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): **August 7, 2014**

AMICUS THERAPEUTICS, INC.
(Exact Name of Registrant as Specified in Its Charter)

Delaware
(State or Other Jurisdiction of Incorporation)

001-33497	**71-0869350**
(Commission File Number)	(IRS Employer Identification No.)
1 Cedar Brook Drive, Cranbury, NJ	**08512**
(Address of Principal Executive Offices)	(Zip Code)

Registrant's telephone number, including area code: **(609) 662-2000**

(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:

o Written communications pursuant to Rule 425 under the Securities Act (17 CFR 230.425)

o Soliciting material pursuant to Rule 14a-12 under the Exchange Act (17 CFR 240.14a-12)

o Pre-commencement communications pursuant to Rule 14d-2(b) under the Exchange Act (17 CFR 240.14d-2(b))

o Pre-commencement communications pursuant to Rule 13e-4(c) under the Exchange Act (17 CFR 240.13e-4(c))

Item 2.02. Results of Operations and Financial Condition.

On August 7, 2014, Amicus Therapeutics, Inc. (the "Company") issued a press release announcing its financial results for the second quarter ended June 30, 2014. A copy of this press release is attached hereto as Exhibit 99.1. The Company will also host a conference call and webcast on August 7, 2014 to discuss its second quarter results of operations. A copy of the conference call presentation materials is also attached hereto as Exhibit 99.2.

In accordance with General Instruction B.2. of Form 8-K, the information in this Current Report on Form 8-K and the Exhibit shall not be deemed "filed" for purposes of Section 18 of the Securities Exchange Act of 1934, as amended (the "Exchange Act"), or otherwise subject to the liability of that section, nor shall it be deemed incorporated by reference in any filing under the Securities Act of 1933, as amended, or the Exchange Act, except as expressly set forth by specific reference in such a filing.

Item 9.01. Financial Statements and Exhibits.

(d) Exhibits: The Exhibit Index annexed hereto is incorporated herein by reference.

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.

Amicus Therapeutics, Inc.

Date: August 7, 2014

By: /s/ William D. Baird III
William D. Baird III
Chief Financial Officer

EXHIBIT INDEX

Exhibit No.	Description
99.1	Press Release dated August 7, 2014
99.2	August 7, 2014 Conference Call Presentation Materials

Amicus Therapeutics Announces Second Quarter 2014
Financial Results and Corporate Updates

Results from Second Phase 3 Fabry Monotherapy Study (Study 012) On Track for 3Q14

Updated Preclinical Data for Next-Generation Pompe ERT Shows Superior Substrate Reduction Compared to Current Standard-of-Care ERT

Completion of $40M At-the-Market (ATM) Equity Financing Extends Cash Runway into 2016

Conference Call and Webcast Today at 5:00pm ET

CRANBURY, NJ, August 7, 2014 — Amicus Therapeutics (Nasdaq: FOLD), a biopharmaceutical company at the forefront of therapies for rare and orphan diseases, today announced financial results for the second quarter ended June 30, 2014. The Company also provided program updates, reiterated full-year 2014 operating expense guidance, and extended cash runway guidance.

John F. Crowley, Chairman and Chief Executive Officer of Amicus Therapeutics, Inc., stated, "We reached a major inflection point for Amicus in the second quarter. Following positive 12- and 24-month data from our first Phase 3 Fabry monotherapy study, or Study 011, we completed the primary treatment period and finalized the statistical analysis plan for our second Phase 3 Fabry monotherapy, or Study 012. In the third quarter we look forward to releasing top-line data from Study 012, in which patients volunteered to switch from ERT to our oral chaperone migalastat as their only therapy for Fabry disease. We also continue to advance development of next-generation ERTs. Additional proof-of-concept data from preclinical studies of our next-generation Pompe ERT continue to show superior substrate reduction compared to standard-of-care ERT, and we remain on track to identify the optimal therapeutic to bring into the clinic next year. Finally, we significantly strengthened our balance sheet with the completion of an ATM financing that has added multiple new top-tier investors to our shareholder base. With the proceeds from this financing, we are well-positioned to execute our operating plan and our commitment to deliver value to our shareholders and, importantly, to these patient communities."

Financial Highlights for Second Quarter Ended June 30, 2014

- Cash, cash equivalents, and marketable securities totaled $78.0 million at June 30, 2014 compared to $82.0 million at December 31, 2013.
- Upon receipt of the final proceeds from the sale of shares under the ATM equity financing, cash was $98.4 million as of July 2, 2014.
- Total 2Q14 operating expenses decreased to $14.7 million compared to $16.0 million for the second quarter of 2013 due to decreases in personnel costs.
- Net cash spend was $12.2 million, compared to $10.6 million for the second quarter 2013.
- Net loss was $14.6 million, or $0.22 per share, compared to a net loss of $15.3 million, or $0.31 per share, for the second quarter 2013.

2014 Financial Guidance

Amicus continues to expect full-year 2014 net cash spend of between $54 million and $59 million. The Company's balance sheet was strengthened with a $40.0 million ATM financing in which a total of 14.3 million shares were sold at various times during the second quarter and early third quarter of 2014. Including the proceeds from this financing, the current cash position is projected to fund the current operating plan into 2016.

CEO Military Duty Leave of Absence

Amicus announced that Chairman and CEO John F. Crowley, a commissioned officer in the United States Navy Reserve, has been ordered to temporary active duty in support of Operation Enduring Freedom (Afghanistan) for a period of approximately 32 weeks beginning by the end of September 2014. Mr. Crowley will remain as Chairman and CEO and a member of the Amicus Board of Directors and expects to continue to advise the company on major strategic and business issues during this leave of absence. Bradley Campbell, Chief Operating Officer, will oversee the company's day to day operations and will chair the company's executive leadership team in Mr. Crowley's absence. Mr. Crowley is expected to return from active duty service and to resume his full responsibilities as Amicus Chairman and CEO in the second quarter of 2015.

Donald J. Hayden, Lead Independent Director, stated, "On behalf of the entire Amicus Board of Directors, we are grateful for John's exemplary leadership of Amicus and for his ongoing service to our country. We are fully supportive of this deployment and look forward to his return full time to the Amicus Chairman and CEO role in the second quarter of 2015. Over the last several years, John has assembled a very experienced and highly capable leadership team and we are confident in their ability to continue to execute on the strategy and vision set by John and the Board of Directors."

Program Updates

Migalastat Monotherapy and Next-Generation Enzyme Replacement Therapy (ERT) for Fabry Disease

Amicus solely owns and controls global development and commercialization of its pharmacological chaperone migalastat HCl ("migalastat") monotherapy and its next-generation ERT (migalastat co-formulated with ERT) for Fabry disease. As a monotherapy, migalastat is designed to bind and stabilize alpha-Gal A enzyme in patients with amenable mutations. In combination with ERT, migalastat is designed to bind and stabilize the infused alpha-Gal A enzyme, independent of a patient's genetic mutation. Between these approaches Amicus believes migalastat has the potential to benefit all patients with Fabry disease.

Migalastat Monotherapy

Migalastat monotherapy is being investigated in two Phase 3 registration studies (Study 011 and Study 012) and an open-label extension study (Study 041) in Fabry patients with amenable mutations. Interim 6-month data and positive 12- and 24-month data from Study 011 have been previously reported.

Study 012 is the first clinical study to compare oral migalastat to standard-of-care ERTs for Fabry disease (Fabrazyme® and Replagal®). The pre-specified co-primary outcome measures of efficacy in Study 012 are the descriptive assessments of comparability of the mean annualized change in measured (iohexol) GFR (mGFR) and estimated GFR (eGFR) for migalastat and ERT in patients with amenable mutations in a GLP-validated, human embryonic kidney cell-based assay ("GLP HEK amenable") .

The primary 18-month treatment period in Study 012 was completed in the second quarter of 2014, and top-line data are on track to be reported in the third quarter of 2014. Pending positive data from Study 012, Amicus expects to submit a marketing application for migalastat monotherapy in Europe. The Company also plans to meet with the Food and Drug Administration in the fourth quarter of this year to discuss the data from both Phase 3 Fabry monotherapy studies to determine the fastest U.S. registration pathway for migalastat.

Next-Generation Fabry ERT

Amicus is developing a next-generation Fabry ERT following positive data from a Phase 2 clinical study (Study 013) of migalastat co-administered with currently approved ERTs for Fabry disease (Fabrazyme® and Replagal®) as well as preclinical studies of migalastat co-formulated with a proprietary investigational ERT for Fabry disease (JCR Pharmaceutical Co Ltd's JR-051).

In the second quarter of 2014, Amicus completed a successful Phase 1 study to assess the pharmacokinetics of an intravenous formulation of migalastat in healthy volunteers to identify the optimal dose for co-formulation with ERT. The Company has also completed manufacturing of drug supply of co-formulated JR-051. Amicus plans to begin the Phase 1/2 clinical study of its next generation ERT pending the outcome of several ongoing business development discussions regarding potential future sources of Fabry enzyme for this next generation product. Amicus expects to provide further updates on the next-generation Fabry ERT development strategy in 2H14.

Next-Generation ERTs for Pompe Disease and MPS I

Amicus also owns exclusive global rights to its next-generation ERTs for Pompe disease and MPS I, as well as all applications of its Chaperone-Advanced Replacement Therapy (CHART™) and enzyme targeting technology platforms. These platform technologies provide a complementary tool set to create next-generation therapies that are designed to enhance tissue uptake of active enzyme, improve lysosomal activity and substrate reduction, and potentially address the tolerability and immunogenicity associated with currently marketed ERTs.

Next-Generation ERT for Pompe Disease

Amicus is advancing a recombinant human acid alpha-glucosidase (rhGAA) for Pompe disease into late preclinical development. This differentiated Pompe ERT, designated AT-B200, has a unique carbohydrate structure and may be further optimized through co-formulation with a pharmacological chaperone to improve enzyme stability and tolerability, and by applying the Company's peptide tagging technology for better targeting. In initial preclinical studies, AT-B200 has shown superior tissue uptake and activity when compared to current standard of care. In longer term preclinical proof-of-concept studies currently underway, AT-B200 has demonstrated superior substrate reduction compared to Lumizyme. Manufacturing scale up activities are on track to provide sufficient supply of AT-B200 for IND-enabling toxicology studies as well as a Phase 1/2 clinical study that is expected to begin in 2015.

Next-Generation ERT for MPS I

Amicus is developing a proprietary human recombinant alpha-L-iduronidase (rhIDUA) enzyme for MPS I. In support of its development of this next-generation ERT, Amicus has received funding of up to $250,000 from a private U.S.-based donor that provides medical research grants to find better treatments and cures for rare genetic disorders, including lysosomal storage diseases.

Novel Small Molecules for Parkinson's Disease

Amicus is developing pharmacological chaperones that target the glucocerobrosidase (GCase) enzyme for the treatment of Parkinson's disease. In September 2014, Amicus and Biogen will conclude their research collaboration on novel small molecules for Parkinson's disease. Amicus' most advanced Parkinson's candidate is AT3375, which was developed outside the collaboration and is wholly-owned by Amicus.

Conference Call and Webcast

Amicus Therapeutics will host a conference call and audio webcast today, August 7, 2014 at 5:00 p.m. ET to discuss second quarter 2014 financial results and program updates. Interested participants and investors may access the conference call at 5:00 p.m. ET by dialing 877-303-5859 (U.S./Canada) or 678-224-7784 (international).

An audio webcast can also be accessed via the Investors section of the Amicus Therapeutics corporate web site at http://www.amicusrx.com, and will be archived for 30 days. Web participants are encouraged to go to the web site 15 minutes prior to the start of the call to register, download and install any necessary software. A telephonic replay of the call will be available for seven days beginning at 8:00 p.m. ET today. Access numbers for this replay are 855-859-2056 (U.S./Canada) and 404-537-3406 (international); participant code 74809147.

About Amicus Therapeutics

Amicus Therapeutics (Nasdaq:FOLD) is a biopharmaceutical company at the forefront of therapies for rare and orphan diseases. The Company is developing novel, first-in-class treatments for a broad range of human genetic diseases, with a focus on delivering new benefits to individuals with lysosomal storage diseases. Amicus' lead programs in development include the small molecule pharmacological chaperone migalastat as a monotherapy for Fabry disease, as well as next-generation enzyme replacement therapy (ERT) products for Fabry disease, Pompe disease, and MPS I.

About Chaperone-Advanced Replacement Therapy (CHART)

The Chaperone-Advanced Replacement Therapy (CHART™) platform combines unique pharmacological chaperones with enzyme replacement therapies (ERTs) for lysosomal storage diseases (LSDs). In a chaperone-advanced replacement therapy, a unique pharmacological chaperone is designed to bind to and stabilize a specific therapeutic enzyme in its properly folded and active form. This proposed CHART mechanism may allow for enhanced tissue uptake of active enzyme, greater lysosomal activity, more reduction of substrate, and lower immunogenicity compared to ERT alone. Improvements in enzyme stability may also enable more convenient delivery of next-generation therapies. Amicus is leveraging the CHART platform to develop proprietary next-generation therapies that consist of lysosomal enzymes co-formulated with pharmacological chaperones.

Forward-Looking Statements

This press release contains, and the accompanying conference call will contain, "forward-looking statements" within the meaning of the Private Securities Litigation Reform Act of 1995 relating to preclinical and clinical development of Amicus' candidate drug products, the timing and reporting of results from preclinical studies and clinical trials evaluating Amicus' candidate drug products, and the projected cash position for the Company. Words such as, but not limited to, "look forward to," "believe," "expect," "anticipate," "estimate," "intend," "potential," "plan," "targets," "likely," "may," "will," "would," "should" and "could," and similar expressions or words identify forward-looking statements. Such forward-looking statements are based upon current expectations that involve risks, changes in circumstances, assumptions and uncertainties. The inclusion of forward-looking statements should not be regarded as a representation by Amicus that any of its plans will be achieved. Any or all of the forward-looking statements in this press release may turn out to be wrong. They can be affected by inaccurate assumptions Amicus might make or by known

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or unknown risks and uncertainties. For example, with respect to statements regarding the goals, progress, timing and outcomes of discussions with regulatory authorities and the potential goals, progress, timing and results of preclinical studies and clinical trials, actual results may differ materially from those set forth in this release due to the risks and uncertainties inherent in the business of Amicus, including, without limitation: the potential that results of clinical or pre-clinical studies indicate that the product candidates are unsafe or ineffective; the potential that it may be difficult to enroll patients in our clinical trials; the potential that regulatory authorities may not grant or may delay approval for our product candidates; the potential that preclinical and clinical studies could be delayed because we identify serious side effects or other safety issues; the potential that we will need additional funding to complete all of our studies and, our dependence on third parties in the conduct of our clinical studies. Further, the results of earlier preclinical studies and/or clinical trials may not be predictive of future results. With respect to statements regarding projections of the Company's cash position, actual results may differ based on market factors and the Company's ability to execute its operational and budget plans. In addition, all forward looking statements are subject to other risks detailed in our Annual Report on Form 10-K for the year ended December 31, 2013. You are cautioned not to place undue reliance on these forward-looking statements, which speak only as of the date hereof. All forward-looking statements are qualified in their entirety by this cautionary statement, and Amicus undertakes no obligation to revise or update this news release to reflect events or circumstances after the date hereof. This caution is made under the safe harbor provisions of Section 21E of the Private Securities Litigation Reform Act of 1995.

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Table 1

Amicus Therapeutics, Inc.
Consolidated Statements of Operations
(Unaudited)
(In thousands, except share and per share amounts)

	Three Months Ended June 30,		Six Months Ended June 30,	
	2013	2014	2013	2014
Revenue:				
Research revenue	$ —	$ 475	$ —	$ 931
Total revenue	$ —	$ 475	$ —	$ 931
Operating Expenses:				
Research and development	$ 10,725	$ 9,978	$ 22,714	$ 19,970
General and administrative	4,830	4,753	9,653	9,929
Changes in fair value of contingent consideration payable	—	(305)	—	200
Restructuring charges	—	(81)	—	(89)
Depreciation and amortization	450	396	889	808
Total operating expenses	16,005	14,741	33,256	30,818
Loss from operations	(16,005)	(14,266)	(33,256)	(29,887)
Other income (expenses):				

Interest income	46	36	111	78
Interest expense	(9)	(374)	(19)	(729)
Change in fair value of warrant liability	619	—	357	—
Other income	—	(10)	—	(19)
Net loss	$ (15,349)	$ (14,614)	$ (32,807)	$ (30,557)
Net loss per common share — basic and diluted	$ (0.31)	$ (0.22)	$ (0.66)	$ (0.46)
Weighted-average common shares outstanding — basic and diluted	49,621,188	67,212,764	49,621,188	65,799,059

Table 2

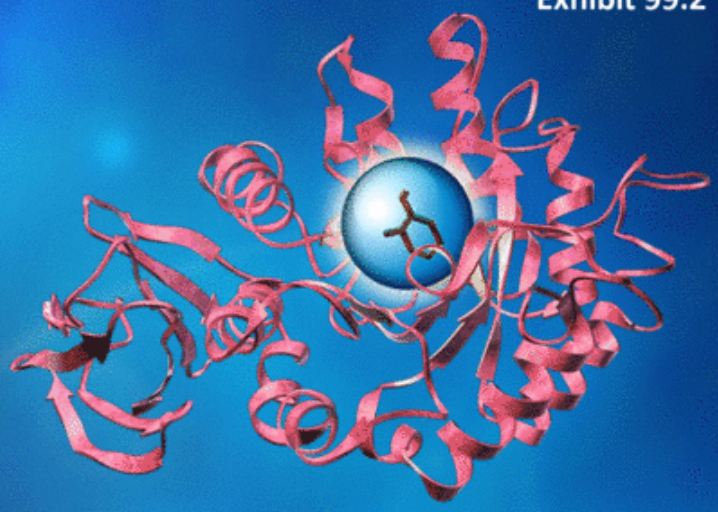
Amicus Therapeutics, Inc.
Consolidated Balance Sheets
(Unaudited)
(in thousands, except share and per share amounts)

	December 31, 2013	June 30, 2014
Assets:		
Current assets:		
Cash and cash equivalents	$ 43,640	$ 41,904
Investments in marketable securities	38,360	36,092
Receivable due from collaboration agreements	1,083	475
Prepaid expenses and other current assets	5,195	951
Total current assets	88,278	79,422
Property and equipment, less accumulated depreciation and amortization of $9,973 and $10,781 at December 31, 2013 and June 30, 2014, respectively	4,120	3,444
In-process research & development	23,000	23,000
Goodwill	11,613	11,613
Other non-current assets	552	515
Total Assets	$ 127,563	$ 117,994
Liabilities and Stockholders' Equity		
Current liabilities:		
Accounts payable and accrued expenses	$ 10,162	$ 9,918
Current portion of secured loan	299	1,253
Total current liabilities	10,461	11,171
Deferred reimbursements	36,677	36,677
Secured loan, less current portion	14,174	13,104
Contingent consideration payable	10,600	10,800
Deferred tax liability	9,186	9,186
Other non-current liability	714	748
Commitments and contingencies		
Stockholders' equity:		
Common stock, $.01 par value, 125,000,000 shares authorized, 61,975,416 shares issued and outstanding at December 31, 2013, 125,000,000 shares authorized, 72,869,861 shares issued and outstanding at June 30, 2014	679	789
Additional paid-in capital	423,593	444,600
Accumulated other comprehensive income	1	(2)
Accumulated deficit	(378,522)	(409,079)
Total stockholders' equity	45,751	36,308
Total Liabilities and Stockholders' Equity	$ 127,563	$ 117,994

FOLD–G

Exhibit 99.2

Amicus Therapeutics



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 **AT**1001-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

Amicus Therapeutics

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

Amicus Therapeutics

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

Amicus Therapeutics

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

s As of Aug 4 2014

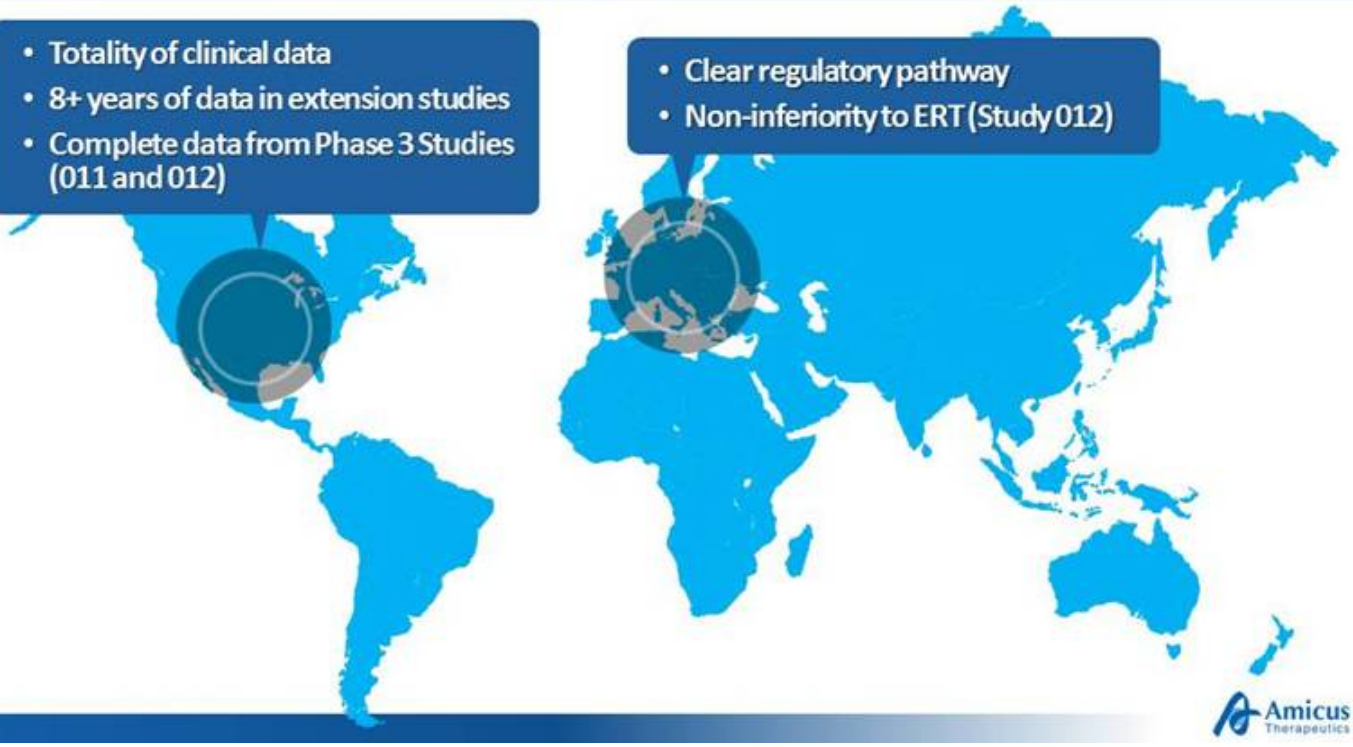
Amicus Therapeutics

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)

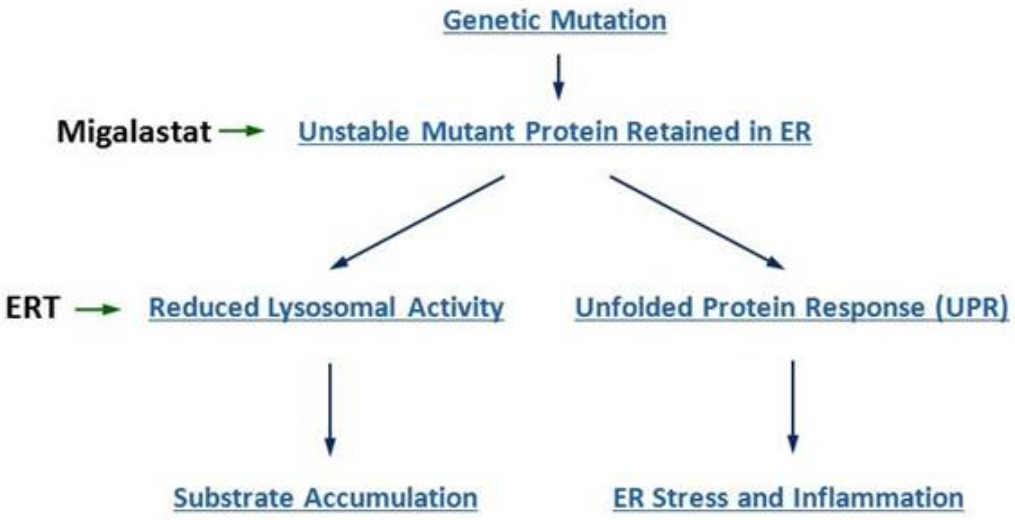
- Clear regulatory pathway
- Non-inferiority to ERT (Study 012)



Amicus Therapeutics

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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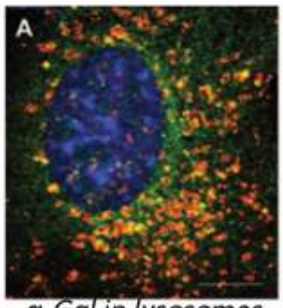
Migalastat Restores Trafficking of Mutant Enzyme

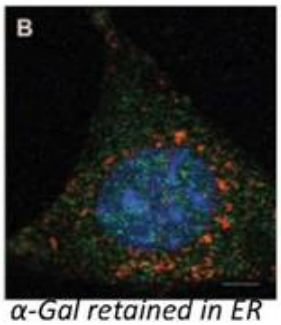
Migalastat Restores Ability of Mutant α-Gal A Retained in ER to Traffic to Lysosomes and Reduces a Marker of ER Stress in a Fabry Mouse Model

Wild-Type Untreated



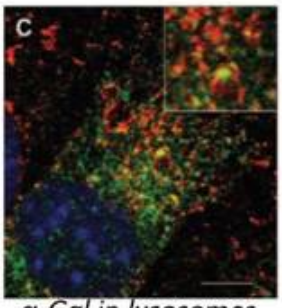
α-Gal in lysosomes

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

Amicus Therapeutics

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

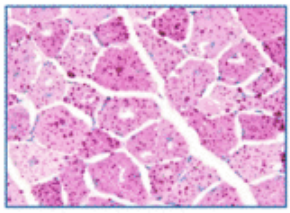
 Sources: Cybulsky 2013; Inagi 2014

Amicus Therapeutics

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



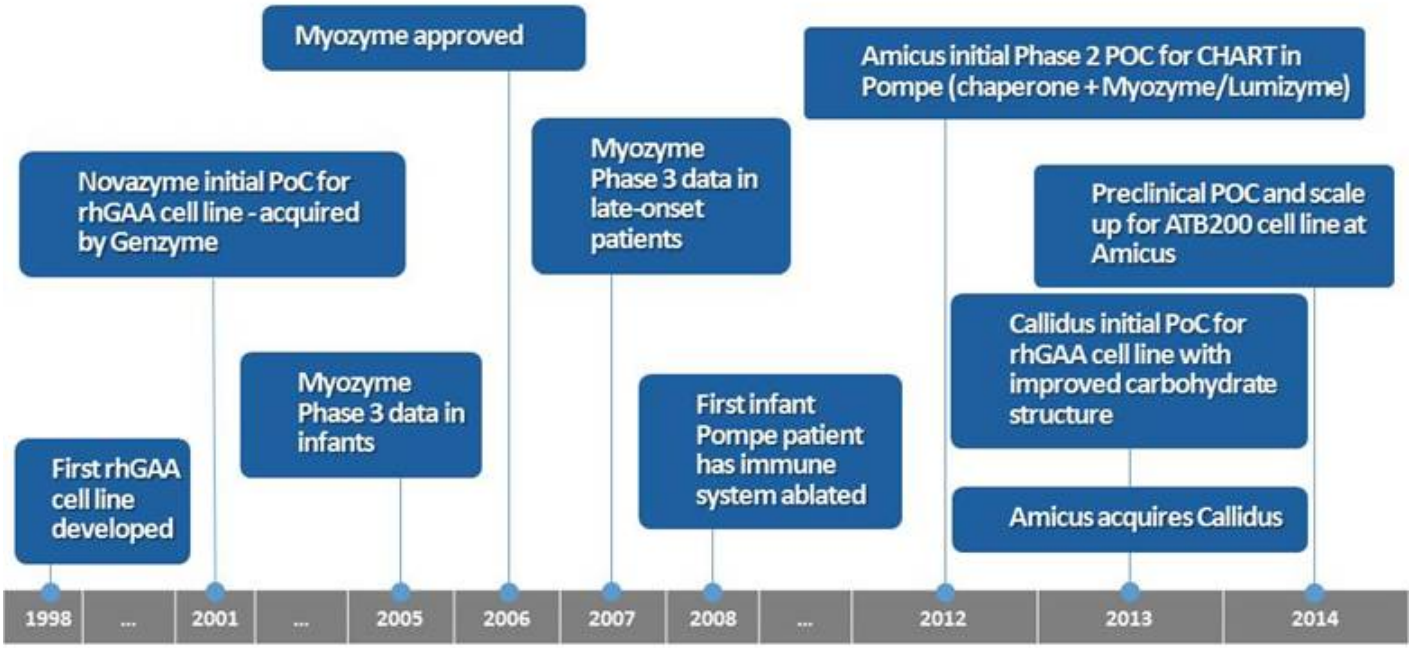
Elevated Glycogen in Muscle

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

Amicus Therapeutics

Select Milestones in Pompe Drug Development

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



Amicus Therapeutics

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]

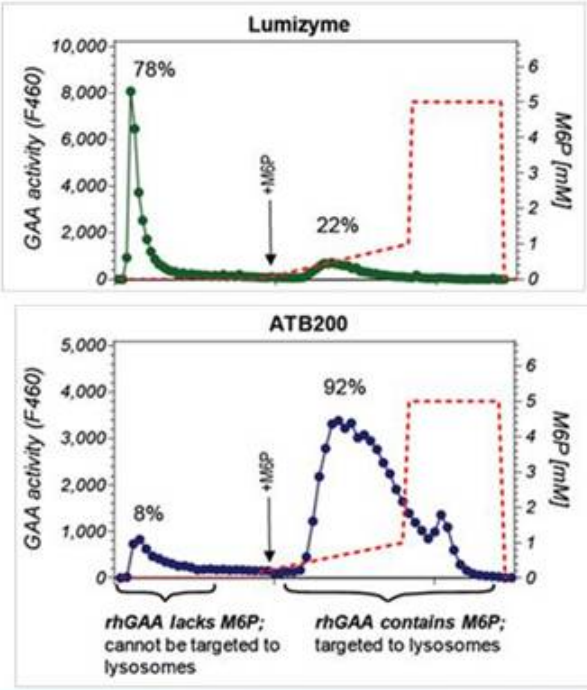
Majority of rhGAA is not delivered to lysosomes[2]

[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.

Amicus Therapeutics

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



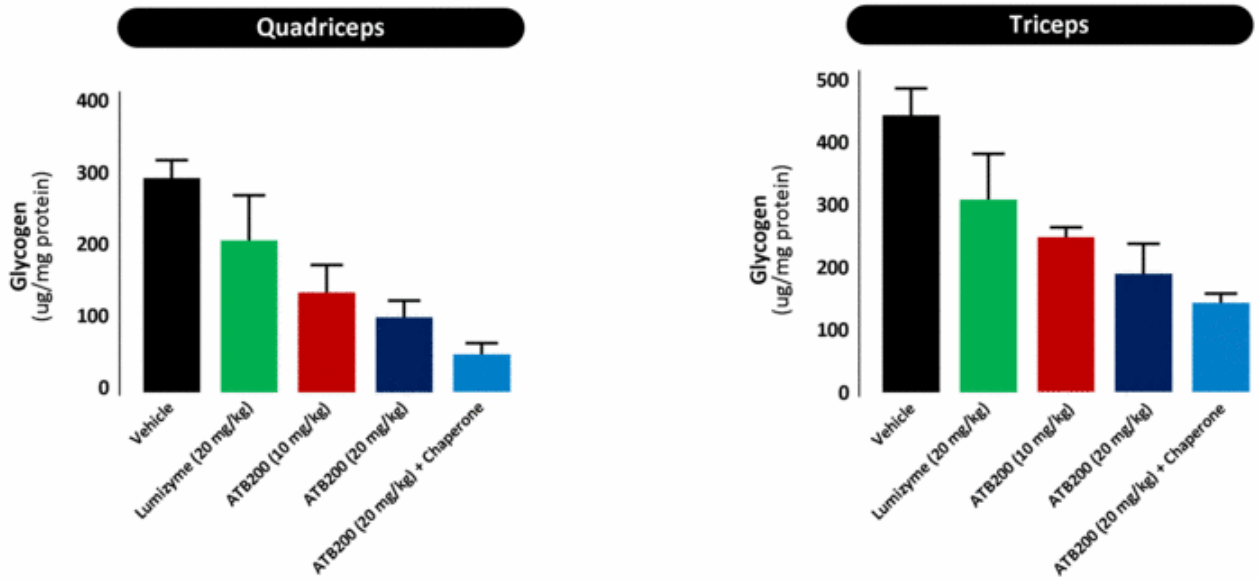
- Developed proprietary cell line for producing rhGAA (designated as ATB200)
- ATB200 has significantly higher M6P content than existing rhGAA ERTs
- ATB200 binds intended M6P receptor substantially better than standard of care ERT

Amicus Therapeutics

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



Amicus Therapeutics

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)

Amicus Therapeutics

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

Amicus Therapeutics

2Q14 Financial Results

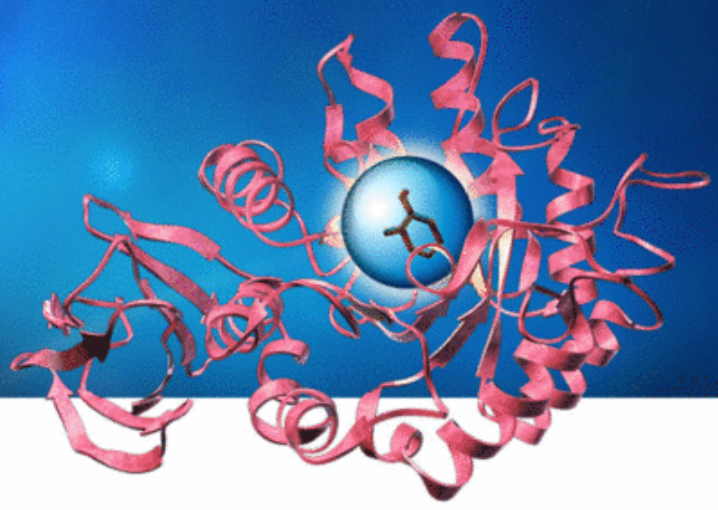
($000s)	June 30, 2014	June 30, 2013
Total Revenue	475	---
Total Operating Expenses	14,741	16,005
Net Loss	(14,614)	(15,349)
Net Loss Per Share	(0.22)	(0.31)

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

Amicus Therapeutics

Amicus Therapeutics



Questions & Answers


Amicus
Therapeutics
2Q14 Financial Results
Conference Call & Webcast
August 7, 2014
at the forefront of therapies
for rare and orphan diseases