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hope through rigorous science

ML BioSolutions

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FORTIFY Phase 3 Interim Analysis Topline Results of BBP-418 in LGMD2I/R9

October 27, 2025

BBP-418 is an investigational therapy and has not been approved for use by any regulatory agency.

Seamus
Living with LGMD2I/R9

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Agenda

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2	**Introduction**	Christine Siu Chief Executive Officer, ML Bio Solutions, a BridgeBio Company
3	**FORTIFY Phase 3 interim analysis topline results**	Douglas Sproule, MD, MSc Chief Medical Officer, ML Bio Solutions, a BridgeBio Company
4	**Glycosylated αDG overview & results**	Uma Sinha, PhD Chief Scientific Officer, BridgeBio
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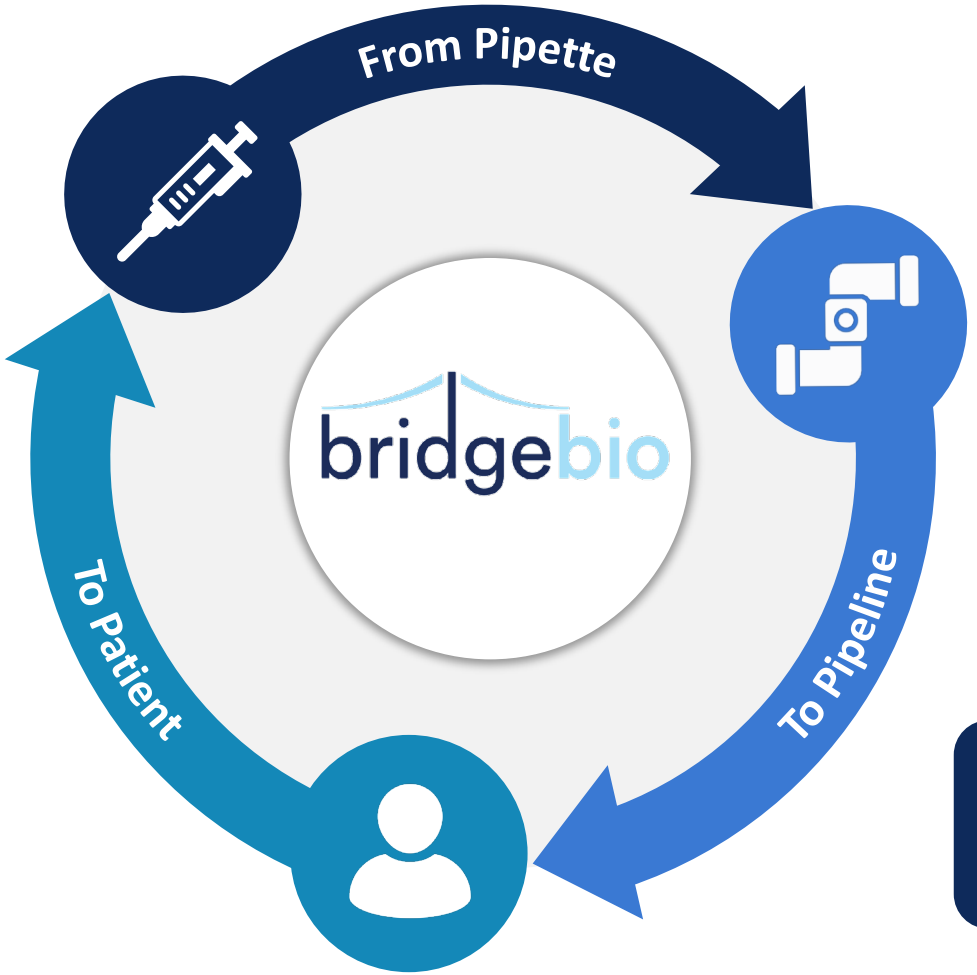
6	**Commercial launch plans**	Matt Outten Chief Commercial Officer, BridgeBio

BBP-418 for Limb-Girdle Muscular Dystrophy Type 2I/R9 (LGMD2I/R9)

A sincere THANK YOU to patients, families, advocates, investigators, clinical research staff, and collaborating research partners

We have built a sustainable, high velocity engine to deliver medicines



Attruby™ (acoramidis) 356 mg tablet

Nulibry® (fosdenopterin) for injection

TRUSELTIQ™ (infigratinib) capsules

Obtained Approval for 3 Medicines

> 2,000 patients in trials	**> 30** trials **> 30** countries **> 500** sites	**3 fully enrolled** pivotal trials	***+ 1 Positive Phase 3 Result***
> 70 papers published	**> 35** academic partnerships	**19** INDs created	**< $10M** and **< 3 yrs** to IND

Two observations stemming from BBP-418

Small molecules can provide an optimal therapeutic profile

- Can provide more **clinical efficacy** than newer technologies
- Often have a more **favorable safety** profile
- More **cost effective**

Delivering meaningful clinical improvement, rather than just delaying progression, is possible

- BBP-418 demonstrated improvement in biomarkers *and* clinical endpoints
- This effect was rapid and seen across subpopulations

Introduction

Christine Siu

Chief Executive Officer

ML Bio Solutions,
a BridgeBio company

BBP-418 is an investigational therapy and has not been approved for use by any regulatory agency.

With BBP-418, we aim to provide a critically needed, first-to-market therapy for individuals living with LGMD2I/R9

“It's **scary to think [about] the future**, and the **hope** of there being something that just allows us to **live our life without** as much **grief and loss** is **exciting for all of us**.”

- Julie

Highly statistically significant results on primary and all key secondary endpoints, including biomarker and clinical measures at 12 months

Primary endpoint (3 months)	p-value
Change from baseline in glycosylated αDG, % of control	p<0.0001

Key secondary endpoints (12 months)	p-value
Change from baseline in glycosylated αDG, % of control	p<0.0001
Change from baseline in serum CK, U/L	p<0.0001
Change from baseline in 100MTT, m/s	p<0.0001
Change from baseline in FVC, % predicted	p=0.0071

αDG = alpha-dystroglycan; CK = Creatine Kinase; 100MTT = 100-meter Timed Test; FVC = Forced Vital Capacity

Unprecedented and consistent improvement across primary and all key secondary efficacy endpoints combined with well-tolerated safety profile

fortify

Ph. 3 FORTIFY interim analysis

Type	Endpoint	Upside case target	Outcome observed
Primary *(3 months)*	Glycosylated αDG	• Statistically significant increase vs. placebo • 1.5x CFB in BBP-418 treated vs. approx. no change in placebo	✓ **Highly statistically significant increase** (**1.8x CFB; absolute increase of 17%** of control) at 3 months ✓ Increase **sustained at 12 months**
	Creatine kinase (CK)	• Average decline of ≥50% CFB in BBP-418 treated	✓ **Highly statistically significant** average **decline of 82%** in BBP-418 treated
Key secondary *(12 months)*	• Ambulatory measures (100MTT) • Pulmonary (FVC)	• Trends in one or more measures favoring BBP-418 treated vs. placebo	✓ **Statistically significant** and **clinically meaningful improvement** in BBP-418 treated ✓ 100MTT: **Increase in velocity** of 0.14 m/s from baseline and 0.27 m/s vs. placebo ✓ FVC: **Increase in ventilatory capacity** of ~3% predicted volume from baseline and a difference of ~5% predicted volume vs. placebo
Safety		• Well-tolerated *(consistent with Ph. 2 results)*	✓ Well-tolerated; consistent with Ph. 2 results

CFB = Change From Baseline; αDG = alpha-dystroglycan; 100MTT = 100-Meter Timed Test; FVC = Forced Vital Capacity

FORTIFY Phase 3 interim analysis topline results

Douglas Sproule, MD, MSc

Chief Medical Officer

ML Bio Solutions,
a BridgeBio company

BBP-418 is an investigational therapy and has not been approved for use by any regulatory agency.

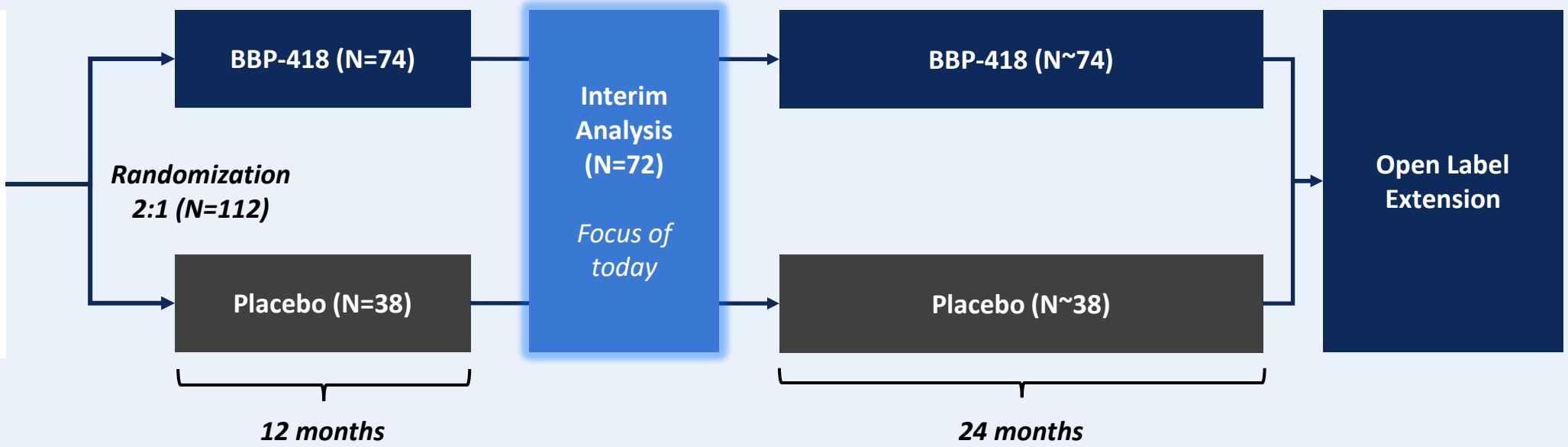
Thank you

to patients and families, advocates, physicians, clinical research staff, and collaborating research partners. We appreciate the LGMD2I/R9 community for all you do on behalf of LGMD patients.

FORTIFY is an ongoing randomized, placebo-controlled Phase 3 study with a planned interim analysis

Screening

- Genetically confirmed, symptomatic LGMD2I/R9
- 12-60 years of age



Interim Endpoints:

- Glycosylated αDG (*primary*)
- Serum creatine kinase (CK)
- Ambulatory measure: 100MTT
- Pulmonary function: FVC

Final Analysis Endpoints:

- NSAD (*primary*)
- Ambulatory measures
  - 10MWT
  - 100MTT
- Pulmonary function: FVC
- Upper limb function: PUL 2.0
- QoL measures

Placebo & BBP-418 arms were stratified by age group (adult vs. pediatric), ambulatory status, and genotype (L276I homozygous vs. Other FKRP genotype)

Note: Ph. 3 trial population limited to 18-60 years in EU region; final analysis exploratory endpoints include cardiac endpoints such as HS Troponin I
Source: Table 14.1.1.1

Glycosylated αDG overview & results

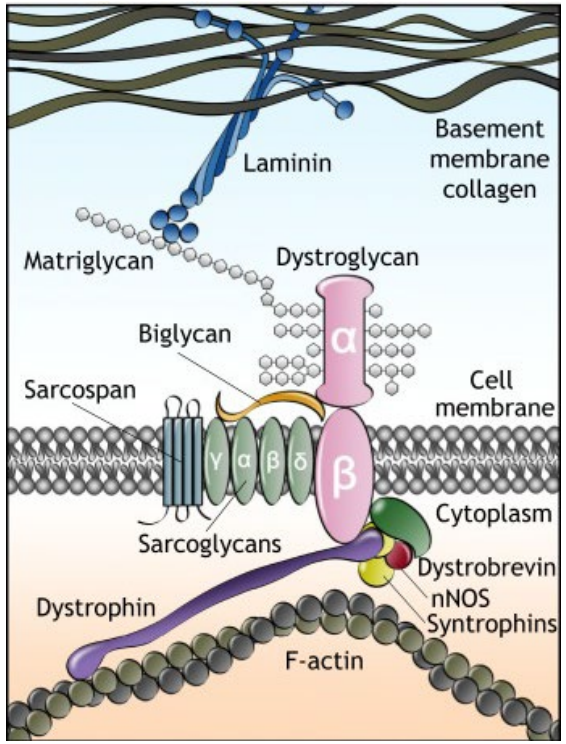
Uma Sinha, PhD

Chief Scientific Officer

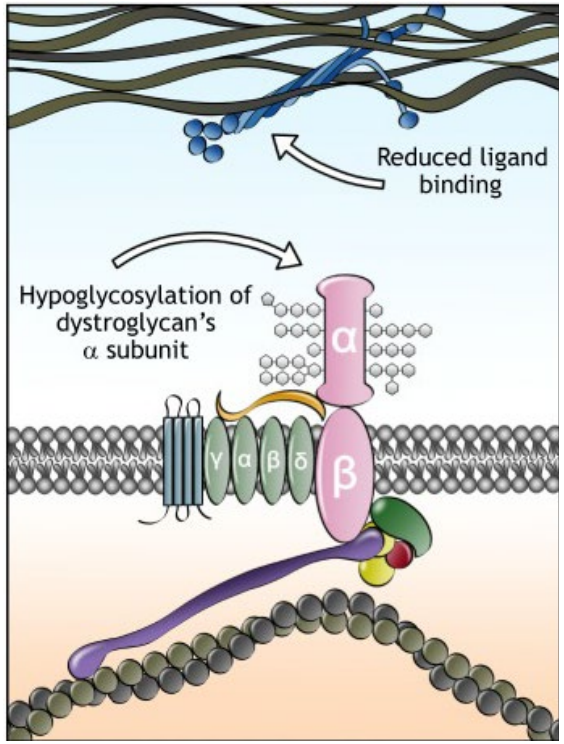
BBP-418 is an investigational therapy and has not been approved for use by any regulatory agency.

LGMD2I/R9 is caused by mutations in FKRP that result in reduced glycosylation of alpha-dystroglycan (αDG), leading to progressive muscle damage

Molecular pathogenesis of LGMD2I/R9



*In **healthy muscle cells**, glycosylated αDG bridges the muscle cell membrane to the extracellular matrix*



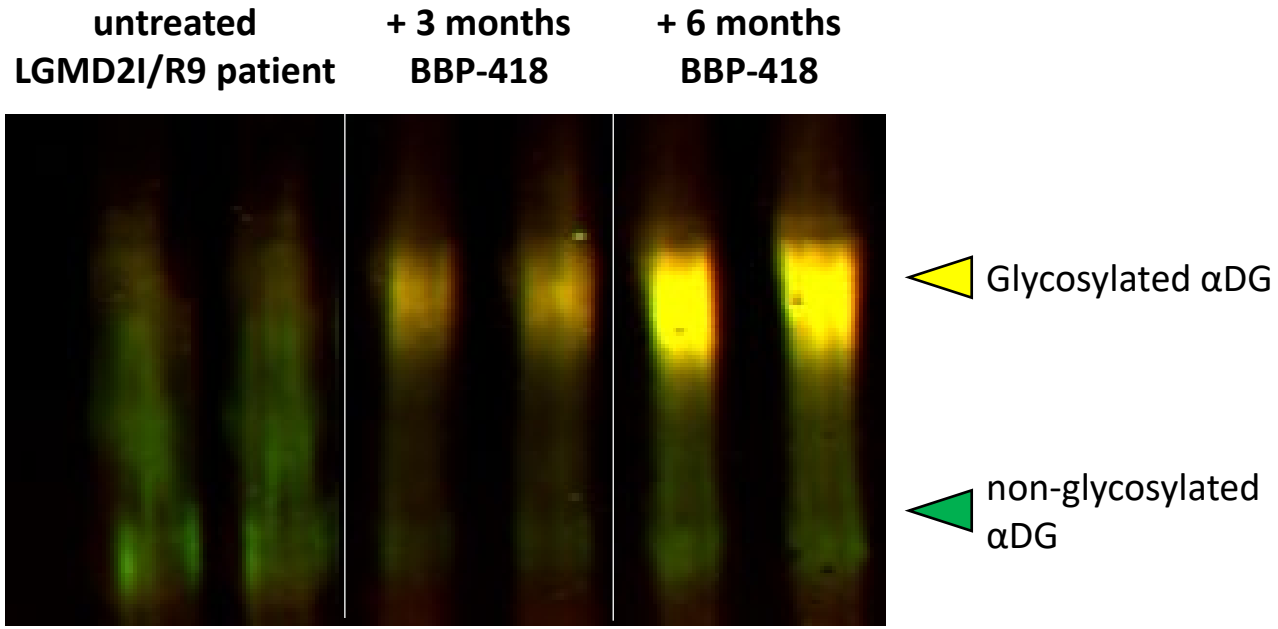
*In untreated **affected muscle cells**, hypoglycosylation of αDG results in loss of αDG-laminin binding*

- In healthy muscle cells, **alpha-dystroglycan (αDG) plays an important role** in linking the muscle cell membrane (sarcolemma) to the extracellular matrix
  - **Glycosylation of αDG is required** to form a glycopeptide chain that binds to laminin in the extracellular matrix
- LGMD2I/R9 is caused by **mutations in *FKRP*** that lead to **reduced function of the FKRP enzyme**, resulting in **decreased glycosylation of alpha-dystroglycan (αDG)**
- **Reduced glycosylation of αDG results in muscular dystrophy**, including the **progressive muscle damage** seen in LGMD2I/R9
- Natural history reflects that LGMD2I/R9 has an established **genotype/phenotype association**
  - LGMD2I/R9 individuals with "other" (non-L276I homozygous) FKRP genotypes, which typically have a **more severe clinical presentation**, have **lower glycosylated αDG levels** relative to L276I homozygous patients

Nickolls AR, Bönnemann CG, Dis Model Mech, 2018; Kanagawa M et al, Cell, 2004

We have developed a proprietary, validated Western Blot assay to accurately and reliably measure glycosylated αDG directly in skeletal muscle tissue

Validated Western Blot evaluates change from baseline in glycosylated alpha-dystroglycan (αDG)[1,2]



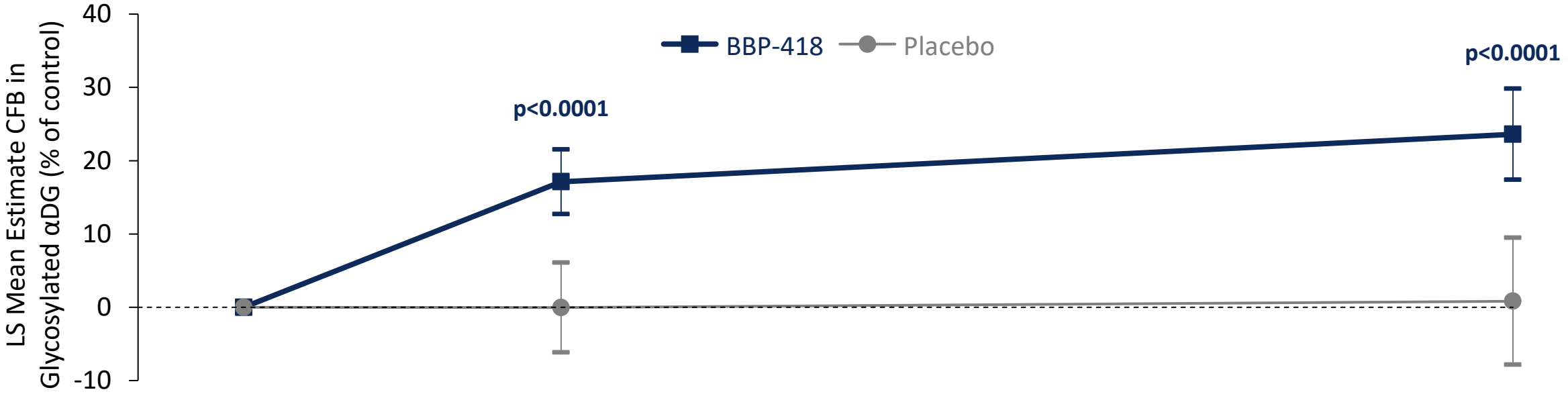
- Yellow signal corresponds to **functional, laminin-binding form of glycosylated αDG**
- Limited glycosylated αDG is seen in untreated LGMD2I/R9 patient muscle biopsy samples at baseline (shown in duplicate)
- **Increase in glycosylated αDG is observed** post treatment with BBP-418

FDA has indicated that our novel approach to measure glycosylated αDG by our validated assay appears reasonable

[1] Blots have been reordered to allow for side-by-side comparison of treatment effect within a patient; [2] See publication for details on this novel, multiplexed assay: Rajasingham et al, J Muscle Res Cell Motil, 2024

Observed statistically significant increase (+17% of control) from baseline in glycosylated αDG for BBP-418 treated, which was sustained at 12 months

Change from baseline in glycosylated αDG (+/- 99% CI)



Timepoint	0 mo.	3 mo.	12 mo.
Cohort		All patients	Interim analysis
BBP-418		17.1% (n=71)	23.6% (n=49)
Placebo		-0.04% (n=36)	0.8% (n=22)

LS Mean Estimate CFB: Least-Squares Mean Estimate Change from Baseline; Comparison of BBP-418 change from baseline to Placebo change from baseline is based on a linear mixed model for repeated measures; n = number of participants with observed value

Source: Table 14.2.1.1.1.2.3a.2

FORTIFY Phase 3 interim analysis topline results *(Continued)*

Douglas Sproule, MD, MSc

Chief Medical Officer

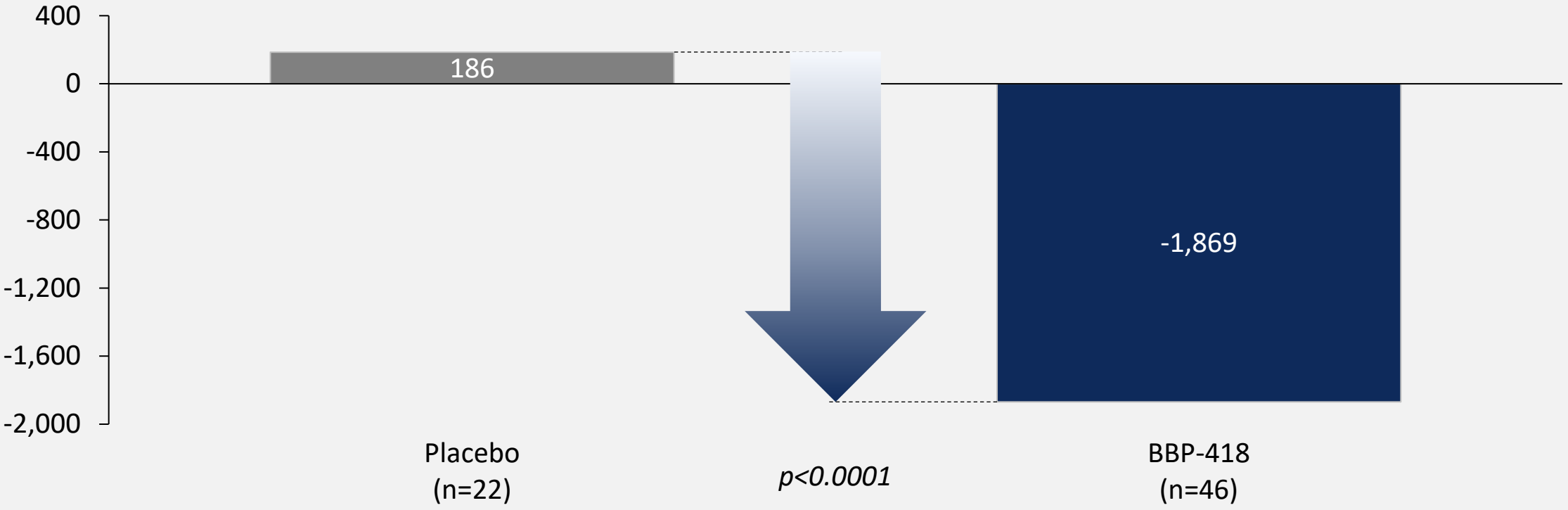
ML Bio Solutions,
a BridgeBio company

BBP-418 is an investigational therapy and has not been approved for use by any regulatory agency.

BBP-418 treated patients experienced a large, statistically significant reduction in serum CK of 82% from baseline at 12 months

Reduction in muscle damage

Change from baseline in serum creatine kinase (U/L)

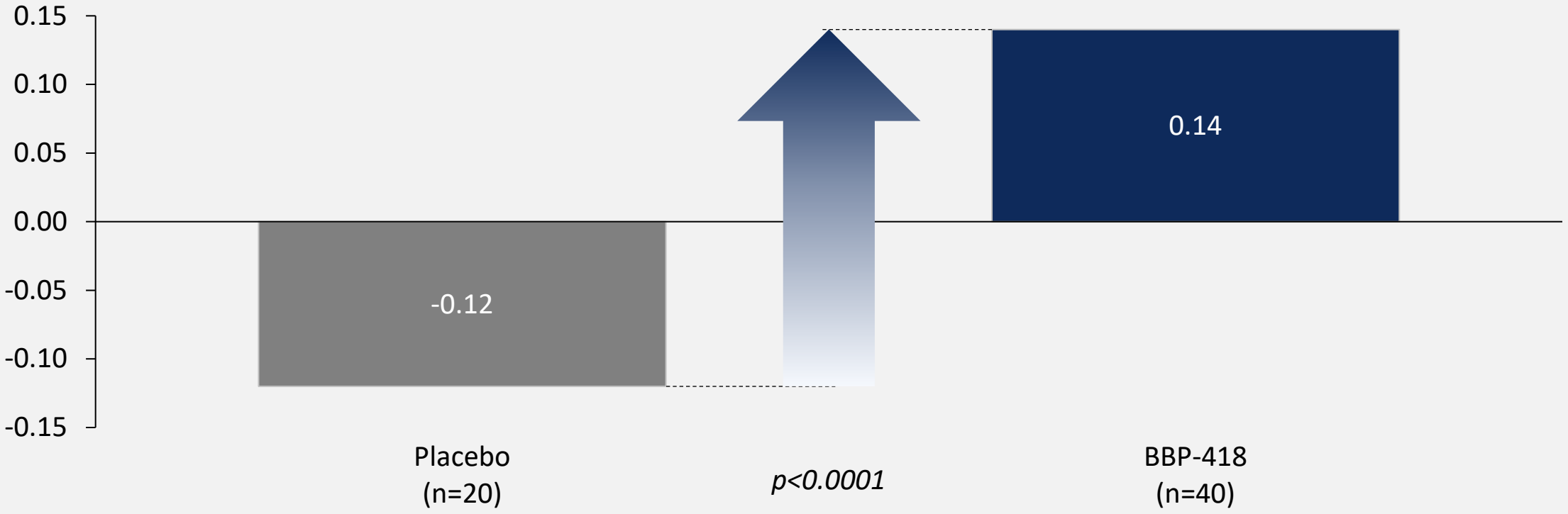


Least-Squares Mean Change from Baseline at 12-month timepoint; Comparison of BBP-418 change from baseline to Placebo change from baseline is based on a linear mixed model for repeated measures; n = number of participants with observed value
Source: Table 14.2.1.2.1.2.1a.2

Difference of 0.27 m/s observed between BBP-418 and placebo arms in 100MTT, translating to difference of ~14 seconds faster

Improved ambulatory function

Change from baseline in 100-meter timed test (m/s)

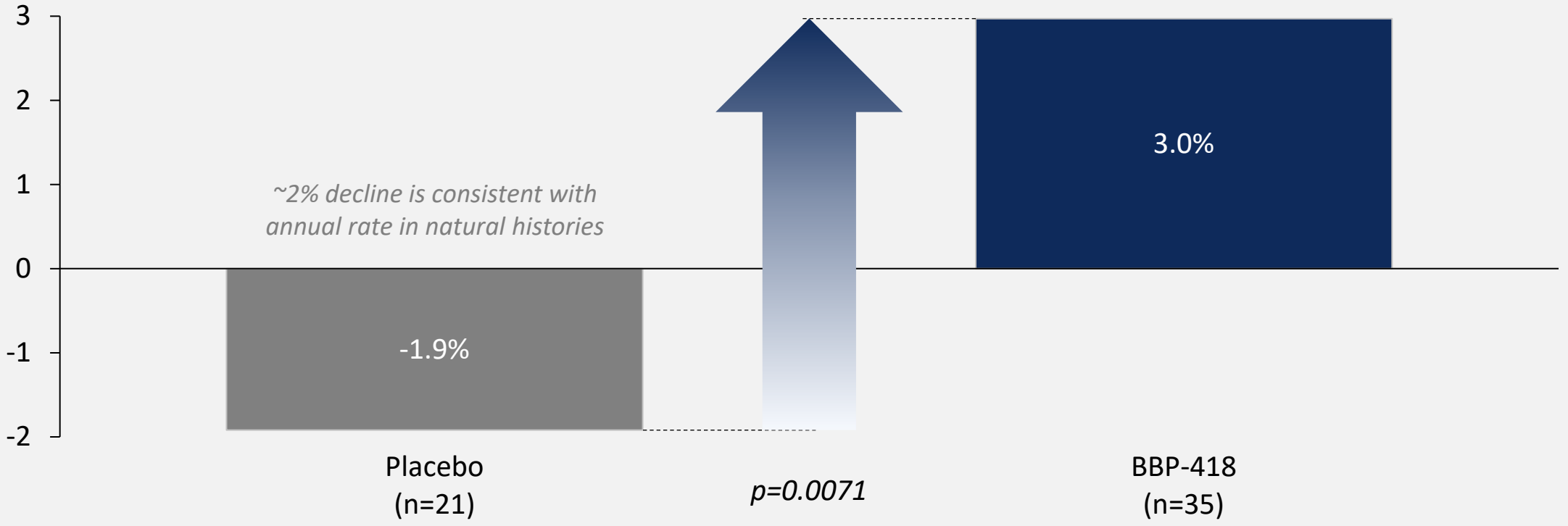


Least-Squares Mean Change from Baseline at 12-month timepoint; Comparison of BBP-418 change from baseline to Placebo change from baseline is based on a linear mixed model for repeated measures; n = number of participants with observed value
Source: Table 14.2.1.3.1.2.2a.2

BBP-418 treated registered ~3% increase in predicted volume from baseline FVC, resulting in a difference of ~5% predicted volume vs. placebo

Improved pulmonary function

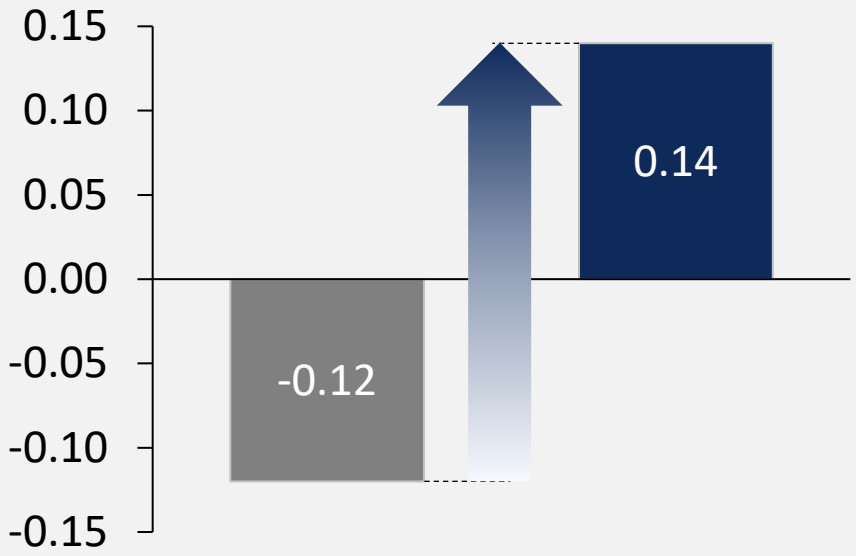
Change from baseline in forced vital capacity, sitting position (% predicted)



Least-Squares Mean Change from Baseline at 12-month timepoint; Comparison of BBP-418 change from baseline to Placebo change from baseline is based on a linear mixed model for repeated measures; n = number of participants with observed value
Source: Table 14.2.1.4.1.2.2a.2

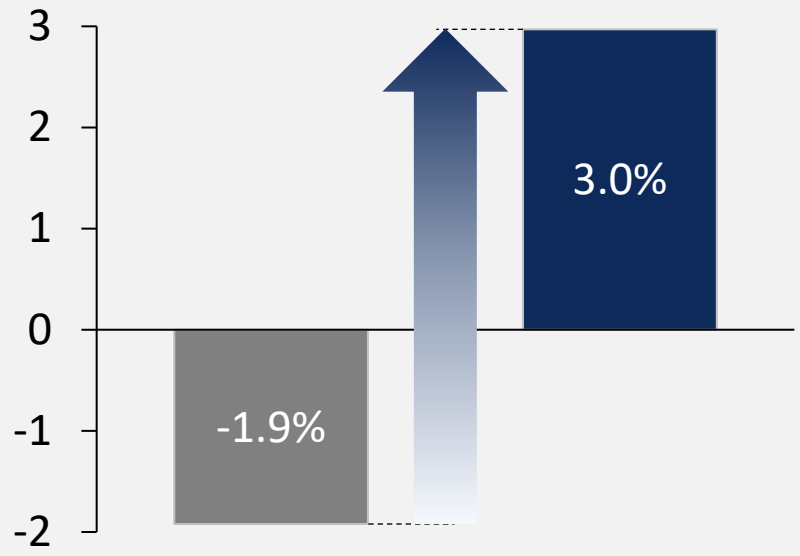
More than stabilization: BBP-418 treated individuals experienced significant, clinically meaningful improvements across clinical endpoints at 12 months

Improved ambulatory function
CFB in 100MTT (m/s)



p<0.0001

Improved pulmonary function
CFB in FVC (% predicted)



p=0.0071

Key: Placebo BBP-418

CFB = Change From Baseline

Least-Squares Mean Change from Baseline at 12-month timepoint; Comparison of BBP-418 change from baseline to Placebo change from baseline is based on a linear mixed model for repeated measures

Source: Table 14.2.1.3.1.2.2a.2; Table 14.2.1.4.1.2.2a.2

Continued highly favorable safety profile of BBP-418

- **No new or unexpected safety findings** have been observed; results consistent with Ph. 2
- **Discontinuation rate was low** overall and **higher in the placebo group**
- **No treatment-related serious TEAEs** were observed

Interim analysis continues to support a favorable risk-benefit profile

Next steps

Topline results from Phase 3 FORTIFY study interim analysis
October 2025

Engage FDA prior to NDA submission
Late 2025 / Early 2026

Present Phase 3 FORTIFY study interim analysis results
MDA Clinical & Scientific Conference
March 2026

File New Drug Application (NDA) with FDA
1H 2026

U.S. approval and commercial launch in LGMD2I/R9
Potential to be granted Priority Review Voucher (PRV)
Late 2026 / Early 2027

Commercial launch plans

Matt Outten

Chief Commercial Officer

BBP-418 is an investigational therapy and has not been approved for use by any regulatory agency.

We will leverage our proven commercial infrastructure to successfully launch BBP-418

Built on the foundation of Attruby's commercial success and tailored to the unique LGMD2I/R9 market opportunity

Established commercial infrastructure
Commercial operational teams already in place

Proven launch playbook
Enabling faster and more efficient mobilization

Ensure broad access
Leverage first-in-class data, and ensure broad access for patients and coverage for all books of business

We will commercialize and launch BBP-418 in the U.S.

Our commercial strategy combines proven rare disease launch capabilities with engagement tailored to neuromuscular specialists and the LGMD2I/R9 community

Core strategies to redefine standard of care in LGMD2I/R9	
	**Position BBP-418 as the standard of care due to transformative clinical outcomes**
	**Drive awareness of LGMD2I/R9 as a genetically distinct, underdiagnosed condition**
	**Build urgency for early diagnosis and initiation of therapy**

Q&A session