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): May 11, 2021

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)**	**(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.

Investor Presentation

On May 11, 2021, Larimar Therapeutics, Inc. (the "***Company***") posted on its website an updated slide presentation, which is attached as Exhibit 99.1 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.

Press Release

On May 11, 2021, the Company issued a press release announcing topline data from the Company's Phase 1 multiple ascending dose (MAD) clinical trial (n=27) evaluating CTI-1601 as a treatment for Friedreich's ataxia (FA) and provided additional updates regarding future activities planned for 2021. A copy of this press release is filed as Exhibit 99.2 hereto and incorporated herein by reference.

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	Larimar Therapeutics, Inc. Corporate Presentation, dated May 11, 2021*
99.2	Press Release of Larimar Therapeutics, Inc., dated May 11, 2021*

* 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: May 11, 2021

Exhibit 99.1

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Phase 1 Topline Data Conference Call

May 11, 2021

Forward Looking Statements

This presentation contains forward-looking statements that are based on the beliefs and assumptions of Larimar Therapeutics, Inc. (the “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 our business, 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 success, cost and timing of the Company’s product development activities, non-clinical studies and clinical trials, including CTI-1601 clinical milestones; that 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 clinical trials, and that clinical trial data are subject to differing interpretations and assessments; the ongoing impact of the COVID-19 pandemic on the Company’s clinical trials, manufacturing, regulatory and nonclinical study timelines, ability to raise additional capital and general economic conditions; the Company’s ability to optimize and scale CTI-1601’s manufacturing process; the Company’s ability to obtain regulatory approval 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 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

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
Has Orphan Drug (US & EU), Rare Pediatric Disease (US), and Fast Track (US) designations for FA

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

Series A investment by Deerfield in Nov. 2016; went public through a reverse merger/PIPE in May 2020
Shareholder base includes high-quality institutional investors

Strong balance sheet
~$81.4 million in cash as of March 31, 2021, with projected runway into 1H 2022

FXN: Frataxin

Executive Summary of Phase 1 POC Data

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 healthy persons)

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POC: Proof-of-concept

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 ~5,000 patients in the U.S. & ~20,000 patients in the EU

>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

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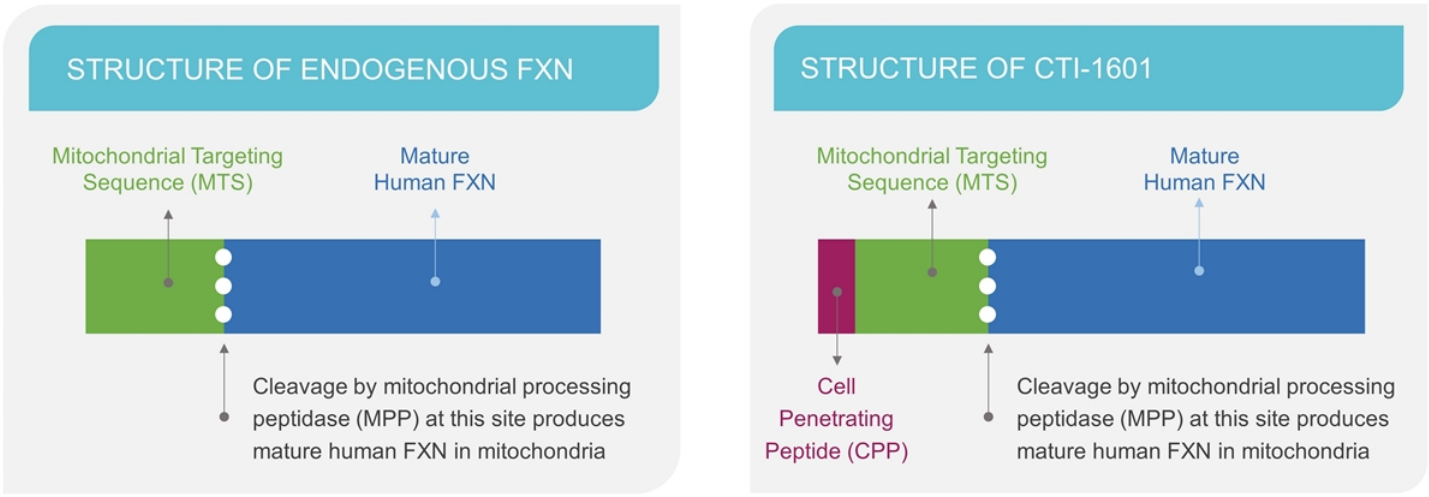
1. E.C. Deutsch et al. Molecular Genetics and Metabolism 101 (2010) 238–245

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

CTI-1601 Maintains the Cleavage Site Between the MTS and Mature Human FXN



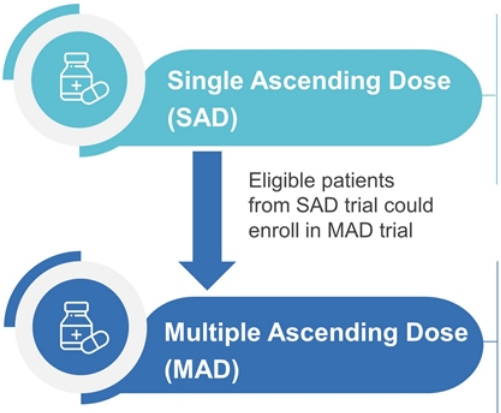
The maintenance of the cleavage site allows the CPP and MTS to be removed by mitochondrial processing peptidase to produce mature human FXN in the mitochondria

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 with analysis ongoing

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 with analysis ongoing

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MAD Trial Patient Enrollment

16 out of 28 patients who participated in the SAD trial enrolled in the MAD trial

MAD Trial Patient Enrollment (n=27)		
Parameter	**Statistic**	**Overall**
Participated in SAD trial?		
Yes	n (%)	16 (59%)
No	n (%)	11 (41%)
Cohort 1 (25 mg) Active vs. Placebo		
Active	n (%)	6 (75%)
Placebo	n (%)	2 (25%)
Cohort 2 (50 mg) Active vs. Placebo		
Active	n (%)	7 (78%)
Placebo	n (%)	2 (22%)
Cohort 3 (100 mg) Active vs. Placebo		
Active	n (%)	7 (70%)
Placebo	n (%)	3 (30%)

Multiple Ascending Dose Study Design

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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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)

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SD: Standard deviation

MAD Trial Patient Disease Characteristics

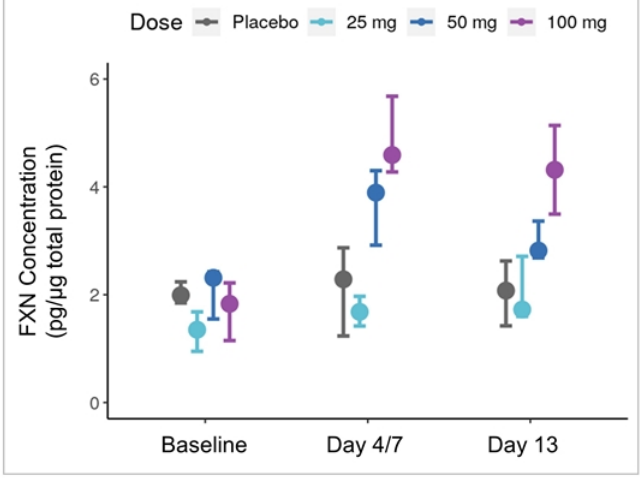
Parameter	Statistic	All placebo (n=7)	25 mg CTI-1601 (n=6)	50 mg CTI-1601 (n=7)	100 mg CTI-1601 (n=7)	All CTI-1601 (n=20)	Overall (n=27)
Age at Symptom Onset							
	Mean	14.1	24.0	19.3	11.9	18.1	17.1
	SD	5.34	14.48	6.21	6.72	10.37	9.39
	Median	15.0	18.0	19.0	10.0	18.0	16.0
	Min, Max	8,23	12,44	8,28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

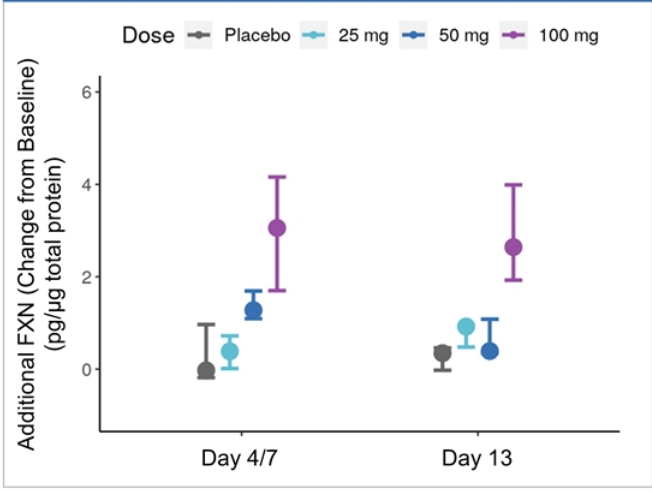
Dose Dependent Increases in FXN Levels Observed in Buccal Cells

Daily SC injections of 100 mg CTI-1601 resulted in an ~2.5 fold increase in FXN levels from baseline

FXN* Levels By Dose Group (Buccal Cells)



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

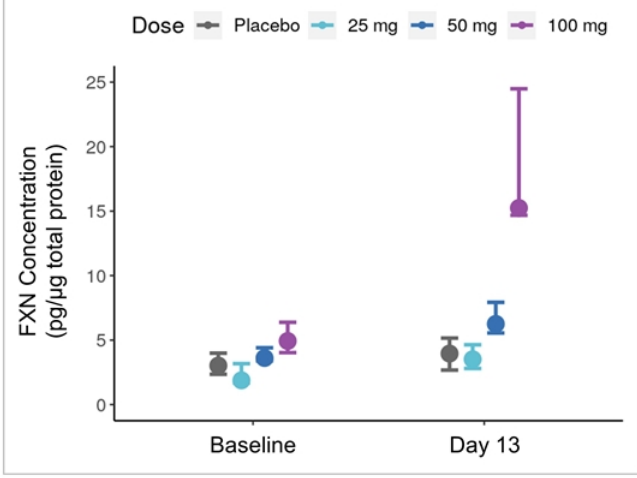


*FXN levels measured via detection of peptide derived from mature FXN; Data represent median and 25th and 75th percentiles; FXN levels from baseline, Day 4, & Day 13 measurements are shown for data derived from the 25 mg cohort; FXN levels from baseline, Day 7 & Day 13 measurements are shown for data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

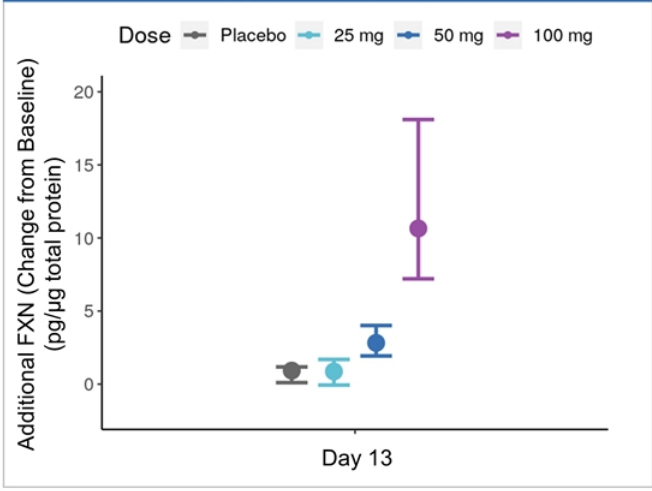
Dose Dependent Increases in FXN Levels Observed in Skin

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)



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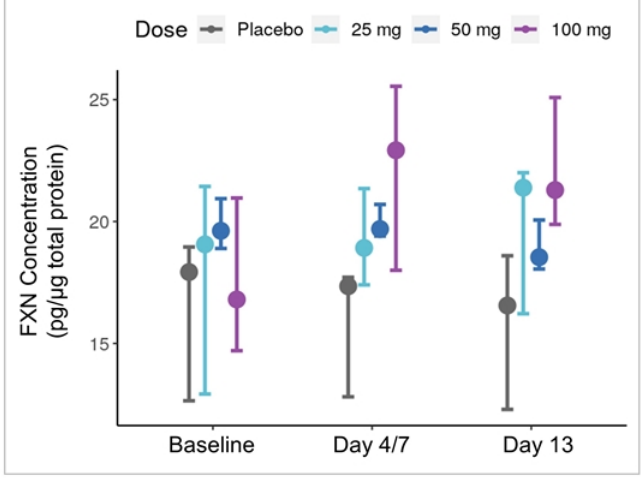
*FXN levels measured via detection of peptide derived from mature FXN; Data represent median and 25th and 75th percentiles

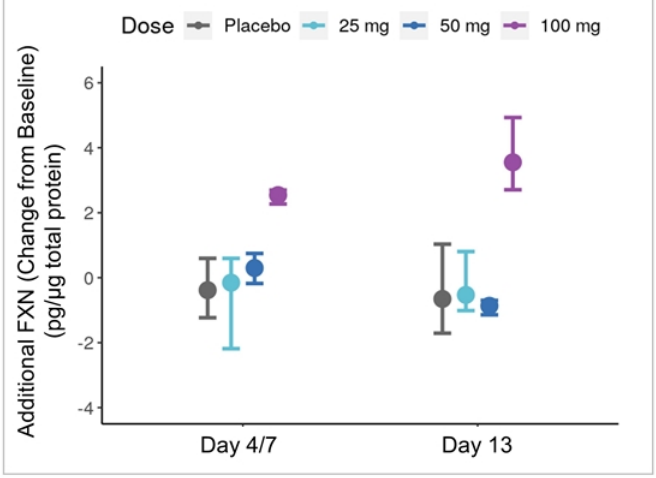
Dose Dependent Increases in FXN Levels Observed in Platelets with Daily Dosing

Daily SC injections of 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)

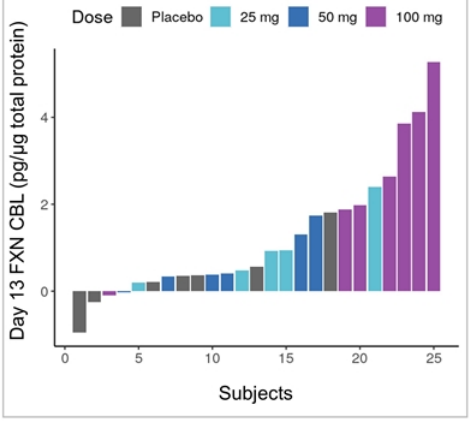


*FXN levels measured via detection of peptide derived from mature FXN; Data represent median and 25th and 75th percentiles; FXN levels from baseline, Day 4, & Day 13 measurements are shown for data derived from the 25 mg cohort; FXN levels from baseline, Day 7 & Day 13 measurements are shown for data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

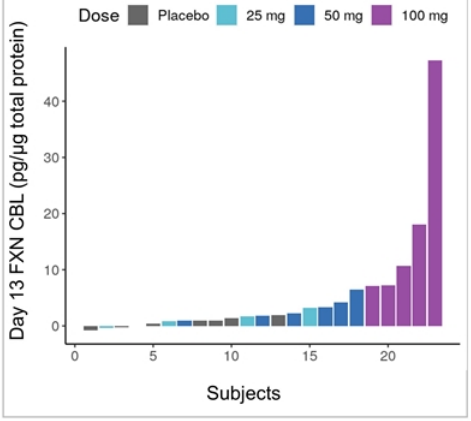
Increases in FXN Correlated with Increasing CTI-1601 Dose

Individual patient data further supports the dose-dependent effects of CTI-1601 in all tissues studied

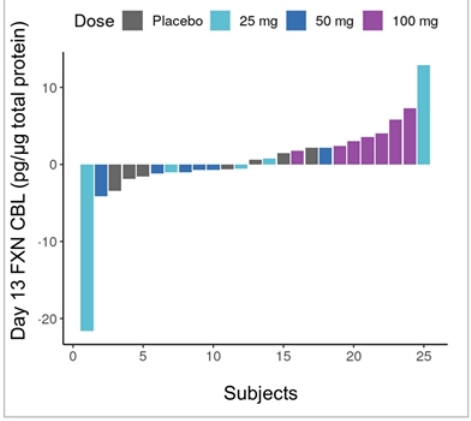
Buccal Cells Day 13 FXN* Change from Baseline



Skin Biopsies# Day 13 FXN* Change from Baseline



Platelets Day 13 FXN* Change from Baseline



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*FXN levels measured via detection of peptide derived from mature FXN; #Two patients in the 100 mg cohort declined skin biopsies
Day 13 observation excluded from one subject in 25 mg group that did not get a Day 13 dose.

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

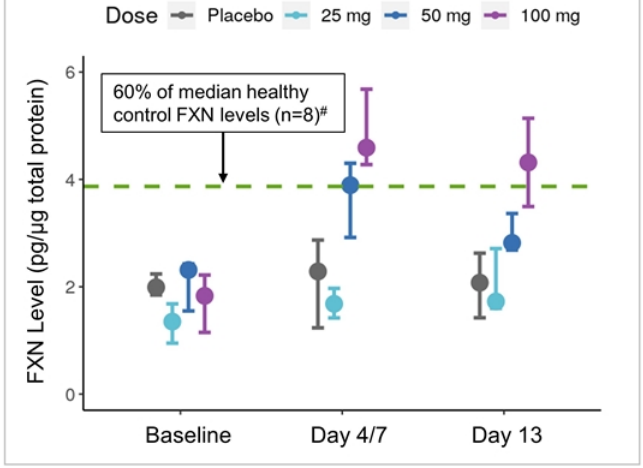
Benchmarking Clinical Relevance

- 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
- Data from additional healthy control buccal cells, skin, and platelets will be collected in a separate non-interventional study

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.

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

Summary of MAD trial safety data:

Repeated doses (25 mg, 50 mg, and 100 mg) of CTI-1601 or placebo were administered subcutaneously. 27 patients were dosed in the trial. 26 patients completed the trial. 1 patient receiving CTI-1601 in Cohort 2 (50 mg) withdrew after experiencing mild/moderate symptoms (nausea and vomiting).

- No serious adverse events (SAEs), important medical events, or treatment-related severe adverse events
- The most common adverse events were mild and moderate injection site reactions (at least one injection site reaction was seen in 43% of placebo patients and in 100% of CTI-1601 patients)
- The number and severity of adverse events did not increase with increasing exposure to CTI-1601

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-160
- Mean half life of CTI-1601 in plasma was approximately 11 hours

Phase 1 Topline Data Demonstrated POC for CTI-1601 in FA

FXN levels in buccal cells & blood have been shown to correlate with disease severity in FA patients[1]

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

The most common AEs were mild and moderate injection site reactions

No SAEs have been reported

Frataxin Measurements

Daily SC injections of CTI-1601 resulted in dose-dependent increases in FXN levels from baseline compared to placebo controls in all evaluated tissues

With daily dosing (50mg and 100mg), achieved median FXN levels that were >60% of the median FXN levels observed in healthy controls

Pharmacokinetic Data

CTI-1601 was quickly absorbed after subcutaneous administration

Dose-proportional increases in exposure observed with increasing doses of CTI-1601

Data support evaluating a once-daily dosing regimen for CTI-1601

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1. Lazaropoulos et al. Ann Clin Transl Neurol. 2015 Aug; 2(8): 831–842; POC: Proof-of-concept; OLE: Open label extension; AE: Adverse events

Upcoming Trials and Regulatory Interactions

Additional analyses from the Phase 1 program planned for presentation at a scientific meeting

Future Planned Trials and Regulatory Interactions Include:

Continued interactions with FDA regarding clinical trials and non-clinical toxicology studies

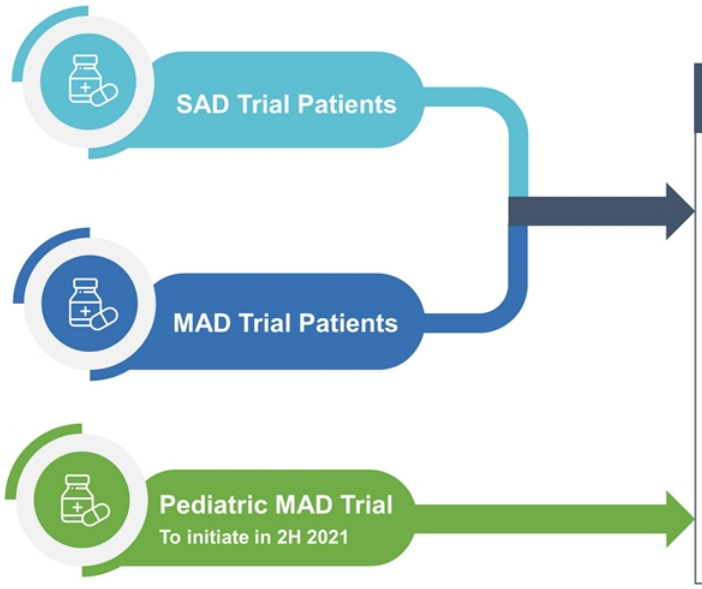
Jive open label extension (OLE) trial for eligible patients who participated in SAD or MAD trials (expected initiation 2H 2021)

MAD trial in patients under 18 years of age (expected initiation 2H 2021). Participants eligible to screen for Jive OLE trial

Global double-blind placebo-controlled pivotal trial (expected initiation as early as 2H 2022)

Expect to Initiate Two Additional Trials in 2H 2021

Patients from SAD, MAD, and pediatric trials are eligible to screen for the Jive open label extension trial



Jive OLE Trial

Dose Level: To be determined based on additional analysis of data from SAD and MAD trials

Treatment Duration: Planned for 24 months with any necessary extensions

Number of Subjects: Up to 50

Dose Regimen: Once-daily doses administered subcutaneously

External Control Arm: Derived from Critical Path Institute Data

1º Endpoint: Safety and tolerability

Key 2º Endpoints: Long-term PD; efficacy assessments

Expected Initiation: 2H 2021

Conclusion

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 healthy persons)

Larimar Therapeutics

Larimar Therapeutics Reports Positive Topline Phase 1 Clinical Trial Data Showing Dose-Dependent Increases in Frataxin Levels in Patients with Friedreich's Ataxia

- Data demonstrate proof-of-concept by showing that daily subcutaneous injections of CTI-1601 for up to 13 days resulted in dose-dependent increases in frataxin levels from baseline compared to placebo in all evaluated tissues

- Data show that frataxin levels achieved in peripheral tissues (buccal cells) following daily 50 mg and 100 mg subcutaneous injections of CTI-1601 were at or in excess of those that would be expected in phenotypically normal heterozygous carriers

- Safety data indicate that repeated subcutaneous injections of CTI-1601 were generally well tolerated at doses up to 100 mg administered daily for 13 days

- Company management to host webcast and conference call today at 8:00 a.m. ET

Bala Cynwyd, PA, May 11, 2021 – Larimar Therapeutics, Inc. ("Larimar") (Nasdaq: LRMR), a clinical-stage biotechnology company focused on developing treatments for Friedreich's ataxia (FA) and other complex rare diseases, today announced topline data from its Phase 1 multiple ascending dose (MAD) clinical trial (n=27) evaluating CTI-1601 as a treatment for FA.

FA patients participating in the trial received subcutaneous injections of CTI-1601 or placebo at increasing dose levels and frequencies over a 13-day period. Patients in Cohort 1 were dosed with 25 mg of CTI-1601 or placebo daily for four days, and then every third day until Day 13. Cohort 2 patients were dosed with 50 mg of CTI-1601 or placebo daily for seven days, and then once every other day until Day 13. Patients in Cohort 3 received daily injections of 100 mg CTI-1601 or placebo for thirteen days.

Data show that repeated subcutaneous administration of CTI-1601 resulted in dose-dependent increases in frataxin (FXN) levels from baseline compared to placebo controls. These dose-dependent increases in frataxin levels were seen in all evaluated tissues (buccal cells, skin biopsies, and platelets) with daily dosing. The median change from baseline in frataxin levels observed for each dosing group and tissue are shown in the table below.

FXN Change from Baseline in Buccal Cells
Units: pg FXN / µg total protein
Data presented as: n median (25th percentile, 75th percentile)

Dose Group	Day 4/7*		Day 13	
Placebo (n=7)	-0.03	(-0.18, 0.97)	0.35	(-0.02, 0.46)
Cohort 1 Active (25 mg, n=6)	0.39	(0.01, 0.72)	0.92	(0.48, 0.94)
Cohort 2 Active (50 mg, n=6)	1.28	(1.09, 1.69)	0.39	(0.35, 1.08)
Cohort 3 Active (100 mg, n=7)	3.06	(1.70, 4.16)	2.64	(1.93, 3.99)

FXN Change from Baseline in Skin Biopsies#
Units: pg FXN / µg total protein
Data presented as: median (25th percentile, 75th percentile)

Dose Group	Day 4/7*	Day 13
Placebo (n=7)	N/A	0.92 (0.10, 1.17)
Cohort 1 Active (25 mg, n=6)	N/A	0.87 (-0.07, 1.69)
Cohort 2 Active (50 mg, n=6)	N/A	2.82 (1.93, 4.01)
Cohort 3 Active (100 mg, n=7)	N/A	10.6 (7.20, 18.1)

FXN Change from Baseline in Platelets
Units: pg FXN / µg total protein
Data presented as: median (25th percentile, 75th percentile)

Dose Group	Day 4/7*		Day 13	
Placebo (n=7)	-0.38	(-1.23, 0.60)	-0.65	(-1.71, 1.03)
Cohort 1 Active (25 mg, n=6)	-0.15	(-2.18, 0.60)	-0.53	(-1.01, 0.80)
Cohort 2 Active (50 mg, n=6)	0.30	(-0.18, 0.75)	-0.87	(-1.14, -0.70)
Cohort 3 Active (100 mg, n=7)	2.54	(2.27, 2.70)	3.55	(2.71, 4.93)

* FXN levels from Day 4 and 13 measurements are shown for data derived from Cohort 1. FXN levels from Day 7 and 13 measurements are shown for data derived from Cohorts 2 and 3. Sample collection days varied in each cohort per the clinical trial protocol.

\# Skin biopsies were not collected on Day 4 or Day 7 per the clinical trial protocol. Skin biopsies were optional per the protocol and were collected in 5 of the 7 patients in Cohort 3.

“The ability of CTI-1601 to elevate frataxin in the evaluated tissues strongly supports its continued clinical evaluation, as frataxin levels in buccal cells and blood have been shown to be associated with disease severity in FA patients,” said Nancy M. Ruiz, MD, Chief Medical Officer of Larimar. “Notably, daily subcutaneous CTI-1601 administration resulted in median frataxin levels in buccal cells that are similar to, and even exceed, the levels we would expect to see in phenotypically normal heterozygous carriers (who have frataxin levels that are approximately 50% of unaffected healthy persons). These findings are exciting and represent a critical step forward in CTI-1601’s development as a frataxin replacement therapy for patients with FA. We believe CTI-1601 has the potential to address a critical unmet need, as FA is caused by patients’ inability to produce sufficient amounts of frataxin and current treatment options for this progressive and devastating disease are limited to symptom management. It is also important to note that CTI-1601 is the only drug candidate that we are aware of in clinical development that is designed to supplement frataxin levels in patients with FA, thus addressing the root cause of the disease.”

Safety data from the trial indicate that repeated subcutaneous injections of CTI-1601 were generally well tolerated at doses up to 100 mg administered daily for 13 days. A summary of the safety data, as well as pharmacokinetic data are shown below.

Safety:

- No serious adverse events (SAEs), important medical events, or treatment-related severe adverse events were reported in the trial
- The most common adverse events were mild and moderate injection site reactions (at least one injection site reaction was seen in 43% of placebo patients and in 100% of CTI-1601 patients)
- The number and severity of adverse events did not increase with increasing exposure to CTI-1601
- Of the 27 patients dosed, 26 completed the trial with one Cohort 2 patient receiving CTI-1601 withdrawing after experiencing mild to moderate nausea and vomiting

Pharmacokinetics:

- CTI-1601 was quickly absorbed after subcutaneous administration
- Dose-proportional increases in exposure were observed with increasing doses of CTI-1601
- The mean half-life of CTI-1601 in plasma was approximately 11 hours

Patients who completed the single ascending dose (SAD) and/or MAD clinical trials are eligible to screen for an open-label extension clinical trial, the Jive study, which Larimar expects to initiate in the second half of 2021. Larimar also expects to initiate a MAD clinical trial in patients under 18 years of age in the second half of 2021. Patients completing this pediatric trial will also be eligible to screen for the Jive study.

Carole Ben-Maimon**,** MD, President and Chief Executive Officer of Larimar, commented, "We are thrilled with these data, which demonstrate proof-of-concept for CTI-1601 and highlight its safety and tolerability profile. Our ability to generate these results amid the pandemic speaks to the commitment of the patients who participated in the trials and the broader FA community. It is also a testament to the great work being done by the Friedreich's Ataxia Research Alliance and the talent of our employees, partners, and investigators. We extend our sincerest thanks to all of these individuals for the important roles they have played in CTI-1601's clinical development to date. We look forward to sharing additional quantitative analyses from our Phase 1 clinical trials at a future scientific meeting. We anticipate this data will inform the design of our planned future clinical trials for CTI-1601."

Conference Call and Webcast

Larimar will host a conference call and webcast today, May 11, 2021 at 8:00 a.m. ET. To access the webcast, please visit this link to the event. To participate by phone please dial 855-327-6837 (domestic) or 631-891-4304 (international) and refer to conference ID 10014696. Following the live event, the archived webcast will be available for 90 days.

About the Phase 1 MAD Clinical Trial

The Phase 1 MAD clinical trial was a double-blind, placebo-controlled, randomized clinical trial designed to assess the safety of subcutaneously administered CTI-1601 versus placebo in adult subjects with FA. The primary objective was to assess the safety and tolerability of multiple ascending doses of CTI-1601 in subjects with FA. Key secondary endpoints included pharmacokinetic and pharmacodynamic analyses following increasing multiple doses of subcutaneously administered CTI-1601. Patients from the MAD clinical trial are eligible to screen for an open-label extension trial that Larimar expects to initiate in the second half of 2021.

About CTI-1601

CTI-1601 is a recombinant fusion protein intended to deliver human frataxin into the mitochondria of patients with Friedreich's ataxia who are unable to produce enough of this essential protein. Currently in Phase 1 clinical trials in the U.S., 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) and orphan drug designation by the European Commission.

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 currently being evaluated in a Phase 1 clinical program in the U.S. as a potential treatment for FA. 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 CTI-1601 and other planned product candidates, Larimar's planned research and development efforts, 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, the success, cost and timing of Larimar's product development activities, non-clinical studies and clinical trials, including CTI-1601 clinical milestones; that 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 clinical trials, and assessments; the ongoing impact of the COVID-19 pandemic on Larimar's future clinical trials, manufacturing, regulatory and nonclinical study timelines, ability to raise additional capital and general economic conditions; Larimar's ability to optimize and scale CTI-1601's manufacturing process; Larimar's ability to obtain regulatory approval 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 the 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 views 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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