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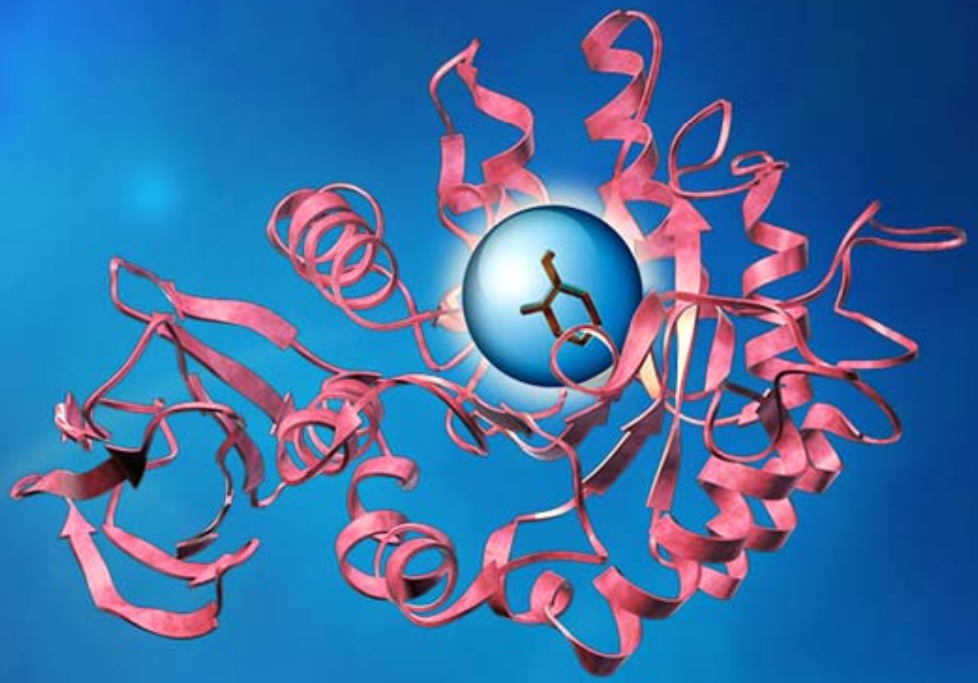

2Q14 Financial Results Conference Call & Webcast

August 7, 2014

at the forefront of therapies for rare and orphan diseases

Safe Harbor

This presentation contains "forward-looking statements" within the meaning of the Private Securities Litigation Reform Act of 1995 relating to business, operations and financial conditions of Amicus including but not limited to preclinical and clinical development of Amicus' candidate drug products, cash runway, ongoing collaborations and the timing and reporting of results from clinical trials evaluating Amicus' candidate drug products. Words such as, but not limited to, "look forward to," "believe," "expect," "anticipate," "estimate," "intend," "plan," "would," "should" and "could," and similar expressions or words, identify forward-looking statements. Although Amicus believes the expectations reflected in such forward-looking statements are based upon reasonable assumptions, there can be no assurance that its expectations will be realized. Actual results could differ materially from those projected in Amicus' forward-looking statements due to numerous known and unknown risks and uncertainties, including the "Risk Factors" described in our Annual Report on Form 10-K for the year ended December 31, 2013. All forward-looking statements are qualified in their entirety by this cautionary statement, and Amicus undertakes no obligation to revise or update this presentation to reflect events or circumstances after the date hereof.

Global Registration Studies

Assembling Robust Dataset to Maximize Chances for Global Approvals of Migalastat Monotherapy for Fabry Patients with Amenable Mutations

The FACETS Study

Fabry **A**T1001 **C**haperone **E**fficacy, **T**herapeutics and **S**afety Study

- Placebo-controlled (6 months)
- 67 patients naïve to ERT
- 6-month surrogate primary endpoint: kidney GL-3 (reported 4Q12)
- 12-month biopsy and 24-month clinical data (reported 2Q14)

THE ATTRACT STUDY

AT1001 **T**herapy Compared to Enzyme **R**eplacement in Fabry Patients with **A**T1001-responsive Mutations: a Global **C**linical **T**rial

- ERT switch study
- 60 patients (1.5:1 randomization)
- 18-month co-primary clinical endpoints of kidney function (mGFR and eGFR)
- Data expected 3Q14

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Recap of Study 011 12- and 24-Month Data: Key Findings

12- and 24-Month Results Released in April 2014 Demonstrated a Clear Efficacy Signal in Fabry Patients with Amenable Mutations

- Subjects who switched from placebo to migalastat after month 6 demonstrated a statistically significant reduction in kidney interstitial capillary GL-3 at month 12 (p=0.013*)
- Subjects who remained on migalastat for 12 months demonstrated a durable reduction in kidney interstitial capillary GL-3
- Reduction in disease substrate also observed in plasma lyso-Gb3 in subjects who switched from placebo to migalastat (p<0.0001**). Subjects who remained on migalastat demonstrated a durable reduction in lyso-Gb3
- Kidney function (estimated glomerular filtration rate (eGFR), iohexol mGFR) remained stable over 18-24 months
- Migalastat was generally safe and well-tolerated
- Of 41 subjects with GLP HEK amenable mutations who completed Study 011, 35 (85%) remain in voluntary extension study (Study 041)

*MMRM, **ANCOVA

Study 012 Definition of Success

With Finalization of Statistical Analysis Plan, Amicus Has Clarified Primary Efficacy Analysis for Study 012

- Descriptive assessment of comparability for migalastat and ERT
- 18-month co-primary endpoints are mean annualized change in mGFR and eGFR measured in 2 ways:
  - 50% overlap in confidence intervals between treatment groups

  AND

  - Mean annualized changes for patients receiving migalastat within 2.2 mL/min/1.73m^2/yr of patients receiving ERT
- Incorporates regulatory feedback

Migalastat Monotherapy Experience

97 Patients Today Take Migalastat HCl as Only Therapy for Fabry Disease[1]

[1] All patients are receiving investigational drug, migalastat HCl, as part of ongoing clinical trials

*Retention defined as # of patients who complete a study and chose to enter extension, e.g., 011 12-mo into 12-mo extension or 011 into 041

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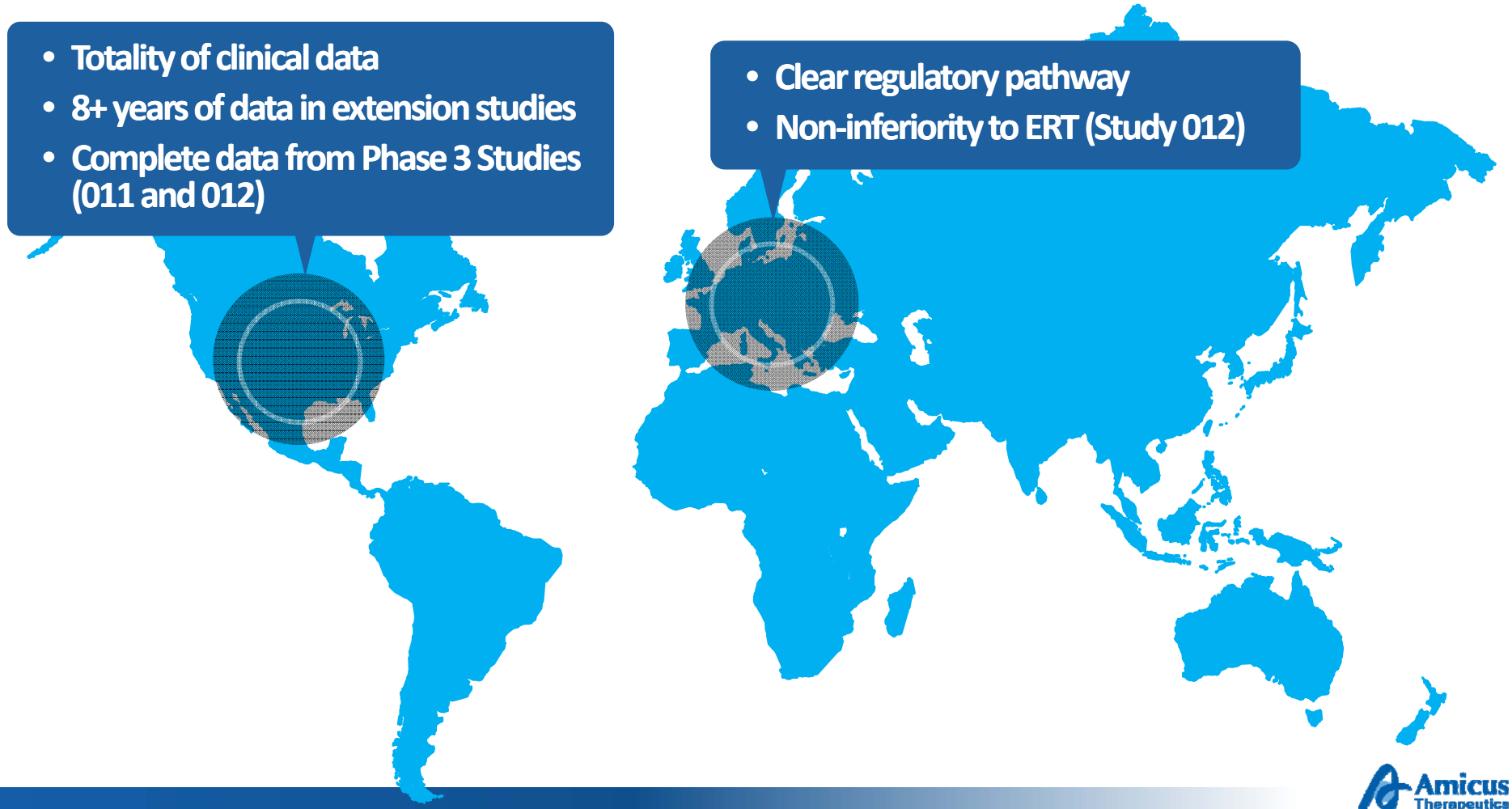
Migalastat Monotherapy: Global Regulatory Strategy

Data from Study 011 (Reported) and Study 012 (Expected 3Q14) to Support Global Approvals of Migalastat Monotherapy for Patients with Amenable Mutations

- Totality of clinical data
- 8+ years of data in extension studies
- Complete data from Phase 3 Studies (011 and 012)

- Clear regulatory pathway
- Non-inferiority to ERT (Study 012)



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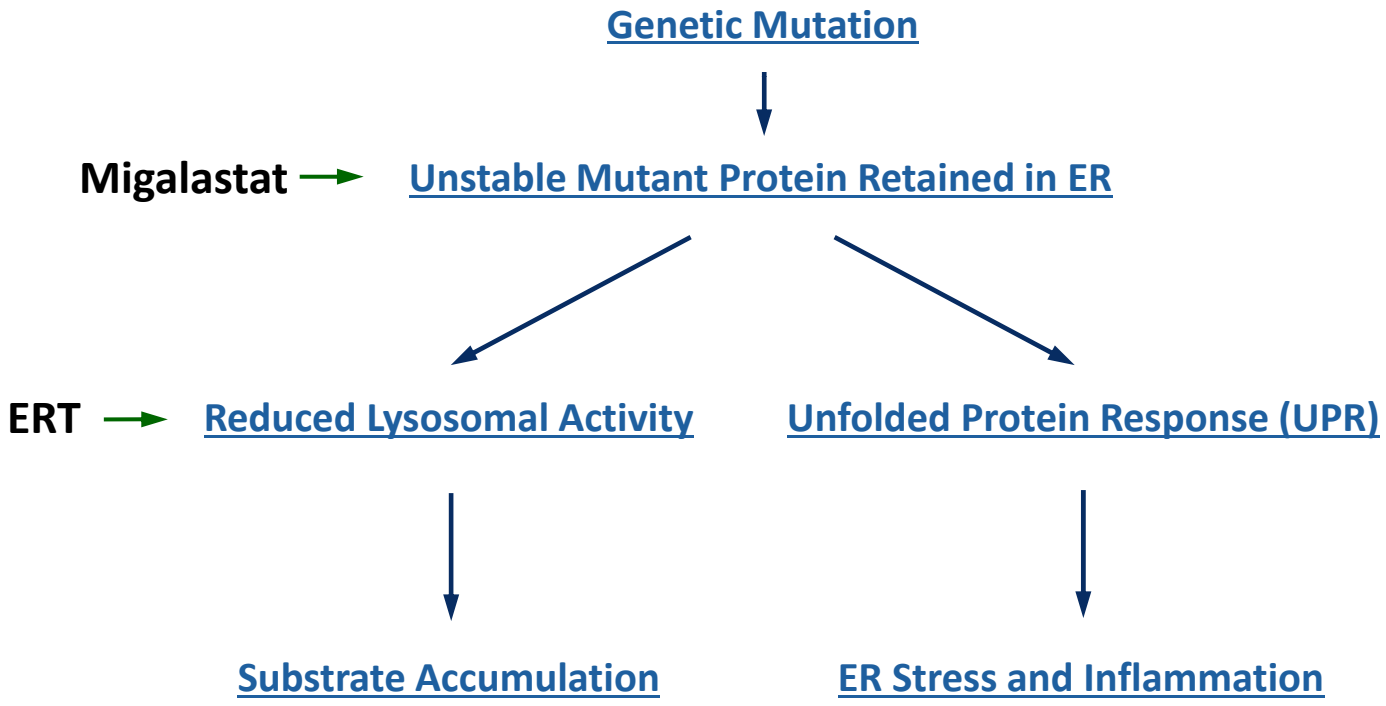
Migalastat MOA: Potential Benefits Beyond Substrate Reduction

Potential to Address Endoplasmic Reticulum (ER) Stress and Cellular Inflammation That May Be Caused by Mutant Enzyme Retained in the ER



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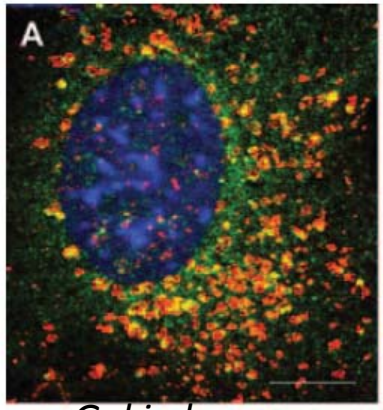
Migalastat Restores Trafficking of Mutant Enzyme

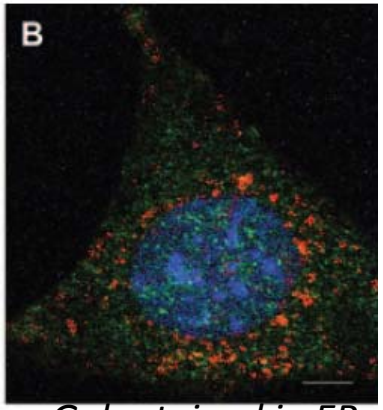
Migalastat Restores Ability of Mutant α-Gal A Retained in ER to Traffic to Lysosomes and Reduces a Marker of ER Stress in a Fabry Mouse Model

Wild-Type Untreated



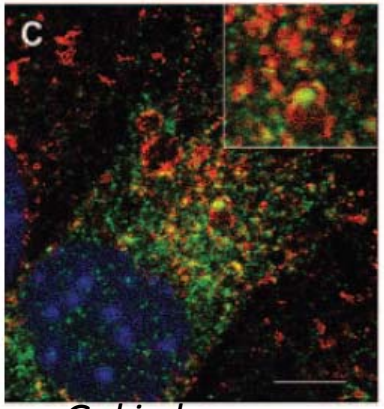
α-Gal in lysosomes

R301Q Untreated



α-Gal retained in ER

R301Q Migalastat-Treated



α-Gal in lysosomes

- Migalastat leads to reduced levels of mutant α-Gal in ER in fibroblast cells
- Migalastat also reduces levels of ER stress marker BiP in white blood cells

Sources: Yam et al 2005; Yam et al 2006

Role of UPR and ER Stress in Nephropathy and Potential Role of Migalastat

Further Research Into the Role of UPR and ER stress in Fabry and the Potential for Migalastat to Address this Pathology Is Ongoing

- There is growing evidence that UPR and ER stress contribute to nephropathy
  - ER stress markers are increased in kidney biopsies from patients with nephropathy
  - Abnormal protein accumulation and ER stress in podocytes and tubular cells can lead to severe proteinuria and tubular apoptosis

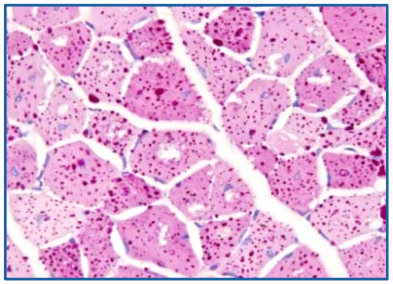
- By chaperoning mutant α-Gal out of the ER, migalastat may reduce UPR and ER stress, potentially addressing aspects of kidney disease not currently addressed by ERT

Sources: Cybulsky 2013; Inagi 2014

Pompe Disease Overview

Severe, progressive, fatal neuromuscular disease

- Deficiency of lysosomal enzyme GAA
- Age of onset ranges from infancy to adulthood
- Glycogen accumulation in muscle tissue
- Incidence 1:28,000[1]
- Current ERT suboptimal



Elevated Glycogen in Muscle

[1]Evidence Report – Newborn Screening for Pompe Disease – June 2013 – HRSA.gov

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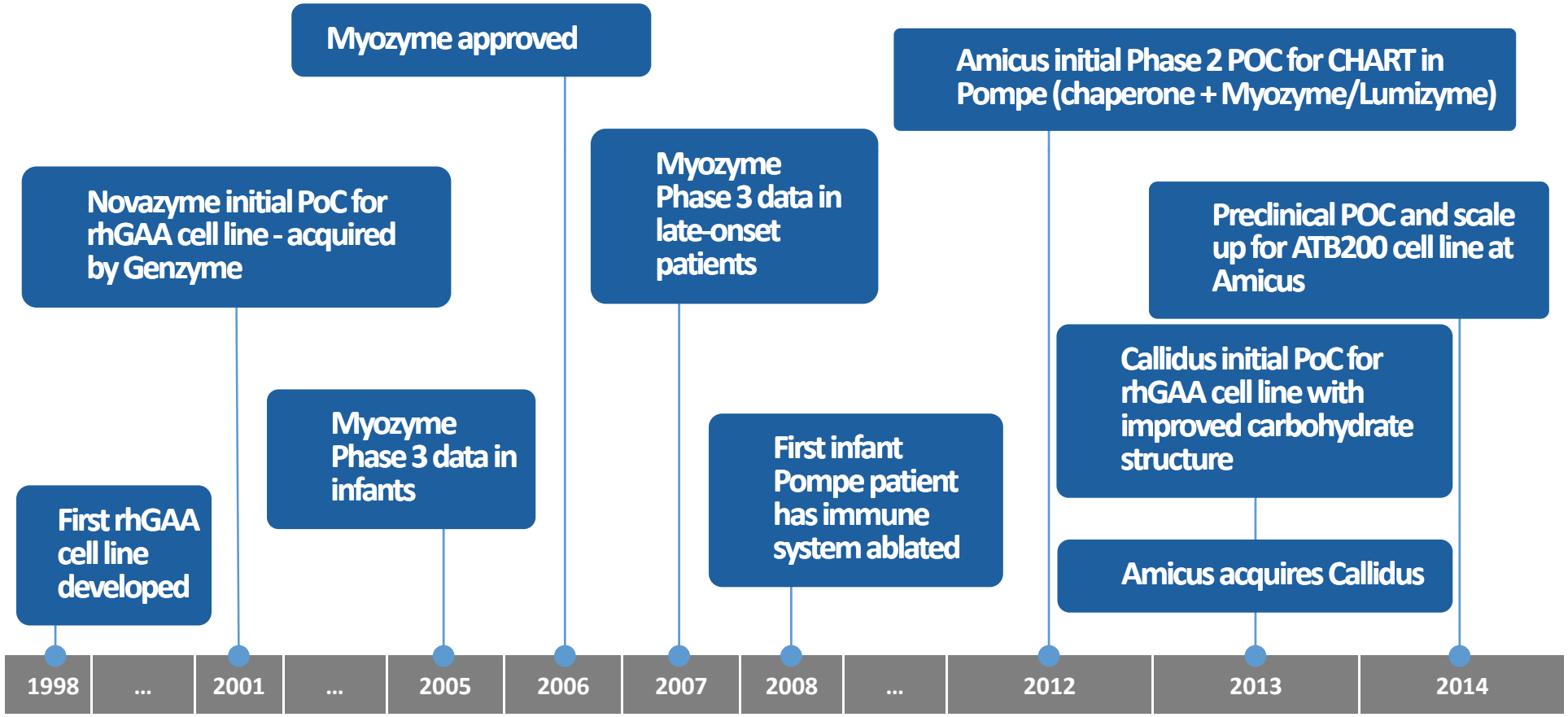
Select Milestones in Pompe Drug Development

16 Years After First Pompe Cell Line, Significant Unmet Medical Needs Remain



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Three Challenges with Pompe ERT

Activity/ Stability

Rapid denaturation of ERT in pH of blood[1]

Tolerability / Immunogenicity

Infusion-associated reactions in ~50% of late-onset patients[3]

High antibody titers shown to affect treatment outcomes[4,5]

Uptake/ Targeting

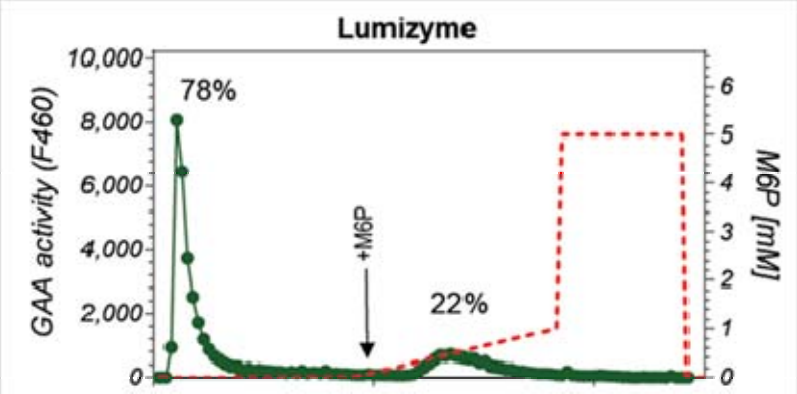
Low M6P receptor uptake into skeletal muscle[2]

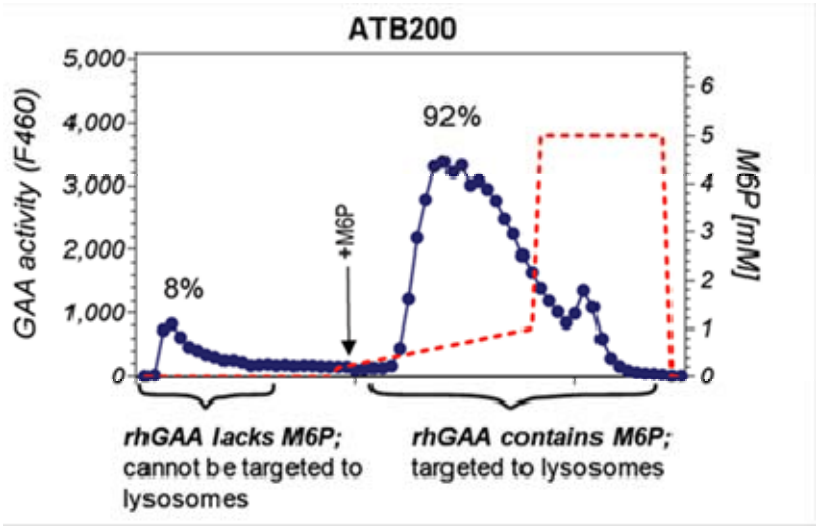
Majority of rhGAA is not delivered to lysosomes[2]

[1]Khanna *et al.,* **PLoS ONE**, 2012; [2]Zhu et al., Amer. Soc. Gene Therapy, 2009 June; [3]Banati *et al.,* **Muscle Nerve**, 2011 Dec. ; [4]Banugaria *et al.*, **Gen. Med.,** 2011 Aug.; [5]de Vries *et al.,* **Mol Genet Metab**., 2010 Dec.

ATB200 rhGAA Contains Higher M6P and Binds M6P Receptor Better Than Myozyme/Lumizyme

Amicus Expertise and Capabilities Enabled Development of Proprietary rhGAA ERT (ATB200) with Optimal Glycosylation for Improved Drug Targeting





- Developed proprietary cell line for producing rhGAA (designated as ATB200)
- ATB200 has significantly higher M6P content than existing rhGAA ERTs
- ATB200 binds intended M6P receptor substantially better than standard of care ERT

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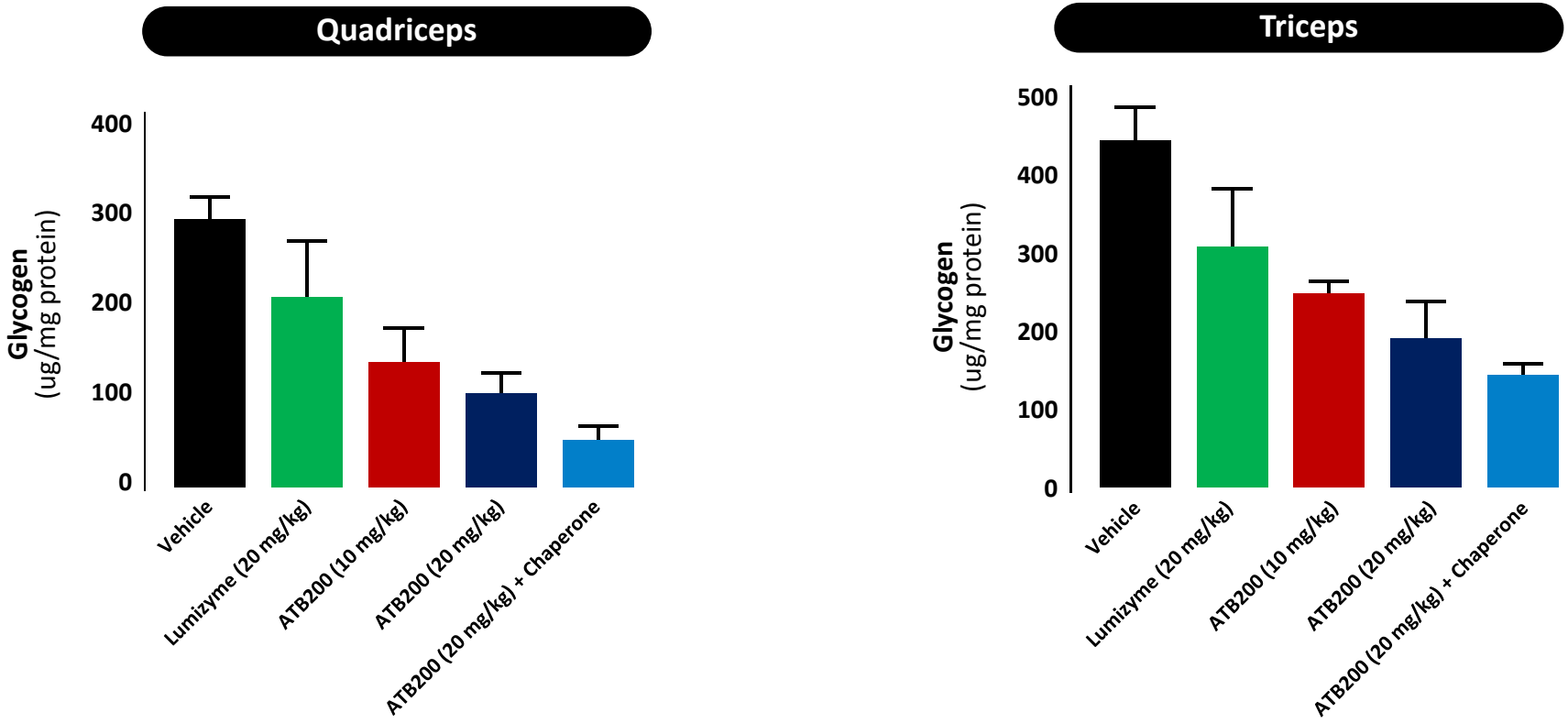
AT-B200: Next-Generation Pompe ERT (rhGAA) Updated Preclinical Proof-of-Concept

AT-B200 Led to Further Glycogen Reduction Compared to Lumizyme in Preclinical Studies in GAA Knock-Out Mice

Residual Muscle Glycogen After ERT



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Program Updates

- Fabry Next-Generation ERT
  - Phase 1 migalastat IV PK study successfully completed
  - Manufacturing of co-formulated JR-051 drug supply complete
  - Phase 1/2 study in Fabry patients will begin pending outcome of ongoing BD discussion on future sources of Fabry enzyme
- Parkinson's
  - Early-stage Biogen collaboration to conclude in September
  - Amicus retains WW rights to most advanced Parkinson's compound (AT3375)

2Q14 Financial Summary

Successful Execution Under ATM Equity Financing Strengthens Balance Sheet and Provides Runway Under Current Operating Plan Into 2016

Financial Position	June 30, 2014	July 2, 2014
Current Cash:	$78.0M	$98.4M
2014 net cash spend:	$54-59M	
Cash runway:	Into 2016	
Capitalization		
Shares outstanding:	72,869,861	78,685,241

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2Q14 Financial Results

($000s)	June 30, 2014	June 30, 2013
Total Revenue	475	---
Total Operating Expenses	14,741	16,005
Net Loss	(14,614)	(15,349)
Net Loss Per Share	(0.22)	(0.31)

Continuity of Leadership at Amicus

- Chairman and CEO John Crowley to take temporary leave of absence (~32 Weeks) to serve active military duty
  - Crowley, a commissioned officer in the United States Navy Reserve, will begin a temporary leave of absence in late September in support of Operation Enduring Freedom (Afghanistan)
  - Crowley will remain CEO and Chairman and expects to continue to advise Amicus on major strategic and business issues during this time
- Bradley Campbell, Chief Operating Officer, will oversee day to day operations and chair executive leadership team in Mr. Crowley's absence
- Mr. Crowley expected to return full-time from active duty service in 2Q15

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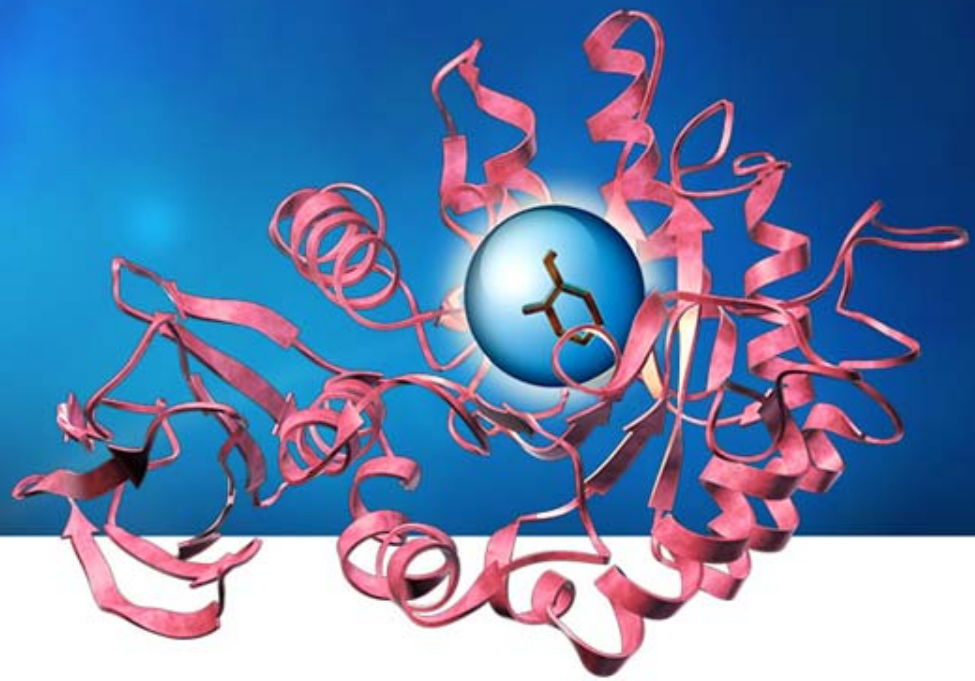

Questions & Answers

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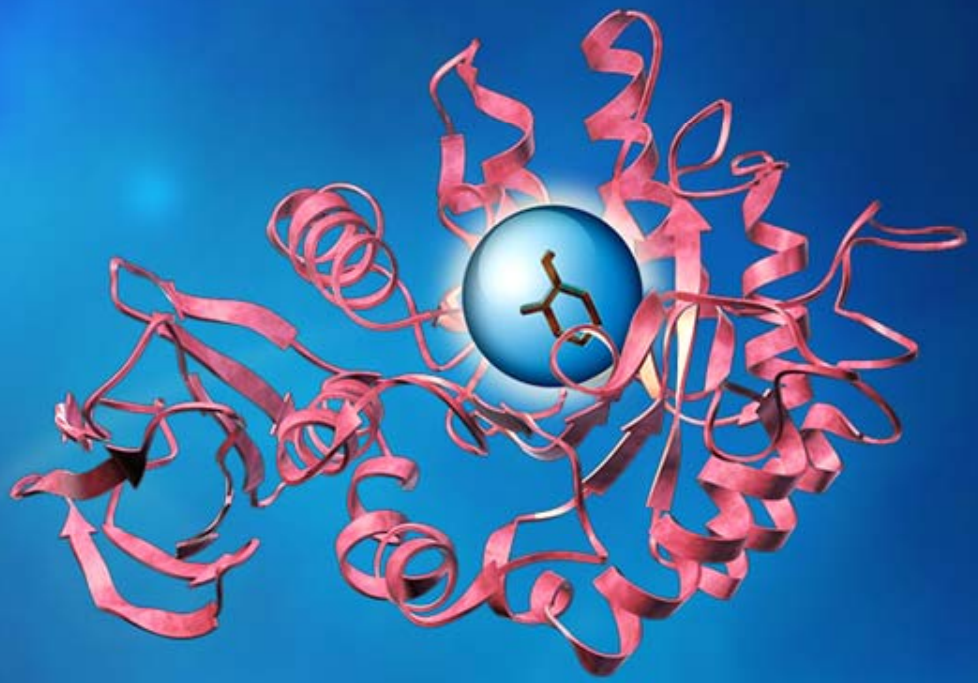

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