progress with all seven sites now activated and interim data planned for the fourth quarter of this year. We plan to initiate a PK run-in study in adolescents with Friedreich’s ataxia (FA) by year-end, with option for study participants to transition to the OLE study after completing the run-in study. Expanding our clinical program into younger patients will allow us to evaluate the effect of nomlabofusp earlier in the disease process which may help further address the effect of the underlying frataxin deficiency in patients with FA.”

Recent Highlights

- Today, Larimar announced it is planning a PK run-in study in adolescents (12 to 17 years of age) and children (2 to 11 years of age) with FA. This study which we plan to initiate by year-end, will initially enroll 12-15 adolescent patients who will be randomized 2:1 to receive either nomlabofusp or placebo daily. Study participants can transition to the OLE study after completing the PK run-in study.

- Today, Larimar announced that all 7 sites of the OLE were activated. The OLE study continues to progress with interim data to be reported in the fourth quarter of the year.
- In June 2024, Larimar entered into an agreement with the Friedreich's Ataxia Research Alliance (FARA) to join the TRACK-FA Neuroimaging Consortium that includes pharmaceutical, biotechnology, academic and clinical partners. The consortium will conduct a natural history study designed to establish disease-specific neuroimaging biomarkers to track disease progression in the brain and spinal cord and provide a basis for utilizing these biomarkers in clinical trials. Using longitudinal data from large cohorts of patients compared to controls, the study will assess changes in areas previously shown to be compromised in individuals with FA. As an industry partner, Larimar will help fund the study and contribute to the study design, research activities, and analysis. Larimar will have access to all study data for use in its regulatory filings, as appropriate.
- In May 2024, Larimar announced that the FDA has selected the nomlabofusp development program as one of a select few programs to participate in the START pilot program. START selection was based on demonstrated development program readiness, including the potential of nomlabofusp to address the serious and unmet medical needs in a rare neurodegenerative condition, alignment of chemistry, manufacturing, and controls (CMC) development timelines with clinical development plans, and a proposed communications plan where enhanced communication could accelerate pivotal study initiation and path to potential BLA submission.

Second Quarter 2024 Financial Results

As of June 30, 2024, the Company had cash, cash equivalents and marketable securities totaling $226.1 million, which provides projected cash runway into 2026.

Second quarter of 2024 compared to the second quarter of 2023

The Company reported a net loss for the second quarter of 2024 of $21.6 million, or $0.34 per share, compared to a net loss of $8.4 million, or $0.19 per share, for the second quarter of 2023.

Research and development expenses for the second quarter of 2024 were $19.7 million, compared to $5.9 million for the second quarter of 2023. This $13.8 million increase is attributable to increased nomlabofusp manufacturing costs of $10.6 million, including costs related to increasing production costs, $2.1 million due to increased clinical trial costs, primarily the OLE and costs associated with the TRACK-FA natural history study and $1.1M of additional costs associated with increasing headcount.

General and administrative expenses were $4.9 million in the second quarter of 2024, compared to $3.7 million in the second quarter of 2023, an increase of $1.2 million. This increase is attributable in part to $0.6 million in increased legal and professional fees, $0.4 million in additional personnel costs driven by increasing headcount and an increase of $0.1 million in increased non-cash stock compensation costs.

Six months ended June 30, 2024 compared to the six months ended June 30, 2023

The Company reported a net loss for the first six months of 2024 of $36.3 million, or $0.62 per share, compared to a net loss of $14.9 million, or $0.34 per share, for the first six months of 2023.

Research and development expenses for the six months ended June 30, 2024 were $32.6 million, compared to $10.4 million for the six months ended June 30, 2023. This $22.2 million increase is attributable to increased nomlabofusp manufacturing costs of $16.3 million, $3.1 million due to increased clinical trial costs, primarily the OLE and costs associated with the TRACK-FA natural history study, $2.0M of additional costs associated with increasing headcount and $0.3 million of increased noncash stock compensation expense.

General and administrative expenses were $8.7 million for the first six months of 2024, compared to $6.8 million for the six months ended June 30, 2023, an increase of $1.9 million. This increase is attributable in part to $0.8 million in increased legal and professional fees, $0.6 million of additional personnel costs driven by increasing headcount and an increase of $0.3 million in increased non-cash stock compensation costs

About Larimar Therapeutics
Larimar Therapeutics, Inc. (Nasdaq: LRMR), is a clinical-stage biotechnology company focused on developing treatments for complex rare diseases. Larimar's lead compound, nomlabofusp, is being developed as a potential treatment for Friedreich's ataxia. Larimar also plans to use its intracellular delivery platform to design other fusion proteins to target additional rare diseases characterized by deficiencies in intracellular bioactive compounds. For more information, please visit: https://larimartx.com.

Forward-Looking Statements
This press release contains forward-looking statements that are based on Larimar's management's beliefs and assumptions and on information currently available to management. All statements contained in this release other than statements of historical fact are forward-looking statements, including but not limited to statements regarding Larimar's ability to develop and commercialize nomlabofusp and other planned product candidates, Larimar's planned research and development efforts, including the timing of its nomlabofusp clinical trials, interactions with the FDA and overall development plan and other matters regarding Larimar's business strategies, ability to raise capital, use of capital, results of operations and financial position, and plans and objectives for future operations.

In some cases, you can identify forward-looking statements by the words "may," "will," "could," "would," "should," "expect," "intend," "plan," "anticipate," "believe," "estimate," "predict," "project," "potential," "continue," "ongoing" or the negative of these terms or other comparable terminology, although not all forward-looking statements contain these words. These statements involve risks, uncertainties and other factors that may cause actual results, performance, or achievements to be materially different from the information expressed or implied by these forward-looking statements. These risks, uncertainties and other factors include, among others, the success, cost and timing of Larimar's product development activities, nonclinical studies and clinical trials, including nomlabofusp clinical milestones and continued interactions with the FDA; that preliminary clinical trial results may differ from final clinical trial results, that earlier non-clinical and clinical data and testing of nomlabofusp may not be predictive of the results or success of later clinical trials, and assessments; that the FDA may not ultimately agree with Larimar's nomlabofusp development strategy; the potential impact of public health crises on Larimar's future clinical trials, manufacturing, regulatory, nonclinical study timelines and operations, and general economic conditions; Larimar's ability and the ability of third-party manufacturers Larimar engages, to optimize and scale nomlabofusp's manufacturing process; Larimar's ability to obtain regulatory approvals for nomlabofusp and future product candidates; Larimar's ability to develop sales and marketing capabilities, whether alone or with potential future collaborators, and to successfully commercialize any approved product candidates; Larimar's ability to raise the necessary capital to conduct its product development activities; and other risks described in the filings made by Larimar with the Securities and Exchange Commission (SEC), including but not limited to Larimar's periodic reports, including the annual report on Form 10-K, quarterly reports on Form 10-Q and current reports on Form 8-K, filed with or furnished to the SEC and available at www.sec.gov. These forward-looking statements are based on a combination of facts and factors currently known by Larimar and its projections of the future, about which it cannot be certain. As a result, the forward-looking statements may not prove to be accurate. The forward-looking statements in this press release represent Larimar's management's views only as of the date hereof. Larimar undertakes no obligation to update any forward-looking statements for any reason, except as required by law.

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(484) 414-2715

Larimar Therapeutics, Inc.
Condensed Consolidated Balance Sheet
(unaudited)

	June 30, 2024	December 31, 2023
Assets		
Current assets:		
Cash and cash equivalents	$ 32,311	$ 26,749
Short-term marketable securities	193,753	60,041
Prepaid expenses and other current assets	5,066	3,385
Total current assets	231,130	90,175
Property and equipment, net	844	684
Operating lease right-of-use assets	3,213	3,078
Restricted cash	1,339	1,339
Other assets	636	659
Total assets	$ 237,162	$ 95,935
Liabilities and Stockholders' Equity		
Current liabilities:		
Accounts payable	$ 2,917	$ 1,283
Accrued expenses	17,246	7,386
Operating lease liabilities, current	992	837
Total current liabilities	21,155	9,506
Operating lease liabilities	4,603	4,709
Total liabilities	25,758	14,215
Commitments and contingencies		
Stockholders' equity:		
Preferred stock; $0.001 par value per share; 5,000,000 shares authorized as of June 30, 2024 and December 31, 2023; no shares issued and outstanding as of June 30, 2024 and December 31, 2023	—	—
Common stock, $0.001 par value per share; 115,000,000 shares authorized as of June 30, 2024 and December 31, 2023; 63,802,517 and 43,909,069 shares issued and outstanding as of June 30, 2024 and December 31, 2023, respectively	64	43
Additional paid-in capital	436,325	270,150
Accumulated deficit	(224,835)	(188,554)

Accumulated other comprehensive gain (loss)	(150)	81
Total stockholders' equity	211,404	81,720
Total liabilities and stockholders' equity	$ 237,162	$ 95,935

Larimar Therapeutics, Inc.

Condensed Consolidated Statements of Operations

(In thousands, except share and per share data)

(unaudited)

	Three Months Ended June 30,		Six Months Ended June 30,	
	2024	2023	2024	2023
Operating expenses:				
Research and development	$ 19,682	$ 5,875	$ 32,621	$ 10,437
General and administrative	4,917	3,745	8,712	6,820
Total operating expenses	24,599	9,620	41,333	17,257
Loss from operations	(24,599)	(9,620)	(41,333)	(17,257)
Other income (expense), net	2,972	1,254	5,052	2,365
Net loss	(21,627)	(8,366)	(36,281)	(14,892)
Net loss per share, basic and diluted	$ (0.34)	$ (0.19)	$ (0.62)	$ (0.34)
Weighted average common shares outstanding, basic and diluted	63,801,792	43,897,603	58,677,749	43,897,603
Comprehensive loss:				
Net loss	$ (21,627)	$ (8,366)	$ (36,281)	$ (14,892)
Other comprehensive gain (loss):				
Unrealized gain (loss) on marketable securities	(125)	12	(231)	43
Total other comprehensive gain (loss)	(125)	12	(231)	43
Total comprehensive loss	$ (21,752)	$ (8,354)	$ (36,512)	$ (14,849)

Exhibit 99.2

Larimar Therapeutics

Corporate Deck

August 2024

Forward-Looking Statements

This presentation contains forward-looking statements that are based on the beliefs and assumptions of Larimar Therapeutics, Inc. ("Company") and on information currently available to management. All statements contained in this presentation other than statements of historical fact are forward-looking statements, including but not limited to Larimar's ability to develop and commercialize nomlabofusp (CTI-1601) and other planned product candidates, Larimar's planned research and development efforts, including the timing of its nomlabofusp clinical trials, expectations with respect to the FDA START pilot program, interactions with the FDA and overall development plan and other matters regarding Larimar's business strategies, ability to raise capital, use of capital, results of operations and financial position, and plans and objectives for future operations.

In some cases, you can identify forward-looking statements by the words "may," "will," "could," "would," "should," "expect," "intend," "plan," "anticipate," "believe," "estimate," "predict," "project," "potential," "continue," "ongoing" or the negative of these terms or other comparable terminology, although not all forward-looking statements contain these words. These statements involve risks, uncertainties and other factors that may cause actual results, performance, or achievements to be materially different from the information expressed or implied by these forward-looking statements. These risks, uncertainties and other factors include, among others, the success, cost and timing of Larimar's product development activities, nonclinical studies and clinical trials, including nomlabofusp clinical milestones and continued interactions with the FDA; that preliminary clinical trial results may differ from final clinical trial results, that earlier non-clinical and clinical data and testing of nomlabofusp may not be predictive of the results or success of later clinical trials, and assessments; that the FDA may not ultimately agree with Larimar's nomlabofusp development strategy; the potential impact of public health crises on Larimar's future clinical trials, manufacturing, regulatory, nonclinical study timelines and operations, and general economic conditions; Larimar's ability and the ability of third-party manufacturers Larimar engages, to optimize and scale nomlabofusp's manufacturing process; Larimar's ability to obtain regulatory approvals for nomlabofusp and future product candidates; Larimar's ability to develop sales and marketing capabilities, whether alone or with potential future collaborators, and to successfully commercialize any approved product candidates; Larimar's ability to raise the necessary capital to conduct its product development activities; and other risks described in the filings made by Larimar with the Securities and Exchange Commission (SEC), including but not limited to Larimar's periodic reports, including the annual report on Form 10-K, quarterly reports on Form 10-Q and current reports on Form 8-K, filed with or furnished to the SEC and available at www.sec.gov. These forward-looking statements are based on a combination of facts and factors currently known by Larimar and its projections of the future, about which it cannot be certain. As a result, the forward-looking statements may not prove to be accurate. The forward-looking statements in this presentation represent Larimar's management's views only as of the date hereof. Larimar undertakes no obligation to update any forward-looking statements for any reason, except as required by law.

Clinical-Stage Novel Protein Replacement Therapy Platform

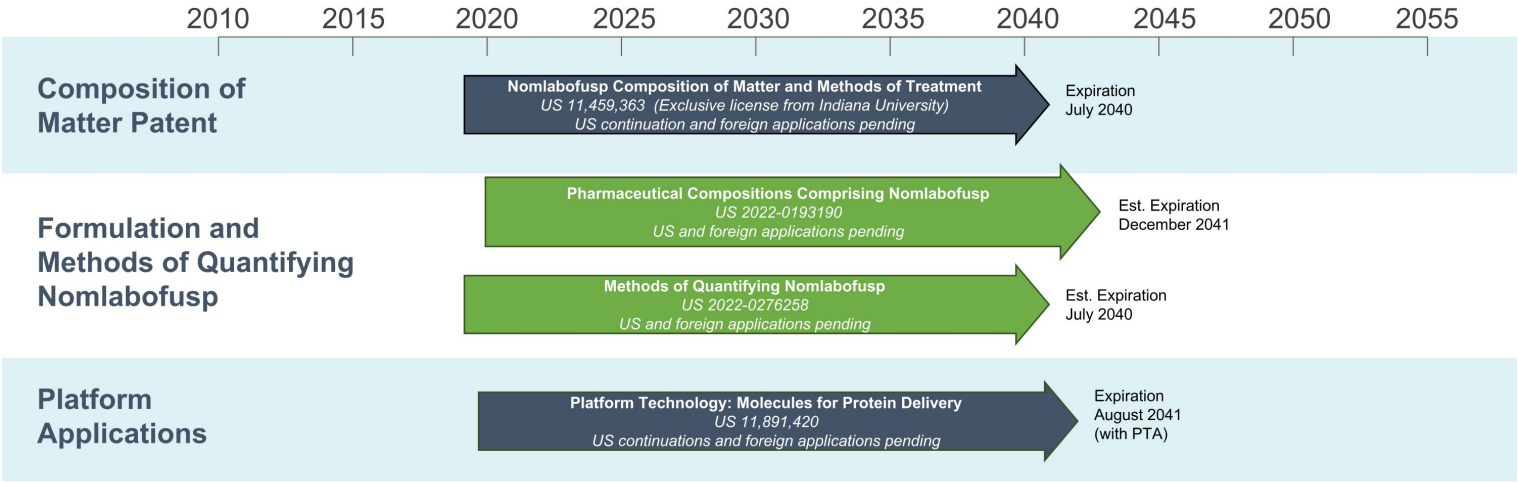
Potential first therapy to increase frataxin levels	Lead candidate nomlabofusp is a recombinant fusion protein designed to directly address frataxin deficiency in patients with FA by delivering the protein to mitochondria. Granted Orphan Drug (US & EU), Rare Pediatric Disease (US), Fast Track (US), & PRIME (EU) designations. **Recently selected by FDA to participate in its START pilot program**
Consistent Phase 1 and Phase 2 findings	Nomlabofusp was generally well tolerated and demonstrated dose-dependent increases in frataxin (FXN) levels from baseline in skin and buccal cells in a completed 4-week placebo-controlled Phase 2 study and a completed multiple ascending dose Phase 1 study
Intend to pursue accelerated approval with FDA	FDA acknowledgement that FXN deficiency appears to be critical to the pathogenic mechanism of FA, and that there continues to be an unmet need for treatments that address the underlying disease pathophysiology. Discussions to support an accelerated approval are ongoing. BLA submission targeted for 2H 2025
Clinical program	**Plans to initiate PK run-in study in adolescents** by end of 2024; transition adolescents into OLE after PK study Dosed first adult patient in OLE study with 25 mg daily dosing in Q1 2024 with interim data expected in Q4 2024 All 7 OLE sites activated; continuing to enroll patients Dose escalation to 50 mg currently planned following further characterization of FXN PD at 25 mg dose
Strong financial foundation	Approximately $226 million in cash and investments as of 6/30/24 which includes $161.8 million in net proceeds raised from a Feb 24 public offering Provides projected cash runway into 2026

Larimar Therapeutics

Nomlabofusp (CTI-1601); FA: Friedreich's ataxia

Larimar Technology is Supported by a Strong IP Portfolio

Granted nomlabofusp (CTI-1601) composition of matter patent extends into 2040



Additional nomlabofusp IP protection

- US and foreign pending applications cover key biomarkers, analytical tools and methods of treatment for additional disease indications for nomlabofusp
- Nomlabofusp should be eligible for **12 years of market exclusivity** upon approval in the US (independent of patents) and at least **10 years of market exclusivity** upon approval in EU (independent of patents)

Granted Pending

Friedreich's Ataxia (FA): A rare and progressive disease

Genetic defect on both alleles lowers frataxin levels

Most patients with FA only produce ~20-40% of normal frataxin levels depending on the tissue, sampling technique, and assay considered*

Affects ~20,000 patients globally

~5,000 patients in the U.S., with most remaining patients in the EU
~70% of patients present before age 14

Progressive disease

Initial symptoms include unsteady posture and frequent falling, and patients are eventually confined to a wheelchair
Life expectancy of 30-50 years with an early death usually caused by heart disease

No approved therapies increase frataxin levels

Only treatment approved for FA does not address frataxin deficiency

Larimar Therapeutics

* E.C. Deutsch et al. Molecular Genetics and Metabolism 101 (2010) 238–245.

Strong Relationship with FARA – Joined FARA's TRACK-FA Neuroimaging Consortium as an Industry Partner

TRACK-FA collects natural history data to establish disease specific neuroimaging biomarkers for potential use in clinical trials. Larimar will have access to all study data for use in regulatory filings, as appropriate

FARA provides industry with several key items

- Assistance with patient recruitment and education
- Access to Global Patient Registry with demographic and clinical information on more than 1,000 FA patients
- Sponsored a Patient-Focused Drug Development Meeting in 2017 resulting in a publication titled "The Voice of the Patient"

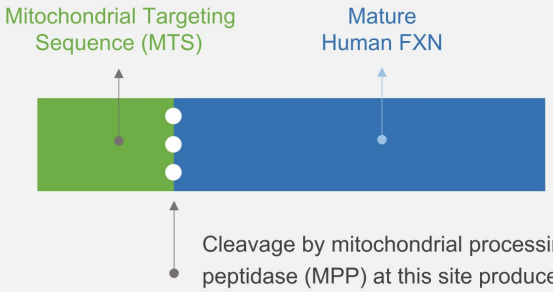
FARA Friedreich's Ataxia Research Alliance

National, non-profit organization dedicated to the pursuit of scientific research leading to treatments and a cure for FA

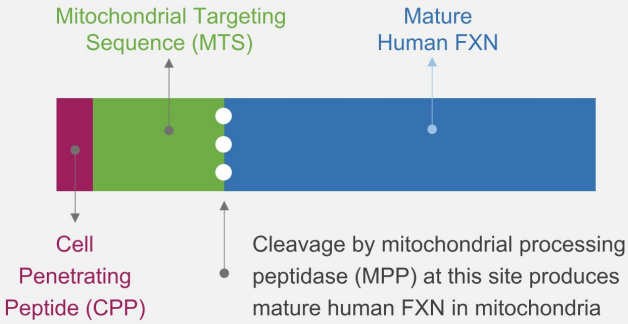
Nomlabofusp is Designed to Deliver Additional Frataxin

Nomlabofusp (CTI-1601) maintains the cleavage site between the MTS and mature human frataxin (FXN)

STRUCTURE OF ENDOGENOUS FXN



STRUCTURE OF NOMLABOFUSP



The presence of the cleavage site allows the CPP and MTS to be removed by mitochondrial processing peptidase to produce mature human FXN in the mitochondria

Larimar Therapeutics

FXN Levels Predict Disease Progression in FA

Lower FXN levels are associated with earlier onset of disease, faster rate of disease progression, and shorter time to loss of ambulation

Median Age of Onset and Rate of Disease Progression in Relation to FXN Levels

FXN Level* (% of Normal Level)	**Age of Onset** (Years)	**FARS**** (Change/Year)
11.2	7	2.9
22.0	11	2.1
31.0	16	2.0
48.7	19	1.6

Adapted from H.L.Plasterer et al. PLoS ONE 2013 8(5):e63958

Median Age of Onset Predicts Time to Loss of Ambulation

Age of Onset (Years)	**Median Time to Loss of Ambulation** (Years)
< 15	11.5
15 to 24	18.3
> 24	23.5

Adapted from C. Rummey et al. EClinicalMedicine. 2020 18:100213

Larimar Therapeutics

*FXN levels measured in peripheral blood mononuclear cells (PBMCs). FXN levels as measured by % of normal demonstrated to be equivalent in PBMCs, buccal cells, and whole blood.
**FARS: Friedreich's ataxia rating score, measures disease progression with a higher score indicating a greater level of disability.

Completed Ph 2 Dose Exploration Study (25 & 50 mg Cohorts)

Goal: Further characterize PK/PD and assess safety to inform long-term dose and dose regimen

28-day Treatment Period - nomlabofusp (CTI-1601) or placebo

1 2 3 4 5 6 7 8 9 10 11 12 13 14 15 16 17 18 19 20 21 22 23 24 25 26 27 28

= Subcutaneous administration of nomlabofusp (CTI-1601) or placebo

= No Administration

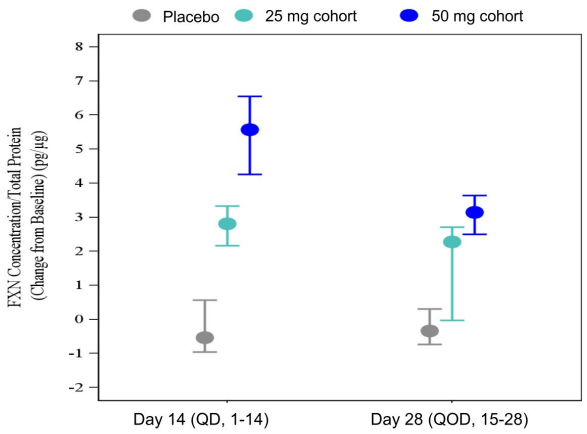
Study Details	
Population	Ambulatory and non-ambulatory Friedreich's ataxia patients ≥18 years of age Nomlabofusp (CTI-1601) treatment naïve or participated (if eligible) in a previous Larimar study
Dose	**Cohort 1:** 25 mg **Cohort 2:** 50 mg
Key Endpoints	Frataxin levels in peripheral tissue, PK, safety and tolerability; other exploratory endpoints include lipids and gene expression levels
Number of Patients	**Cohort 1:** Enrolled 13 participants (9 on nomlabofusp; 4 on placebo) **Cohort 2:** Enrolled 15 participants (10 on nomlabofusp; 5 on placebo)
Key Results	Generally well tolerated; most common adverse events were mild and moderate injection site reactions Dose dependent increases of frataxin levels in tissues tested (skin and buccal cells) Baseline FXN levels in skin cells in the 50 mg cohort were < 17% of the average of healthy volunteers. After daily dosing for 14 days, FXN levels increased to 33% to 59% of the average of the healthy volunteers

Larimar Therapeutics

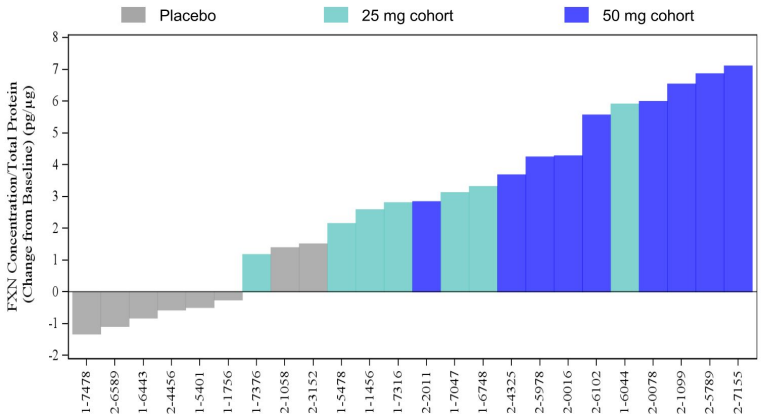
Dose-Dependent Increase in FXN Levels in Skin Cells

Participants dosed daily for 14 days, then every other day until day 28

Skin Cells
FXN Levels* Change from Baseline**



FXN Levels* in Skin Cells
Change from Baseline at Day 14



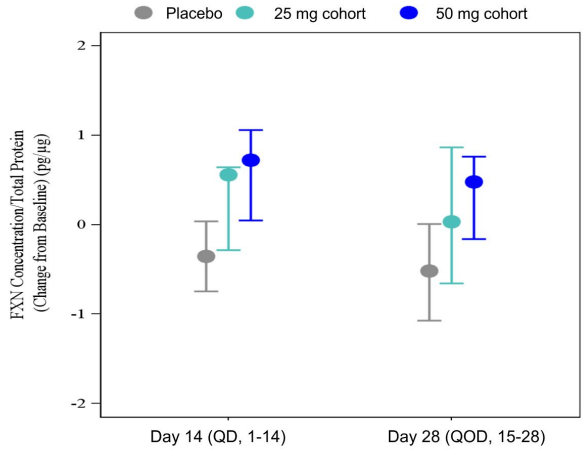
Larimar Therapeutics

*FXN levels measured via detection of peptide derived from mature FXN; FXN concentrations are normalized to total cellular protein content in each sample. Data represent median and 25th and 75th percentiles. Only participants with quantifiable levels at both baseline and Day 14 are included in the figures.
**Median baseline FXN levels in patients were 3.5 pg/µg for the placebo, 3.7 pg/µg for the 25 mg cohort and 2.1 pg/µg for the 50 mg cohort.

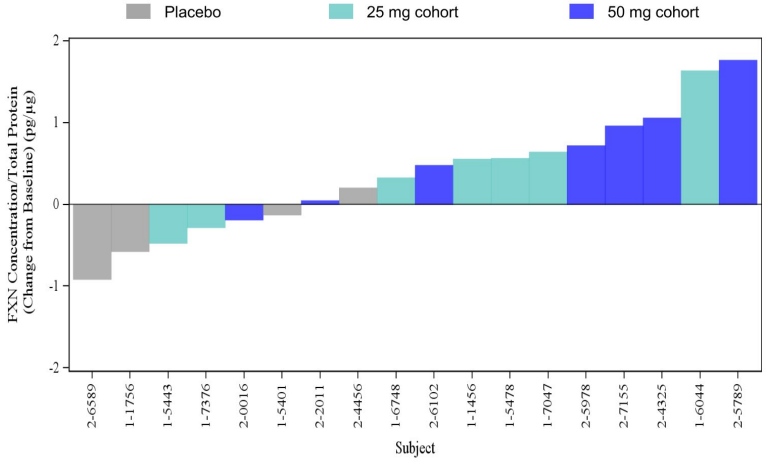
Dose-Dependent Increase in FXN Levels in Buccal Cells

Participants dosed daily for 14 days, then every other day until day 28

Buccal Cells
FXN Levels* Change from Baseline**

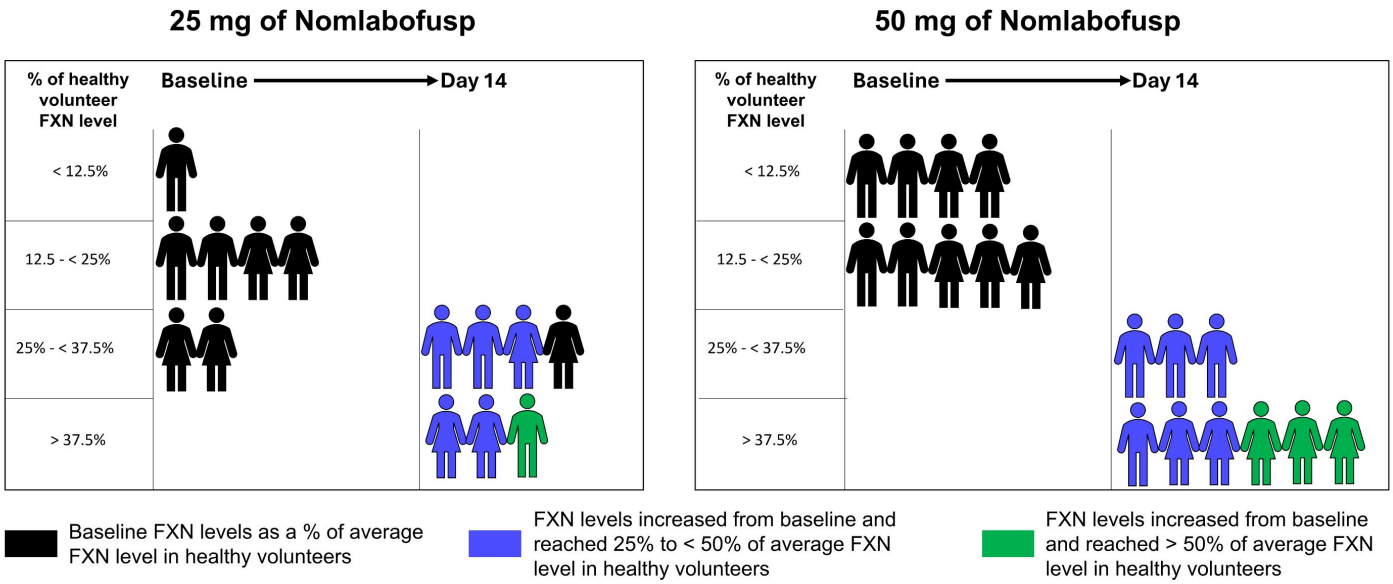


FXN Levels* in Buccal Cells
Change from Baseline at Day 14



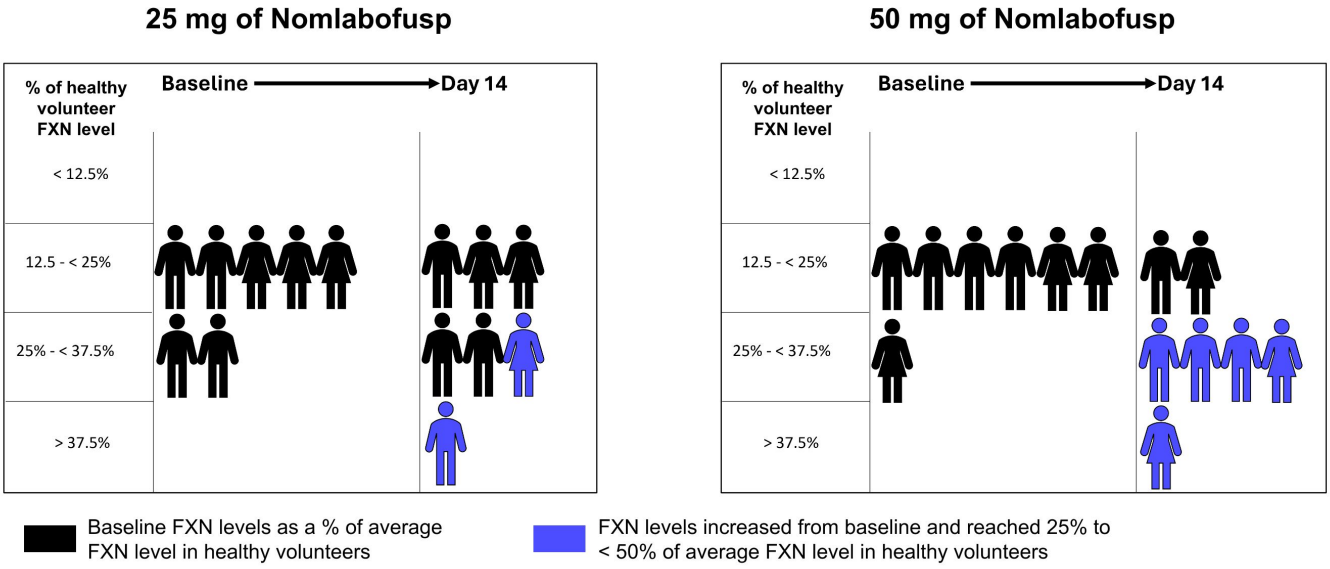
*FXN levels measured via detection of peptide derived from mature FXN; FXN concentrations are normalized to total cellular protein content in each sample. Data represent median and 25th and 75th percentiles. Only participants with quantifiable levels at both baseline and Day 14 are included in the figures.
**Median baseline FXN level in patients were 2.1 pg/µg for the placebo, 1.8 pg/µg for the 25 mg cohort and 1.6 pg/µg for the 50 mg cohort.

Skin Cell FXN Levels Achieve Higher % of Healthy Volunteers* Following 14 days of Daily Nomlabofusp



Only participants with quantifiable levels at baseline and day 14 are included in the figures.
*% of healthy volunteer FXN level is calculated by dividing each participant's FXN level by the average FXN level (16.34 pg/µg) from the noninterventional healthy volunteer study (N=60).

Buccal Cell FXN Levels Achieve Higher % of Healthy Volunteers* Following 14 days of Daily Nomlabofusp



Only participants with quantifiable levels at baseline and day 14 are included in the figures.
*% of healthy volunteer FXN level is calculated by dividing each participant's FXN level by the average FXN level (8.24 pg/µg) from Larimar's noninterventional healthy volunteer study (N=60).

Nomlabofusp: Predictable Pharmacokinetics

1. Quick absorption after subcutaneous administration
2. Dose-proportional increases in exposure observed
3. Pharmacokinetic profile consistent with Phase 1 studies

Open-label Extension Study: Dosed first patient in Q1 2024

Preliminary interim data expected in Q4 2024

Key Eligibility Criteria

Previous participation in Phase 1 or Phase 2 trials

Daily subcutaneous injection of 25 mg nomlabofusp;
self-administered or by a caregiver
Plan to increase dose to 50 mg daily

- All 7 sites activated
- First patient dosed in March 2024
- Continuing to enroll patients
- Amending study to include adolescents (12-17 yrs) and children (2-11 yrs) after completion of PK run-in study

Key Study Objectives

- Safety and tolerability
- Long-term PK
- Dose escalation to 50 mg currently planned following further characterization of FXN pharmacodynamics at 25 mg dose
- Tissue FXN concentrations and potential use as surrogate endpoint to support accelerated approval
- Clinical efficacy measures compared to the matched set of untreated patients from FACOMS* database

Screening Period ≤ 42 days** → Treatment Period Planned for ≥ 1 year → Potential extensions

*FACOMS: Friedreich's Ataxia Clinical Outcome Measures Study.
**Estimated screening period may be extended for those study participants who have not been on a stable regimen of omaveloxolone for at least six months.

Nomlabofusp Clinical Development Plan

Intend to pursue accelerated approval pathway with potential BLA submission targeted for 2H 2025
Recently selected by FDA to participate in its START pilot program

Ongoing open-label extension study with 25 mg daily dosing for eligible patients who participated in SAD, MAD, and/or four-week dose exploration studies

Interim data expected Q4 2024

Plans to Initiate PK run-in study in adolescents (12-17 yrs) before year end 2024, followed by children (2-11 yrs) in 1H 2025

Participants completing the PK run-in study eligible to transition into OLE after the PK run-in study is completed

Planned global double-blind placebo-controlled registration/confirmatory study targeted to be initiated by mid- 2025*

BLA submission targeted for 2H 2025

Larimar Therapeutics

*Company initiated discussions with FDA on the potential use of FXN levels to support accelerated approval. Also, the Company is planning discussions with regulators and investigators outside the U.S. to expand clinical program to international geographies.

Nomlabofusp is a Competitively Differentiated Treatment Approach*

$7.3B Acquisition supports the **robust market potential** for FA treatments

REATA PHARMACEUTICALS Biogen

Nomlabofusp is a potential **first-and-only protein replacement therapy** designed to address the underlying cause of FA

Approach	Product	Company	Mechanism of Action	Clinical Status
Protein replacement	**Nomlabofusp (CTI-1601)**	**Larimar**	**Recombinant frataxin protein**	**Phase II**
Mitochondrial Oxidative Stress Modifier	Omaveloxolone (SKYCLARYS™)	Reata Pharma/Biogen	Nrf2 Activator	Approved (US and EU)
	Vatiquinone	PTC Therapeutics	15-Lipoxygenase Inhibitor	Phase III
Gene Expression Regulator	DT-216P2 (new formulation)	Design Therapeutics	GeneTAC	Pre-clinical
Gene Therapy	LX2006	Lexeo Therapeutics	Frataxin Gene Replacement	Phase I/II

Larimar Therapeutics

*Competitive landscape focuses on clinical-stage, industry-sponsored programs from public companies

Positive Ph2 Data, OLE Escalating to 50 mg & Initiating in Adolescents

Consistent Ph 1 and Ph 2 Findings	Nomlabofusp is generally well tolerated at doses tested up to 4 weeks Dose-dependent increases in FXN levels from baseline in evaluated tissues (skin and buccal cells) Baseline FXN levels in skin cells in the 50 mg cohort were < 17% of the average of healthy volunteers. After daily dosing for 14 days, FXN levels increased to 33% to 59%
Clinical & Regulatory Updates	Plans to Initiate PK run-in study in adolescents by end of 2024; transition adolescents into OLE after PK study Pursuing clinical sites in the U.S., Europe, the U.K., Canada. and Australia for planned initiation of registration/confirmatory study targeted for mid- 2025 Selected by FDA to participate in its START pilot program Initiated discussions with FDA regarding use of FXN as a surrogate endpoint to support accelerated approval
2024/2025 Milestones	**Q4 2024:** Initiate PK run-in study in adolescents (ages 12-17 years old) **Q4 2024:** Interim data from OLE study **Q4 2024:** Final Phase 2 data planned to be presented at a conference **1H 2025:** Initiate PK run-in study in children (ages 2-11 years old) **Mid 2025:** Initiate Global confirmatory/registration study **2H 2025:** BLA submission; intend to pursue accelerated approval

Clinical-Stage Novel Protein Replacement Therapy Platform

Potential first therapy to increase frataxin levels	Lead candidate nomlabofusp is a recombinant fusion protein designed to directly address frataxin deficiency in patients with FA by delivering the protein to mitochondria. Granted Orphan Drug (US & EU), Rare Pediatric Disease (US), Fast Track (US), & PRIME (EU) designations. **Recently selected by FDA to participate in its START pilot program**
Consistent Phase 1 and Phase 2 findings	Nomlabofusp was generally well tolerated and demonstrated dose-dependent increases in frataxin (FXN) levels from baseline in skin and buccal cells in a completed 4-week placebo-controlled Phase 2 study and a completed multiple ascending dose Phase 1 study
Intend to pursue accelerated approval with FDA	FDA acknowledgement that FXN deficiency appears to be critical to the pathogenic mechanism of FA, and that there continues to be an unmet need for treatments that address the underlying disease pathophysiology. Discussions to support an accelerated approval are ongoing. BLA submission targeted for 2H 2025
Clinical program	**Plans to initiate PK run-in study in adolescents** by end of 2024; transition adolescents into OLE after PK study Dosed first adult patient in OLE study with 25 mg daily dosing in Q1 2024 with interim data expected in Q4 2024 All 7 OLE sites activated; continuing to enroll patients Dose escalation to 50 mg currently planned following further characterization of FXN PD at 25 mg dose
Strong financial foundation	Approximately $226 million in cash and investments as of 6/30/24 which includes $161.8 million in net proceeds raised from a Feb 24 public offering Provides projected cash runway into 2026

Larimar Therapeutics

Nomlabofusp (CTI-1601); FA: Friedreich's ataxia


Larimar
Therapeutics
Larimar Therapeutics
August 2024 Corporate Deck
THANK YOU

Larimar Therapeutics

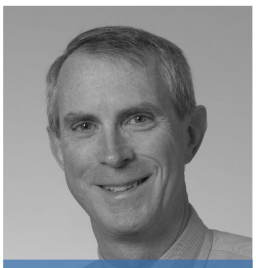
Appendix

Scientific Advisory Board

Giovanni Manfredi, MD, PhD

Finbar and Marianne Kenny Professor in Clinical and Research Neurology at Weill Cornell Medicine.

Professor of Neuroscience at Weill Cornell Medicine.



Mark Payne, MD

Co-founder of Chondrial Therapeutics, which became Larimar Therapeutics, Inc.

Professor of Pediatrics at Indiana University School of Medicine

Marni J. Falk, MD

Executive Director of the Mitochondrial Medicine Frontier Program at The Children's Hospital of Philadelphia (CHOP)

Professor in the Division of Human Genetics, Department of Pediatrics at University of Pennsylvania Perelman School of Medicine

Jill Ostrem, MD

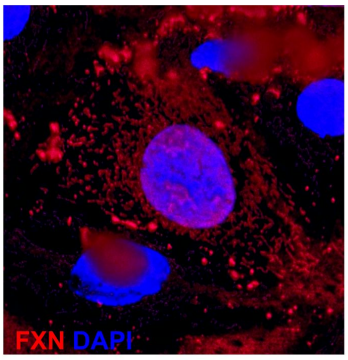
Medical Director and Division Chief of the University of California San Francisco (UCSF) Movement Disorders and Neuromodulation Center.

Carlin and Ellen Wiegner Endowed Professor of Neurology

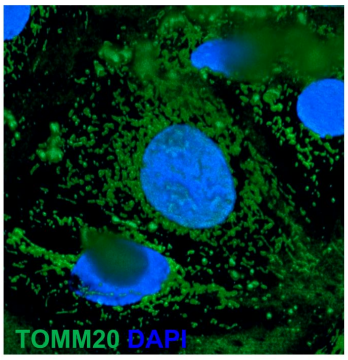
Mitochondrial Localization and Preclinical Data

Nomlabofusp Transduction of Cells In Vitro Leads to hFXN Located in Mitochondria

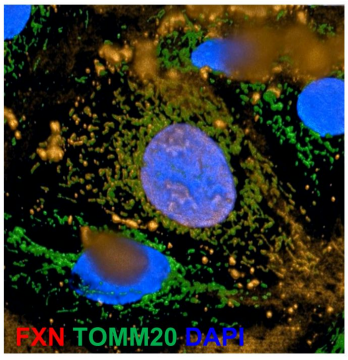
FXN staining



TOMM20 (mitochondria) staining



FXN co-localizes with TOMM20



- Rat cardiomyocytes (H9C2) were transduced with nomlabofusp
- Cells were fixed and analyzed by immunofluorescence microscopy to detect the presence of human frataxin (hFXN) and TOMM20 (a mitochondrial outer membrane protein)
- Nuclei were stained with DAPI

Larimar Therapeutics

Nomlabofusp Extends Survival in FXN-deficient KO Mice

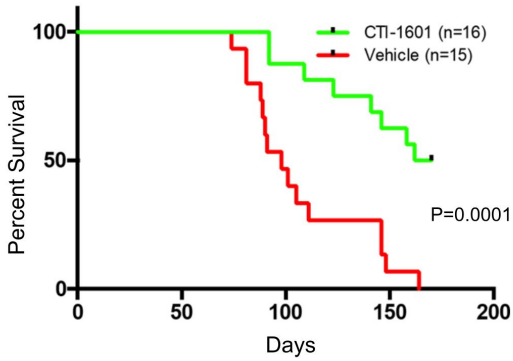
Initial proof-of-concept for FXN replacement therapy in cardiac mouse model of FA

Median survival of MCK-Cre FXN-KO mice

- 166 days (nomlabofusp) vs. 98 days (Vehicle)
- Nomlabofusp administered 10 mg/kg SC every other day

Survival beyond vehicle mean (107.5 days)

- 87.5% (nomlabofusp) vs. 33% (Vehicle)
- Demonstrates that nomlabofusp is capable of delivering sufficient amounts of FXN to mitochondria



Nomlabofusp (CTI-1601) rescues a severe disease phenotype in a well-characterized cardiac mouse model of FA

Larimar Therapeutics

Nomlabofusp Prevents Development of Ataxic Gait in Neurologic KO Mouse Model

In-Vivo Efficacy Data in Pvalb-Cre FXN-KO Mouse Model

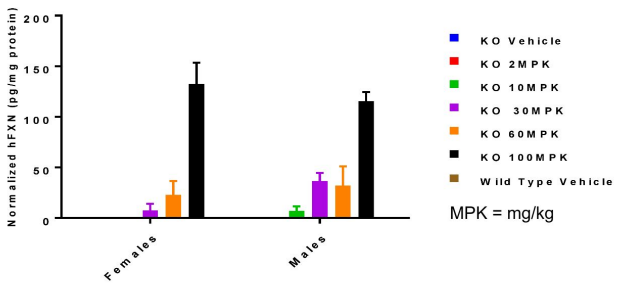
Single dose level: 10 mg/kg nomlabofusp or vehicle given intraperitoneally three times per week

- hFXN replacement with nomlabofusp **prevents development of ataxic gait**
- Nomlabofusp-treated mice **survive longer** than untreated mice
- Human frataxin **present in brain, dorsal root ganglia and spinal cord** demonstrating central nervous system penetration

Nomlabofusp Delivers hFXN to Mitochondria and Restores SDH Activity in KO Mice

Study Design – Cardiac and skeletal muscle FXN knockout mice (MCK-CRE) were treated at varying SQ doses of nomlabofusp every other day for two weeks at Jackson Laboratories (Bar Harbor, ME). After dosing, animals were sacrificed, and heart and skeletal muscle were evaluated for hFXN concentration in mitochondrial extracts and SDH activity was assessed.

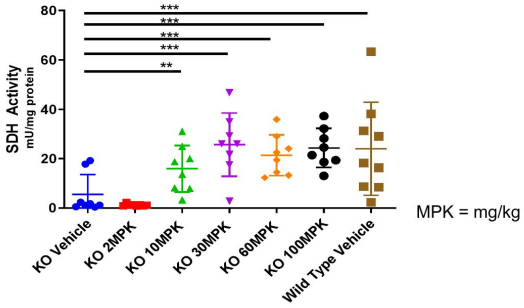
Mitochondrial FXN (Heart)



Mitochondria hFXN concentration increases dose-dependently

Given subcutaneously, nomlabofusp functionally replaces hFXN in mitochondria of KO mice

SDH Activity (Muscle)



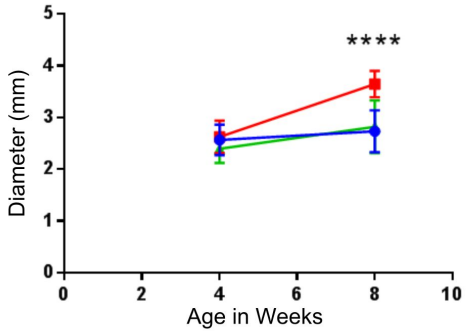
Succinate dehydrogenase (SDH) activity, which is indicative of mitochondrial function, increases in a dose-dependent manner after administration of nomlabofusp; activity plateaus at 30 mg/kg and is equivalent to activity in wild type

Larimar Therapeutics

Nomlabofusp Prevents Left Ventricle Dilation in KO Mice

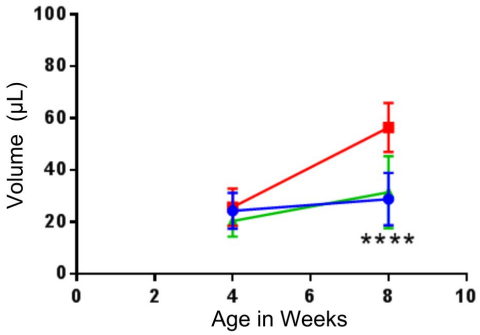
Study Design – Cardiac and skeletal muscle FXN knockout mice (MCK-CRE) were treated at 10 mg/kg every other day at Jackson Laboratories (Bar Harbor, ME). Echocardiograms were performed pre-dose and post dose.

Left Ventricle Internal Diameter (Systole)



Left ventricular (LV) volume increases in systole in untreated mice by 8 weeks (after 4 weeks of dosing with vehicle), but remains similar to wildtype when treated with nomlabofusp (10 mg/kg every other day)

Left Ventricle Volume (Systole)



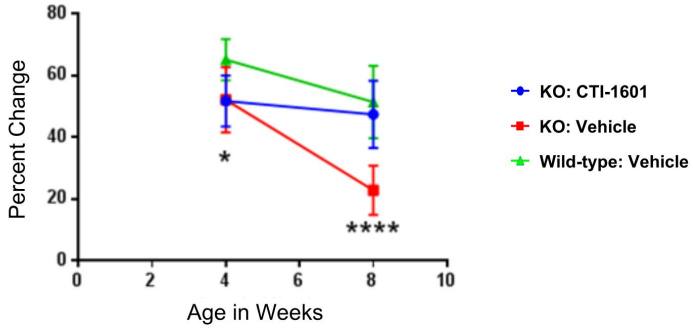
Nomlabofusp-treated mice have similar LV volume as wild type; echocardiogram shows significant differences between vehicle and nomlabofusp treated (10 mg/kg every other day) KO mice

Larimar Therapeutics

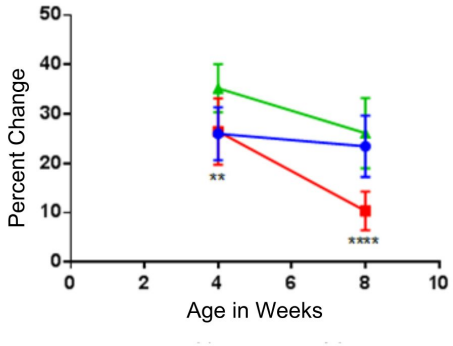
Nomlabofusp Preserves Left Ventricle Function in KO Mice

Study Design – Cardiac and skeletal muscle FXN knockout mice (MCK-CRE) were treated at 10 mg/kg every other day at Jackson Laboratories (Bar Harbor, ME). Echocardiograms were performed pre-dose and post dose.

Left Ventricle Ejection Function



Left Ventricle Fractional Shortening



Left ventricular (LV) function drops significantly in vehicle treated mice by Week 8

Nomlabofusp-treated (10 mg/kg every other day) mice have similar LV function as wildtype; echocardiogram shows significant differences between vehicle and nomlabofusp treated KO mice

Larimar Therapeutics

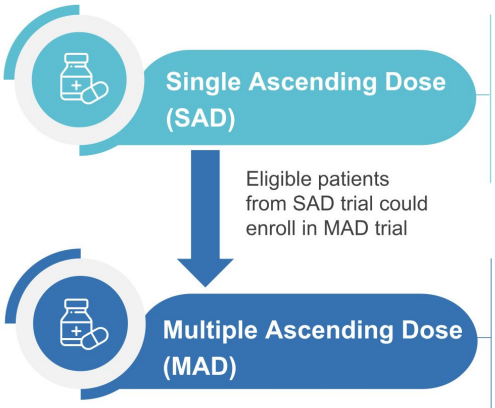
Phase 1 Clinical Data

CTI-1601: Phase 1 Clinical Program in Patients with FA

Program consisted of double-blind, placebo controlled single- and multiple-ascending dose trials

Phase 1 Development Plan

- Two double-blind, placebo-controlled dosing trials in patients with FA
- Patient dosing began December 2019
- Safety Review Committee assessed all blinded data between each cohort to ensure patient safety



Number of subjects: 28
Dose levels: 25 mg, 50 mg, 75 mg and 100 mg (subcutaneous administration)
Treatment Duration: 1 day
1º Endpoint: Safety and tolerability
2º Endpoints: PK; PD; FXN levels; multiple exploratory
Status: Complete

Number of Subjects: 27
Dose Range: 25 mg, 50 mg, 100 mg (subcutaneous administration)
Treatment Regimen: Multiple increasing doses administered subcutaneously over 13 days
1º Endpoint: Safety and tolerability
2º Endpoints: PK; PD; FXN levels (buccal cells, platelets, optional skin biopsies); multiple exploratory
Status: Complete

Larimar Therapeutics

Completed Phase 1 Multiple Ascending Dose Study

Treatment Schedules for Each Cohort- nomlabofusp (CTI-1601) or placebo

Cohort 1 (25 mg; n = 8)

13-day Treatment Period

1	2	3	4	5	6	7	8	9	10	11	12	13	14

= Administration of nomlabofusp or placebo

= No Administration

Cohort 2 (50 mg; n = 9)

13-day Treatment Period

1	2	3	4	5	6	7	8	9	10	11	12	13	14

= Administration of nomlabofusp or placebo

= No Administration

Cohort 3 (100 mg n = 10)

13-day Treatment Period

1	2	3	4	5	6	7	8	9	10	11	12	13	14

= Administration of nomlabofusp or placebo

= No Administration

FXN Level Sampling Days Presented for Each Cohort

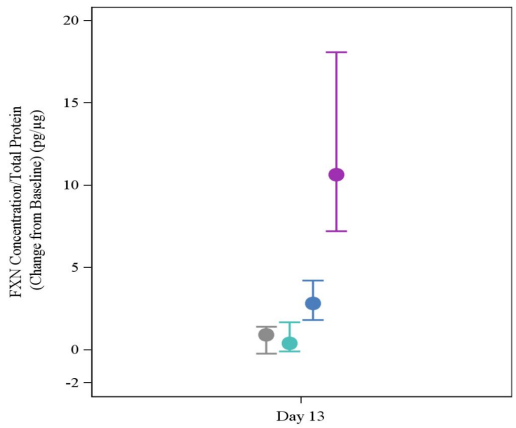
Cohort 1 Sampling Days	
Buccal Cells	Baseline, Day 4, Day 13
Skin	Baseline, Day 13
Platelets	Baseline, Day 4, Day 13

Cohort 2 Sampling Days	
Buccal Cells	Baseline, Day 7, Day 13
Skin	Baseline, Day 13
Platelets	Baseline, Day 7, Day 13

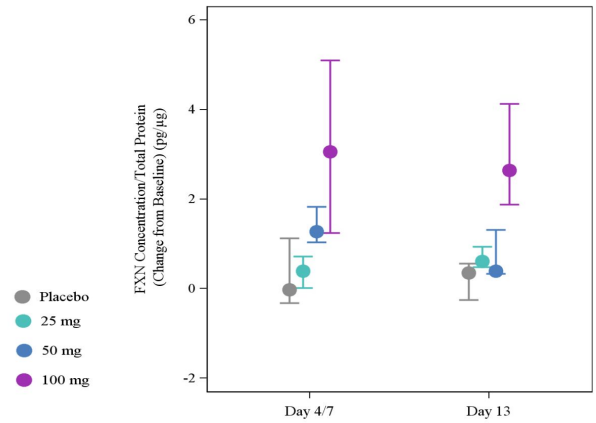
Cohort 3 Sampling Days	
Buccal Cells	Baseline, Day 7, Day 13
Skin	Baseline, Day 13
Platelets	Baseline, Day 7, Day 13

Dose Dependent Increases in FXN Levels Observed in Skin and Buccal Cells in Phase 1

FXN* Change from Baseline By Dose Group (Skin Cells)



FXN* Change from Baseline By Dose Group (Buccal Cells)



Placebo: Participants randomized to placebo in each cohort
25 mg: Dosed daily for 4 days, every third day thereafter
50 mg: Dosed daily for 7 days, every other day thereafter
100 mg: Dosed daily for 13 days

*FXN levels measured via detection of peptide derived from mature FXN; FXN concentrations are normalized to total cellular protein content in each sample; Data represent median and 25th and 75th percentiles; FXN levels from Day 4, & Day 13 measurements are shown for data derived from the 25 mg cohort; FXN levels from Day 7 & Day 13 measurements are shown for data derived from the 50 & 100 mg cohorts;

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MAD Trial Patient Demographics

Parameter	Statistic	All placebo (n=7)	25 mg CTI-1601 (n=6)	50 mg CTI-1601 (n=7)	100 mg CTI-1601 (n=7)	All CTI-1601 (n=20)	Overall (n=27)
Sex							
Male	n (%)	5 (71.4)	3 (50.0)	4 (57.1)	3 (42.9)	10 (50.0)	15 (55.6)
Female	n (%)	2 (28.6)	3 (50.0)	3 (42.9)	4 (57.1)	10 (50.0)	12 (44.4)
Age (years)							
	Mean	25.7	39.7	34.7	28.0	33.9	31.7
	SD	6.37	16.59	9.03	8.96	12.13	11.40
	Median	23	37	36	24	34	28
	Min, Max	20,36	21,65	19,47	20,44	19,65	19,65
Race							
White	n (%)	6 (85.7)	6 (100.0)	6 (85.7)	6 (85.7)	18 (90.0)	24 (88.9)
Asian	n (%)	0	0	1 (14.3)	1 (14.3)	2 (10.0)	2 (7.4)
American Indian	n (%)	1 (14.3)	0	0	0	0	1 (3.7)
Ethnicity							
Hispanic/Latino	n (%)	2 (28.6)	0	0	0	0	2 (7.4)
Not Hispanic/Latino	n (%)	5 (71.4)	6 (100.0)	7 (100.0)	7 (100.0)	20 (100.0)	25 (92.6)

SD: Standard deviation

MAD Trial Patient Disease Characteristics

Parameter	Statistic	All placebo (n=7)	25 mg CTI-1601 (n=6)	50 mg CTI-1601 (n=7)	100 mg CTI-1601 (n=7)	All CTI-1601 (n=20)	Overall (n=27)
Age at Symptom Onset							
	Mean	14.1	24.0	19.3	11.9	18.1	17.1
	SD	5.34	14.48	6.21	6.72	10.37	9.39
	Median	15.0	18.0	19.0	10.0	18.0	16.0
	Min, Max	8,23	12,44	8,28	5,22	5,44	5,44
Age at Diagnosis							
	Mean	18.3	31.5	26.4	15.9	24.3	22.7
	SD	7.87	19.88	4.28	8.21	13.24	12.23
	Median	20.0	25.5	28.0	13.0	27.0	21.0
	Min, Max	9,32	14,64	17,30	5,27	5,64	5,64
Assistive Device							
Walker	n (%)	0	2 (33.3)	3 (42.9)	0	5 (25.0)	5 (18.5)
Wheelchair	n (%)	4 (57.1)	3 (50.0)	1 (14.3)	6 (85.7)	10 (50.0)	14 (51.9)
Other	n (%)	1 (14.3)	0	1(14.3)	0	1 (5.0)	2 (7.4)
None	n (%)	2 (28.6)	1 (16.7)	2 (28.6)	1 (14.3)	4 (20.0)	6 (22.2)

SD: Standard deviation

PK analyses support evaluating once-daily and every-other-day dosing regimens for CTI-1601

Summary of MAD Trial PK Analyses

- CTI-1601 was quickly absorbed after subcutaneous administration
- Dose-proportional increases in exposure observed with increasing doses of CTI-1601
- Mean half life of CTI-1601 in plasma was approximately 11 hours
- CTI-1601 appeared to be at or close to steady state exposure after 13 days of dosing 100 mg once daily

Phase 2 Demographic/ Disease Characteristics and Additional Data

Demographics – Phase 2 Trial

	25 mg Cohort			50 mg Cohort		
	Placebo *N = 4*	Nomlabofusp *N = 9*	Overall *N = 13*	Placebo *N = 5*	Nomlabofusp *N = 10*	Overall *N = 15*
Age at Screening (Years)						
Mean (SD)	34.0 (9.20)	37.8 (14.93)	36.6 (13.16)	28.6 (4.67)	28.1 (11.00)	28.3 (9.17)
Median	33	31	31	27	24	26
Q1, Q3	27, 42	27, 42	27, 42	26, 30	21, 32	21, 32
Min, Max	25, 45	25, 69	25, 69	24, 36	19, 54	19, 54
Sex n (%)						
Male	2 (50.0)	5 (55.6)	7 (53.8)	1 (20.0)	4 (40.0)	5 (33.3)
Female	2 (50.0)	4 (44.4)	6 (46.2)	4 (80.0)	6 (60.0)	10 (66.7)
Previously Treated with Nomlabofusp n (%)						
Yes	1 (25.0)	3 (33.3)	4 (30.8)	0	1 (10.0)	1 (6.7)
No	3 (75.0)	6 (66.7)	9 (69.2)	5 (100.0)	9 (90.0)	14 (93.3)

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Disease Characteristics – Phase 2 Study

	25 mg Cohort			50 mg Cohort		
	Placebo *N = 4*	Nomlabofusp *N = 9*	Overall *N = 13*	Placebo *N = 5*	Nomlabofusp *N = 10*	Overall *N = 15*
Age at Symptom Onset (Years)						
Mean (SD)	14.5 (4.93)	13.0 (10.47)	13.5 (8.77)	15.2 (7.26)	13.7 (8.37)	14.2 (7.78)
Median	14.5	10	11	14	12.5	14
Q1, Q3	11, 19	8, 13	9, 15	11, 16	7, 18	7, 18
Min, Max	9, 20	5, 38	5, 38	8, 27	5, 30	5, 30
Age at Diagnosis (Years)						
Mean (SD)	17.5 (5.57)	18.6 (11.20)	18.2 (9.58)	18.6 (6.80)	16.6 (8.03)	17.3 (7.46)
Median	16.5	16	16	19	13.5	14
Q1, Q3	14, 22	14, 20	14, 20	13, 20	10, 21	12, 21
Min, Max	12, 25	5, 42	5, 42	12, 29	9, 30	9, 30
Time Since Diagnosis (Years)						
Mean (SD)	16.1 (5.97)	18.5 (11.52)	17.8 (9.94)	9.5 (3.72)	11.9 (7.05)	11.1 (6.10)
Median	13.42	14.32	13.5	11	11.26	11
Q1, Q3	12.9, 19.3	12.8, 21.6	12.8, 21.6	5.8, 11.3	7.4, 15.3	5.8, 15.2
Min, Max	12.5, 25.0	5.4, 45.0	5.4, 45.0	5.6, 14.0	2.3, 25.1	2.3, 25.1

Absolute Increases in Skin FXN Levels

Dose response in tissue FXN concentrations and increases from baseline after dosing

Day 14 Skin FXN Levels			
Dose	Visit	Absolute Values (pg/µg)	
		Median	Mean
25 mg	Baseline	3.70	3.38
	Day 14	5.53	6.40
	Change from Baseline	**2.81**	**3.02**
50 mg	Baseline	2.12	2.08
	Day 14	7.40	7.32
	Change from Baseline	**5.57**	**5.24**

Day 28 Skin FXN Levels			
Dose	Visit	Absolute Values (pg/µg)	
		Median	Mean
25 mg	Baseline	3.70	3.38
	Day 28	4.39	4.80
	Change from Baseline	**2.28**	**1.41**
50 mg	Baseline	2.12	2.08
	Day 28	5.23	5.24
	Change from Baseline	**3.14**	**3.17**

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Only participants with quantifiable levels at baseline and day 14 and day 28 are included in the tables.

Absolute Increases in Buccal FXN Levels

Dose response in tissue FXN concentrations and increases from baseline after dosing

Day 14 Buccal FXN Levels			
Dose	Visit	Absolute Values (pg/µg)	
		Median	Mean
25 mg	Baseline	1.78	1.80
	Day 14	2.24	2.22
	Change from Baseline	**0.56**	**0.42**
50 mg	Baseline	1.61	1.69
	Day 14	2.44	2.38
	Change from Baseline	**0.72**	**0.69**

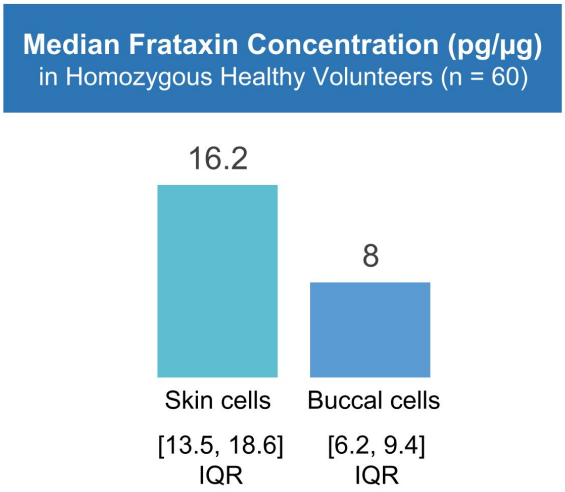
Day 28 Buccal FXN Levels			
Dose	Visit	Absolute Values (pg/µg)	
		Median	Mean
25 mg	Baseline	1.70	1.65
	Day 28	1.73	1.76
	Change from Baseline	**0.03**	**0.11**
50 mg	Baseline	1.76	1.77
	Day 28	2.15	2.15
	Change from Baseline	**0.48**	**0.38**

Only participants with quantifiable levels at baseline and day 14 and day 28 are included in the tables.

Non-Interventional Study Data

CLIN-1601-002: Top-line Non-interventional Study Results

Non-interventional study measured FXN in homozygous healthy volunteers



Most patients with FA only produce ~20-40%[1] of normal frataxin levels depending on the tissue, sampling technique, and assay considered

Lower FXN levels seen with typical onset[2] (5 to 15 years of age)

Higher FXN levels seen with late onset[2] (after 25 years of age)

Heterozygous carriers who show no signs of disease have buccal cell FXN levels of ~50% of unaffected healthy persons[1]

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FXN concentrations were measured in skin and buccal cells from 60 homozygous healthy volunteers utilizing the same sampling technique and assay as clinical trials of nomlabofusp; FXN levels measured via detection of peptide derived from mature FXN; FXN concentrations normalized to total cellular protein content in each sample. 1. E.C. Deutsch et al. Molecular Genetics and Metabolism 101 (2010) 238–245. 2. Friedreich's Ataxia Research Alliance

FDA START Pilot Program

Nomlabofusp Selected by FDA for START Pilot Program

Highlights FDA commitment to augment formal meetings with more rapid, ad-hoc communications to accelerate program development of rare diseases

START Pilot Program Support for Clinical Trials Advancing Rare Disease Therapeutics	**CDER Selection Based On**
A new milestone-driven program launched by the FDA in September 2023 Designed to **accelerate development of novel therapies** intended to address unmet medical needs in **rare diseases** **7 novel drugs selected** • 3 products by CDER (nomlabofusp) for rare neurodegenerative conditions • 4 products by CBER for cell and gene therapy	Demonstrated development **program readiness** (e.g., sponsors who demonstrate the ability to move the program towards a marketing application) Potential to address serious and unmet medical need in a **rare neurodegenerative condition** **Alignment of CMC** development timelines with clinical development plans Proposed plan where **enhanced communication can improve efficiency of product development**

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FDA: Food and Drug Administration; CDER: Center for Drug Evaluation and Research; CBER: Center for Biologics Evaluation and Research; CMC: Chemistry, Manufacturing, and Controls