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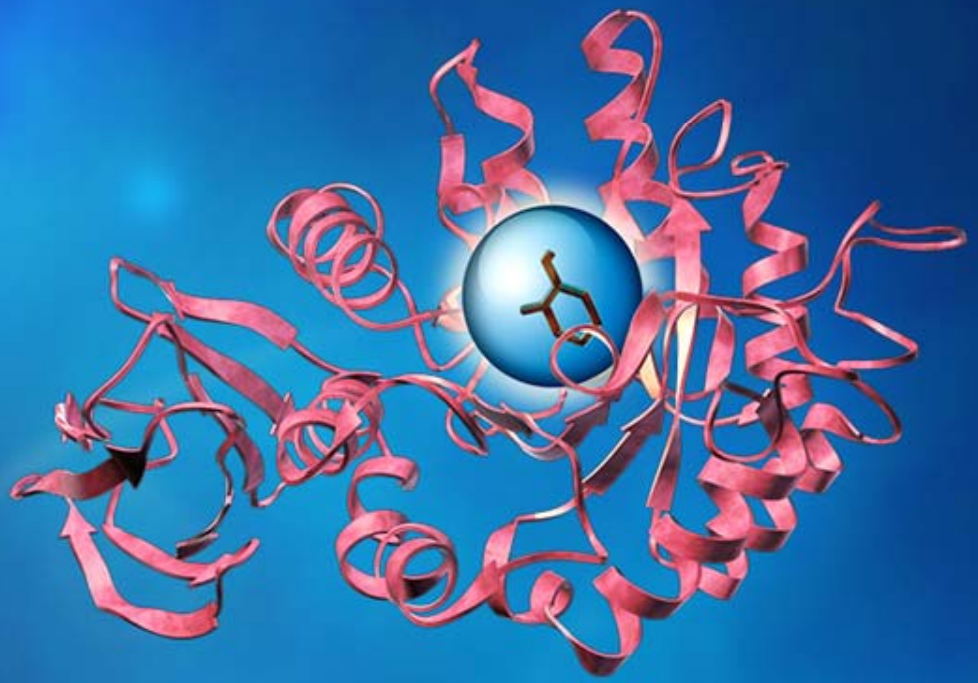


1Q14 Financial Results Conference Call & Webcast

May 5, 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.

Agenda

- Opening Remarks
- Summary of Phase 3 Monotherapy Study (Study 011) 12 and 24 Month Results
- Update on Next-Generation ERTs in Fabry, Pompe and MPS I
- Financial Results
- Q&A

Migalastat Monotherapy: Study 011 12- and 24-Month Data - Key Findings

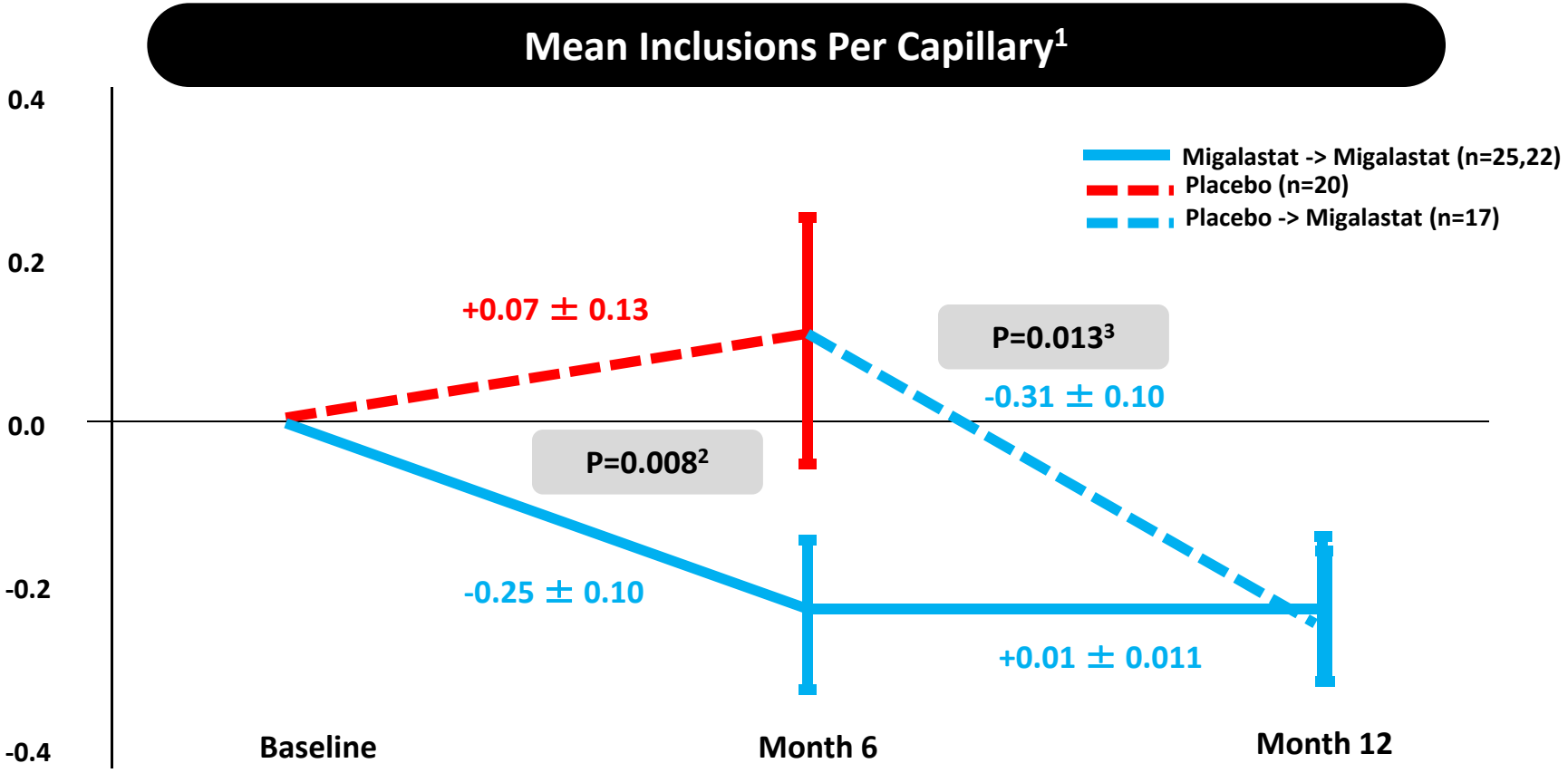
Migalastat Demonstrated Statistically Significant and Durable Substrate Reductions on 12-Month Pre-Specified Primary Analysis in Fabry Patients with Amenable Mutations

- 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

12-Month Pre-Specified Primary Analysis

Statistically Significant Mean Change in Kidney Interstitial Capillary GL-3 in Patients Switching from Placebo to Migalastat HCl (GLP HEK Amenable)*



*All patients with evaluable paired biopsies and amenable GLA mutations in GLP-validated HEK assay – post hoc at month 6 and pre-specified at month 12 [1]Data points are baseline corrected; represent mean ± standard error (SEM) change from baseline in the mean number of GL-3 inclusions per capillary after 6 months of treatment with migalastat or placebo. [2]Analysis of covariance (ANCOVA) model with covariate adjustment for baseline value and factors for treatment group and treatment by baseline interaction. P-value corresponding to least-square mean difference between migalastat and placebo is displayed. [3]MMRM Pbo change M6 to M12.

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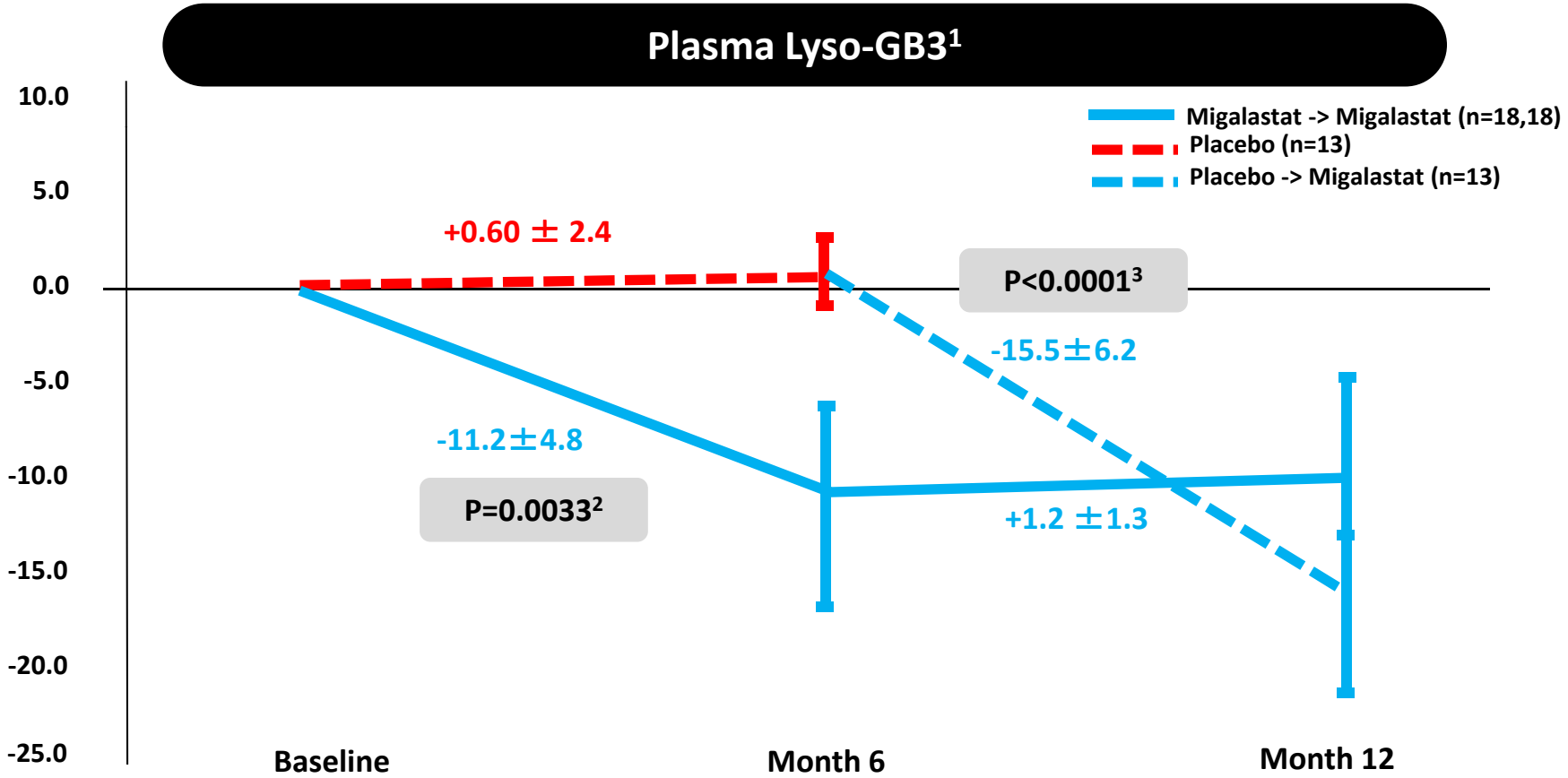
Disease Substrate in Plasma (Plasma Lyso-GB3)

Statistically Significant Reduction in Plasma Lyso-GB3 at Month 6 and Month 12 Following Treatment with Migalastat (GLP HEK Amenable)*



*Patients with amenable GLA mutations in GLP-validated HEK assay [1]Baseline corrected. Error bars are SEM [2]ANCOVA comparing migalstat to placebo in Stage 1 [3]ANCOVA comparing change from month 6 to month 12 in subjects switching from placebo to migalastat

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Kidney Function: Annualized Glomerular Filtration Rate (GFR)

GFR Remained Stable Over 18-24 Months (GLP HEK Amenable)*

Annualized GFR (ml/min/m^2/yr) at Month 18 or 24[1]			
GFR Measure	**N***	**Mean**	**(SEM)**
eGFR (CKD-EPI)	41	-0.30	(0.66)
eGFR (MDRD)	41	0.79	(1.03)
mGFR (iohexol)	37	-1.51	(1.33)

*Patients with amenable GLA mutations in GLP-validated HEK assay

[1]24 Months of Data in Subjects Treated with Migalastat from Baseline, 18 Months of Data in Subjects Switched from Placebo to Migalastat After 6 Months

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Safety Summary

Migalastat Generally Safe and Well Tolerated

Most Common Treatment Emergent Adverse Events (≥ 10% of Subjects)

Adverse event	Baseline to Month 6		Months 7-12		Open-Label Extension (Months 13-24)	
	Placebo* (n=33)	Migalastat (n=34)	Placebo-Migalastat* (n=30)	Migalastat (n=33)	Placebo-Migalastat* (n=28)	Migalastat (n=29)
Any Event	91%	91%	80%	79%	86%	83%
Headache	21%	35%			11%	10%
Fatigue	12%	12%				
Nausea	9%	12%				
Nasopharyngitis	6%	15%				
Paresthesia	12%	9%				
Procedural Pain			10%	12%		
Proteinuria					18%	14%
Bronchitis					11%	10%

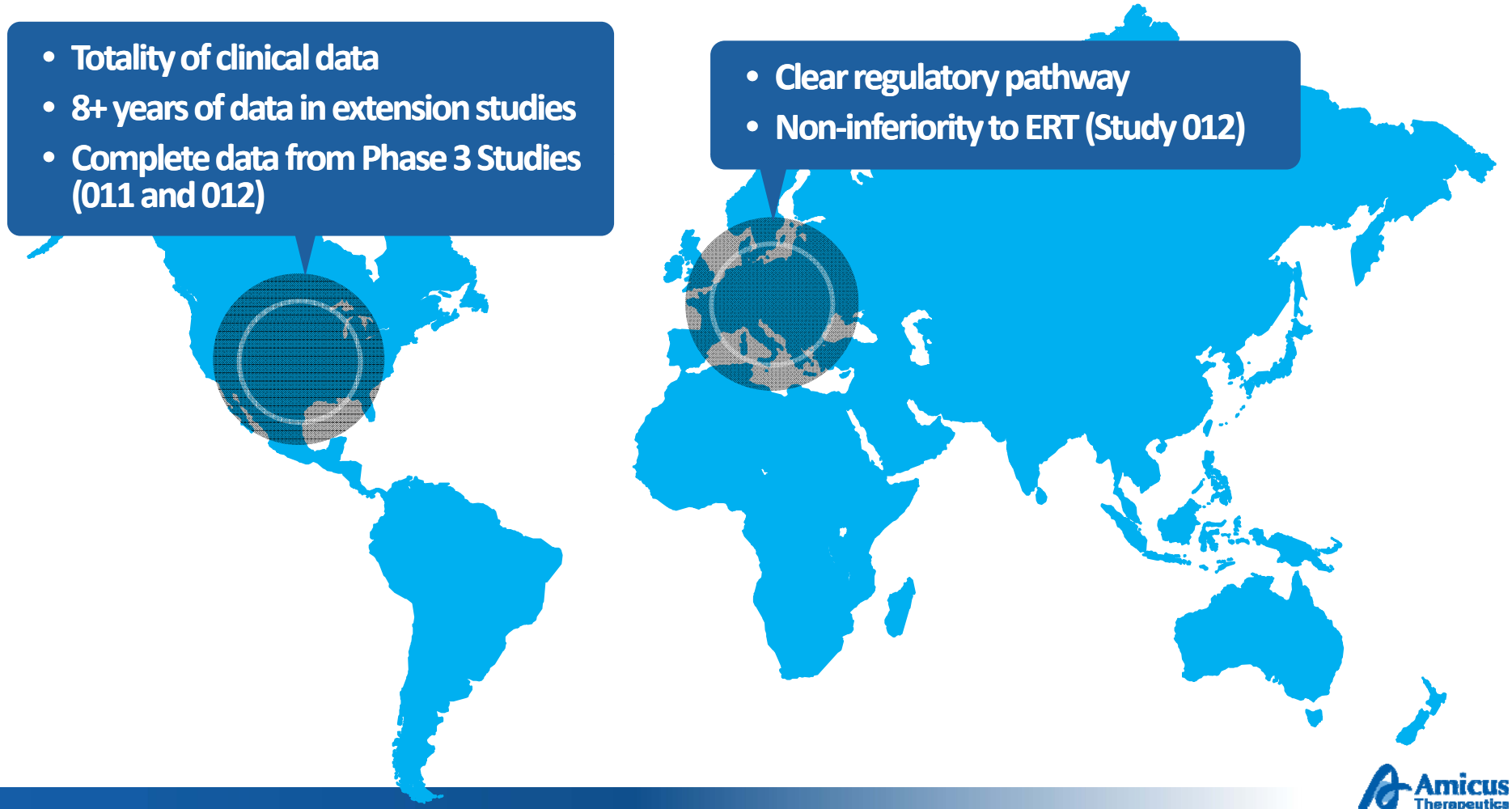
*Subjects Received Placebo from Baseline to Month 6, Switched to Migalastat After Month 6

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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3-in-3 Strategy: Pathway to Clinic

Executing Strategy to Advance 3 Next-Generation ERTs into Clinic in Next 3 Years with Lead Programs in Fabry, Pompe and MPS I

Milestones	Fabry Next-Generation ERT	
1H14	Phase 1 study initiation of IV migalastat in healthy volunteers	✓
2H14	**Phase 1/2 study initiation**	
Milestones	**Pompe Next-Generation ERT**	
1Q14	Initial preclinical proof-of-concept presented at LDN WORLD	✓
Ongoing	Longer-term preclinical proof-of-concept studies to optimize product for clinic with better tissue uptake and enzyme stability	✓
Ongoing	Manufacturing scale-up activities	✓
2H14	**Selection of final drug candidate for IND-enabling studies**	
2015	**Phase 1/2 study initiation**	

Current Financial Picture

Financial Position	
March 31 cash:	$71.6M
2014 net cash spend:	$54-59M
Cash runway:	2H15
Capitalization	
Shares outstanding:	64,366,088

1Q14 Financial Results

	March 31, 2014	March 31, 2013
Total Revenue	456	---
Total Operating Expenses	16,077	17,251
Net Loss	(15,943)	(17,458)
Net Loss Per Share	(0.25)	(0.35)

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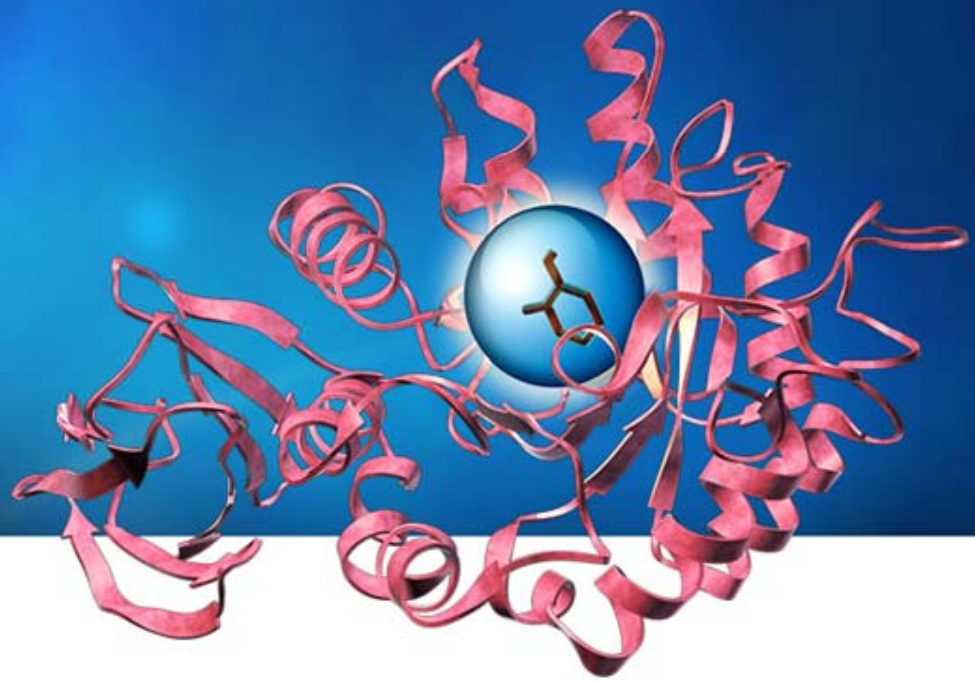

Questions & Answers

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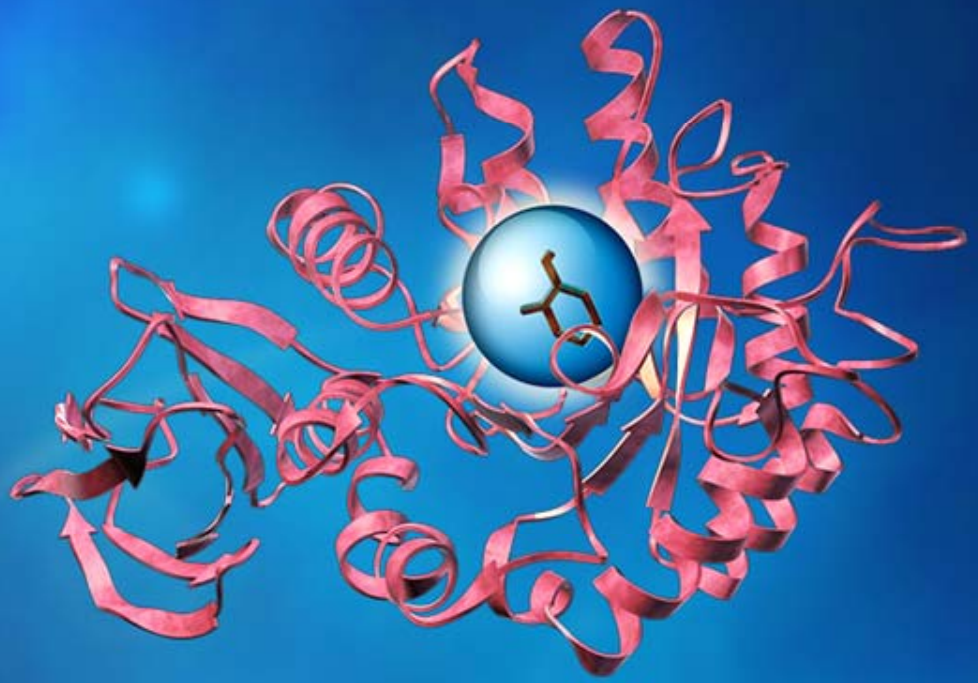

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