UNITED STATES
SECURITIES AND EXCHANGE COMMISSION
Washington, DC 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): September 14, 2022

Larimar Therapeutics, Inc.
(Exact name of registrant as specified in its charter)

Delaware	**001-36510**	**20-3857670**
(State or other jurisdiction of incorporation or organization)	**(Commission File Number)**	**(I.R.S. Employer Identification No.)**

Three Bala Plaza East. Suite 506
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 (see General Instruction A.2. below):

☐ 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 8.01 Other Events.

Press Release

On September 14, 2022, Larimar Therapeutics, Inc. (the "Company") issued a press release announcing that the U.S. Food and Drug Administration ("FDA") has cleared the initiation of the 25 mg cohort of a Phase 2, four-week, placebo-controlled, dose exploration trial of CTI-1601 in Friedreich's ataxia patients, removing the full clinical hold on the Company's CTI-1601 program and imposing a partial clinical hold. A copy of this press release is filed as Exhibit 99.1 hereto and incorporated herein by reference.

Investor Presentation

On September 14, 2022, the Company updated information reflected in a slide presentation, which is filed 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 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 of Larimar Therapeutics, Inc., dated September 14, 2022*
99.2	Larimar Therapeutics, Inc. Corporate Presentation, dated September 14, 2022*
104	Cover Page Interactive Data File (embedded within the Inline XBRL document)

* Filed 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.

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

Date: September 14, 2022

Larimar Therapeutics Announces FDA Clearance to Initiate the 25 mg Cohort of a Phase 2 Dose Exploration Trial of CTI-1601 in Friedreich's Ataxia Patients

- Initiation of the Phase 2 trial is expected in Q4 2022, with top-line data expected in 2H 2023

- Company management hosting webcast and conference call today at 8:30 a.m. ET

Bala Cynwyd, PA, September 14, 2022 – Larimar Therapeutics, Inc. ("Larimar") (Nasdaq: LRMR), a clinical-stage biotechnology company focused on developing treatments for complex rare diseases, today announced that the U.S. Food and Drug Administration (FDA) has cleared the initiation of the 25 mg cohort of a Phase 2, four-week, placebo-controlled, dose exploration trial of CTI-1601 in Friedreich's ataxia (FA) patients. In a written communication to Larimar, the FDA indicated it was lifting its full clinical hold on the CTI-1601 program and imposing a partial hold. The design of the upcoming Phase 2 trial is identical to the design proposed by Larimar, with the exception of a requirement for the FDA to review data from the 25 mg cohort prior to escalating the dose in the second cohort. Larimar expects to begin the Phase 2 trial in Q4 2022, with top-line data expected in 2H 2023.

"We thank the FDA for their engagement and are pleased with their decision to clear CTI-1601's return to the clinic," said Carole Ben-Maimon**,** MD, President and Chief Executive Officer of Larimar. "Given the strength of our Phase 1 data and the urgent need for a disease-modifying FA therapy, we believe today's news is an important event for not only Larimar, but for the entire FA community. We are now working expeditiously to initiate our Phase 2 dose exploration trial next quarter. We anticipate that the results of this trial will provide crucial safety, pharmacokinetic, and pharmacodynamic data that will inform the design of future studies."

The CTI-1601 program was placed on a clinical hold by the FDA following the Company's notification to the agency of 3 mortalities out of a total of 34 animals in a 26-week non-human primate (NHP) toxicology study designed to support extended dosing of patients with CTI-1601. All 3 of these NHPs were in the study's two highest dose groups and all NHPs in the two lower dose groups survived to the end of the study. The FDA's decision to allow the upcoming CTI-1601 Phase 2 trial to proceed follows Larimar's submission of a complete response with detailed analyses from Larimar's NHP toxicology studies and Phase 1 clinical trials.

Larimar's upcoming Phase 2 trial is designed to further characterize CTI-1601's safety, pharmacodynamic (PD), and pharmacokinetic (PK) profiles to provide information about the preferred long-term dose and dose regimen. Eligible patients will include ambulatory and non-ambulatory individuals with FA who are at least 18 years old. Patients may be CTI-1601 treatment naïve or have previously participated in Larimar's Phase 1 single- or multiple ascending dose trials.

Patients enrolled into the Phase 2 trial will be randomized 2:1 to receive CTI-1601 or placebo. The trial is designed to enroll approximately 24 – 30 total patients across two cohorts, with the first cohort of 12 – 15 patients evaluating a 25 mg dose of CTI-1601. Patients will receive CTI-1601 or placebo daily via subcutaneous injections for the first 14 days, and then every other day until day 28. Key endpoints will include safety assessments, measures of frataxin levels and other PD markers (e.g., lipid profiles and gene expression data) in peripheral tissues, as well as PK

assessments. Dose escalation to 50 mg in the second cohort will be contingent on the FDA's agreement based on its review of the data from the trial's first cohort, and on the review by the trial's independent data monitoring committee.

Nancy M. Ruiz, MD, Chief Medical Officer of Larimar, added, "Our Phase 2 dose exploration trial is designed to build upon the positive findings of our previously completed multiple ascending dose trial. In the multiple ascending dose trial, daily subcutaneous injections of 50 mg of CTI-1601 increased frataxin levels in the buccal cells of FA patients, with the levels achieved exceeding those we would expect to see in phenotypically normal heterozygous carriers. This was a promising finding, as FA is caused by insufficient frataxin production and frataxin levels in buccal cells have been shown to correlate with neurological function in patients. By exploring extended daily dosing at 25 mg before potentially escalating to 50 mg in our upcoming trial, we aim to determine if lower doses for a longer period of time can also drive relevant increases in peripheral frataxin, allowing us to better understand CTI-1601's minimum effective dose. In addition, two weeks of daily dosing followed by two weeks of every-other-day dosing will provide valuable data to support the PK/PD models that will help inform the design of subsequent studies."

Previously completed Phase 1 single- and multiple-ascending dose (MAD) clinical trials evaluated the safety, PK, and PD profiles of CTI-1601 administered subcutaneously at doses up to 100 mg daily for up to 13 days. No serious adverse events, important medical events, or treatment-related severe adverse events were reported in the trials. The most common adverse events were mild and moderate injection site reactions, which all resolved without intervention. Except for injection site reactions, the number and severity of adverse events did not increase with increasing exposure to CTI-1601. Data from cohorts 2 and 3 of the MAD trial also showed that subcutaneous injections of 50 or 100 mg of CTI-1601, administered daily for at least seven days, resulted in frataxin levels in buccal cells that were at or in excess of those that would be expected in phenotypically normal heterozygous carriers. Cohort 1 of the MAD trial, which evaluated a 25 mg dose, explored a daily dosing regimen for only four days. In contrast, the 25 mg cohort of the upcoming Phase 2 trial will explore 14 days of daily dosing followed by 14 days of every-other-day dosing.

Conference Call and Webcast

Larimar will host a conference call and webcast today, September 14, 2022, at 8:30 a.m. ET. To access the webcast, please visit this link to the event. To participate by phone please dial 1-877-407-9716 (domestic) or 1-201-493-6779 (international) and refer to conference ID 13732889. Following the live event, the archived webcast will be available on the "Investors" page of the Larimar website.

About CTI-1601

CTI-1601 is a recombinant fusion protein intended to deliver human frataxin to the mitochondria of patients with Friedreich's ataxia who are unable to produce enough of this essential protein. CTI-1601 has been granted Rare Pediatric Disease designation, Fast Track designation and Orphan Drug designation by the U.S. Food and Drug Administration (FDA), Orphan Drug Designation by the European Commission, and a PRIME designation by the European Medicines Agency.

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, CTI-1601, 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 Larimar's expectations regarding its ability to resolve the partial clinical hold imposed by the FDA related to CTI-1601, Larimar's ability to develop and commercialize CTI-1601 and other planned product candidates, Larimar's planned research and development efforts, including the timing of its CTI-1601 clinical development plan and other matters regarding Larimar's business strategies, 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, Larimar's ability to successfully engage with the FDA and satisfactorily respond to requests from the FDA for further information and data regarding the CTI-1601 clinical trial including the FDA review of data from cohort one from the Phase 2 dose escalation trial and FDA 's agreement to escalate the dosing in cohort two, the timing and outcomes of Larimar's interactions with the FDA concerning the partial clinical hold, the success, cost and timing of Larimar's product development activities, nonclinical studies and clinical trials, including CTI-1601 clinical milestones; that preliminary clinical trial results may differ from final clinical trial results, that earlier non-clinical and clinical data and testing of CTI-1601 may not be predictive of the results or success of later clinical trials, and assessments; the ongoing impact of the COVID-19 pandemic on Larimar's future clinical trials, manufacturing, regulatory, nonclinical study timelines and operations, and the potential impact of the Russian invasion of Ukraine on Larimar's ability to raise additional capital and general economic conditions; Larimar's ability and the ability of third-party manufacturers Larimar engages, to optimize and scale CTI-1601's manufacturing process; Larimar's ability to obtain regulatory approvals for CTI-1601 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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Company Contact:

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Chief Financial Officer
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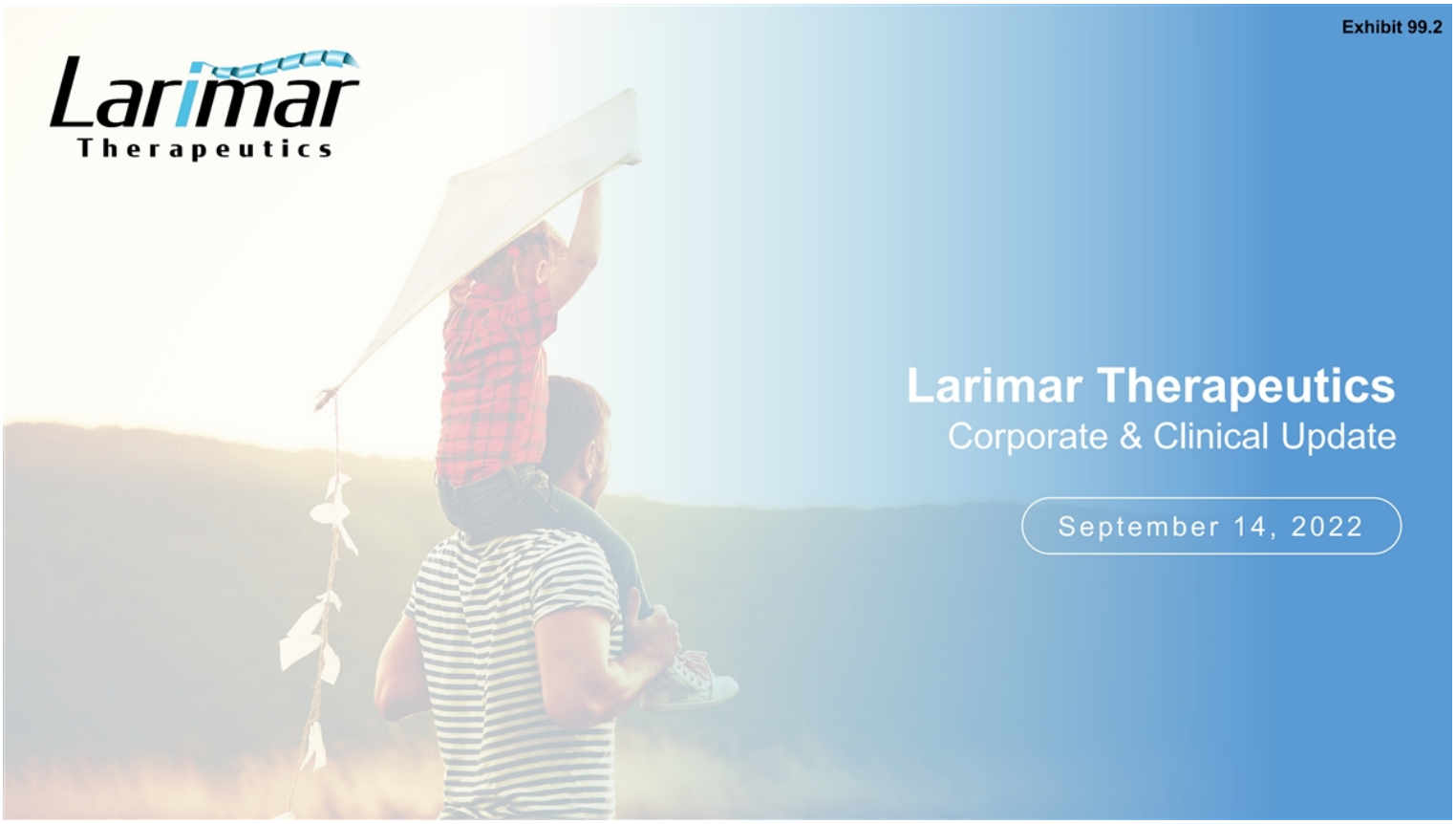
Exhibit 99.2



Larimar Therapeutics

Corporate & Clinical Update

September 14, 2022

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 statements regarding the expectations and assumptions regarding the future of the Company’s business, including the Company’s ability to develop and commercialize CTI-1601 and other planned product candidates, the Company’s planned research and development efforts, and other matters regarding the Company’s business strategies, 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 Company’s ability to successfully engage with the FDA and satisfactorily respond to requests from the FDA for further information and data regarding the CTI-1601 clinical trial including the FDA review of data from cohort one from the Phase 2 dose escalation trial and FDA ‘s agreement to escalate the dosing in cohort two , the timing and outcomes of the Company’s interactions with the FDA concerning the partial clinical hold, the success, cost and timing of the Company’s product development activities, nonclinical studies and clinical trials, including CTI-1601 clinical milestones; that preliminary clinical trial results may differ from final clinical trial results, that earlier non-clinical and clinical data and testing of CTI-1601 may not be predictive of the results or success of later clinical trials, and assessments; the ongoing impact of the COVID-19 pandemic on the Company’s future clinical trials, manufacturing, regulatory, nonclinical study timelines and operations, and the potential impact of the Russian invasion of Ukraine on the Company’s ability to raise additional capital and general economic conditions; the Company’s ability and the ability of third-party manufacturers the Company engages, to optimize and scale CTI-1601’s manufacturing process; the Company’s ability to obtain regulatory approvals for CTI-1601 and future product candidates; the Company’s ability to develop sales and marketing capabilities, whether alone or with potential future collaborators, and to successfully commercialize any approved product candidates; the Company’s ability to raise the necessary capital to conduct its product development activities; and other risks described in the filings made by the Company with the Securities and Exchange Commission (SEC), including but not limited to the Company’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 the Company 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. These forward-looking statements are based on information currently available to us, and we assume no obligation to update any forward-looking statements, except as required by law.

Investment Highlights: CTI-1601 Cleared for Return to Clinic

Clinical-stage biotechnology company with a novel protein replacement therapy platform
Focused on addressing unmet needs in Friedreich's ataxia (FA) and other complex rare diseases based on a platform technology backed by a strong intellectual property portfolio

Lead candidate: CTI-1601, a recombinant fusion protein designed to deliver frataxin to mitochondria
Orphan Drug (US & EU), Rare Pediatric Disease (US), Fast Track (US), & PRIME (EU) designations for FA

Double-blind, placebo-controlled Phase 1 proof-of-concept trials in FA patients complete
Data show dose dependent increases in frataxin (FXN) levels from baseline compared to placebo in all evaluated tissues with daily dosing & that CTI-1601 was generally well tolerated when dosed for up to 13 days

FDA clearance to initiate a placebo-controlled, Phase 2, 4-week dose exploration study in FA patients
FDA lifted full clinical hold on CTI-1601 and imposed a partial hold, thereby clearing advancement to Phase 2 Cohort 1 to evaluate 25 mg dose; dose escalation contingent on FDA review of cohort 1 data
Study is expected to initiate in Q4 2022, with top-line data from both cohorts in 2H 2023

Strong financial foundation with projected cash runway into 2H 2024
Today announced pricing of an underwritten offering expected to provide $70M in gross proceeds[1]
High-quality institutional investor base includes founding investor Deerfield Management

Larimar Therapeutics

1. Represents gross proceeds before deducting the underwriting discounts and commissions and other offering expenses. Additional details available in a press release issued by Larimar on September 14, 2022

Friedreich's Ataxia (FA)

Rare and Progressive Disease

Caused by genetic defect resulting in low levels of frataxin

- Patients with FA only produce ~20-40% of normal frataxin levels depending on the tissue, sampling technique, and assay considered[1]
- Affects ~20,000 patients globally, with ~5,000 patients in the U.S. and majority of the remaining patients in the EU

Approximately 70% of patients present before age 14

- Initial symptoms may include unsteady posture, frequent falling and progressive difficulty in walking. By the time symptoms occur, heart damage may have already occurred. Progressive disease: symptoms worsen and patients are eventually confined to a wheelchair with speech becoming hesitant and jerky (often referred to as "scanning of speech")

Life expectancy of 30-50 years

- Early death usually caused by heart disease

No approved therapies available

- Current treatment options are limited to symptom management

LRMR continues to have a strong relationship with Friedreich's Ataxia Research Alliance

- Dedicated FA patient advocacy group focused on treatments for FA

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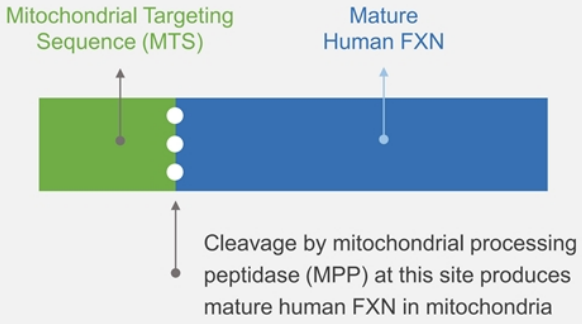
1. E.C. Deutsch et al. Molecular Genetics and Metabolism 101 (2010) 238–245

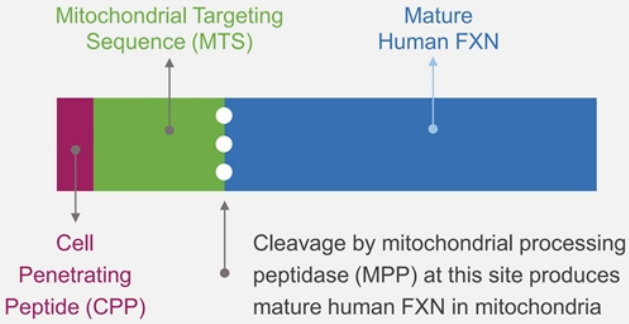
CTI-1601 is Designed to Deliver Additional Frataxin (FXN)

CTI-1601 maintains the cleavage site between the MTS and mature human FXN

STRUCTURE OF ENDOGENOUS FXN



STRUCTURE OF CTI-1601



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

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Upcoming Phase 2, Four-week Dose Exploration Study

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

Treatment Schedule

28-day Treatment Period

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

= Administration of CTI-1601 or placebo

= No Administration

Study Details	
Population	Ambulatory and non-ambulatory Friedreich's ataxia patients ≥18 years of age. CTI-1601 treatment naïve or participated (if eligible) in a previous Larimar study.
Dose	**Cohort 1:** 25 mg **Cohort 2:** Dose escalation to 50 mg contingent on a review of Cohort 1 data by FDA and IDMC
Key Endpoints	Frataxin levels in peripheral tissue, PK, PD, safety and tolerability. PD endpoints include lipid profiles and gene expression data.
Number of Patients	~24-30 patients total across Cohorts 1 and 2 randomized 2:1 to receive CTI-1601 or placebo.
Timing	Study initiation expected in Q4 2022. Top-line data from both cohorts expected in 2H 2023.

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IDMC: Independent data monitoring committee

Completed Multiple Ascending Dose Study

Treatment Schedules for Each Cohort

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 CTI-1601 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 CTI-1601 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 CTI-1601 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

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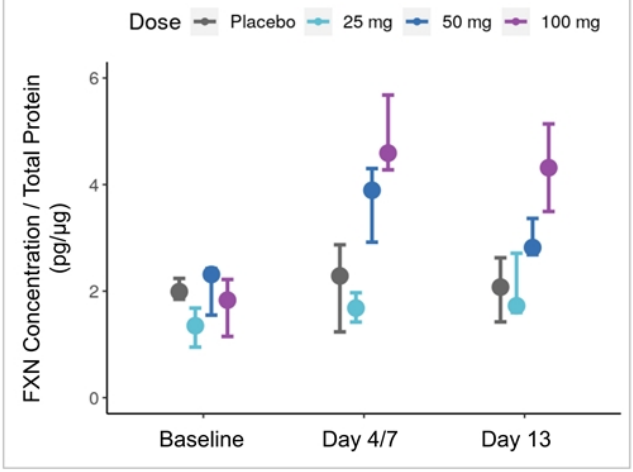
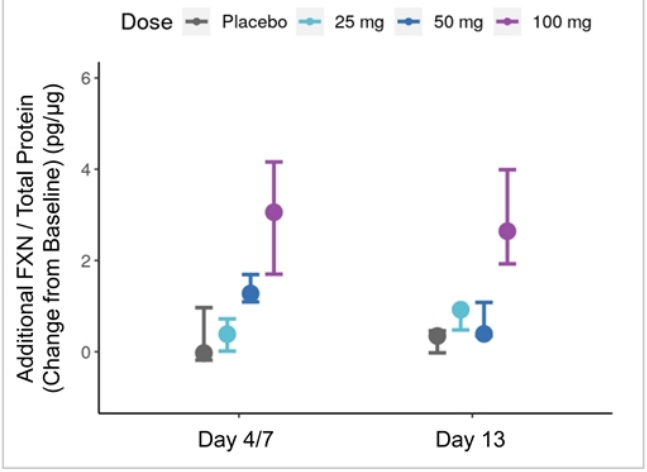
Dose Dependent Increases in FXN Levels Observed in Buccal Cells

FXN* Levels By Dose Group (Buccal Cells)



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



*FXN levels measured via detection of peptide derived from mature FXN; Data represent median and 25th and 75th percentiles; FXN levels from baseline, Day 4, & Day 13 measurements are shown for data derived from the 25 mg cohort; FXN levels from baseline, Day 7 & Day 13 measurements are shown for data derived from the 50 & 100 mg cohorts; Sample collection days varied in each cohort per the trial protocol

Data Compare Favorably to FXN Levels Expected in Heterozygous Carriers

Achieved median FXN levels that were >60% of the median FXN levels observed in healthy controls

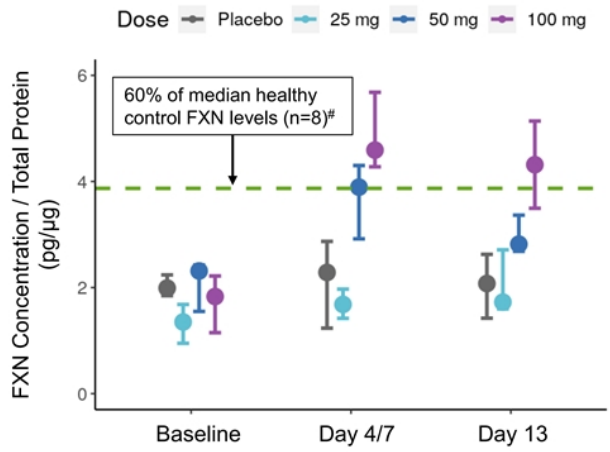
Benchmarking Clinical Relevance

- FXN levels in buccal cells and blood have been shown to correlate with neurological function in FA patients[1]
- Patients with FA only produce ~20-40% of normal frataxin levels depending on the tissue considered[2]
- Heterozygous carriers who show no signs of disease have FXN levels of ~50% of unaffected healthy persons[2]

Comparison to Healthy Controls

- FXN levels were measured in buccal cells from 8 healthy controls using the same assay and sampling technique employed in the Phase 1 MAD trial
- With daily administration, patients in Cohorts 2 & 3 of the Phase 1 MAD trial achieved median buccal cell FXN levels that were >60% of the median FXN levels observed in healthy controls

FXN* Levels By Dose Group (Buccal Cells)



*FXN levels measured via detection of peptide derived from mature FXN; #Data on file; Data represent median and 25th and 75th percentiles ; FXN levels from baseline, Day 4, & Day 13 measurements are shown for data derived from the 25 mg cohort; FXN levels from baseline, Day 7 & Day 13 measurements are shown for data derived from the 50 & 100 mg cohorts; Sample collection days varied in each cohort per the trial protocol. 1. Lazaropoulos et al. Ann Clin Transl Neurol. 2015 Aug; 2(8): 831–842; 2. E.C. Deutsch et al. Molecular Genetics and Metabolism 101 (2010) 238–245.

Clinical & Non-clinical Safety Data Support Initiation of the 4-Week, Phase 2 Dose Exploration Study's 25 mg Cohort

FDA cleared Phase 2 study's initiation following review of clinical and non-clinical data

SUMMARY OF MULTIPLE-ASCENDING DOSE (MAD) TRIAL SAFETY DATA

Repeated SC injections of CTI-1601 appear to be generally well tolerated at doses up to 100 mg administered daily for 13 days.

- No serious adverse events (SAEs), important medical events, or treatment-related severe adverse events were observed.
- Most common AEs were mild and moderate injection site reactions (ISR). At least one ISR was seen in 43% of patients receiving placebo, and all patients receiving CTI-1601 experienced ISRs. Most ISRs resolved within an hour after injection, and all ISRs resolved without intervention. There were no study discontinuations due to ISRs.
- Except for ISRs, the number and severity of AEs did not increase with increasing exposure to CTI-1601.
- Accumulation of CTI-1601 was not observed at the doses and dose regimens studied.

SUMMARY OF NON-HUMAN PRIMATE (NHP) DATA

- The clinical hold was put in place following deaths that occurred during the 26-week toxicology study in 3 out of a total of 34 NHPs. All 3 of these NHPs were in the two highest dose groups. All NHPs in the two lower dose groups survived to the end of the 26-week toxicology study.
- Based on AUC, C_{max}, and C_{trough} from the Phase 1 studies at the 25 mg and 50 mg levels, and the no observed adverse effect levels from the 4-, 13-, and 26-week toxicology studies, the safety margins calculated for CTI-1601 are generally greater than 10.
- Though the precise mechanism of toxicity in NHPs was not determined, we believe the toxicity was associated with accumulation and high levels of exposure as demonstrated by the safety margins. We believe the presence of persistent edema at the injection sites in some NHPs may explain the accumulation associated with adverse events, as well as higher plasma levels of CTI-1601. In the clinic, injection sites will be closely monitored and we intend to avoid the use of injection sites where persistent edema is present.

SC: Subcutaneous

Upcoming CTI-1601 Trials

Future Planned Trials Include:

Phase 2, four-week dose exploration study intended to identify dose and dose regimen for long-term studies. Expected to begin in Q4 2022.

Jive OLE trial for eligible patients who participated in SAD, MAD, and/or four-week dose exploration studies. Expected to begin in 2H 2023.

MAD trial in patients 2 to 17 years of age. Participants eligible to screen for Jive OLE trial. Expected to begin in 2H 2023.

Global double-blind placebo-controlled pivotal trial.

**Dose escalation in the Phase 2 trial and initiation of the Jive, pediatric MAD trials and global pivotal trial will be subject to the FDA lifting the partial clinical hold.*
OLE: Open-label extension

Summary: CTI-1601 Advancing to Phase 2 Trial

CTI-1601	Designed to address the root cause of Friedreich's ataxia by delivering mature FXN to mitochondria.
Clinical POC Data	Daily dosing of 50 mg or 100 mg of CTI-1601 for at least 7 days resulted in buccal cell FXN levels that met or exceeded those expected in phenotypically normal heterozygous carriers.
Next Steps	Initiate in Q4 2022 a Phase 2 dose exploration study in Friedreich's ataxia patients. Cohort 1 to evaluate 25 mg dose; dose escalation contingent on FDA review of cohort 1 data. Top-line data from both cohorts expected in 2H 2023.

Larimar Therapeutics

POC: Proof-of-concept

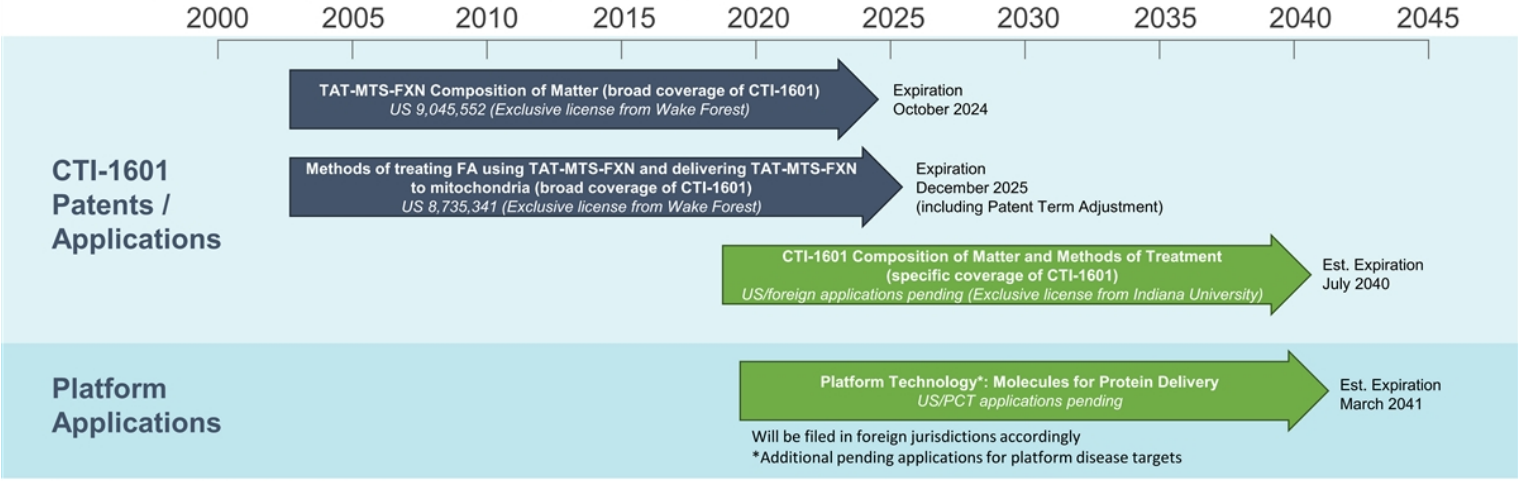

Larimar Therapeutics
Larimar Therapeutics
Corporate & Clinical Update
THANK YOU

Larimar Therapeutics

Appendix

Platform Technology is Supported by a Strong IP Portfolio

Pending CTI-1601 patent application, if issued, extends IP into 2040



Additional CTI-1601 IP protection

- CTI-1601 pending applications cover key biomarkers, analytical tools and quantification methods
- CTI-1601 is 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)

Larimar Therapeutics

Granted Pending

Phase 1 Top-line Data Demonstrated POC for CTI-1601 in FA

Safety	CTI-1601 appears to be generally well tolerated at doses up to 100 mg administered daily for 13 days
Pharmacodynamics	Daily dosing of CTI-1601 resulted in dose-dependent increases in FXN levels from baseline compared to placebo controls in all evaluated tissues
Pharmacokinetics	Pharmacokinetic analyses support evaluating a once-daily dosing regimen for CTI-1601
Conclusion	Daily subcutaneous (SC) administration of 50mg and 100mg doses of CTI-1601 resulted in FXN levels in buccal cells that are at, or in excess of, those we would expect to see in phenotypically normal heterozygous carriers (who have FXN levels of ~50% of unaffected persons)

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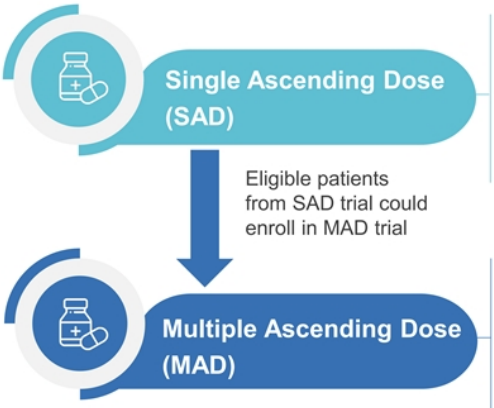
POC: Proof-of-concept

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

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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

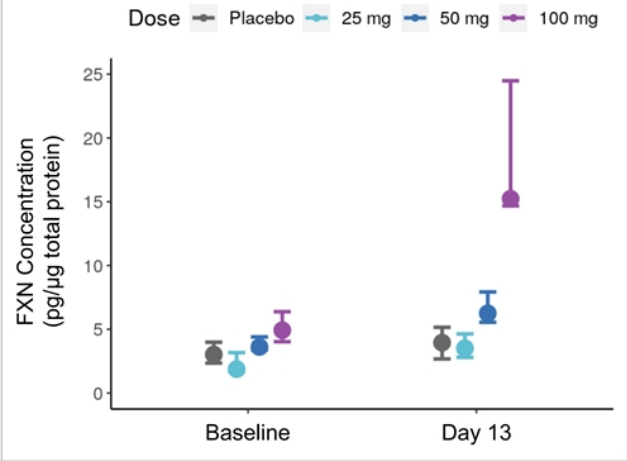
Parameter	Statistic	All placebo (n=7)	25 mg CTI-1601 (n=6)	50 mg CTI-1601 (n=7)	100 mg CTI-1601 (n=7)	All CTI-1601 (n=20)	Overall (n=27)
Age at Symptom Onset							
	Mean	14.1	24.0	19.3	11.9	18.1	17.1
	SD	5.34	14.48	6.21	6.72	10.37	9.39
	Median	15.0	18.0	19.0	10.0	18.0	16.0
	Min, Max	8,23	12,44	8,28	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

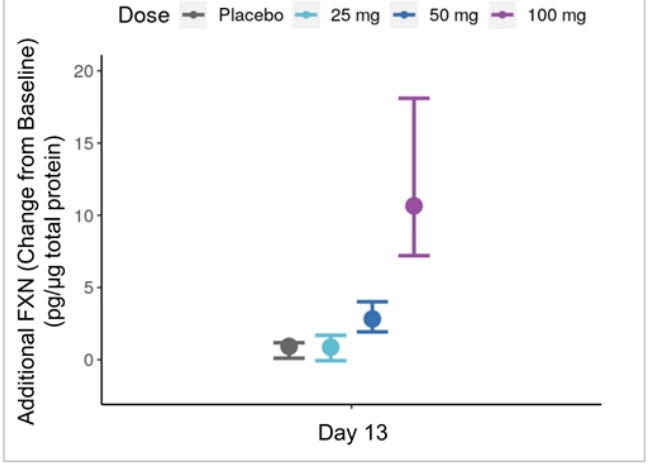
Dose Dependent Increases in FXN Levels Observed in Skin

Daily SC injections of 100 mg CTI-1601 resulted in an ~3 fold increase in FXN levels from baseline

FXN* Levels By Dose Group (Skin Biopsies)



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

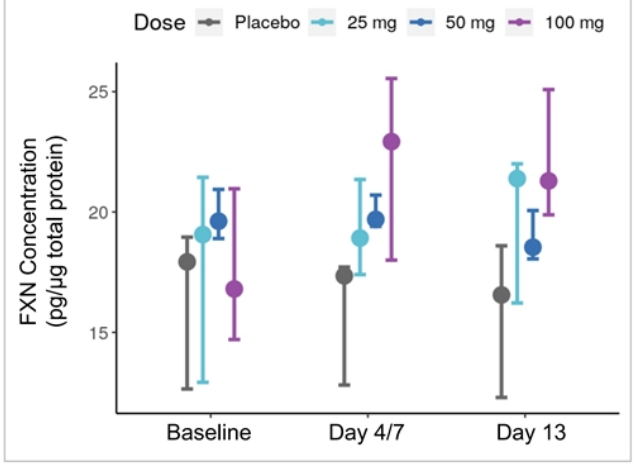


*FXN levels measured via detection of peptide derived from mature FXN; Data represent median and 25th and 75th percentiles

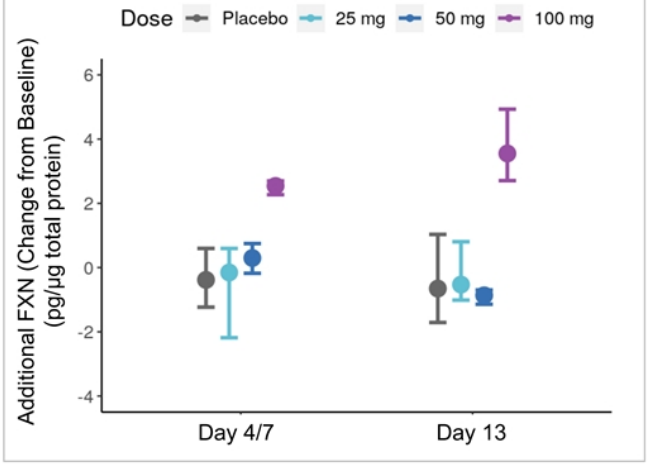
Dose Dependent Increases in FXN Levels Observed in Platelets with Daily Dosing

Daily SC injections of 100mg CTI-1601 resulted in increases in FXN levels from baseline compared to placebo

FXN* Levels By Dose Group (Platelets)



FXN* Change from Baseline By Dose Group (Platelets)



*FXN levels measured via detection of peptide derived from mature FXN; Data represent median and 25th and 75th percentiles; FXN levels from baseline, Day 4, & Day 13 measurements are shown for data derived from the 25 mg cohort; FXN levels from baseline, Day 7 & Day 13 measurements are shown for data derived from the 50 & 100 mg cohorts; Sample collection days varied in each cohort per the trial protocol

PK analyses support evaluating a once-daily dosing regimen for CTI-1601

Summary of 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 appears to be at or close to steady state exposure after 13 days of dosing 100 mg once daily

Leadership Team

Carole Ben-Maimon, MD
Chief Executive Officer

Impax teva

Michael Celano
Chief Financial Officer

OraSure Technologies Kensey Nash

Nancy Ruiz, MD, FACP, FIDSA
Chief Medical Officer

Schering-Plough MERCK

Jennifer Johansson, JD
VP, Regulatory Affairs & Counsel

Andrx SQUIRE PATTON BOGGS

Mohamed Hamdani
VP, Biometrics

Apellis Shire

David Bettoun, PhD
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VP, Finance & Operations

teva Cell Pathways, Inc.

Noreen Scherer
VP, Clinical Operations

Shire Wyeth Pharmaceuticals

Francis Michael Conway
VP, Controller

TELA BIO stryker

Strong Relationship with FARA

Company has strong relationship with Friedreich's Ataxia Research Alliance (FARA)

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

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

Scientific Advisory Board

Russell Clayton, DO (Chairman)

Former Chief Medical Officer at Alcresta Therapeutics, a medical device company

Former Senior Vice President of Research and Development at Discovery Labs, a pharmaceutical and medical device company

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

CTI-1601: Positive Mouse Model Data Support Development

Proof-of-Concept Demonstrated In Mouse Models of FA

Cardiac Knock Out Mouse Model Studies
(MCK-Cre FXN KO Mouse)

- Extended survival
- Demonstrated ability to deliver hFXN to mitochondria
- Increased in a dose dependent manner, succinate dehydrogenase (SDH) activity. SDH is an FXN dependent enzyme, whose activity is indicative of mitochondrial function
- Prevented left ventricle dilation and maintained function

Neurologic Knock Out Mouse Model Study
(Pvalb-CRE FXN KO Mouse)

- Prevented development of ataxic gait
- Showed that treated mice survive longer than untreated mice
- Demonstrated CNS penetration, as hFXN was present in brain, dorsal root ganglia & spinal cord

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CTI-1601 Extends Survival in FXN-deficient KO Mice

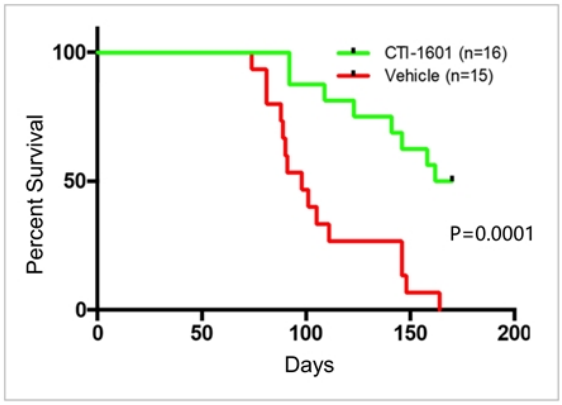
Initial Proof-of-Concept for FXN Replacement Therapy in Cardiac Mouse Model of FA

Median Survival of MCK-Cre FXN-KO Mice

- 166 days (CTI-1601) vs. 98 days (Vehicle)
- CTI-1601 was administered 10 mg/kg SC every other day

Survival beyond vehicle mean (107.5 days)

- 87.5% (CTI-1601) vs. 33% (Vehicle)
- Demonstrates that CTI-1601 is capable of delivering sufficient amounts of FXN to mitochondria



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

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CTI-1601 Prevents The Development of Ataxic Gait in KO mice

In-Vivo Efficacy Data in Neurologic KO Mouse Model

Pvalb-Cre FXN-KO mouse

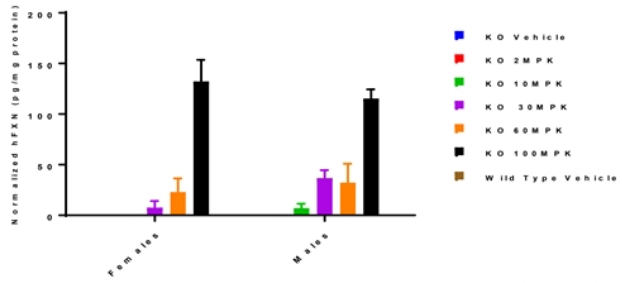
Single dose level: 10 mg/kg CTI-1601 or vehicle given intraperitoneally three times per week

- hFXN replacement with CTI-1601 **prevents the development of ataxic gait**
- CTI-1601-treated mice **survive longer** than untreated mice
- Human frataxin **present in brain, dorsal root ganglia and spinal cord** demonstrating central nervous system penetration

CTI-1601 Delivers hFXN to Mitochondria in KO Mice

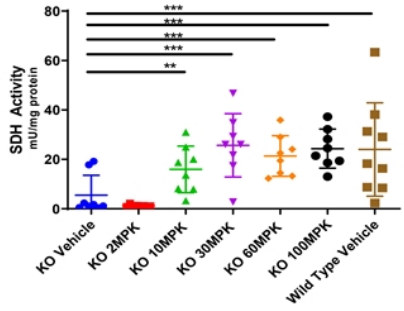
- hFXN concentration within mitochondria increases in a dose-dependent manner
- Given subcutaneously, CTI-1601 functionally replaces hFXN in mitochondria of KO mice
- Succinate dehydrogenase (SDH) activity, which is indicative of mitochondrial function, increases in a dose-dependent manner after administration of CTI-1601; activity plateaus at 30 mg/kg and is equivalent to activity in wild type animals
- Demonstrated normalization of gene expression in cardiac tissue

Normalized Mitochondrial FXN (Heart)



MPK = mg/kg

Normalized SDH Activity (Muscle)



MPK = mg/kg

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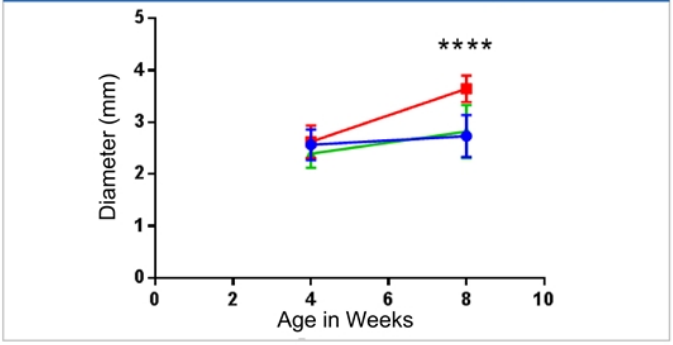
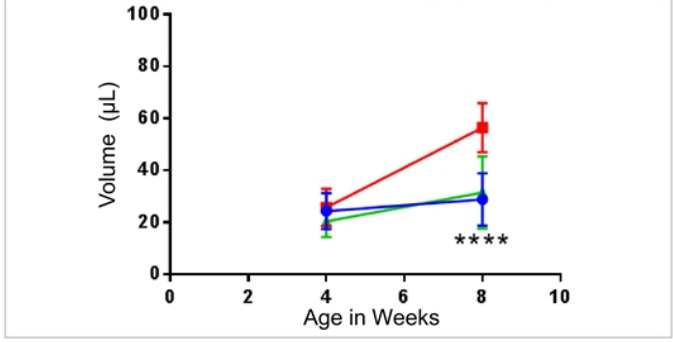
CTI-1601 Prevents Left Ventricle Dilation in KO Mice

- 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 CTI-1601 (10 mg/kg every other day)
- CTI-1601-treated mice have similar LV volume as healthy controls; echocardiogram shows significant differences between vehicle and CTI-1601 treated (10 mg/kg every other day) KO mice

Left Ventricle Internal Diameter (Systole)



Left Ventricle Volume (Systole)



KO: CTI-1601 KO: Vehicle Wild-type: Vehicle

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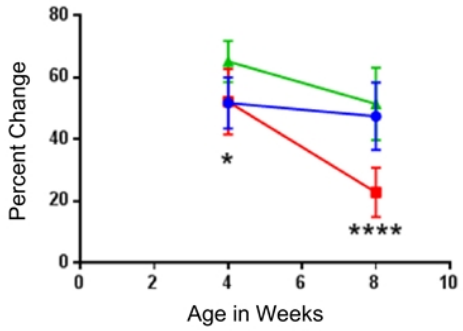
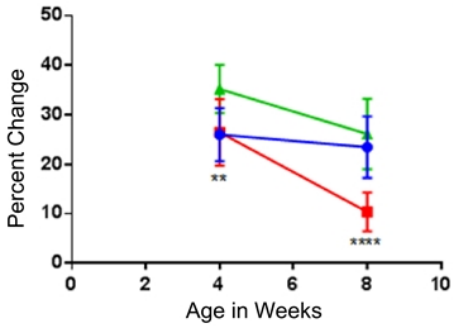
CTI-1601 Preserves Left Ventricle Function in KO Mice

- Left ventricular (LV) function drops significantly in vehicle treated mice by week 8
- CTI-1601-treated (10 mg/kg every other day) mice have similar LV as healthy controls; echocardiogram shows significant differences between vehicle and CTI-1601 treated KO mice

Left Ventricle Ejection Function



Left Ventricle Fractional Shortening



KO: CTI-1601 KO: Vehicle Wild-type: Vehicle

Larimar Therapeutics