



DEDICATED TO ADVANCING
CARE FOR PATIENTS WITH
KIDNEY DISEASE

CAUTIONARY NOTE ON FORWARD-LOOKING STATEMENTS

Statements in this presentation regarding Akebia's strategy, plans, prospects, expectations, beliefs, intentions or goals are forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995, as amended, including but not limited to statements regarding the expected period of time our cash resources and estimated product revenue will fund operations; the timing, availability and presentation of clinical trial data and results; the commercial potential, growth potential and market opportunity for our product and, if approved, our product candidates; our strategy, mission and vision; potential for our product candidates to set a new standard of care; the potential benefits of our product candidates; the timing of enrollment, including full enrollment, of our clinical trials; the target enrollments of our clinical trials; the assessments and evaluations we expect from our clinical programs; the potential to be a partner of choice for innovation in renal; and exploring co-development potential for vadadustat and Auryxia. The terms "estimate," "expect," "growth," "momentum," "mission" "opportunity," "positioned," "potential," "vision" and similar expressions are intended to identify forward-looking statements, although not all forward-looking statements contain these identifying words. Each forward-looking statement is subject to risks and uncertainties that could cause actual results to differ materially from those expressed or implied in such statement, including the actual product revenues for Auryxia; the timing of generic entrants for Auryxia, vadadustat or any other product candidates; the rate of enrollment in clinical studies of vadadustat; the risk that clinical trials may not be successful; the risk that existing preclinical and clinical data may not be predictive of the results of ongoing or later clinical trials; manufacturing risks; the quality and manner of the data that will result from clinical studies of vadadustat; the actual funding

required to develop and commercialize Akebia's product candidates and operate the company, and the actual expenses associated therewith; efficacy, safety and tolerability of our products and product candidates; the risk that clinical studies need to be discontinued for any reason, including for safety, tolerability, enrollment, manufacturing or economic reasons; early termination of any of Akebia's collaborations or license agreements, and the parties' ability to satisfy their obligations under such agreements; the timing and content of decisions made by regulatory, judicial or similar authorities; the timing of any additional studies initiated for vadadustat; the actual time it takes to initiate and complete research and clinical studies; the success of competitors in developing product candidates for diseases for which Akebia is currently developing its product candidates; the scope, timing, and outcome of any ongoing legal proceedings; changes in the economic and financial conditions of the businesses of Akebia and its partners; and Akebia's ability to obtain, maintain and enforce patent and other intellectual property protection for Auryxia, vadadustat and its other product candidates. Other risks and uncertainties include those identified under the heading "Risk Factors" in Akebia's Quarterly Report on Form 10-Q for the quarter ended March 31, 2019, filed with the SEC, and other filings that Akebia may make with the SEC in the future. These forward-looking statements (except as otherwise noted) speak only as of the date of this presentation, and Akebia does not undertake, and specifically disclaims, any obligation to update any forward-looking statements contained in this presentation. Vadadustat is an investigational drug and has not yet been approved by the U.S. Food and Drug Administration (FDA) or any regulatory authority.



Fully Integrated Biotech Company Focused On Kidney Disease

J o h n P . B u t l e r , P r e s i d e n t a n d C E O

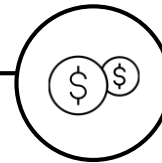
FDA - APPROVED

Product revenue in two FDA-approved indications with substantial growth potential



CASH RESOURCES

Expect cash resources¹ to fund operations into Q3 2020. \$168 million cash position².



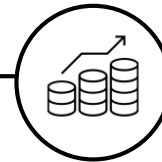
MULTIPLE CLINICAL CATALYSTS

Multiple clinical catalysts over next 12 to 18 months for Phase 3 product candidate with multi-billion-dollar global market opportunity



COMMERCIAL OPERATIONS

Experienced nephrology focused commercial sales and marketing organization



¹ Includes prepaid quarterly committed cost share funding from Akebia's collaborators | ² Cash, cash equivalents and available for sale securities as of 3/31/19.



Significant Strategic Partnerships





OUR VISION

Improve the Health of
Patients with Kidney
Disease through Better
Disease Management
and Novel Therapeutics



CKD On Dialysis

CKD Non-Dialysis

C O M M E R C I A L I Z E D

**HYPERPHOSPHATEMIA
IN DIALYSIS**

Auryxia®
(Ferric Citrate)
FDA approved in two indications

**IRON DEFICIENCY ANEMIA
IN NON-DIALYSIS**

**ANEMIA DUE TO
CKD IN DIALYSIS**

Vadadustat
(An Investigational HIF PHl inhibitor)
In global Phase 3 with read-outs
beginning 2019

**ANEMIA DUE TO CKD IN
NON-DIALYSIS**

D E V E L O P M E N T P R O G R A M



Auryxia

 **Akebia**
THERAPEUTICS



Dialysis

AURYXIA OPPORTUNITY IN
THE HYPERPHOSPHATEMIA
MARKET:

Patient Dissatisfaction With
Standard Of Care And Nephrologist
Positive Perception



Approximately 20% of
Patients Discontinue
Use of Renvela¹



65%

Cite Pill
Burden



42%

Cite Tolerability
Issues



Nephrologists View
Auryxia's Profile Favorably
Compared to Other Binders



Nephrologists
anticipate
prescribing

35%

more Auryxia 6
months from now²



When Asked:

"If you yourself (nephrologist)
had to take a phosphate binder,
which would you choose?"³

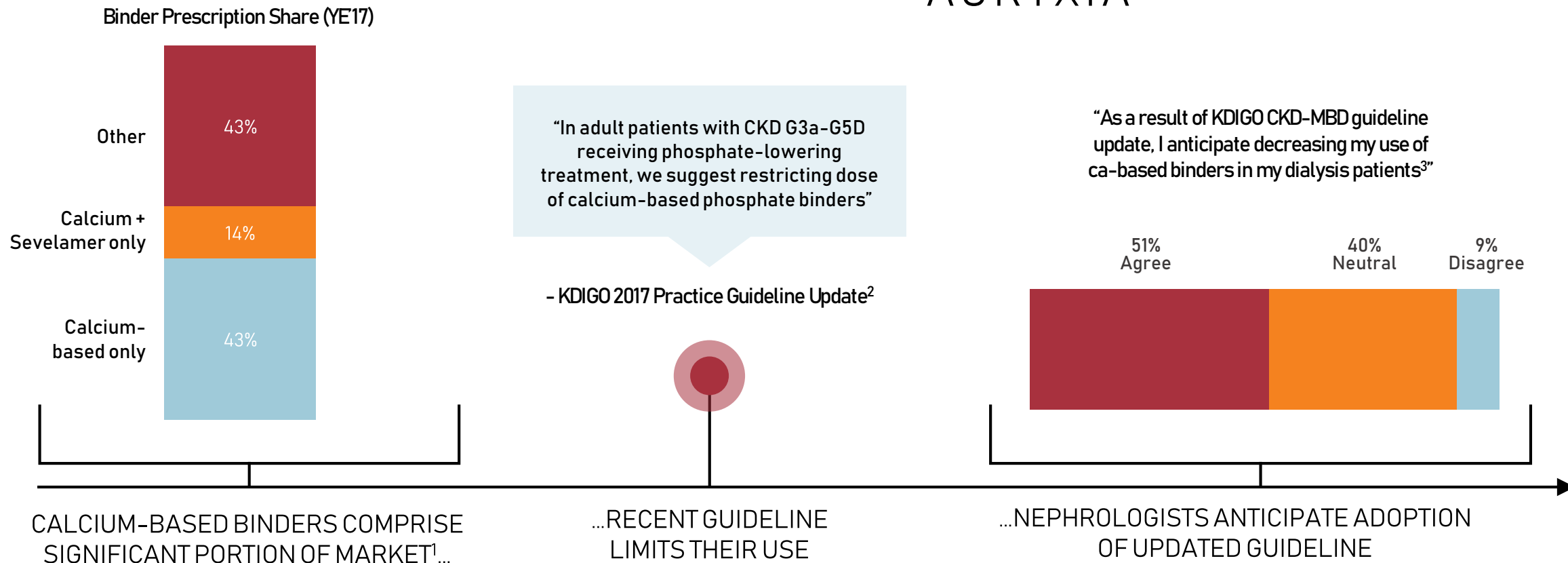
35%

Chose Auryxia

SOURCES: 1 Spherix Global Insights Bone and Mineral Metabolism, quantitative market research survey, Q4 2018, n=195 nephrologists, "Considering your use of Auryxia, Velphoro and Renvela, what percent of all the patients you prescribed these agents for in the past six months have since discontinued the brand (e.g. have been switched to a different brand or discontinued altogether)?", "To what extent are the following reasons that patients typically discontinue AURYXIA, VELPHORO, RENVELA 1= Rarely/Never, 5= Very Frequently/ Almost Always"; 2 Auryxia ATU Q4 2018, n = 104 nephrologists.; 3 Spherix Global Insights Bone and Mineral Metabolism, quantitative market research survey, Q4 2018, n=195 nephrologists.

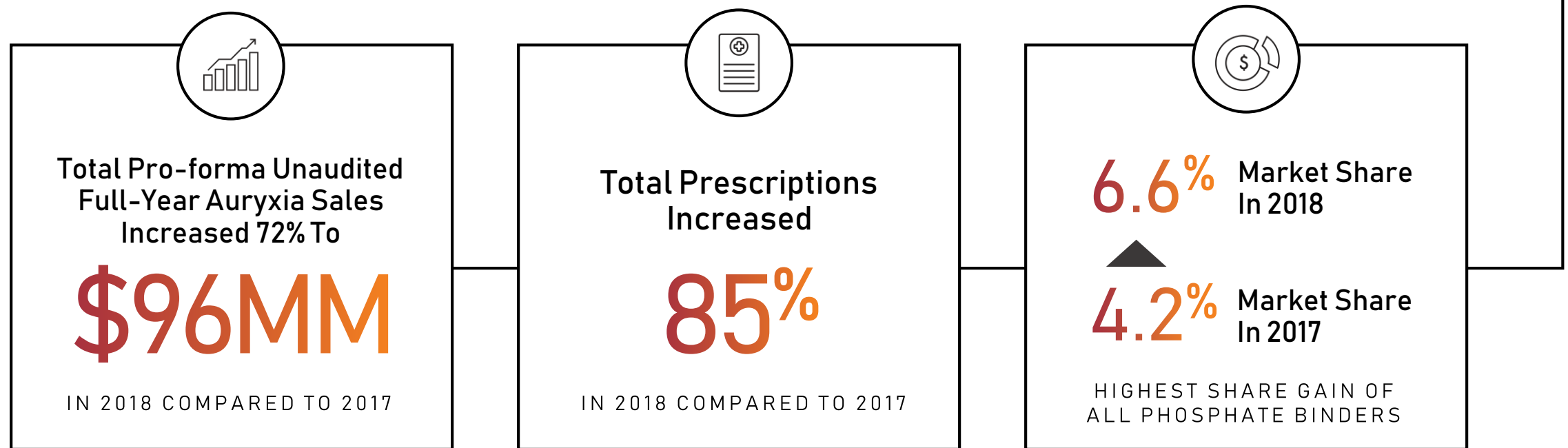
Dialysis

KDIGO TREATMENT GUIDELINE UPDATE FAVORABLE FOR AURYXIA



SOURCES: 1 Phosphate Binder Use, by type. DOPPS Practice Monitor. <https://www.dopps.org/dpm/DPMSlideBrowser.aspx>. Accessed 12.21.2018.; 2 KDIGO 2017 Clinical Practice Guideline Update for the Diagnosis, Evaluation, Prevention, and Treatment of Chronic Kidney Disease–Mineral and Bone Disorder (CKD-MBD), *Kidney International Supplements* (2017) 7, 1–59.; 3 Spherix Global Insights Bone and Mineral Metabolism, quantitative market research survey, Q4 2018, n=195 nephrologists, “Please rate your level of agreement with the following statement: As a result of KDIGO CKD-MBD guideline update, I anticipate decreasing my use of ca-based binders in my dialysis patients”.

AURYXIA: SOLID PERFORMANCE



Well Positioned to Drive Continued Growth in 2019

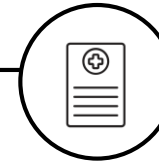
Source: IQVIA NPA, Dialysis SPTRx Data *2018 Annual Auryxia revenue is pro-forma unaudited combined full-year Auryxia sales including sales recorded by Keryx prior to the merger on December 12, 2018.

AURYXIA: IMPROVING Q2FY19 OUTLOOK



Highest prescription
demand in first
4 WEEKS
of any quarter since
Auryxia launch

May 9, 2019 Q1FY19 Press
Release



Highest prescription
demand in first
8 WEEKS
of any quarter since
Auryxia launch

June 4, 2019 Jefferies
Conference Presentation

Well Positioned to Drive Continued Growth in 2019



Non-Dialysis

AURYXIA OPPORTUNITY IN THE
IRON DEFICIENCY ANEMIA (IDA)
MARKET:

Nephrologists Believe Auryxia Has
Strong Potential In IDA



Nephrologist Perspective On
Auryxia to Potentially
Address The Unmet Need¹

48% Very likely to use after
traditional oral iron failure

44% Very likely to use in place
of traditional oral iron

Nephrologist Perspective
On How Auryxia Compares
to Traditional Oral Iron²

● Ferrous sulfate is significantly better (1, 2) ● Ferrous sulfate and Auryxia are similar (3) ● Auryxia is significantly better (4, 5)

GI Tolerability*



Improvement in TSAT**



Improvement in Hb



Disclaimer: Not Based On Head-to-head Data

* Gastrointestinal; ** Transferrin saturation SOURCES: 1 Auryxia ATU market research survey Q3 2018 (n= 102 nephrologists).; 2 Auryxia ATU market research survey Q4 2018 (n= 74 nephrologists), only polled nephrologists currently using Auryxia.

Non-Dialysis

DR. GEOFF BLOCK ET AL
PRESENTED RESULTS FROM
AN INVESTIGATOR-
SPONSORED STUDY OF FERRIC
CITRATE AT ERA-EDTA 2018

- Single-center, open-label, randomized trial comparing ferric citrate (FC) and standard of care in subjects with advanced non-dialysis CKD (Stage 4/5)
- Assessed hypothesis that “provision of fixed dose FC to subjects with advanced CKD, independent of serum phosphate or degree of anemia, would improve multiple biochemical aspects simultaneously and reduce the need for exogenous ESA or intravenous (IV) iron”¹
- Baseline characteristics were comparable with the exception of diabetes (FC 47%, SOC 77%, p=0.001)¹
- Accepted for publication

A Randomized Trial of Ferric Citrate in Advanced Chronic Kidney Disease

Geoffrey A Block, Martha Persky, Gerard Smits, Laura Kooienga, Rupal Mehta, Tamara Isakova, Myles Wolf and Glenn Chertow

“The data from this study suggest that administering ferric citrate to late-stage pre-dialysis patients not only improves biochemical parameters associated with chronic kidney disease, but also has the potential to delay the need for dialysis,” said Geoffrey Block, M.D., Director of Clinical Research at Denver Nephrology.

“With the impact of ferric citrate across multiple aspects of CKD, it is worth further investigation to determine which of these many factors is contributing to the reduced risk of renal replacement therapy observed in this study.”

*Keryx Biopharmaceuticals, Inc. (now a wholly owned subsidiary of Akebia) provided funding and study medication for this investigator initiated study.; SOURCE 1 Geoffrey Block, Martha Block, Gerard Smits, Laura Kooienga, Rupal Mehta, Tamara Isakova, Myles Wolf, Glenn Chertow. Randomized trial of the effects of ferric citrate in patients with advanced chronic kidney disease. ERA-EDTA 2018.

Vadadustat



A woman with short dark hair, wearing safety glasses and a white lab coat over a pink shirt, is smiling while using a blue pipette in a laboratory. Another person with blonde hair is partially visible in the foreground on the right. The background shows laboratory equipment and shelves.

Vadadustat

AN INVESTIGATIONAL
ORAL HIF PH INHIBITOR
DESIGNED TO STIMULATE
ENDOGENOUS EPO
PRODUCTION

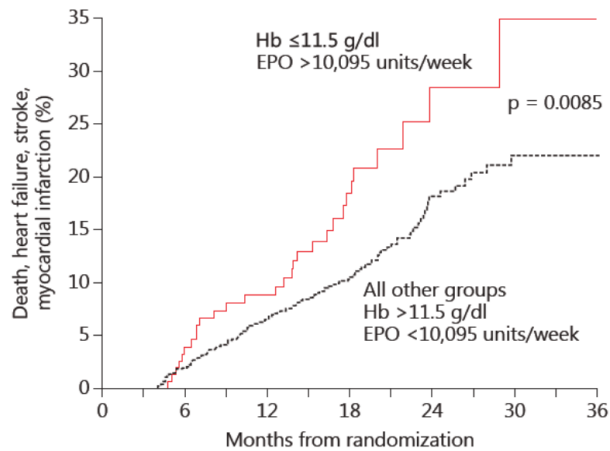
with the potential to establish
a new standard of care

Vadadustat now in Phase 3 cardiovascular outcomes trials for anemia due to CKD in non-dialysis and dialysis subjects. Vadadustat is an investigational HIF PH inhibitor that is not approved by the FDA or any other regulatory authority.

Vadadustat is an oral HIF PH inhibitor designed to stimulate endogenous EPO production, with the potential to increase hemoglobin while avoiding supra-physiological EPO levels

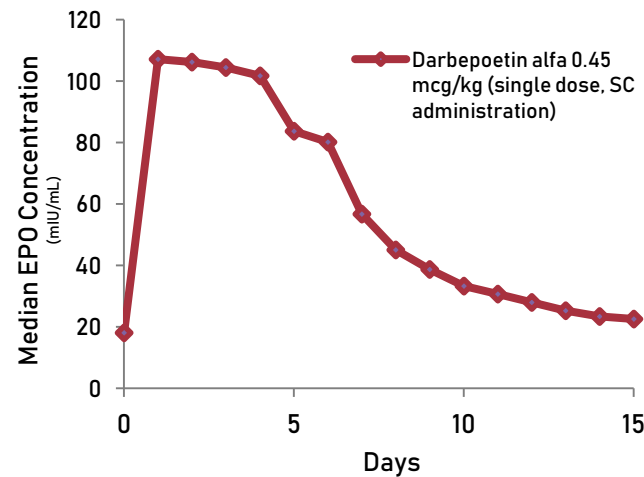
HIGH EPO LEVEL IS
ASSOCIATED WITH
INCREASED CV RISK

Kaplan-Meier Survival Curves¹
Death, Heart Failure, Stroke, Myocardial Infarction



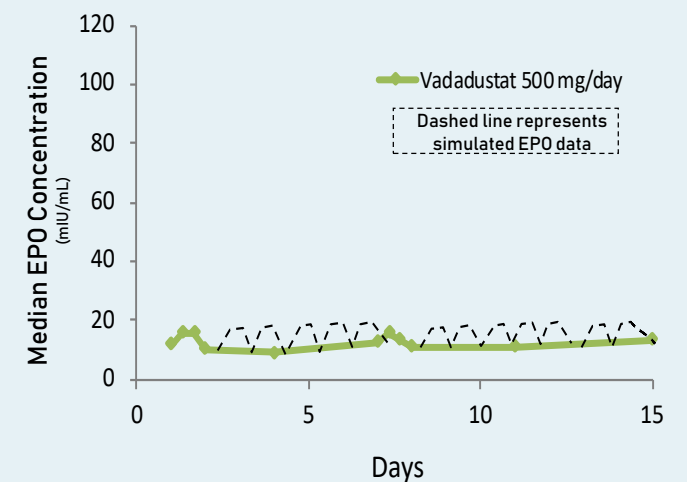
SoC WITH INJECTABLE
ESAs RESULT IN SUPRA-
PHYSIOLOGICAL EPO LEVELS

EPO vs Time with Darbepoetin Alfa (SC)
PK-PD Model in CKD Subjects²



VADADUSTAT, ORAL HIF PH
INHIBITOR, AVOIDED SUPRA-
PHYSIOLOGICAL EPO LEVELS

EPO vs Time with vadadustat (Oral)
Phase 1 Study in Healthy Volunteers³



SOURCES: 1 McCullough P.A., et al. Am J Nephrol 2013;37:549-558 (DOI:10.1159/000351175); Permission granted by S. Karger AG, Basel.; 2 Doshi S et al. Journal of Clinical Pharmacology, 2010;50:75S-90S. Original figure redrawn to depict darbepoetin alfa serum concentration (ng/mL/(mcg/kg)) converted to mIU/mL. Data from 6 clinical studies conducted with extensive PK sampling in CKD patients following subcutaneous (SC) administration of a single dose or first dose of a monthly dosing regimen ranging from 0.4-0.6mcg/kg, dose normalized to 0.45 mcg/kg.; 3 Akebia Therapeutics, Inc. Data on File (2010). Data from Phase 1 study in healthy volunteers with vadadustat once daily dosing. Pre-dose EPO concentrations were evaluated on Days 1, 4, 7, 11, 15 and 22. Post-dose data to assess acute rise in EPO following vadadustat dosing was only completed on Day 1 and Day 7 (8 and 16 hours post-dose). Dashed line represents estimated EPO levels based on post-dose data from Day 1 and Day 7.

Non-Dialysis

GLOBAL PHASE 3
PROGRAM FOR
VADADUSTAT FOR
NON-DIALYSIS

NDA/MAA Core Package

Cardiovascular Outcomes PRO₂TECT Phase 3 Studies
Anemia due to CKD in Non-Dialysis Dependent Subjects

PRO₂TECT

CORRECTION

Not ESA Treated
Vadadustat vs
Darbepoetin Alfa

N ≈ 3,700

PRO₂TECT

CONVERSION

ESA Treated
Vadadustat vs
Darbepoetin Alfa

PRIMARY EFFICACY ENDPOINT:
Change in hemoglobin (Hb) from baseline

PRIMARY SAFETY ENDPOINT:
Major Adverse Cardiovascular Events (MACE)



Non-Dialysis

GLOBAL PHASE 3
PROGRAM FOR
VADADUSTAT FOR
NON-DIALYSIS

Additional Non-Primary Endpoints

INDIVIDUAL MACE COMPONENTS

CHRONIC HEART FAILURE

THROMBOTIC EVENTS

HOSPITALIZATIONS

HB EXCURSIONS

TIME IN RANGE (HB)

IV IRON USE

CKD PROGRESSION

Dialysis

GLOBAL PHASE 3
PROGRAM FOR
VADADUSTAT
FOR DIALYSIS

NDA/MAA Core Package

Cardiovascular Outcomes INNO₂VATE Phase 3 Studies
Anemia due to CKD in Dialysis Subjects

INNO₂VATE

CORRECTION
CONVERSION

New-Onset Dialysis
Vadadustat vs
Darbepoetin Alfa

N ≈ 3,900

INNO₂VATE

CONVERSION

ESA Treated
Vadadustat vs
Darbepoetin Alfa


PRIMARY EFFICACY ENDPOINT:
Change in hemoglobin (Hb) from baseline

PRIMARY SAFETY ENDPOINT:
Major Adverse Cardiovascular Events (MACE)



Dialysis

GLOBAL PHASE 3
PROGRAM FOR
VADADUSTAT
FOR DIALYSIS



Additional Ongoing/Planned Studies to Support Value Proposition

FO₂WARD-2

Phase 2, Open-label, efficacy, safety, PK/PD in DD¹-CKD, control arm epoetin alfa, modified once daily (QD) and Three Times a Week (TIW) dosing, and ESA Hyporesponders,

EXPLOR₂E

Open-label, Sponsor-Blind, parallel arm study evaluating efficacy and safety of Modified QD dosing of vadadustat compared to ESA

TRILOR₂GY-2

Open-label, Sponsor-Blind, parallel arm study evaluating efficacy and safety of TIW dosing of vadadustat compared to ESA

¹Dialysis-dependent

Dialysis



Unique market dynamics with treatment adoption driven by protocols



Agreement with Vifor (International) Ltd. positions vadadustat for rapid uptake in Fresenius LDO subject to FDA approval*



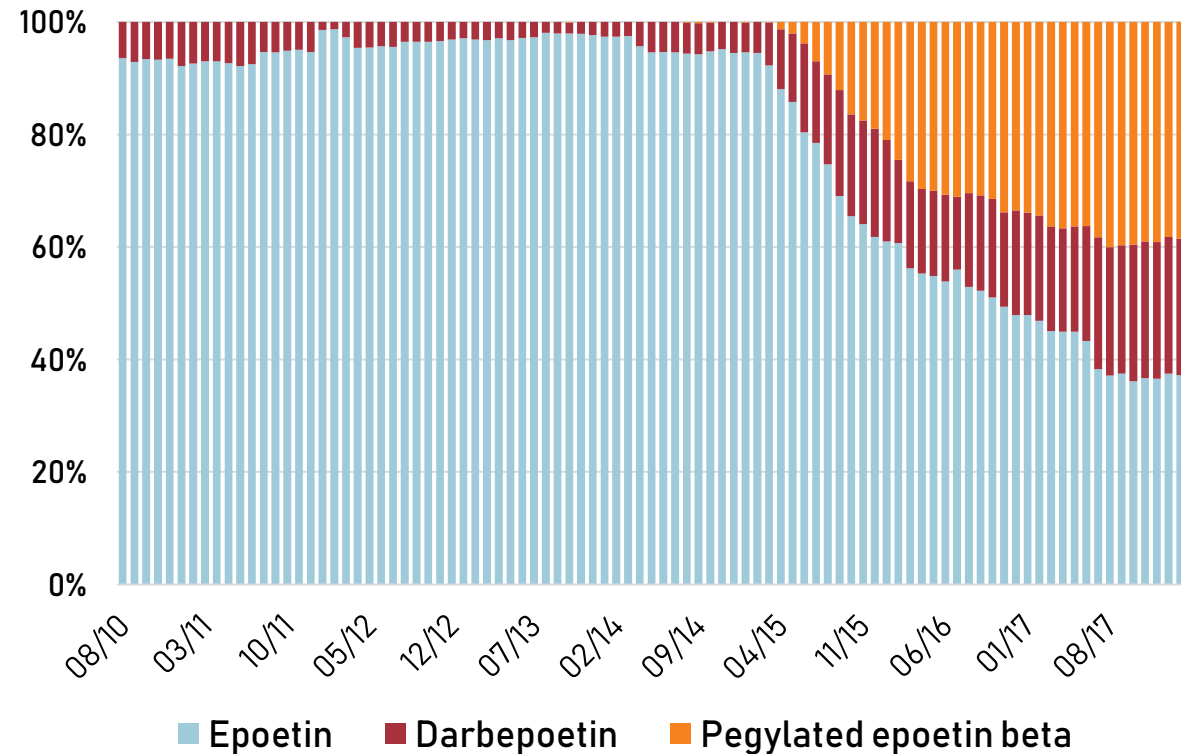
Recent amendment expands agreement, facilitating access to up to 60% of U.S dialysis patients



TDAPA² creates major additional opportunity for value creation for Akebia

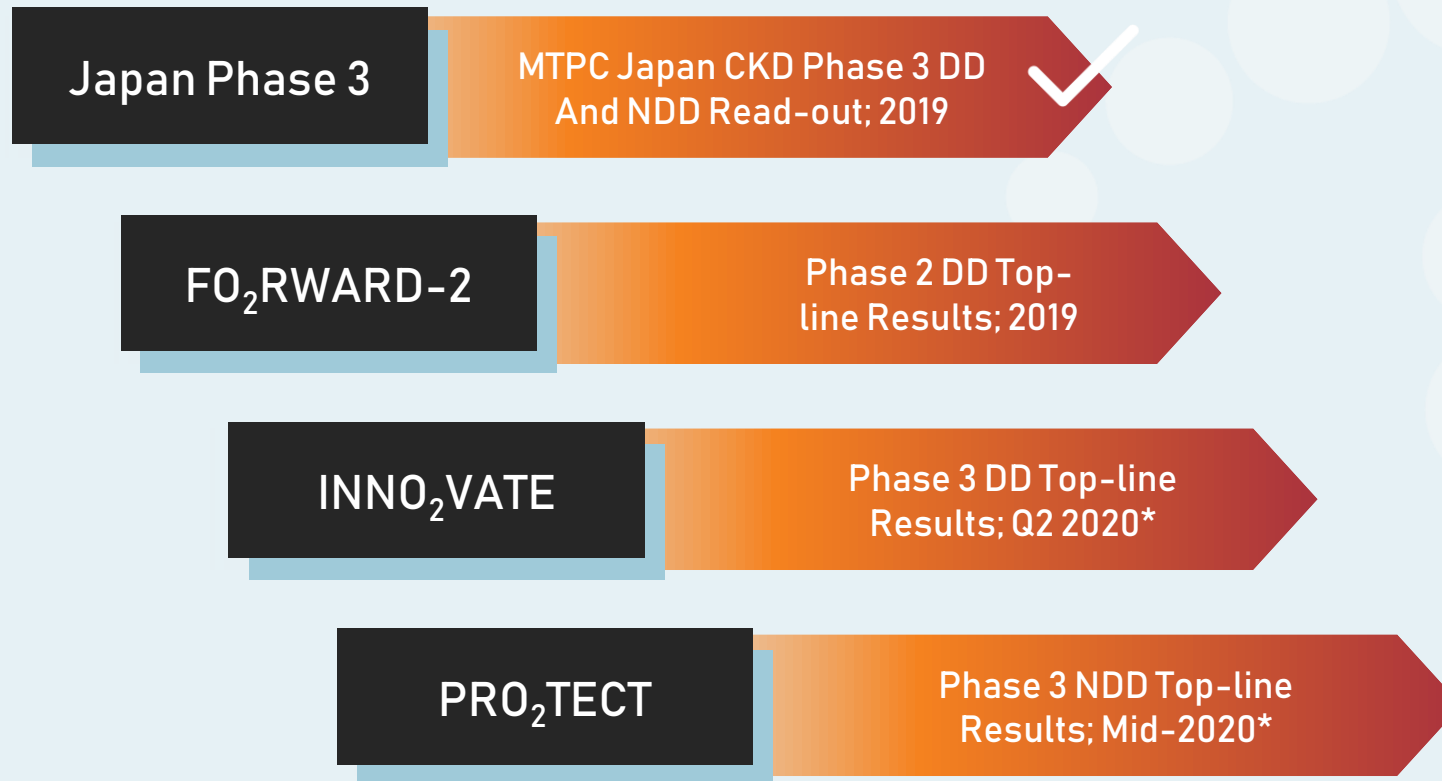
STRONG DISTRIBUTION CHANNEL TO U.S. DIALYSIS NETWORK

Mircera Uptake into Fresenius Dialysis Centers¹



*Subject to the earlier of reimbursement under TDAPA (defined below) or inclusion in the ESRD bundle. SOURCES: 1 ESA Use, by type. DOPPS Practice Monitor. <https://www.dopps.org/dpm/DPMSlideBrowser.aspx>. Accessed 12/19/2018.; 2 TDAPA: Transitional drug add-on payment adjustment, CMS Ruling CMS-1691-F. Medicare Program; End-Stage Renal Disease Prospective Payment System, Payment for Renal Dialysis Services Furnished to Individuals with Acute Kidney Injury, End-Stage Renal Disease Quality Incentive Program, Durable Medical Equipment, Prosthetics, Orthotics and Supplies (DMEPOS) Competitive Bidding Program (CBP) and Fee Schedule Amounts, and Technical Amendments to Correct Existing Regulations Related to the CBP for Certain DMEPOS.

Expected Near-Term Clinical Catalysts



*Subject to accrual of MACE events.



THANK YOU | QUESTIONS?