FGCL-3019-079 OVERVIEW

Open-Label, Single-Arm Phase 2 Study in Non-Ambulatory Subjects with Duchenne Muscular Dystrophy

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PROJECT PARENT MUSCULAR DYSTROPHY 2018 ANNUAL CONFERENCE

FibroGen

Pamrevlumab Background

Pamrevlumab (FG-3019): a novel investigational agent for treating fibrotic and fibro-proliferative diseases

- Pamrevlumab is a recombinant human monoclonal antibody that inhibits the activity of connective tissue growth factor (CTGF), a critical mediator in the progression of fibrosis and related serious diseases. Pamrevlumab has been studied in several indications including pancreatic ductal adenocarcinoma (PDAC) and idiopathic pulmonary fibrosis (IPF)
- 11 Phase 1 and Phase 2 clinical trials completed or ongoing
- Approximately 530 patients have received pamrevlumab
- About half of the patients have received treatment for >6 months

Pancreatic cancer

- Completed Phase 1/ 2 trial in 75 patients showing dose-related improvement in survival (*J Cancer Clin Trials 2017, 2:1)*
- Ongoing Phase 1/ 2 trial in 37 patients with locally advanced unresectable pancreatic cancer
- All patients completed treatment and are currently in long-term follow-up

Pamrevlumab Background

Idiopathic Pulmonary Fibrosis (IPF)

- Completed a one-year, open-label Phase 2 trial in 89 patients showing reversal of lung fibrosis and improved lung function in some patients (*Eur Respir J 2016; 47: 1481–1491*)
- Completed randomized, placebo-controlled Phase 2b trial with 103 patients (PRAISE study)
- Pamrevlumab did better than placebo at preserving lung function according to FVC measurements
- Study results presented at the ERS International Congress 2017 and the ATS International Conference in 2018

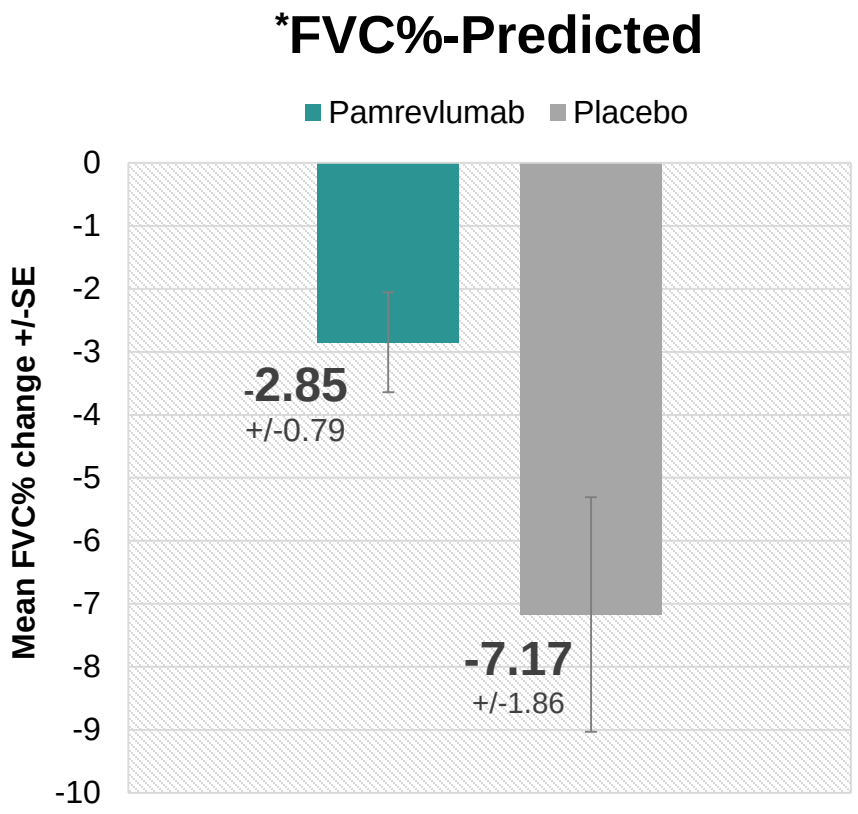
Safety

- No safety signals to date that might prevent further development of the drug
- Well tolerated and no dose-limiting toxicities observed to date

PRAISE in IPF

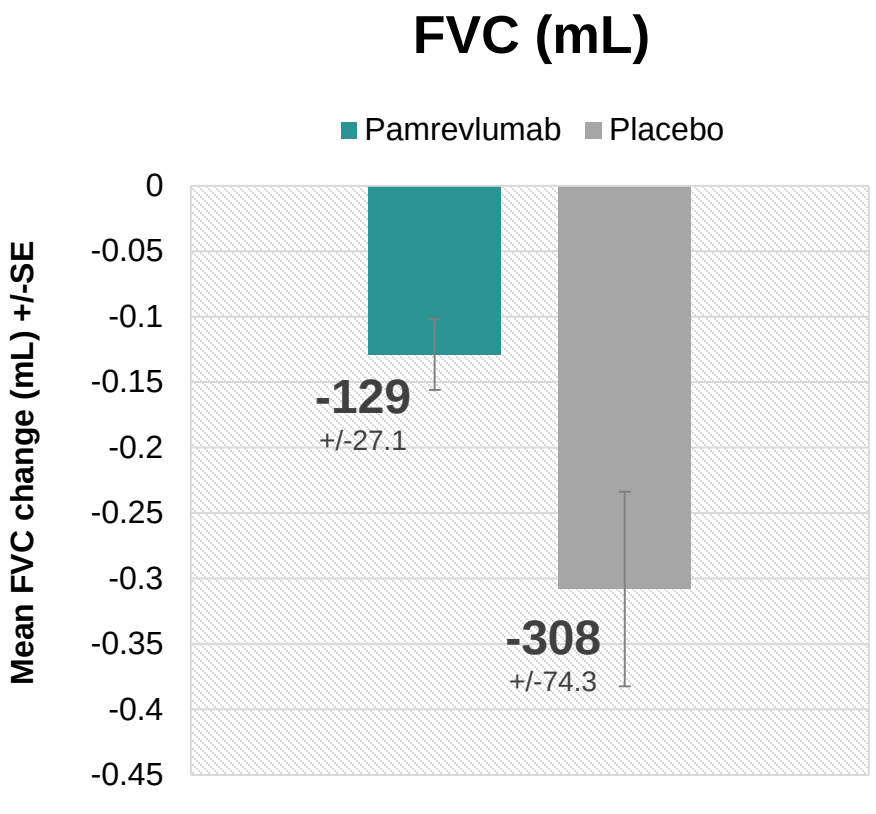
Randomized, Double-Blind, Placebo-Controlled Phase 2 Study to Evaluate the Safety and Efficacy of Pamrevlumab (FG-3019) in Patients with Idiopathic Pulmonary Fibrosis

Efficacy Analysis from PRAISE, Primary Endpoint*: Mean Change from Baseline at Week 48 in FVC Random Coefficients Model (Linear Slope Method)



p-value = 0.0331

FVC%-Predicted Difference: 4.33%
Relative Difference: 60%



p-value = 0.0249

Absolute FVC Difference: 178mL
Relative Difference: 58%

Safety Summary

- The majority of the treatment emergent adverse events were mild to moderate in severity and were ≤ Grade 3
- Treatment emergent serious adverse events were infrequent during the study and mostly respiratory-related
- Pamrevlumab infusions were generally well tolerated, mild to moderate in severity (no serious AEs observed) and did not lead to study discontinuations
- In general, pamrevlumab was well tolerated in IPF and warrants further clinical investigation

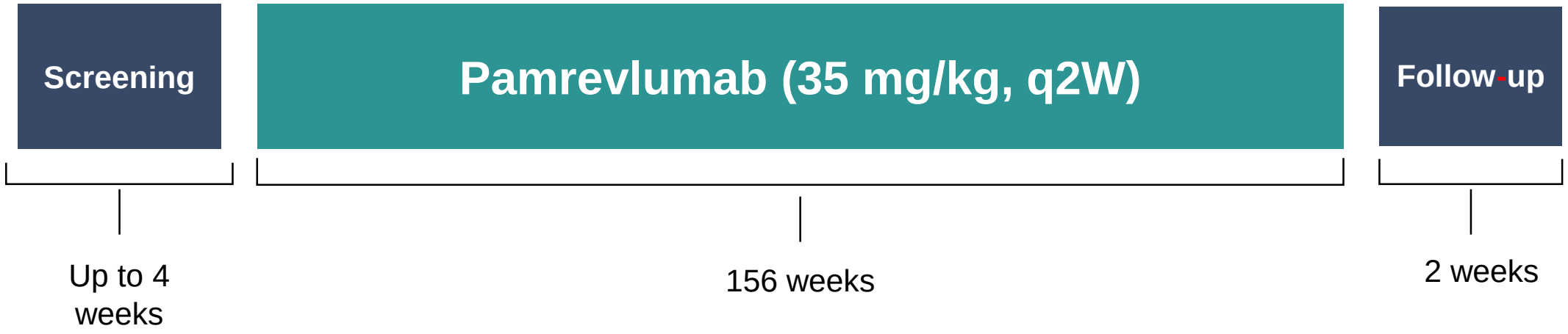
STUDY FGCL-3019-079

Open-Label, Single-Arm Phase 2 Study in Non-Ambulatory Subjects with Duchenne Muscular Dystrophy

Study FGCL-3019-079 Design

- An open-label, single-arm Phase 2 study in non-ambulatory DMD patients
- Each subject will receive IV infusions of pamrevlumab (35 mg/kg every 2 weeks, not to exceed 150 cc/hour) for up to 156 weeks
- All subjects are closely monitored for safety

DMD 079- Study (N=21)



Key Inclusion Criteria

- At least 12 years of age
- Non-ambulatory
- Brooke Score for Arms and Shoulders ≤5
- Diagnosis of DMD by medical history and confirmed Duchenne mutation in available genetic testing using a validated genetic test
- Able to perform spirometry
- Able to undergo cardiac and extremity (upper arm) MRI
- Percent predicted FVC between 40 and 90, inclusive
- At least one historical FVC% predicted value within 18 months of baseline
- Left ventricular ejection fraction ≥45% as determined by cardiac MRI at screening or within 3 months prior to Day 0
- Patients currently receiving heart failure cardiac medications must achieve a stable regimen for at least 3 months prior to screening
- On a stable dose of corticosteroids for a minimum of 6 months, with no substantial change in dosage for a minimum of 3 months

Key Exclusion Criteria

- Requires ≥16 hours continuous ventilation
- Anticipated spine surgery within 156 weeks
- Severe uncontrolled heart disease including any of the following:
  - Need for IV diuretics or inotropic support within 3 months prior to screening
  - Hospitalization for a heart failure exacerbation or arrhythmia in last 3 months
  - Arrhythmia requiring anti-arrhythmic therapy
- Hospitalization due to respiratory failure in the last 6 weeks
- Poorly controlled asthma or underlying lung disease
- BMI ≥40 kg/m2 or weight >117 kg
- Exposure to another investigational drug or approved product for DMD within 28 days prior to start of study treatment (or 5 half-lives, whichever is longer) with the exception of deflazacort

Status of Study 079

- Enrollment is complete, with 21 subjects currently on study
- 10 U.S. sites
- An interim analysis will be conducted after at least 10 to 12 subjects have completed 52 weeks of treatment
- Summary of patient exposure to pamrevlumab in days:

Statistics	Study 079 (N=21)
Mean (SD) (days)	326.4 (243.0)
Range (days)	29, 872
PEY* (years)	18.77
*PEY = All patients' duration on treatment/365.25	

- Currently, 16 subjects have been on the study for more than 6 months and 3 subjects >2 years

Site ID	All Subjects (N=21) n (%)
7901	1 (4.8)
7903	4 (19.0)
7904	4 (19.0)
7905	1 (4.8)
7908	4 (19.0)
7910	2 (9.5)
7912	2 (9.5)
7913	1 (4.8)
7920	1 (4.8)
7921	1 (4.8)
All Sites	21 (100.0)

Summary of Safety

- All subjects are currently on the study
- No death on the study
- 90% of subjects had TEAEs, none of the adverse events led to treatment discontinuation
- All TEAEs were mild to moderate in severity and were graded 1-3 per CTCAE. There were no grade >4 adverse events reported to date
- All vitals and labs were within accepted ranges for patients on the study

QUESTIONS?

Thank you