Second Quarter 2016 Financial Results

August 4, 2016

Cautionary Note Regarding Forward-Looking Statements

This presentation and various remarks we make during this presentation contain forward-looking statements within the meaning of The Private Securities Litigation Reform Act of 1995, including those regarding Agios' expectations and beliefs about: the potential of IDH1/IDH2 and pyruvate kinase-R mutations as therapeutic targets; the potential benefits of Agios' product candidates targeting IDH1/IDH2 or pyruvate kinase-R mutations or other genetic mutations, including AG-221, AG-120, AG-881, AG-348 and AG-519; its plans and timelines for the clinical development of AG-221, AG-120, AG-881, AG-348 and AG-519; its plans regarding future data presentations; its financial guidance regarding the amount of cash, cash equivalents and marketable securities that the company will have as of December 31, 2016; and the potential benefit of its strategic plans and focus. The words "anticipate," "believe," "estimate," "expect," "intend," "may," "plan," "predict," "project," "would," "could," "potential," "possible," "hope" and similar expressions are intended to identify forward-looking statements, although not all forward-looking statements contain these identifying words. Such statements are subject to numerous important factors, risks and uncertainties that may cause actual events or results to differ materially from Agios' current expectations and beliefs. For example, there can be no guarantee that any product candidate Agios is developing will successfully commence or complete necessary preclinical and clinical development phases, or that development of any of Agios' product candidates will successfully continue. There can be no guarantee that any positive developments in Agios' business will result in stock price appreciation. Management's expectations and, therefore, any forward-looking statements in this presentation or the various remarks made during this presentation could also be affected by risks and uncertainties relating to a number of other important factors, including: Agios' results of clinical trials and preclinical studies, including subsequent analysis of existing data and new data received from ongoing and future studies; the content and timing of decisions made by the U.S. FDA and other regulatory authorities, investigational review boards at clinical trial sites and publication review bodies; Agios' ability to obtain and maintain requisite regulatory approvals and to enroll patients in its planned clinical trials; unplanned cash requirements and expenditures; competitive factors; Agios' ability to obtain, maintain and enforce patent and other intellectual property protection for any product candidates it is developing; Agios' ability to maintain key collaborations, such as its agreements with Celgene; and general economic and market conditions. These and other risks are described in greater detail under the caption "Risk Factors" included in Agios' Quarterly Report on Form 10-Q for the quarter ended March 31, 2016, and other filings that Agios may make with the Securities and Exchange Commission in the future. Any forward-looking statements contained in this presentation or in remarks made during this presentation speak only as of the date hereof, and Agios expressly disclaims any obligation to update any forward-looking statements, whether as a result of new information, future events or otherwise, except as required by law.

Agios Conference Call Participants

Prepared Remarks

Introduction

- RENEE LECK, Sr. Manager, Investor & Public Relations

Second Quarter Highlights

- DAVID SCHENKEIN, M.D., Chief Executive Officer

Clinical Development Progress

- CHRIS BOWDEN, M.D., Chief Medical Officer

Second Quarter 2016 Financial Results

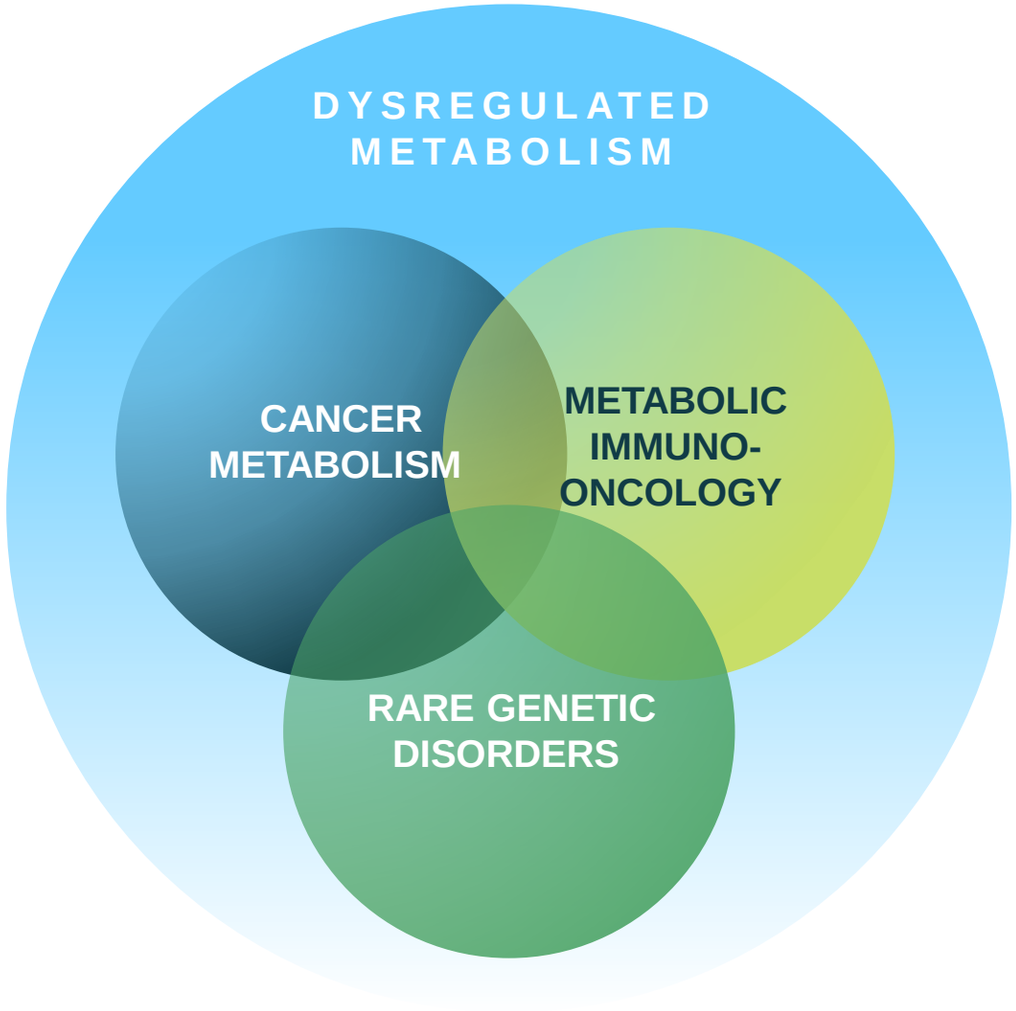
- GLENN GODDARD, Senior Vice President, Finance

Second Quarter Highlights

David Schenkein, M.D., Chief Executive Officer

agios

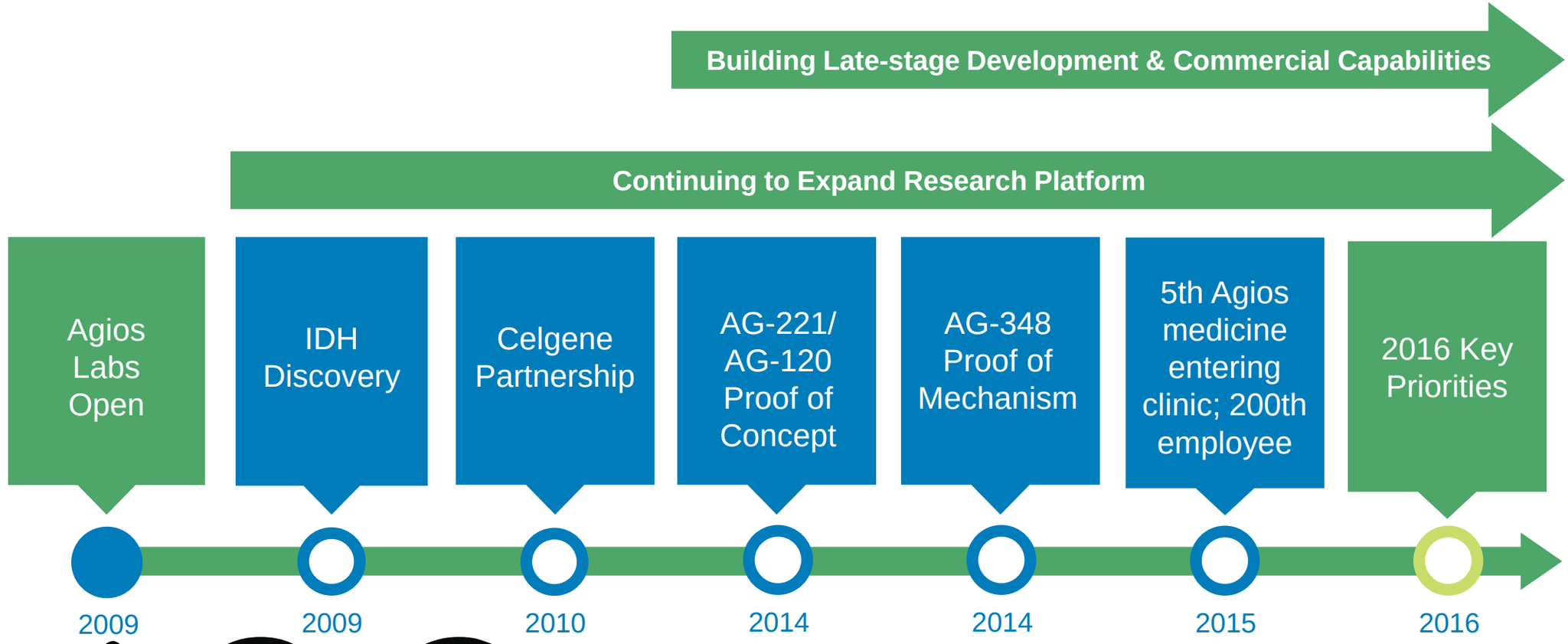
We Are Driven By a Clear Vision and Values



agios

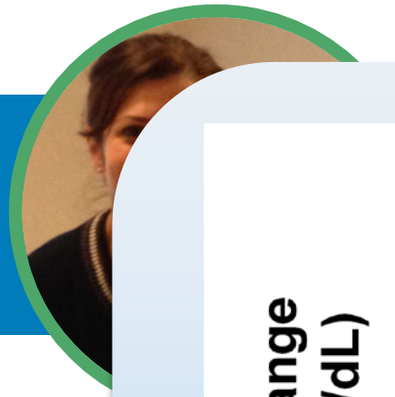
Agios is passionately committed to applying our scientific leadership in the field of cellular metabolism to transform the lives of patients with cancer and rare genetic disorders.

Building a Great Sustainable Biopharmaceutical Company



agios

Our 2016 Key Priorities: Maturing into a Late-stage Company



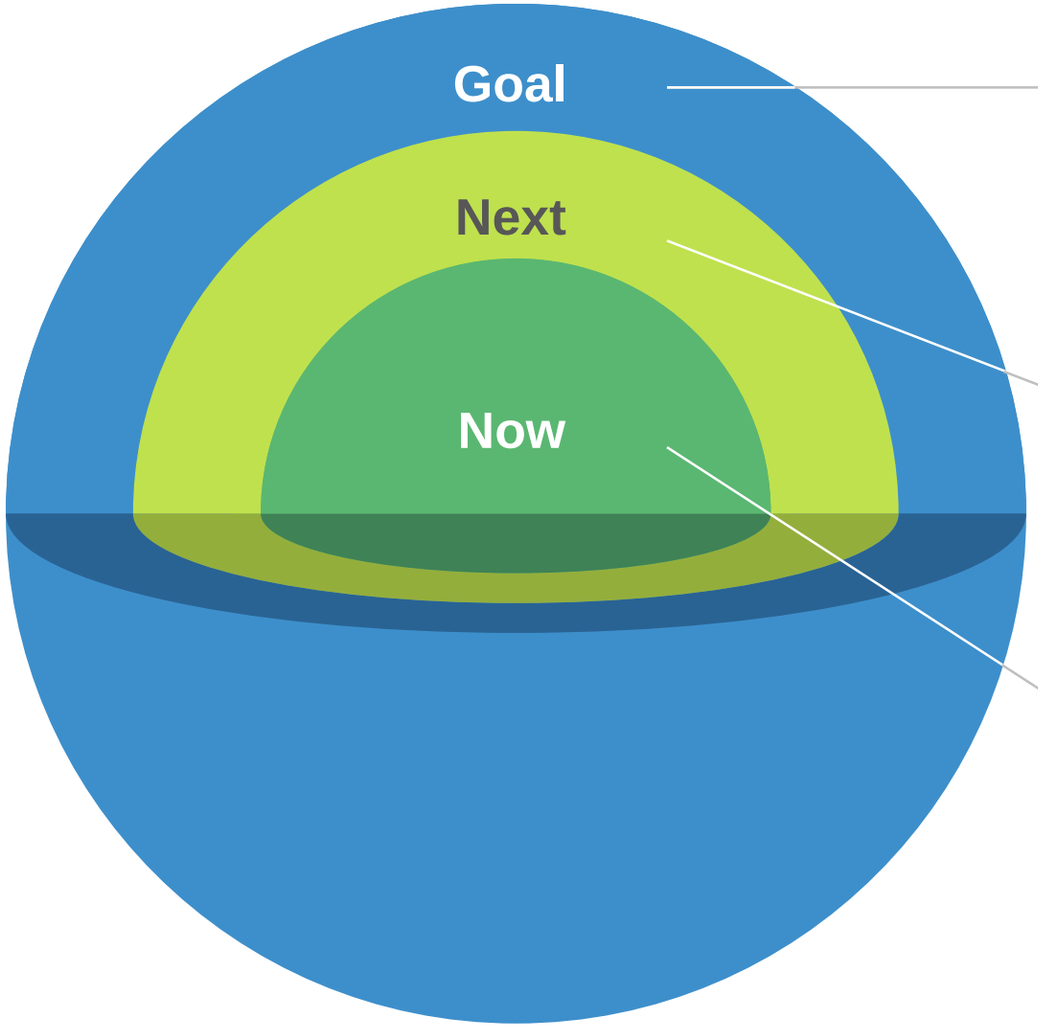
al

De
in

clinical
rch program

What's Possible for IDHm Patients

A Roadmap for Speed and Breadth



All IDHm patients screened and treated with an IDHm inhibitor for the entire course of their disease

Solid tumors
Frontline AML
Combination trials
Maintenance
MDS
Other hematologic malignancies

Relapsed/
Refractory AML

Agios / Celgene Collaboration Highlights

New Collaboration

- New strategic collaboration to discover, develop and commercialize metabolic immuno-oncology therapies
- Agios received $200 million upfront payment, extends runway to mid-2018

Amended Rights to 2010 Agreement

- 50/50 worldwide rights to two cancer metabolism discovery programs, including an MTAP program, moved under new research collaboration
- Agios gained global rights for AG-120

Clinical Development Progress

Chris Bowden, M.D., Chief Medical Officer

agios

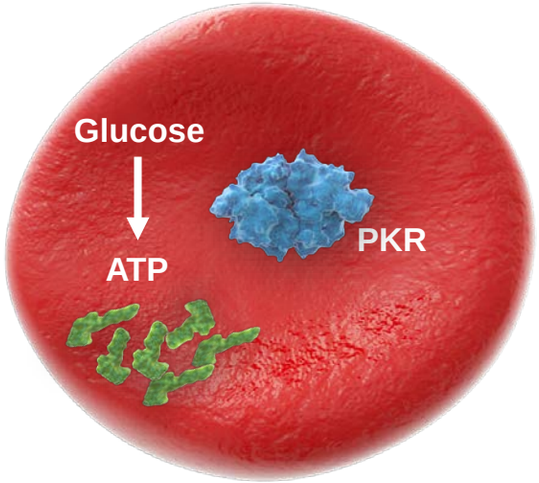
PK Deficiency Is a Rare Genetic Disease that Affects Red Blood Cells

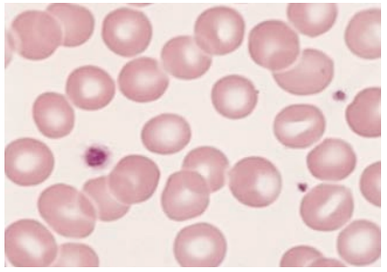
Rare genetic disease of erythrocyte pyruvate kinase

- PK deficiency often presents at birth with jaundice and can cause lifelong hemolytic anemia and associated morbidities.

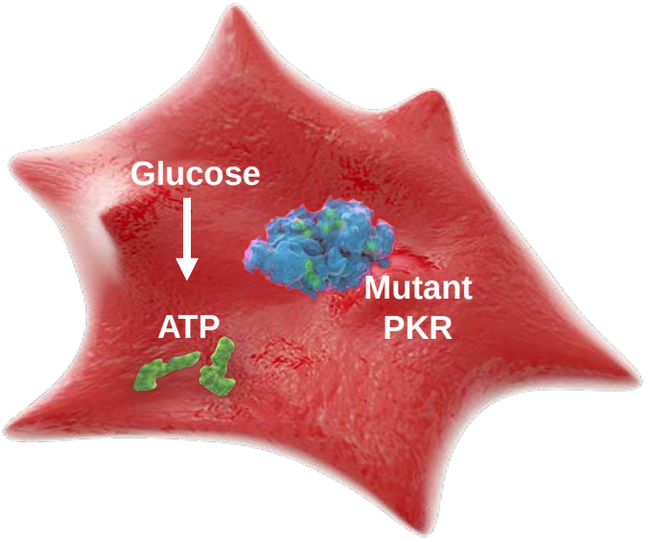
PKR regulates a crucial step in red blood cell metabolism & causes premature death of these cells

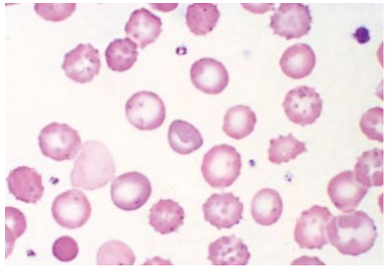
Healthy





PK Deficiency

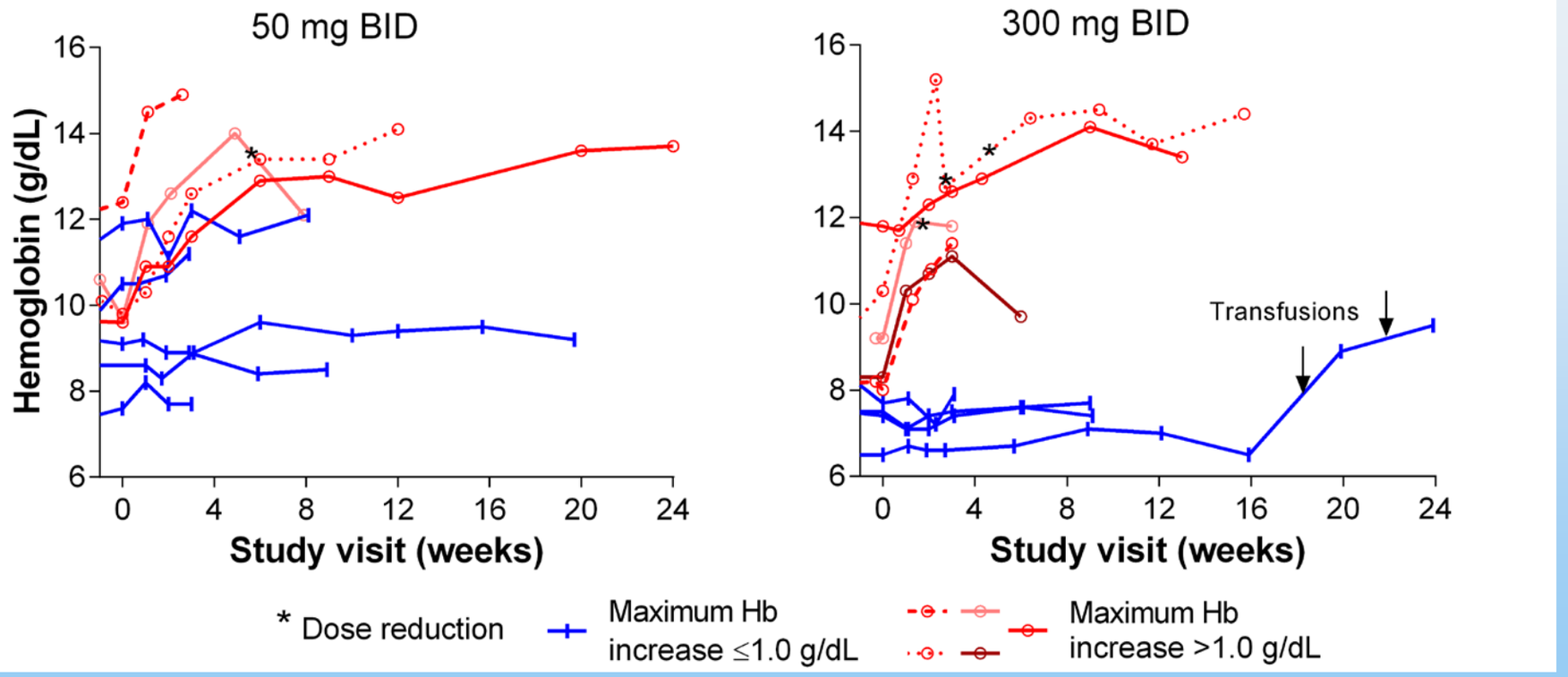




AG-348 Has Achieved Proof-of-Concept in PK Deficiency

Hemoglobin Increases Are Robust, Rapid and Sustained

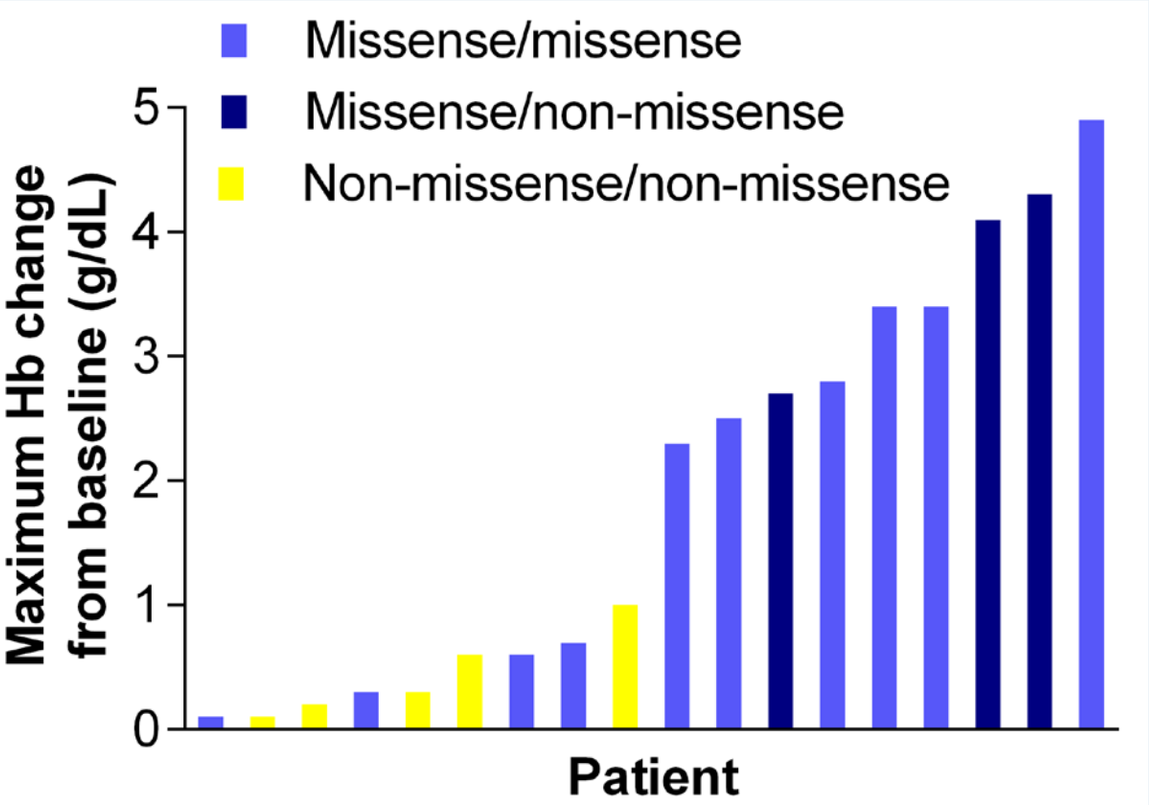
- Robust increases in hemoglobin over 1.0 g/dL reported in 9 of 18 patients
  - Mean maximum increase was 3.4 g/dL (range 2.3–4.9 g/dL)
  - Median time to a Hb increase >1.0 g/dL was 1.9 weeks (range 1.1–9.1 weeks)
- Well-tolerated safety profile with up to 6 months twice daily dosing



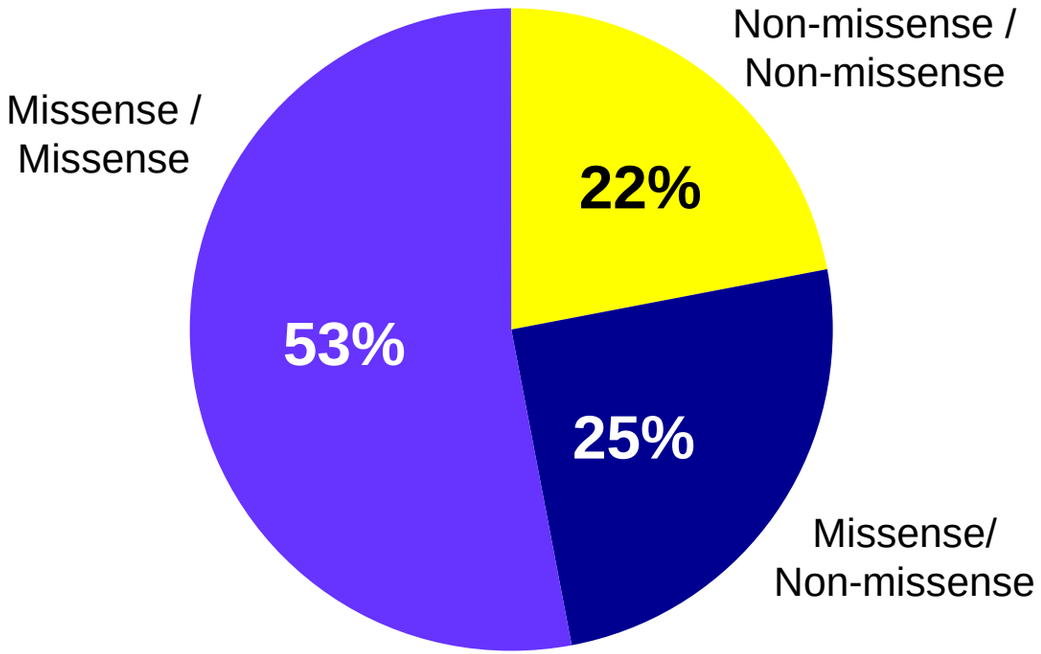
AG-348 DRIVE PK Data presented at EHA, 6/11/16

Relationship Between Genotype and Hemoglobin Change

9 out of 13 patients with at least one missense mutation had an increase in Hb >1.0 g/dL

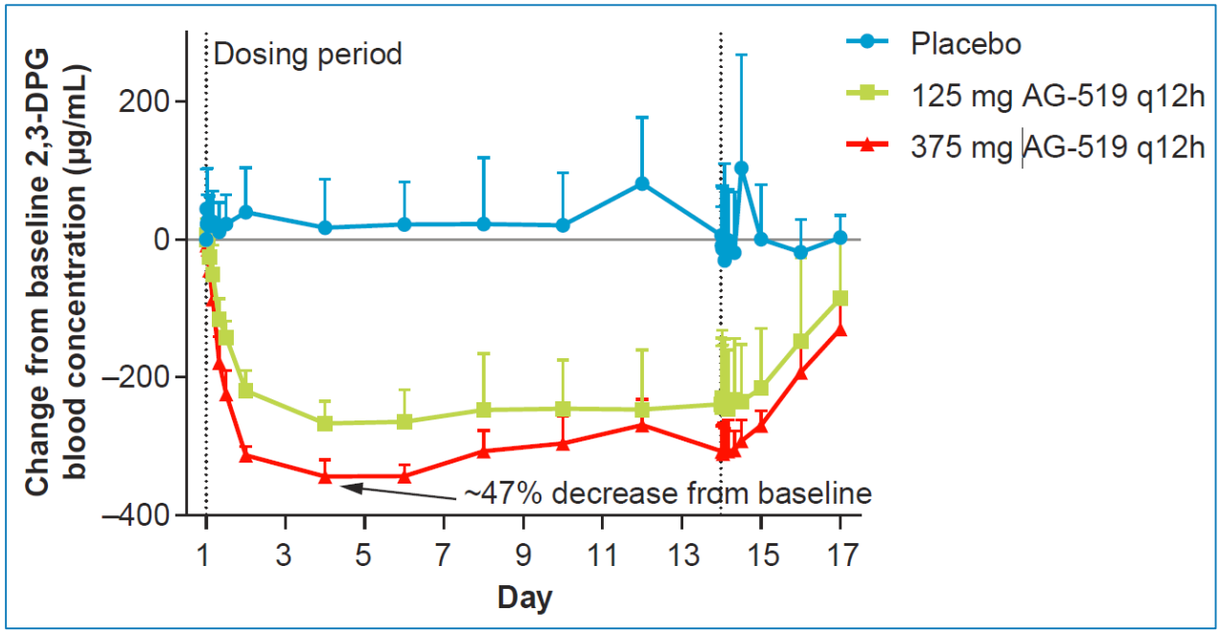


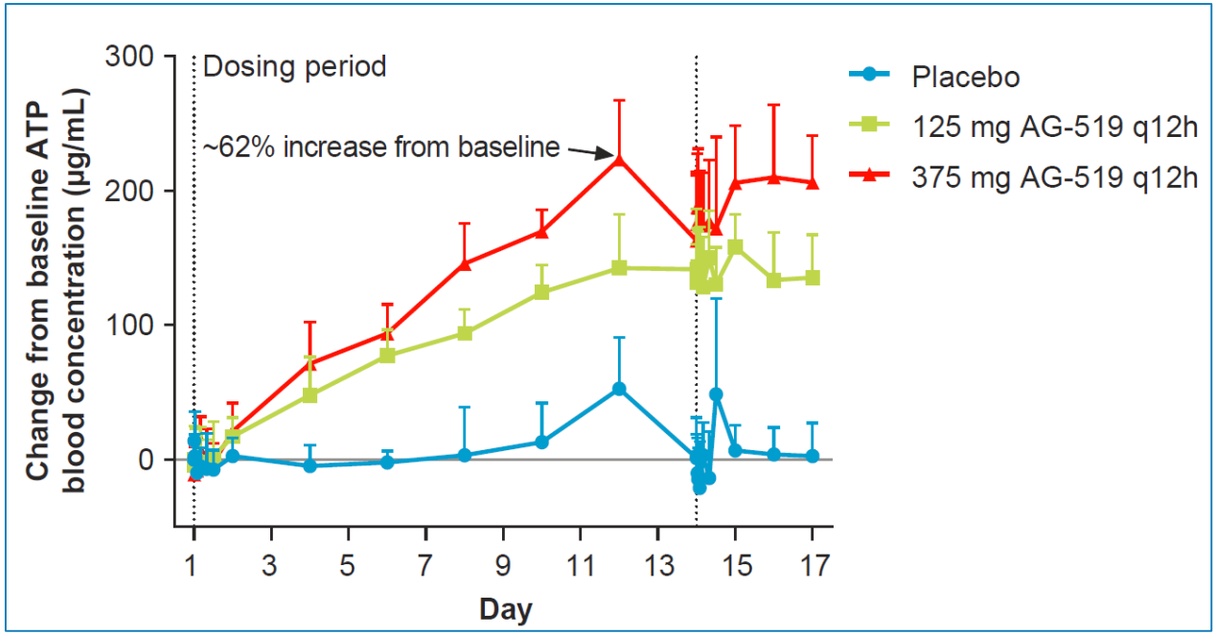
Type of PKR mutations found in 74 unrelated PK deficiency cases enrolled in the natural history study



AG-348 DRIVE PK Data presented at EHA, 6/11/16

AG-519 Has Demonstrated Proof-of-Mechanism

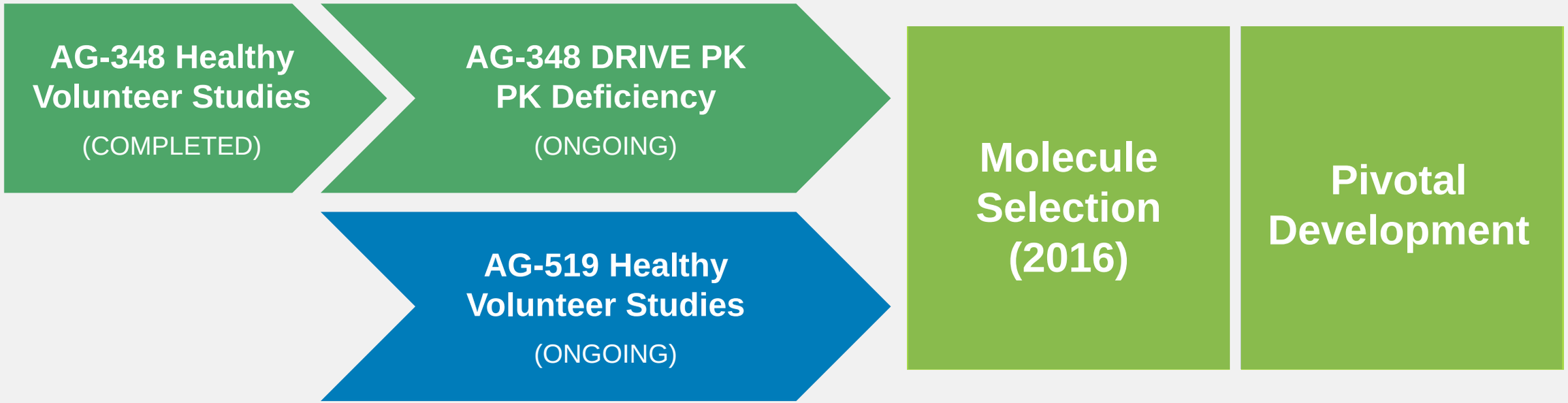




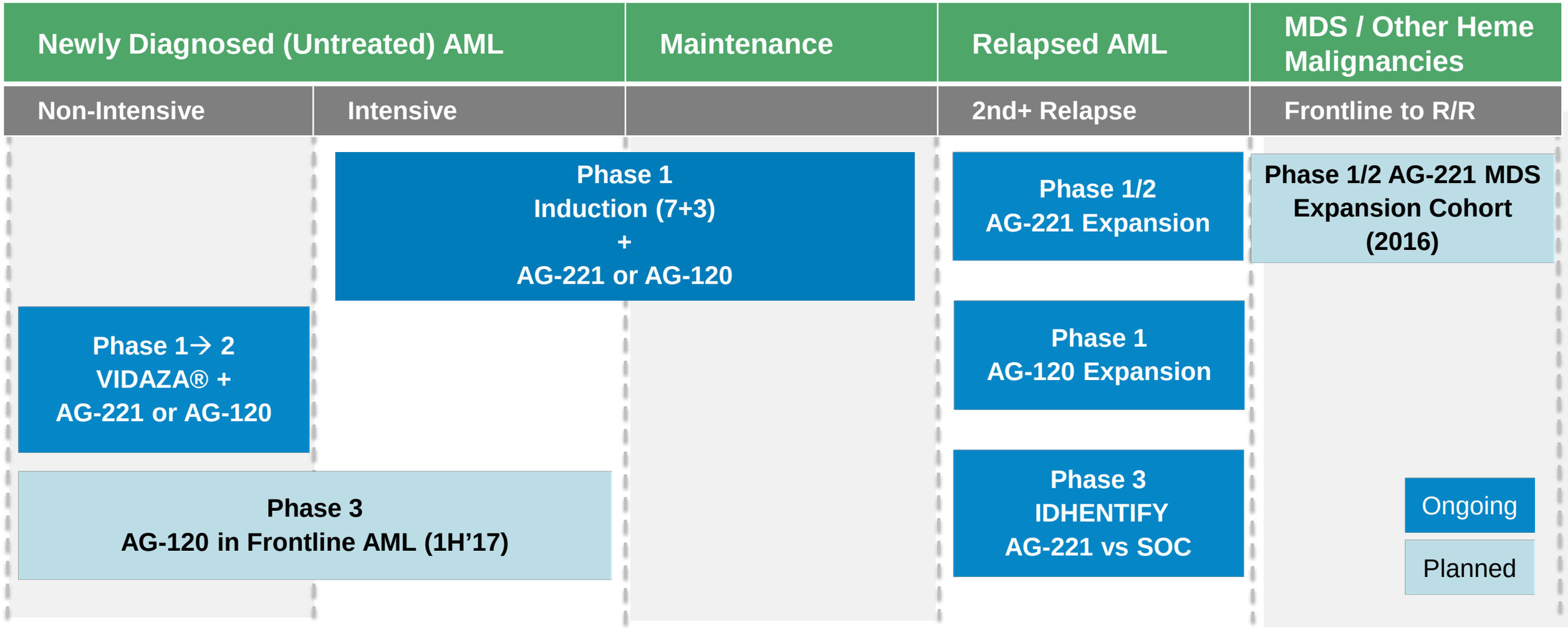
- Rapid 2,3-DPG lowering
- Substantial elevation of ATP levels, peaking at Day 12
- PK/PD profile comparable to AG-348
- Favorable safety profile, majority of AEs mild or moderate

Data presented at EHA, 6/10-11/16

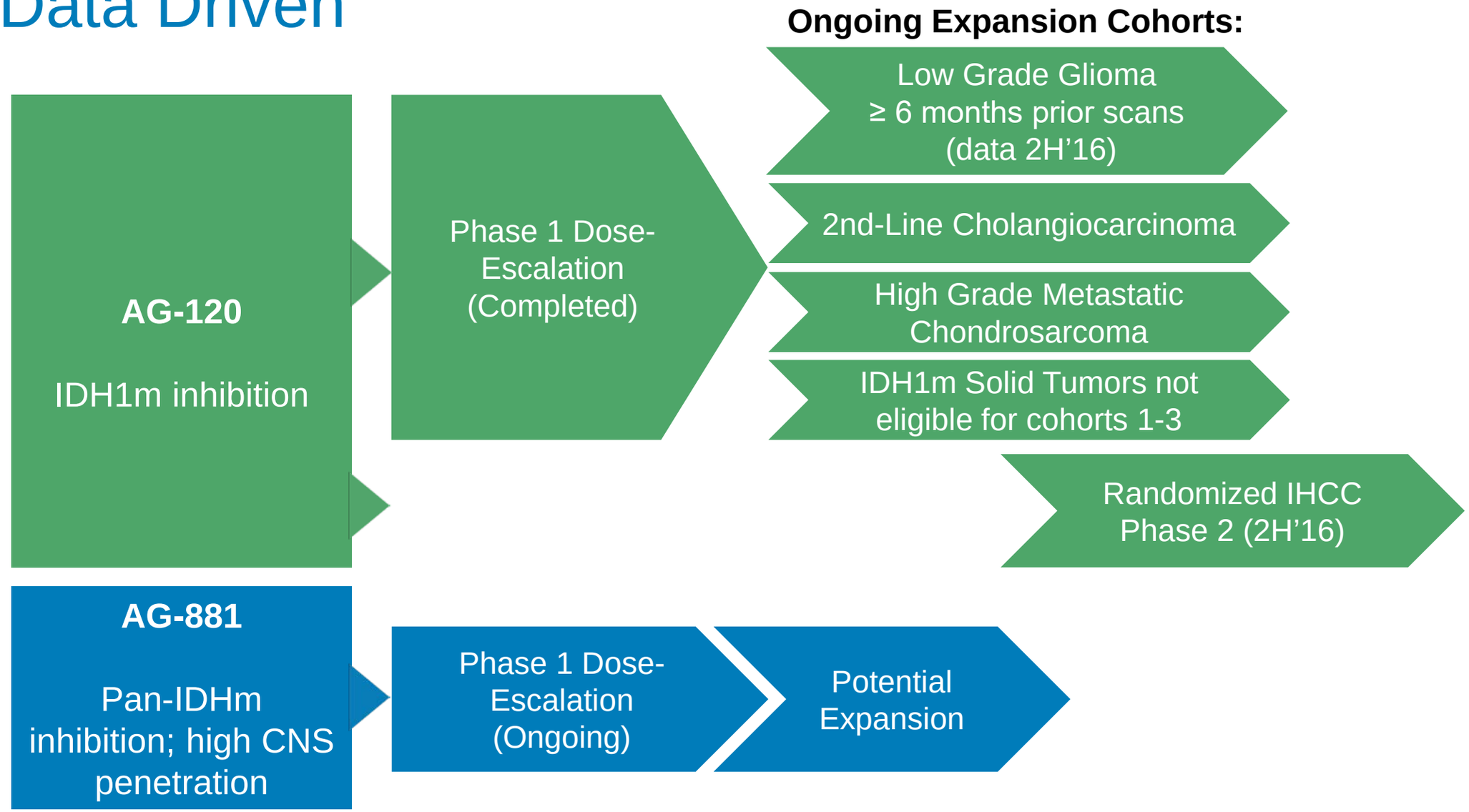
Proof-of-Concept Achieved, Moving Toward Pivotal Development



Targeting Multiple Lines of Treatment in IDHm AML and Other Hematologic Malignancies



Clinical Development Path in IDH1m Solid Tumors Will Be Data Driven



Second Quarter 2016 Financial Results

Glenn Goddard, Senior Vice President, Finance

agios

Second Quarter 2016 Financial Results

Balance Sheet	June 30, 2016	December 31, 2015
Cash, cash equivalents and marketable securities	$512M	$376M
Total Assets	$558M	$420M

Statement of Operations	Three Months Ended June 30, 2016	Three Months Ended June 30, 2015
Collaboration Revenue	$7M	$13M
Research & Development Expense (1)	$51M	$36M
General & Administrative Expense	$13M	$9M

Note 1 (R&D expenses): R&D expense are presented net of amounts received from Celgene for reimbursement of certain development costs incurred on Celgene's behalf related to AG-221, AG-120 and AG-881. The R&D expense reported for the three months ended June 30, 2016 and 2015 are presented net of $5.9 million and $4.5 million, respectively, of reimbursements received.

2016 Milestone Progress

IDHm Inhibitors

- ✓ Complete enrollment in AG-221 expansion arm
- Complete enrollment in AG-120 expansion arm in 2H
- Initiate MDS expansion arm for AG-221
- First data from AG-120 dose-expansion cohort in low grade glioma expected 2H
- Initiate randomized Phase 2 study of AG-120 in cholangiocarcinoma in 2H

PKR Activators

- ✓ Present first data from AG-348 Phase 2 DRIVE PK and AG-519 Phase 1 healthy volunteer studies at EHA in June
- Present new findings from Natural History Study of PK deficiency in 2H
- Outline clinical development plans for PKR activators in beta-thalassemia in 2H

Research

- ✓ Published on MTAP cancer metabolism program
- Initiate preclinical development activities for MTAP cancer metabolism program