

Epicrispr Reports Positive Six-Month Data at The World Muscle Society Meeting Demonstrating Increased Lean Muscle Volume and Muscle Strength in FSHD Patients Treated with EPI-321

Lean Muscle Volume Increased Across Total, Upper- and Lower-Body Composite Measures at Month 6

Strength Gains on Quantitative Muscle Testing Track with Muscle Volume Increases, Including in Upper-Body Muscle Groups Commonly Affected Early in FSHD

SAN FRANCISCO--([BUSINESS WIRE](#))-- [Epicrispr Biotechnologies](#), a clinical-stage company pioneering epigenetic therapies, today announced updated six-month clinical data from the ongoing open-label, first-in-human Phase 1/2 study evaluating EPI-321 in patients with facioscapulohumeral muscular dystrophy (FSHD). This updated data from the first six patients enrolled in the study were presented at the 31st Annual Congress of the World Muscle Society (WMS 2026) in Hiroshima, Japan.

At six months, whole-body MRI showed increases in lean muscle volume across total-, upper- and lower-body composite muscle groups – a reversal of the declines seen in the untreated digital twin comparator built from longitudinal FSHD natural history data. The greatest difference in lean muscle volume was observed in the upper body where FSHD symptoms typically emerge first, with a 3.0% increase for EPI-321 treated patients versus a predicted decline of 3.5% in the matched digital twin for these 6 patients.

The muscle volume gains were accompanied by improvements in muscle strength. On quantitative muscle testing (QMT), EPI-321 treated patients improved upper body QMT by 4.1 percent versus a 0.3 percent decline that was observed in a matched external comparator arm from the ReSolve FSHD natural history study.

At the individual muscle level, across all six patients, EPI-321 treatment resulted in an 8.1 percent increase in elbow extension strength, accompanied by a 3.2 percent increase in elbow extensor lean muscle volume compared to declines of 1.1 percent and 9.0 percent respectively that were seen in the matched digital twin and ReSolve FSHD natural history study comparators – one of several instances in which strength and MRI findings moved in the same direction for the same muscle group.

EPI-321 continues to demonstrate a manageable safety and tolerability profile. No EPI-321 related serious adverse events were observed.

“These updated results across the six patients build on the encouraging data we initially observed in the first three patients and further strengthen our confidence in the potential of EPI-321 to address the underlying biology of FSHD,” said Amber Salzman, Ph.D., Chief Executive Officer, Epicrispr Biotechnologies. “As additional patients have reached six months of follow-up, we continue to see a manageable safety profile and encouraging evidence of increased lean muscle volume, including in muscle groups commonly affected by FSHD. We look forward to presenting twelve-month data from all patients in this study in the second half of 2027.”

“These updated six-month data are encouraging, particularly given the progressive nature of FSHD and the muscle loss patients experience over time,” said Richard Roxburgh, MB ChB, FRACP, PhD Associate Professor of Medicine at the University of Auckland and a principal investigator for the EPI-321 clinical trial. “Seeing evidence of increased lean muscle volume builds on the data observed in the first three patients. While these remain early data and longer follow-up in more patients is needed, the consistency of the findings supports continued evaluation of EPI-321 and its potential to alter the course of FSHD.”

The ongoing open-label Phase 1/2 study (NCT06907875) is evaluating the safety, tolerability, biological activity and preliminary efficacy of EPI-321 in adults with FSHD. The study is now fully enrolled with six patients treated with a single intravenous (IV) infusion at each of two dose levels of EPI-321 (2×10^{13} vg/kg and 4×10^{13} vg/kg).

About EPI-321

EPI-321 is an investigational epigenetic therapy that aims to address the underlying molecular mechanisms of FSHD with a one-time dose. Following intravenous administration, EPI-321 is directed to muscle tissue within a single AAV

vector, which has been clinically validated for muscle delivery. Preclinical studies on EPI-321 have demonstrated its ability to robustly suppress pathological expression of the DUX4 gene and reduce muscle cell death. EPI-321 previously reported interim data from the Phase 1/2 trial, including statistically significant increases in whole-body lean muscle volume measured by MRI, favorable changes in circulating biomarkers consistent with DUX4 suppression, favorable strength and functional outcomes, and a manageable safety profile.

About Facioscapulohumeral Muscular Dystrophy (FSHD)

Facioscapulohumeral muscular dystrophy (FSHD) is one of the most common forms of muscular dystrophy, affecting more than an estimated 1 million people worldwide. FSHD is a progressive genetic neuromuscular disease characterized by the gradual weakening and loss of skeletal muscle, often beginning in the face, shoulders, and upper arms before progressing to other parts of the body. The disease is caused by aberrant expression of DUX4, a gene that is normally silenced in healthy muscle tissue but becomes activated in individuals with FSHD, leading to muscle damage, inflammation, and progressive muscle degeneration. There are currently no approved disease-modifying therapies for FSHD.

About Epicrispr Biotechnologies

Epicrispr Biotechnologies is a biotechnology company pioneering gene-modulating therapies, leading with treatments for neuromuscular diseases. The company's proprietary Gene Expression Modulation System (GEMS) enables precise and durable epigenetic modulation of gene expression, unlocking first-in-class treatments for previously untreatable conditions. Epicrispr's lead program, EPI-321 is in clinical trials for FSHD, and the company is advancing additional gene-modulating therapies. Learn more at www.epicrispr.com or follow us on [LinkedIn](#).

Contacts

Investor
Benson Cheng
benson.cheng@epic-bio.com

Media
Kimberly Ha
KKH Advisors
917-291-5744
kimberly.ha@kkhadvisors.com

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